

The GALE ENCYCLOPEDIA *of Medicine*

FOURTH EDITION

VOLUME

5

P-S



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MEDICINE

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LAURIE J. FUNDUKIAN, EDITOR



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A

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Abscess incision and drainage
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Achondroplasia
Acid phosphatase test
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Actinomycosis
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Antimyocardial antibody test	Asperger syndrome	
Antinausea drugs	Aspergillosis	
Antinuclear antibody test	Aspirin	

B

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Bacillary angiomatosis
Bacteremia
Bacterial vaginosis
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Balance and coordination tests
Balanitis
Balantidiasis
Balloon valvuloplasty
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Barbiturate-induced coma
Barbiturates
Bariatric surgery
Barium enema
Bartholin's gland cyst
Bartonellosis
Basal cell carcinoma
Battered child syndrome
Bedbug infestation
Bedsores
Bedwetting

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Bejel	Borderline personality disorder	Cancer therapy, supportive
Bence Jones protein test	Botulinum toxin injections	Cancer vaccines
Bender-Gestalt test	Botulism	Candidiasis
Benzodiazepines	Bowel preparation	Canker sores
Bereavement	Bowel resection	Carbohydrate intolerance
Beriberi	Bowel training	Carbon monoxide poisoning
Berylliosis	Brain abscess	Carcinoembryonic antigen test
Beta ₂ -microglobulin test	Brain biopsy	Carcinogens
Beta blockers	Brain tumor	Cardiac blood pool scan
Bile duct cancer	Breast biopsy	Cardiac catheterization
Biliary atresia	Breast cancer	Cardiac rehabilitation
Binge eating	Breast implants	Cardiac tamponade
Biofeedback	Breast reconstruction	Cardiomyopathy
Bipolar disorder	Breast reduction	Cardiopulmonary resuscitation
Birth defects	Breast self-examination	Cardioversion
Birthmarks	Breast ultrasound	Carotid sinus massage
Bites and stings	Breastfeeding	Carpal tunnel syndrome
Black lung disease	Breast-feeding problems	Cataract surgery
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Bladder stones	Bronchiectasis	Cataracts
Bladder training	Bronchiolitis	Catatonia
Blastomycosis	Bronchodilators	Catecholamines tests
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Bone grafting	Calcium	Cervicitis
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Bone scan	Cancer therapy, definitive	

C

C-reactive protein
Caffeine
Calcium
Calcium channel blockers
Campylobacteriosis
Cancer
Cancer therapy, definitive

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Charcot Marie Tooth disease	Cognitive-behavioral therapy	Coronary stenting
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Chelation therapy	Cold sores	Corticosteroids, dermatologic
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Chemotherapy	Colitis	Corticosteroids, systemic
Chest drainage therapy	Colon cancer	Cortisol tests
Chest physical therapy	Colonic irrigation	Cosmetic dentistry
Chest x ray	Colonoscopy	Costochondritis
Chickenpox	Color blindness	Cough
Child abuse	Colostomy	Cough suppressants
Childbirth	Colposcopy	Couvade syndrome
Childhood obesity	Coma	Cox-2 inhibitors
Children's health	Common cold	Craniopharyngioma
Chiropractic	Common variable immunodeficiency	Craniosacral therapy
Chlamydial pneumonia	Complement deficiencies	Craniotomy
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Cholangitis	Computed tomography scans	Creatinine test
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Cholera	Conduct disorder	Crohn's disease
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Cholesterol-reducing drugs	Congenital brain defects	Cryptococcosis
Cholinergic drugs	Congenital heart disease	Cryptosporidiosis
Chondromalacia patellae	Congenital hip dysplasia	CT-guided biopsy
Choriocarcinoma	Congenital lobar emphysema	Culture-fair test
Chorionic villus sampling	Congenital ureter anomalies	Cushing's syndrome
Chronic fatigue syndrome	Congestive cardiomyopathy	Cutaneous larva migrans
Chronic granulomatous disease	Congestive heart failure	Cutaneous T-cell lymphoma
Chronic kidney failure	Conjunctivitis	Cutis laxa
Chronic obstructive pulmonary disease	Constipation	Cyanosis
Circumcision	Contact dermatitis	Cyclic vomiting syndrome
Cirrhosis	Contractures	Cyclosporiasis
Cleft lip and palate	Cooling treatments	Cystectomy
Clenched fist injury	Coombs' tests	Cystic fibrosis
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Clubfoot	Corneal abrasion	Cystitis
Cluster headache	Corneal transplantation	Cystometry
Coagulation disorders	Corneal ulcers	Cystoscopy
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Coccyx injuries		

D

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Death
Debridement
Decompression sickness
Decongestants
Deep vein thrombosis
Defibrillation
Dehydration
Delayed hypersensitivity skin test
Delirium
Delusions
Dementia
Dengue fever
Dental fillings
Dental implants
Dental sealants
Dental trauma
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Diabetes mellitus
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Diabetic ketoacidosis
Diabetic neuropathy
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Diarrhea
Diets
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DiGeorge syndrome
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Diphtheria
Discoid lupus erythematosus
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Dissociative disorders
Distal pancreatectomy
Diuretics
Diverticulosis and diverticulitis
Dizziness
Doppler ultrasonography
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Drug metabolism/interactions
Drug overdose
Drug therapy monitoring
Drugs used in labor
Dry mouth
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Dysentery
Dysfunctional uterine bleeding
Dyslexia
Dysmenorrhea
Dyspareunia
Dyspepsia
Dysphasia

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Electromyography
Electronic fetal monitoring
Electrophysiology study of the heart
Elephantiasis
Embolism
Emergency contraception
Emphysema
Empyema
Encephalitis
Encopresis
Endarterectomy
Endocarditis
Endometrial biopsy
Endometrial cancer
Endometriosis
Endorectal ultrasound
Endoscopic retrograde cholangiopancreatography
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Enlarged prostate
Enterobacterial infections
Enterobiasis
Enterostomy
Enterovirus infections
Enzyme therapy
Eosinophilic pneumonia
Epidermolysis bullosa
Epididymitis
Epiglottitis
Epilepsy
Episiotomy
Epstein-Barr virus
Epstein-Barr virus test
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Erectile dysfunction treatment
Erysipelas
Erythema multiforme
Erythema nodosum
Erythroblastosis fetalis
Erythrocyte sedimentation rate

E

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Ear, nose, and throat surgery
Ear surgery
Eating disorders
Echinacea
Echinococcosis
Echocardiography
Ectopic pregnancy
Eczema
Edema
Edwards' syndrome
Ehlers-Danlos syndrome
Ehrlichiosis
Elder abuse
Electric shock injuries
Electrical nerve stimulation
Electrical stimulation of the brain
Electrocardiography
Electroconvulsive therapy
Electroencephalography
Electrolyte disorders
Electrolyte supplements

Erythromycins and macrolide antibiotics	Fetal hemoglobin test	Gallstones
Erythropoietin test	Fever	Gamma globulin
Escherichia coli	Fever evaluation tests	Gamma knife surgery
Esophageal atresia	Fever of unknown origin	Ganglion
Esophageal cancer	Fibrin split products	Gangrene
Esophageal disorders	Fibrinogen test	Gas embolism
Esophageal function tests	Fibroadenoma	Gastrectomy
Esophageal pouches	Fibrocystic condition of the breast	Gastric acid determination
Esophagogastroduodenoscopy	Fibromyalgia	Gastric bypass
Evoked potential studies	Fifth disease	Gastric emptying scan
Exercise	Filariasis	Gastrinoma
Exophthalmos	Finasteride	Gastritis
Expectorants	Fingertip injuries	Gastroenteritis
External sphincter electromyography	First aid	Gastroesophageal reflux disease
Extracorporeal membrane oxygenation	Fish and shellfish poisoning	Gastrostomy
Eye and orbit ultrasounds	Fistula	Gaucher disease
Eye cancer	Flesh-eating disease	Gay and lesbian health
Eye examination	Flower remedies	Gender identity disorder
Eye glasses and contact lenses	Fluke infections	Gene therapy
Eye muscle surgery	Fluoroquinolones	General adaptation syndrome
Eyelid disorders	Folic acid	General surgery

F

Facelift
Factitious disorders
Failure to thrive
Fainting
Familial Mediterranean fever
Familial polyposis
Family therapy
Fanconi's syndrome
Fasciotomy
Fasting
Fatigue
Fatty liver
Fecal incontinence
Fecal occult blood test
Feldenkrais method
Female genital mutilation
Female orgasmic disorder
Female sexual arousal disorder
Fetal alcohol syndrome

Fetal alcohol syndrome	Fetal hemoglobin test
Fever	Fever evaluation tests
Fever of unknown origin	Fever of unknown origin
Fibrin split products	Fibrinogen test
Fibrinogen test	Fibroadenoma
Fibroadenoma	Fibrocystic condition of the breast
Fibromyalgia	Fibromyalgia
Fifth disease	Fifth disease
Filariasis	Filariasis
Finasteride	Finasteride
Fingertip injuries	Fingertip injuries
First aid	First aid
Fish and shellfish poisoning	Fish and shellfish poisoning
Fistula	Fistula
Flesh-eating disease	Flesh-eating disease
Flower remedies	Flower remedies
Fluke infections	Fluke infections
Fluoroquinolones	Fluoroquinolones
Folic acid	Folic acid
Folic acid deficiency anemia	Folic acid deficiency anemia
Follicle-stimulating hormone test	Follicle-stimulating hormone test
Folliculitis	Folliculitis
Food allergies	Food allergies
Food poisoning	Food poisoning
Foot care	Foot care
Foreign objects	Foreign objects
Fracture repair	Fracture repair
Fractures	Fractures
Fragile X syndrome	Fragile X syndrome
Friedreich's ataxia	Friedreich's ataxia
Frostbite and frostnip	Frostbite and frostnip
Fugu poisoning	Fugu poisoning

G

Galactorrhea	Galactosemia
Galactosemia	Gallbladder cancer
Gallbladder cancer	Gallbladder nuclear medicine scan
Gallbladder nuclear medicine scan	Gallbladder x rays
Gallbladder x rays	Gallium scan of the body
Gallium scan of the body	Gallstone removal
Gallstone removal	Gallstones
Gallstones	Gamma globulin
Gamma globulin	Gamma knife surgery
Gamma knife surgery	Ganglion
Ganglion	Gangrene
Gangrene	Gas embolism
Gas embolism	Gastrectomy
Gastrectomy	Gastric acid determination
Gastric acid determination	Gastric bypass
Gastric bypass	Gastric emptying scan
Gastric emptying scan	Gastrinoma
Gastrinoma	Gastritis
Gastritis	Gastroenteritis
Gastroenteritis	Gastroesophageal reflux disease
Gastroesophageal reflux disease	Gastrostomy
Gastrostomy	Gaucher disease
Gaucher disease	Gay and lesbian health
Gay and lesbian health	Gender identity disorder
Gender identity disorder	Gene therapy
Gene therapy	General adaptation syndrome
General adaptation syndrome	General surgery
General surgery	Generalized anxiety disorder
Generalized anxiety disorder	Genetic counseling
Genetic counseling	Genetic testing
Genetic testing	Genital herpes
Genital herpes	Genital warts
Genital warts	Germ cell tumors
Germ cell tumors	Gestalt therapy
Gestalt therapy	Gestational diabetes
Gestational diabetes	GI bleeding studies
GI bleeding studies	Giardiasis
Giardiasis	Ginkgo biloba
Ginkgo biloba	Ginseng
Ginseng	Glaucoma
Glaucoma	Glomerulonephritis
Glomerulonephritis	Glucose-6-phosphate dehydrogenase deficiency
Glucose-6-phosphate dehydrogenase deficiency	Gluten-free diet
Gluten-free diet	Glycogen storage diseases
Glycogen storage diseases	Glycosylated hemoglobin test
Glycosylated hemoglobin test	Goiter
Goiter	Gonorrhea
Gonorrhea	Goodpasture's syndrome
Goodpasture's syndrome	Gout
Gout	Gout drugs
Gout drugs	Graft-vs.-host disease

H

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Group therapy	Hematocrit	Homocysteine
Growth hormone tests	Hemochromatosis	Hookworm disease
Guided imagery	Hemoglobin electrophoresis	Hormone replacement therapy
Guillain-Barré syndrome	Hemoglobin test	Hospital-acquired infections
Guinea worm infection	Hemoglobinopathies	HPV vaccination
Gulf War syndrome	Hemolytic-uremic syndrome	Human-potential movement
Gynecomastia (male breast enlargement)	Hemolytic anemia	Human bite infections
	Hemophilia	Human chorionic gonadotropin pregnancy test
	Hemophilus infections	Human leukocyte antigen test
	Hemoptysis	Human papilloma virus
	Hemorrhagic fevers	Huntington's disease
	Hemorrhoids	Hydatidiform mole
	Hepatitis A	Hydrocelectomy
	Hepatitis, alcoholic	Hydrocephalus
	Hepatitis, autoimmune	Hydronephrosis
	Hepatitis B	Hydrotherapy
	Hepatitis C	Hyperaldosteronism
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	Hepatitis E	Hypercholesterolemia
	Hepatitis G	Hypercoagulation disorders
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	Herbalism, Western	Hyperkalemia
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	Hernia	Hyperopia
	Hernia repair	Hyperparathyroidism
	Herniated disk	Hyperpigmentation
	Hiatal hernia	Hypersensitivity pneumonitis
	Hiccups	Hypersplenism
	High-risk pregnancy	Hypertension
	Hirschsprung's disease	Hyperthyroidism
	Hirsutism	Hypertrophic cardiomyopathy
	Histiocytosis X	Hyphema
	Histoplasmosis	Hypnotherapy
	Hives	Hypoactive sexual desire disorder
	Hodgkin's lymphoma	Hypocalcemia
	Holistic medicine	Hypochondriasis
	Holter monitoring	Hypoglycemia
	Holtzman ink blot test	Hypogonadism
	Homeopathic medicine	Hypokalemia
	Homeopathic medicine, acute prescribing	Hypolipoproteinemia

Hyponatremia
Hypoparathyroidism
Hypophysectomy
Hypopituitarism
Hypospadias and epispadias
Hypotension
Hypothermia
Hypothyroidism
Hypotonic duodenography
Hysterectomy
Hysteria
Hysterosalpingography
Hysteroscopy
Hysterosonography

I

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Idiopathic primary renal hematuric/proteinuric syndrome
Idiopathic thrombocytopenic purpura
Ileus
Immobilization
Immune complex test
Immunodeficiency
Immunoelectrophoresis
Immunoglobulin deficiency syndromes
Immunologic therapies
Immunosuppressant drugs
Impacted tooth
Impedance phlebography
Impetigo
Implantable cardioverter-defibrillator
Impotence
Impulse control disorders
In vitro fertilization
Inclusion conjunctivitis
Incompetent cervix
Indigestion
Indium scan of the body
Induction of labor

Infant massage
Infection control
Infectious arthritis
Infectious disease
Infectious mononucleosis
Infertility
Infertility drugs
Infertility therapies
Influenza
Influenza vaccination
Inhalants and related disorders
Inhalation therapies
Insecticide poisoning
Insomnia
Insulin resistance
Intermittent claudication
Intermittent explosive disorder
Intersex states
Interstitial microwave thermal therapy
Intestinal obstructions
Intestinal polyps
Intrauterine growth retardation
Intravenous rehydration
Intravenous urography
Intussusception
Ipecac
Iron deficiency anemia
Iron tests
Irritable bowel syndrome
Ischemia
Isolation
Itching
IUD

J

Japanese encephalitis
Jaundice
Jaw wiring
Jet lag
Jock itch
Joint biopsy
Joint fluid analysis
Joint replacement
Juvenile arthritis

K

Kaposi's sarcoma
Kawasaki syndrome
Keloids
Keratitis
Keratosis pilaris
Kidney biopsy
Kidney cancer
Kidney disease
Kidney function tests
Kidney nuclear medicine scan
Kidney stones
Kidney transplantation
Kidney, ureter, and bladder x-ray study
Kinesiology, applied
Klinefelter syndrome
Knee injuries
Kneecap removal
KOH test
Korsakoff's syndrome
Kyphosis

L

Labyrinthitis
Laceration repair
Lacrimal duct obstruction
Lactate dehydrogenase isoenzymes test
Lactate dehydrogenase test
Lactation
Lactic acid test
Lactose intolerance
Laminectomy
Laparoscopy
Laryngeal cancer
Laryngectomy
Laryngitis
Laryngoscopy
Laser surgery
Late effects of childhood cancer and its treatment
Laxatives
Lead poisoning

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Leeches	Lyme disease	Mediterranean diet
Legionnaires' disease	Lymph node biopsy	Medullary sponge kidney
Leishmaniasis	Lymphadenitis	Melioidosis
Leprosy	Lymphangiography	Memory loss
Leptospirosis	Lymphedema	Ménière's disease
Lesch-Nyhan syndrome	Lymphocyte typing	Meningitis
Leukemia stains	Lymphocytic choriomeningitis	Meningococcemia
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Leukemias, chronic	Lymphogranuloma venereum	Men's health
Leukocytosis	Lysergic acid diethylamide (LSD)	Menstrual disorders
Leukotriene inhibitors		Mental retardation
Lice infestation		Mental status examination
Lichen planus		Mercury poisoning
Lichen simplex chronicus		Mesothelioma
Life support		Metabolic acidosis
Light therapy		Metabolic alkalosis
Lipase test		Methadone
Lipidoses		Methamphetamine
Lipoproteins test		Methemoglobinemia
Liposuction		Microphthalmia and anophthalmia
Listeriosis		Mifepristone
Lithotripsy		Migraine headache
Liver biopsy		Mineral deficiency
Liver cancer		Mineral toxicity
Liver disease		Minerals
Liver encephalopathy		Minnesota multiphasic personality inventory (MMPI-2)
Liver function tests		Minority health
Liver nuclear medicine scan		Minoxidil
Liver transplantation		Miscarriage
Lobectomy		Mitral valve insufficiency
Low back pain		Mitral valve prolapse
Low sugar diet		Mitral valve stenosis
Lower esophageal ring		Moles
Lumbar puncture		Monkeypox
Lumpectomy		Monoamine oxidase inhibitors
Lung abscess		Mood disorders
Lung biopsy		Motion sickness
Lung cancer, non-small cell		Movement disorders
Lung cancer, small cell		Movement therapy
Lung diseases due to gas or chemical exposure		MRSA infections
Lung perfusion and ventilation scan		Mucopolysaccharidoses
Lung surgery		Mucormycosis
Lung transplantation		Multiple chemical sensitivity

M

Macular degeneration
Magnesium imbalance
Magnetic field therapy
Magnetic resonance imaging
Malabsorption syndrome
Malaria
Malignant lymphomas
Malignant melanoma
Malingering
Mallet finger
Mallory-Weiss syndrome
Malnutrition
Malocclusion
MALT lymphoma
Mammography
Mania
Marfan syndrome
Marijuana
Marriage counseling
Marshall-Marchetti-Krantz procedure
Massage therapy
Mastectomy
Mastitis
Mastocytosis
Mastoidectomy
Mastoiditis
Maternal to fetal infections
Maxillofacial trauma
Measles
Meckel's diverticulum
Mediastinoscopy

Multiple-gated acquisition (MUGA) scan	Nasal trauma	Occupational therapy
Multiple endocrine neoplasia syndromes	Nasogastric suction	Oil spills: health effects
Multiple myeloma	Nasopharyngeal culture	Oligomenorrhea
Multiple personality disorder	Naturopathic medicine	Omega-3 fatty acids
Multiple pregnancy	Nausea and vomiting	Onychomycosis
Multiple sclerosis	Near-drowning	Oophorectomy
Mumps	Necrotizing enterocolitis	Ophthalmoplegia
Munchausen syndrome	Neonatal jaundice	Oppositional defiant disorder
Muscle relaxants	Nephrectomy	Optic atrophy
Muscle spasms and cramps	Nephritis	Optic neuritis
Muscular dystrophy	Nephrotic syndrome	Oral contraceptives
Mushroom poisoning	Nephrotoxic injury	Oral hygiene
Music therapy	Neuralgia	Orbital and periorbital cellulitis
Mutism	Neuroblastoma	Orchitis
Myasthenia gravis	Neuroendocrine tumors	Organ donation
Mycetoma	Neurofibromatosis	Organic food
Mycobacterial infections, atypical	Neurogenic bladder	Orthodontics
Mycoplasma infections	Neurolinguistic programming	Orthopedic surgery
Myelodysplastic syndrome	Neurological exam	Orthostatic hypotension
Myelofibrosis	Neurosurgery	Osteoarthritis
Myelography	Neutropenia	Osteochondroses
Myers-Briggs type indicator	Nicotine and related disorders	Osteogenesis imperfecta
Myocardial biopsy	Night terrors	Osteomyelitis
Myocardial resection	Nitrogen narcosis	Osteopathy
Myocarditis	Nocardiosis	Osteopetroses
Myoglobin test	Nongonococcal urethritis	Osteoporosis
Myomectomy	Non-nucleoside reverse transcriptase inhibitors	Ostomy
Myopathies	Nonsteroidal anti-inflammatory drugs	Otitis externa
Myopia	Noroviruses	Otitis media
Myositis	Nosebleed	Otosclerosis
Myotonic dystrophy	Numbness and tingling	Ototoxicity
Myringotomy and ear tubes	Nutrition	Ovarian cancer
Myxoma	Nutrition through an intravenous line	Ovarian cysts
	Nutritional supplements	Ovarian torsion
	Nystagmus	Overactive bladder
		Overhydration
		Oxygen/ozone therapy

N

Nail-patella syndrome
Nail removal
Narcolepsy
Narcotics
Nasal irrigation
Nasal packing
Nasal papillomas
Nasal polyps

O

Obesity
Obesity surgery
Obsessive-compulsive disorder
Obstetrical emergencies
Occupational asthma

P

Pacemakers
Paget's disease of bone
Paget's disease of the breast
Pain
Pain management
Palliative care

Palpitations	Peroxisomal disorders	Pneumonia
Pancreas transplantation	Personality disorders	Pneumothorax
Pancreatectomy	Pervasive developmental disorders	Poison ivy and poison oak
Pancreatic cancer, endocrine	Pet therapy	Poisoning
Pancreatic cancer, exocrine	Peyronie's disease	Polarity therapy
Pancreatitis	Pharmacogenetics	Polio
Panic disorder	Phenylketonuria	Polycystic kidney disease
Pap test	Pheochromocytoma	Polycystic ovary syndrome
Papilledema	Phimosis	Polycythemia vera
Paracentesis	Phlebotomy	Polydactyly and syndactyly
Paralysis	Phobias	Polyglandular deficiency syndromes
Paranoia	Phosphorus imbalance	Polyhydramnios and oligohydramnios
Parathyroid hormone test	Photodynamic therapy	Polymyalgia rheumatica
Parathyroid scan	Photorefractive keratectomy and laser-assisted in-situ keratomileusis	Polymyositis
Parathyroidectomy	Photosensitivity	Polysomnography
Paratyphoid fever	Phototherapy	Porphyrias
Parkinson's disease	Physical allergy	Portal vein bypass
Parotidectomy	Physical examination	Positron emission tomography (PET)
Paroxysmal atrial tachycardia	Physical therapy	Post-concussion syndrome
Parrot fever	Pica	Post-traumatic stress disorder
Partial thromboplastin time	Pickwickian syndrome	Postmenopausal bleeding
Paruresis	Piercing and tattoos	Postpartum depression
Patau syndrome	Pilates	Postpolio syndrome
Patent ductus arteriosus	Pinguecula and pterygium	Prader-Willi syndrome
Pellagra	Pinta	Precocious puberty
Pelvic exam	Pituitary dwarfism	Preeclampsia and eclampsia
Pelvic fracture	Pituitary tumors	Pregnancy
Pelvic inflammatory disease	Pityriasis rosea	Premature ejaculation
Pelvic relaxation	Placenta previa	Premature labor
Pelvic ultrasound	Placental abruption	Premature menopause
Penicillins	Plague	Premature rupture of membranes
Penile cancer	Plasma renin activity	Prematurity
Penile prostheses	Plasmapheresis	Premenstrual dysphoric disorder
Percutaneous transhepatic cholangiography	Plastic, reconstructive, and cosmetic surgery	Premenstrual syndrome
Perforated eardrum	Platelet aggregation test	Prenatal surgery
Perforated septum	Platelet count	Preparing for surgery
Pericardiocentesis	Platelet function disorders	Prepregnancy counseling
Pericarditis	Pleural biopsy	Presbyopia
Perinatal infection	Pleural effusion	Priapism
Periodic paralysis	Pleurisy	Prickly heat
Periodontal disease	Pneumococcal pneumonia	Primary biliary cirrhosis
Peripheral neuropathy	Pneumocystis pneumonia	Proctitis
Peripheral vascular disease	Pneumonectomy	Progressive multifocal leukoencephalopathy
Peritonitis		
Pernicious anemia		

Progressive supranuclear palsy
 Prolactin test
 Prolonged QT syndrome
 Prophylaxis
 Prostate biopsy
 Prostate cancer
 Prostate ultrasound
 Prostatectomy
 Prostate-specific antigen test
 Prostatitis
 Protease inhibitors
 Protein components test
 Protein electrophoresis
 Protein-energy malnutrition
 Prothrombin time
 Proton pump inhibitors
 Provence (sipuleucel-T)
 Pseudogout
 Pseudomonas infections
 Pseudoxanthoma elasticum
 Psoriasis
 Psoriatic arthritis
 Psychiatric confinement
 Psychoanalysis
 Psychological tests
 Psychosis
 Psychosocial disorders
 Psychosurgery
 Psychotherapy
 Ptosis
 Puberty
 Puerperal infection
 Pulmonary alveolar proteinosis
 Pulmonary artery catheterization
 Pulmonary edema
 Pulmonary embolism
 Pulmonary fibrosis
 Pulmonary function tests
 Pulmonary hypertension
 Pulmonary valve insufficiency
 Pulmonary valve stenosis
 Pyelonephritis
 Pyloric stenosis
 Pyloroplasty
 Pyruvate kinase deficiency

Q

Q fever
 Qigong

R

Rabies
 Radial keratotomy
 Radiation injuries
 Radiation therapy
 Radical neck dissection
 Radioactive implants
 Rape and sexual assault
 Rashes
 Rat-bite fever
 Raynaud's disease
 Recompression treatment
 Rectal cancer
 Rectal examination
 Rectal polyps
 Rectal prolapse
 Recurrent miscarriage
 Red blood cell indices
 Red reflex testing
 Reflex sympathetic dystrophy
 Reflex tests
 Reflexology
 Rehabilitation
 Reiki
 Reiter's syndrome
 Relapsing fever
 Relapsing polychondritis
 Renal artery occlusion
 Renal artery stenosis
 Renal tubular acidosis
 Renal vein thrombosis
 Renovascular hypertension
 Respiratory acidosis
 Respiratory alkalosis
 Respiratory distress syndrome
 Respiratory failure
 Respiratory syncytial virus infection
 Restless legs syndrome

Restrictive cardiomyopathy
 Reticulocyte count
 Retinal artery occlusion
 Retinal detachment
 Retinal hemorrhage
 Retinal vein occlusion
 Retinitis pigmentosa
 Retinoblastoma
 Retinopathies
 Retrograde cystography
 Retrograde ureteropyelography
 Retrograde urethrography
 Retropubic suspension
 Reye's syndrome
 Rheumatic fever
 Rheumatoid arthritis
 Rhinitis
 Rhinoplasty
 Riboflavin deficiency
 Rickets
 Rickettsialpox
 Ringworm
 Rocky Mountain spotted fever
 Rolfering
 Root canal treatment
 Rosacea
 Roseola
 Ross River Virus
 Rotator cuff injury
 Rotavirus infections
 Roundworm infections
 Rubella
 Rubella test

S

Sacroiliac disease
 Salivary gland scan
 Salivary gland tumors
 Salmonella food poisoning
 Salpingectomy
 Salpingo-oophorectomy
 Sarcoidosis
 Sarcomas
 Saw palmetto
 Scabies

Scarlet fever	Shingles	Spinal cord injury
Scars	Shock	Spinal cord tumors
Schistosomiasis	Shortness of breath	Spinal instrumentation
Schizoaffective disorder	Shy-Drager syndrome	Spinal stenosis
Schizophrenia	Shyness	Spirometry
Sciatica	Sick sinus syndrome	Splenectomy
Scleroderma	Sickle cell disease	Splenic trauma
Sclerotherapy for esophageal varices	Sideroblastic anemia	Sporotrichosis
Scoliosis	Sudden infant death syndrome	Sports injuries
Scrotal nuclear medicine scan	Sigmoidoscopy	Sprains and strains
Scrotal ultrasound	Sildenafil citrate	Sputum culture
Scrub typhus	Silicosis	Squamous cell carcinoma of the skin
Scurvy	Single photon emission computed tomography	St. John's wort
Seasonal affective disorder	Sinus endoscopy	Stanford-Binet intelligence scales
Seborrheic dermatitis	Sinusitis	Stapedectomy
Secondary polycythemia	Situs inversus	Staphylococcal infections
Sedation	Sitz bath	Staphylococcal scalded skin syndrome
Seizure disorder	Sjogren's syndrome	Starvation
Selective serotonin reuptake inhibitors	Skin biopsy	Stem cell transplantation
Self-mutilation	Skin cancer, non-melanoma	Steroids
Semen analysis	Skin culture	Stillbirth
Seniors' health	Skin grafting	Stockholm syndrome
Sensory integration disorder	Skin lesion removal	Stomach cancer
Sepsis	Skin lesions	Stomach flushing
Septic shock	Skin pigmentation disorders	Stomachache
Septoplasty	Skin resurfacing	Stomatitis
Serum sickness	Skull x rays	Stool culture
Severe acute respiratory syndrome (SARS)	Sleep apnea	Stool fat test
Severe combined immunodeficiency	Sleep deprivation	Stool O and P test
Sex hormones tests	Sleep disorders	Strabismus
Sex reassignment surgery	Sleeping sickness	Strep throat
Sex therapy	Small intestine biopsy	Streptococcal antibody tests
Sexual abuse	Smallpox	Streptococcal infections
Sexual addiction	Smelling disorders	Stress
Sexual dysfunction	Smoke inhalation	Stress reduction
Sexual perversions	Smoking-cessation drugs	Stress test
Sexually transmitted diseases	Smoking	Stridor
Sexually transmitted diseases cultures	Snoring	Stroke
Shaken baby syndrome	Sodium	Stuttering
Shiatsu	Somatoform disorders	Subacute sclerosing panencephalitis
Shigellosis	Sore throat	Subarachnoid hemorrhage
Shin splints	South American blastomycosis	Subdural hematoma
	Speech disorders	Substance abuse and dependence
	Speech therapy	Sudden cardiac death

Suicide	Thoracoscopy	Transfusion
Sulfonamides	Threadworm infection	Transhepatic biliary catheterization
Sunburn	Throat culture	Transient ischemic attack
Sunscreens	Thrombocytopenia	Transplant surgery
Superior vena cava syndrome	Thrombocytosis	Transposition of the great arteries
Surfactant	Thrombolytic therapy	Transurethral bladder resection
Swallowing disorders	Thrombophlebitis	Transvaginal ultrasound
Swollen glands	Thymoma	Transverse myelitis
Sydenham's chorea	Thyroid biopsy	Traumatic amputations
Sympathectomy	Thyroid cancer	Traveler's diarrhea
Syphilis	Thyroid function tests	Tremors
Systemic lupus erythematosus	Thyroid hormones	Trench fever
<hr/>		
T		
Tai chi	Thyroid nuclear medicine scan	Trichinosis
Tanning	Thyroid ultrasound	Trichomoniasis
Tapeworm diseases	Thyroidectomy	Tricuspid valve insufficiency
Tardive dyskinesia	Thyroiditis	Tricuspid valve stenosis
Tarsorrhaphy	Tilt table test	Trigeminal neuralgia
Tay-Sachs disease	Tinnitus	Trigger finger
Technetium heart scan	Tissue typing	Triglycerides
Teeth whitening	Tonsillectomy and adenoidectomy	Triglycerides test
Teething	Tonsillitis	Triple screen
Temporal arteritis	Tooth decay	Tropical spastic paraparesis
Temporomandibular joint disorders	Tooth extraction	Troponins test
Tendinitis	Tooth replacements and restorations	Tubal ligation
Tennis elbow	Toothache	Tube compression of the esophagus and stomach
Tensilon test	Topical anesthesia	Tube feedings
Tension headache	TORCH test	Tuberculin skin test
Testicular cancer	Torticollis	Tuberculosis
Testicular self-examination	Total parenteral nutrition	Tularemia
Testicular surgery	Tourette syndrome	Tumor markers
Testicular torsion	Toxic epidermal necrolysis	Tumor removal
Tetanus	Toxic shock syndrome	Turner syndrome
Tetracyclines	Toxoplasmosis	2,3-diphosphoglycerate test
Tetralogy of Fallot	Trabeculectomy	Typhoid fever
Thalassemia	Tracheoesophageal fistula	Typhus
Thallium heart scan	Tracheotomy	Tzanck preparation
Thematic apperception test	Trachoma	
Therapeutic baths	Traction	
Therapeutic touch	Traditional Chinese medicine	
Thoracentesis	Trager psychophysical integration	
Thoracic outlet syndrome	Trans fatty acids	
Thoracic surgery	Transcranial Doppler ultrasonography	

U

- Ulcer surgery
- Ulcers (digestive)
- Ultraviolet light treatment
- Umbilical cord blood banking
- Umbilical hernia repair

Undernutrition
Undescended testes
Upper GI exam
Ureteral stenting
Urethritis
Uric acid tests
Urinalysis
Urinary anti-infectives
Urinary catheterization
Urinary diversion surgery
Urinary incontinence
Urinary tract infection
Urine culture
Urine flow test
Uterine fibroid embolization
Uterine fibroids
Uveitis

V

Vaccination
Vaginal pain
Vaginismus
Vagotomy
Valsalva maneuver
Valvular heart disease
Varicose veins
Vascular disease
Vascular surgery
Vasculitis
Vasectomy
Vasodilators
Vegetarianism
Vegetative state
Velopharyngeal insufficiency
Vena cava filter
Venography

Venous access
Venous insufficiency
Ventricular aneurysm
Ventricular assist device
Ventricular ectopic beats
Ventricular fibrillation
Ventricular septal defect
Ventricular shunt
Ventricular tachycardia
Vesicoureteral reflux
Vibriosis
Vision training
Visual impairment
Vitamin A deficiency
Vitamin B6 deficiency
Vitamin D deficiency
Vitamin E deficiency
Vitamin K deficiency
Vitamin tests
Vitamin toxicity
Vitamins
Vitiligo
Vitrectomy
Vocal cord nodules and polyps
Vocal cord paralysis
Vomiting
Von Willebrand disease
Vulvar cancer
Vulvodynia
Vulvovaginitis

W

Waldenström's
macroglobulinemia
Warts
Wechsler intelligence test

Wegener's granulomatosis
Weight loss drugs
West Nile virus
Wheezing
Whiplash
White blood cell count and
differential
Whooping cough
Wilderness medicine
Wilms' tumor
Wilson disease
Wiskott-Aldrich syndrome
Withdrawal syndromes
Wolff-Parkinson-White
syndrome
Women's health
Wound culture
Wound flushing
Wounds

X

X-linked agammaglobulinemia
X rays of the orbit

Y

Yaws
Yellow fever
Yersiniosis
Yoga

Z

Zellweger syndrome
Zoonosis

PLEASE READ—IMPORTANT INFORMATION

The *Gale Encyclopedia of Medicine, Fourth Edition* is a health reference product designed to inform and educate readers about a wide variety of health topics such as diseases, disorders and conditions, treatments and diagnostic tests, diets, alternative treatments, and prevention. Gale, Cengage Learning believes the product to be comprehensive, but not necessarily definitive. It is intended to supplement, not replace, consultation with a physician or other healthcare practitioners. While Gale, Cengage Learning has made substantial efforts to provide information that is accurate, comprehensive, and up-to-date, Gale, Cengage Learning

makes no representations or warranties of any kind, including without limitation, warranties of merchantability or fitness for a particular purpose, nor does it guarantee the accuracy, comprehensiveness, or timeliness of the information contained in this product. Readers should be aware that the universe of medical knowledge is constantly growing and changing, and that differences of opinion exist among authorities. Readers are also advised to seek professional diagnosis and treatment for any medical condition, and to discuss information obtained from this book with their healthcare provider.

INTRODUCTION

The *Gale Encyclopedia of Medicine 4 (GEM4)* is a one-stop source for medical information on common medical disorders, conditions, tests, treatments, drugs, and other health-related topics, including high-profile diseases such as AIDS, Alzheimer's disease, cancer, and heart disease. This encyclopedia avoids medical jargon and uses language that laypersons can understand, while still providing thorough coverage of each topic. The *Gale Encyclopedia of Medicine 4* fills a gap between basic consumer health resources, such as single-volume family medical guides, and highly technical professional materials.

Scope

More than 1,800 full-length articles are included in the *Gale Encyclopedia of Medicine 4*, including disorders/conditions, tests/procedures, and treatments/therapies. Many common drugs are also covered, with generic drug names appearing first and brand names following in parentheses—e.g., acetaminophen (Tylenol). Prominent individuals in medicine are highlighted as sidebar biographies that accompany the main topical essays. Articles follow a standardized format that provides information at a glance. Rubrics include:

Disorders/Conditions	Tests/Treatments
Definition	Definition
Demographics	Purpose
Description	Precautions
Causes and symptoms	Description
Diagnosis	Preparation
Treatment	Aftercare
Prognosis	Risks
Prevention	Normal/abnormal results
Resources	Resources
Key terms	Key terms

In recent years, there has been a resurgence of interest in holistic medicine that emphasizes the connection between mind and body. Aimed at achieving

and maintaining good health rather than just eliminating disease, this approach has come to be known as alternative medicine. The *Gale Encyclopedia of Medicine 4* includes a number of essays on alternative therapies, ranging from traditional Chinese medicine to homeopathy and from meditation to aromatherapy. In addition to full essays on alternative therapies, the encyclopedia features specific **Alternative treatment** sections for diseases and conditions that may be helped by complementary therapies. The *Gale Encyclopedia of Medicine 4* also includes entries on diets, nutrition, and general wellness.

Inclusion Criteria

A preliminary list of diseases, disorders, tests, and treatments was compiled from a wide variety of sources, including professional medical guides and textbooks as well as consumer guides and encyclopedias. The general advisory board, made up of public librarians, medical librarians, and consumer health experts, evaluated the topics and made suggestions for inclusion. The list was sorted by category and sent to *GEM4* medical advisors, certified physicians with various medical specialities, for review. Final selection of topics to include was made by the medical advisors in conjunction with the Gale, Cengage Learning editor.

About the Contributors

The essays were compiled by experienced medical writers, including physicians, pharmacists, nurses, and other health care professionals. *GEM4* medical advisors reviewed the completed essays to ensure that they are appropriate, up to date, and medically accurate.

How to Use this Book

The *Gale Encyclopedia of Medicine 4* has been designed with ready reference in mind.

- Straight **alphabetical arrangement** allows users to locate information quickly.
- Bold-faced terms function as **print hyperlinks** that point the reader to related entries in the encyclopedia.
- **Cross-references** placed throughout the encyclopedia direct readers to where information on subjects without entries can be found. Synonyms and acronyms are also cross-referenced.
- Lists of **key terms** are provided where appropriate to define unfamiliar terms or concepts. A **glossary** of key terms is also included at the back of Volume 6.
- Valuable **contact information** for organizations and support groups is included with each entry. The appendix

contains an extensive list of organizations arranged in alphabetical order.

- The **resources section** directs users to additional sources of medical information on a topic.
- A comprehensive **general index** allows users to easily target detailed aspects of any topic, including Latin names.

Graphics

The *Gale Encyclopedia of Medicine 4* is enhanced with 765 images, including photos, charts, tables, and detailed illustrations.

ADVISORY BOARD

An advisory board comprised of medical specialists from a variety of backgrounds provided invaluable assistance in the formulation of this encyclopedia. This advisory board performed a myriad of duties, from defining the scope of coverage to reviewing individual entries for accuracy and accessibility. We would therefore like to express our sincere thanks and appreciation for all of their contributions.

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P

PAC see **Atrial ectopic beats**

Pacemaker implantation see **Pacemakers**

Pacemakers

Definition

A pacemaker is a surgically implanted electronic device that regulates a slow or erratic heartbeat.

Purpose

Pacemakers are implanted to regulate irregular contractions of the heart (arrhythmia). They are most frequently prescribed to speed the heartbeat of patients who have a heart rate well under 60 beats per minute (severe symptomatic bradycardia). They are also used in some cases to slow a fast heart rate (tachycardia).

Precautions

The symptoms of **fatigue** and lightheadedness that are characteristic of bradycardia can also be caused by a number of other medical conditions, including anemia. Certain prescription medications can also slow the heart rate. A doctor should take a complete medical history and perform a full physical work-up to rule out all non-cardiac causes of bradycardia.

Patients with cardiac pacemakers should not undergo a **magnetic resonance imaging** (MRI) procedure. Devices that emit electromagnetic waves (including magnets) may alter pacemaker programming or functioning. A 1997 study found that cellular phones often interfere with pacemaker programming and cause irregular heart rhythm. However, advances in pacemaker design and materials have greatly reduced the risk of pacemaker interference from electromagnetic fields.

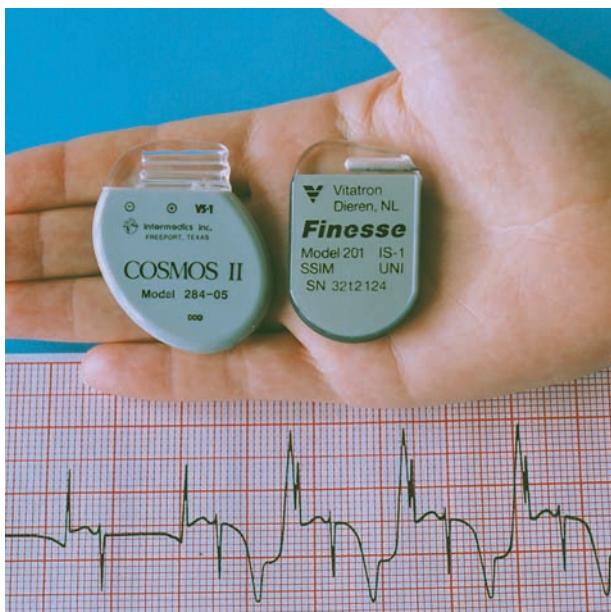
Description

Approximately 500,000 Americans have an implantable permanent pacemaker device. A pacemaker implantation is performed under **local anesthesia** in a hospital by a surgeon assisted by a cardiologist. An insulated wire called a lead is inserted into an incision above the collarbone and guided through a large vein into the chambers of the heart. Depending on the configuration of the pacemaker and the clinical needs of the patient, as many as three leads may be used in a pacing system. Current pacemakers have a double, or bipolar, electrode attached to the end of each lead. The electrodes deliver an electrical charge to the heart to regulate heartbeat. They are positioned on the areas of the heart that require stimulation. The leads are then attached to the pacemaker device, which is implanted under the skin of the patient's chest.

Patients undergoing surgical pacemaker implantation usually stay in the hospital overnight. Once the procedure is complete, the patient's vital signs are monitored and a **chest x ray** is taken to ensure that the pacemaker and leads are properly positioned.

Modern pacemakers have sophisticated programming capabilities and are extremely compact. The smallest weigh less than 13 grams (under half an ounce) and are the size of two stacked silver dollars. The actual pacing device contains a pulse generator, circuitry programmed to monitor heart rate and deliver stimulation, and a lithiumiodide battery. Battery life typically ranges from seven to 15 years, depending on the number of leads the pacemaker is configured with and how much energy the pacemaker uses. When a new battery is required, the unit can be exchanged in a simple outpatient procedure.

A temporary pacing system is sometimes recommended for patients who are experiencing irregular heartbeats as a result of a recent **heart attack** or other acute medical condition. The implantation procedure for the pacemaker leads is similar to that for a



Pacemakers like these are usually implanted under the skin below the collarbone. The pacemaker is connected to the heart by a wire inserted into a major vein in the neck and guided down into the heart. (Eamonn McNulty/Photo Researchers, Inc.)

permanent pacing system, but the actual pacemaker unit housing the pulse generator remains outside the patient's body. Temporary pacing systems may be replaced with a permanent device at a later date.

Preparation

Patients being considered for pacemaker implantation will undergo a full battery of cardiac tests, including an electrocardiogram (ECG) or an electrophysiological study or both to fully evaluate the bradycardia or tachycardia.

Patients are advised to abstain from eating 6–8 hours before the surgical procedure. The patient is usually given a sedative to help him or her relax for the procedure. An intravenous (IV) line will also be inserted into a vein in the patient's arm before the procedure begins in case medication or blood products are required during the insertion.

Aftercare

Pacemaker patients should schedule a follow-up visit with their cardiologist approximately six weeks after the surgery. During this visit, the doctor will make any necessary adjustments to the settings of the pacemaker. Pacemakers are programmed externally with a handheld electromagnetic device. Pacemaker

KEY TERMS

Electrocardiogram (ECG)—A recording of the electrical activity of the heart. An ECG uses externally attached electrodes to detect the electrical signals of the heart.

Electrophysiological study—A test that monitors the electrical activity of the heart in order to diagnose arrhythmia. An electrophysiological study measures electrical signals through a cardiac catheter that is inserted into an artery in the leg and guided up into the atrium and ventricle of the heart.

Embolism—A blood clot, air bubble, or clot of foreign material that blocks the flow of blood in an artery. When an embolism blocks the blood supply to a tissue or organ, the tissue the artery feeds dies (infarction). Without immediate and appropriate treatment, an embolism can be fatal.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct images of internal structures.

batteries must be checked regularly. Some pacing systems allow patients to monitor battery life through a special telephone monitoring service that can read pacemaker signals.

Risks

Because pacemaker implantation is an invasive surgical procedure, internal bleeding, infection, hemorrhage, and **embolism** are all possible complications. Infection is more common in patients with temporary pacing systems. Antibiotic therapy given as a precautionary measure can reduce the risk of pacemaker infection. If infection does occur, the entire pacing system may have to be removed.

The placing of the leads and electrodes during the implantation procedure also presents certain risks for the patient. The lead or electrode could perforate the heart or cause scarring or other damage. The electrodes can also cause involuntary stimulation of nearby skeletal muscles.

A complication known as *pacemaker syndrome* develops in approximately 7% of pacemaker patients with single-chamber pacing systems. The syndrome is characterized by the low blood pressure and **dizziness** that are symptomatic of bradycardia. It can usually be

corrected by the implantation of a dual-chamber pacing system.

Normal results

Pacemakers that are properly implanted and programmed can correct a patient's arrhythmia and resolve related symptoms.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

Paula Anne Ford-Martin

Packed cell volume see **Hematocrit**

Packed red blood cell volume see
Hematocrit



Paget's disease of bone

Definition

Paget's disease of bone (*osteitis deformans*) is the abnormal formation of bone tissue that results in weakened and deformed bones.

Description

Named for Sir James Paget (1814–1899), this disease affects 1–3% of people over 50 years of age, but affects over 10% of people over 80 years of age. Paget's disease can affect one or more bones in the body. Most often, the pelvis, bones in the skull, the long bones (the large bones that make up the arms and legs), and the collarbones are affected by Paget's disease. In addition, the joints between bones (the knees or elbows, for example) can develop arthritis because of this condition.

Paget's disease is characterized by changes in the normal mechanism of bone formation. Bone is a living material made by the body through the continual processes of formation and breakdown (resorption). The combination of these two actions is called remodeling and is used by the body to build bone tissue that is strong and healthy. Strong bones are formed when bone tissue is made up of plate-shaped crystals of **minerals** called hydroxyapatite. Normal wear and tear on the skeletal system is repaired throughout life by the ongoing process of remodeling. In fact, the entire human skeleton is remodeled every five years.

This woman's legs are bowed due to Paget's disease.

(Custom Medical Stock Photo, Inc. Reproduced by permission.)

Healthy bone tissue has an ordered structure that gives the bone its strength. Bones affected by Paget's disease, however, have a structure that is disorganized. This disorganized structure weakens the diseased bone and makes people suffering from this disease more likely to have **fractures**. These fractures are slow to heal.

Paget's disease of bone is most commonly found in Europe, England, Australia, New Zealand, and North America. In these areas, up to 3% of all people over 55 years of age are affected with the disease. It is interesting to note that Paget's disease is rare in Asia, possibly showing that this disease may affect some ethnic groups and geographic areas more than others.

Causes and symptoms

The cause of Paget's disease is not known. Various viruses have been suggested to be involved in this disease, but the relationship between viral infections

and Paget's disease remains uncertain. There also may be a genetic component to this disease since it may appear in more than one person within the same family.

Paget's disease usually begins without any symptoms. And, in its early stages, the symptoms that do occur are often confused with symptoms of arthritis. However, as the disease progresses, bone and joint **pain** develop. A unique feature of Paget's disease is the enlargement of areas of affected bone. This type of enlargement is clearly identifiable on an x ray.

If the bones of the skull are affected by Paget's disease, enlargement of the skull can occur and may result in a loss of hearing. When the long bones in the legs are affected, they can become bent under the body's weight because of their weakness. Little or no injury to a bone can cause fractures in the weakened bones. Fractures that occur when no traumatic injury is present are known as spontaneous fractures.

Although rare, bone **cancer** occurs in less than 1% of patients with Paget's disease. Such cancer is often accompanied by an abrupt increase in the intensity of pain at the diseased site. Unfortunately, this type of cancer has a poor prognosis; the average survival time from the onset of symptoms is generally one to three years.

Diagnosis

Paget's disease is often found when an individual is having x rays taken for medical reasons unrelated to this bone disease. A diagnosis of Paget's disease can also be made when higher than normal levels of a chemical called alkaline phosphatase are found in the blood. Alkaline phosphatase is a substance involved in the bone formation process, so if its levels are abnormally high this indicates that the balance between bone formation and resorption is upset.

Treatment

Treatment, given only when symptoms are present, consists of the following types:

Drugs

Paget's disease is most often treated with drug therapy, with bone pain lessening within weeks of starting the treatment. While non-steroidal anti-inflammatory drugs can reduce bone pain, two additional categories of drugs are used to treat this disease.

HORMONE TREATMENT. Calcitonin, a hormone which is made naturally by the thyroid gland, is used to treat Paget's disease. This chemical rapidly decreases

the amount of bone breakdown or loss (resorption). After approximately two to three weeks of treatment with extra calcitonin, bone pain lessens and new bone tissue forms. Calcitonin is commonly given as daily injections for one month, followed by three injections each week for several additional months. The total dose of calcitonin given to an individual depends upon the amount of disease present and how well the individual's condition responds to the treatment.

Although calcitonin is effective in slowing the progression of Paget's disease, the favorable effects of the drug do not continue for very long once administration of the drug is stopped. In addition, some temporary side effects can occur with this drug. **Nausea** and flushing are the most common side effects and have been found in 20–30% of individuals taking calcitonin. **Vomiting**, **diarrhea**, and abdominal pain can also occur, but these effects are also temporary. A form of calcitonin taken nasally causes fewer side effects, but requires higher doses because less of the drug reaches the diseased bone.

BISPHOSPHONATES. The bisphosphonate group of drugs are drugs that bind directly to bone minerals because of their specific chemical structure. Once bound to the bone, these drugs inhibit bone loss by reducing the action of bone cells that normally degrade bone during the remodeling process. Unlike treatment with calcitonin, the positive effects of increased bone formation and reduced pain can continue for many months or even years after bisphosphonate treatment is stopped. Bisphosphonates are considered the treatment of choice for Paget's disease and are usually given for 3–6 months at a time.

Bisphosphonate drugs suitable for the treatment of Paget's disease are alendronate, clodronate, etidronate, pamidronate, risedronate, and tiludronate. The main side effects of these drugs include a flu-like reaction (pamidronate), gastrointestinal disturbances (alendronate, clodronate), and abnormal bone formation (etidronate, when taken in high doses). Risedronate is the newest of these drugs. It is about 1,000 times more potent than etidronate and 3 to 5 times more potent than alendronate. Because of the greater potency of this drug, lower doses and a shorter duration of treatment are required. This leads to fewer side effects with similar, or better, clinical results in the patient.

Surgery

Treatment of Paget's disease usually begins with drug therapy. However, various surgical treatments

KEY TERMS

Bisphosphonate—A class of drugs used to treat Paget's disease. These drugs bind to the minerals in bone tissue and lessen the amount of bone loss associated with Paget's disease.

Calcitonin—A naturally occurring hormone made by the thyroid gland that can be used as a drug to treat Paget's disease.

Remodeling—The ongoing process of bone formation and breakdown that results in healthy bone development.

can also be used to treat skeletal conditions that occur in patients with Paget's disease.

In patients with severe arthritis of the hip or knee, a **joint replacement** operation can be beneficial. However, in addition to the malformation of bone tissue caused by this condition, there are greater numbers of blood vessels that form in the diseased bone relative to a healthy bone. This makes surgery on bones affected with Paget's disease more difficult.

Prognosis

There is no cure for Paget's disease. However, the development of potent bisphosphonate drugs like risedronate has resulted in the ability to slow the progress of the disease.

Paul A. Johnson, Ed.M.

Paget's disease of the breast

Definition

Paget's disease of the breast is a rare form of **breast cancer** which makes up approximately 1–4 % of all breast tumors. While sharing its name with **Paget's disease of bone**, these are two medically unrelated conditions. They are simply named after the same doctor who first described them.

Description

Paget's disease of the breast is generally associated with an underlying breast **cancer**. It is generally seen

in people between the ages of 40 and 80 years. Cases in men have been identified, but they are extremely rare.

Paget's disease of the breast may also be called **mammary Paget's disease (MPD)**. There is a much rarer form of this disease called **extramammary Paget's disease (EMPD)**. MPD affects the breast nipple and is also called **Paget's disease of the nipple**. EMPD can affect the skin of the external genital tissues in both women and men, as well as the skin of the eyelids and external ear canal. MPD is believed to develop from a tumor growth within the milk ducts of the breast. EMPD may represent a spreading (metastasis) of MPD to other parts of the body.

Causes and symptoms

The cause of Paget's disease of the breast is unknown, but it is usually associated with an underlying cancer of the breast.

The symptoms of Paget's disease of the breast include:

- red scaly patches of skin on the nipple and sometimes also on the dark area of skin around the nipple (areola)
- crusting, bleeding, or ulceration of the skin of the affected area
- a discharge of fluid from the nipple
- a turning inward (inversion) of the nipple

In approximately 30–40% of cases of Paget's disease of the breast, there is also a detectable lump in the breast.

Diagnosis

Paget's disease of the breast is often confused with other skin conditions, such as **eczema**, **dermatitis**, or **psoriasis**. These misdiagnoses often lead to delays in appropriate treatment. Misdiagnosis is more common when both breast are affected and no lump in the breast is detected. When only one breast is affected, or when the presence of a lump in the breast is also detected, a correct initial diagnosis is more likely to occur.

Once Paget's disease of the breast is suspected, it can be definitively confirmed by biopsy of the affected tissue. In this procedure, a small piece of the affected skin and the underlying tissue is removed and sent to a laboratory for examination under a microscope. The shape and other characteristics of the cells in the biopsied sample will allow the laboratory personnel to determine if the sample is affected with Paget's disease of the breast, or some other condition.

KEY TERMS

Metastasis—The spread of a cancer from one part of the body (where the cancer originated) to another part of the body.

Ulceration—The formation of an ulcer, a patch of tissue that is discontinuous with the surrounding tissue because the tissue within the ulcer has decayed or died and been swept away.

Topical steroid creams are usually used to treat eczema, dermatitis, and psoriasis. These creams will have no effect on the skin conditions caused by Paget's disease of the breast.

Treatment

Surgery is the main treatment for Paget's disease of the breast. Removal of the breast (**mastectomy**) may be recommended if the cancer is seen in a wide area away from the nipple or appears to be deep into the breast tissue. Breast conservation surgery, aimed at keeping as much of the breast as possible, may be recommended in cases where the disease is diagnosed early enough and the cancer has not spread far from the surface of the nipple.

Some people will require further treatment after surgery. This treatment may include **radiation therapy**, **chemotherapy**, or a combination of both. Radiation therapy involves using high-energy x rays to destroy any cancer cells that may remain after surgical removal of the primary tumor. Radiation therapy is most common after breast conservation surgery. Chemotherapy involves the use of medicinal drugs to destroy the growth of any cancer cells that may remain after removal of the primary cancer. Chemotherapy treatments are most common after mastectomy.

Alternative treatment

Alternative treatments for Paget's disease of the breast include: the use of cartilage from cows or sharks; a diet known as Gerson therapy; administration of the chemicals hydrazine sulfate or laetrile; and, the injection of solutions derived from the mistletoe plant.

Prognosis

The prognosis for Paget's disease of the breast depends on the underlying cancer that is causing this

condition and whether or not this cancer has spread (metastasized) to other parts of the body.

Prevention

Because the cause of Paget's disease of the breast is not known, prevention of this disease is not possible. In instances where this condition arises from other underlying cancers of the breast, it may be possible to prevent Paget's disease of the breast from occurring if the underlying cause is diagnosed and successfully treated prior to the development of Paget's disease of the breast.

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Ruth, Laura. *Paget's Disease: A Rare Form of Breast Cancer*. May 12, 2001. <http://users.cnmnetwork.com/~lrs1/paget.htm>.

ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329, (800) 227-2345, <http://www.cancer.org>.

National Breast Cancer Coalition, 1101 17th Street, NW, Suite 1300, Washington, DC, 20036, (202) 296-7477, (202) 265-6854, (800) 622-2838, <http://stopbreastcancer.org>.

Paul A. Johnson, Ed.M.

Pain

Definition

Pain is an unpleasant feeling that is conveyed to the brain by sensory neurons. The discomfort signals actual or potential injury to the body. However, pain is more than a sensation, or the physical awareness of pain; it also includes perception, the subjective interpretation of the discomfort. Perception gives information on the pain's location, intensity, and something about its nature. The various conscious and unconscious responses to both sensation and perception, including

the emotional response, add further definition to the overall concept of pain.

Description

Pain arises from any number of situations. Injury is a major cause, but pain may also arise from an illness. It may accompany a psychological condition, such as depression, or may even occur in the absence of a recognizable trigger.

Acute pain

Acute pain often results from tissue damage, such as a skin burn or broken bone. Acute pain can also be associated with headaches or **muscle cramps**. This type of pain usually goes away as the injury heals or the cause of the pain (stimulus) is removed.

To understand acute pain, it is necessary to understand the nerves that support it. Nerve cells, or neurons, perform many functions in the body. Although their general purpose, providing an interface between the brain and the body, remain constant, their capabilities vary widely. Certain types of neurons are capable of transmitting a pain signal to the brain.

As a group, these pain-sensing neurons are called nociceptors, and virtually every surface and organ of the body is wired with them. The central part of these cells is located in the spine, and they send threadlike projections to every part of the body. Nociceptors are classified according to the stimulus that prompts them to transmit a pain signal. Thermoreceptive nociceptors are stimulated by temperatures that are potentially tissue damaging. Mechanoreceptive nociceptors respond to a pressure stimulus that may cause injury. Polymodal nociceptors are the most sensitive and can respond to temperature and pressure. Polymodal nociceptors also respond to chemicals released by the cells in the area from which the pain originates.

Nerve cell endings, or receptors, are at the front end of pain sensation. A stimulus at this part of the nociceptor unleashes a cascade of neurotransmitters (chemicals that transmit information within the nervous system) in the spine. Each neurotransmitter has a purpose. For example, substance P relays the pain message to nerves leading to the spinal cord and brain. These neurotransmitters may also stimulate nerves leading back to the site of the injury. This response prompts cells in the injured area to release chemicals that not only trigger an immune response, but also influence the intensity and duration of the pain.

Chronic and abnormal pain

Chronic pain refers to pain that persists after an injury heals, **cancer** pain, pain related to a persistent or degenerative disease, and long-term pain from an unidentifiable cause. It is estimated that one in three people in the United States will experience chronic pain at some point in their lives. Of these people, approximately 50 million are either partially or completely disabled.

Chronic pain may be caused by the body's response to acute pain. In the presence of continued stimulation of nociceptors, changes occur within the nervous system. Changes at the molecular level are dramatic and may include alterations in genetic transcription of neurotransmitters and receptors. These changes may also occur in the absence of an identifiable cause; one of the frustrating aspects of chronic pain is that the stimulus may be unknown. For example, the stimulus cannot be identified in as many as 85% of individuals suffering lower back pain.

Scientists have long recognized a relationship between depression and chronic pain. In 2004, a survey of California adults diagnosed with major depressive disorder revealed that more than one-half of them also suffered from chronic pain.

Other types of abnormal pain include allodynia, hyperalgesia, and phantom limb pain. These types of pain often arise from some damage to the nervous system (neuropathic). Allodynia refers to a feeling of pain in response to a normally harmless stimulus. For example, some individuals who have suffered nerve damage as a result of viral infection experience unbearable pain from just the light weight of their clothing. Hyperalgesia is somewhat related to allodynia in that the response to a painful stimulus is extreme. In this case, a mild pain stimulus, such as a pin prick, causes a maximum pain response. Phantom limb pain occurs after a limb is amputated; although an individual may be missing the limb, the nervous system continues to perceive pain originating from the area.

Causes and symptoms

Pain is the most common symptom of injury and disease, and descriptions can range in intensity from a mere ache to unbearable agony. Nociceptors have the ability to convey information to the brain that indicates the location, nature, and intensity of the pain. For example, stepping on a nail sends an information-packed message to the brain: the foot has experienced a puncture wound that hurts a lot.

Pain perception also varies depending on the location of the pain. The kinds of stimuli that cause a pain response on the skin include pricking, cutting, crushing, burning, and freezing. These same stimuli would not generate much of a response in the intestine. Intestinal pain arises from stimuli such as swelling, inflammation, and distension.

Diagnosis

Pain is considered in view of other symptoms and individual experiences. An observable injury, such as a broken bone, may be a clear indicator of the type of pain a person is suffering. Determining the specific cause of internal pain is more difficult. Other symptoms, such as **fever** or **nausea**, help narrow down the possibilities. In some cases, such as lower back pain, a specific cause may not be identifiable. Diagnosis of the disease causing a specific pain is further complicated by the fact that pain can be referred to (felt at) a skin site that does not seem to be connected to the site of the pain's origin. For example, pain arising from fluid accumulating at the base of the lung may be referred to the shoulder.

Since pain is a subjective experience, it may be very difficult to communicate its exact quality and intensity to other people. There are no diagnostic tests that can determine the quality or intensity of an individual's pain. Therefore, a medical examination will include a lot of questions about where the pain is located, its intensity, and its nature. Questions are also directed at what kinds of things increase or relieve the pain, how long it has lasted, and whether there are any variations in it. An individual may be asked to use a pain scale to describe the pain. One such scale assigns a number to the pain intensity; for example, 0 may indicate no pain, and 10 may indicate the worst pain the person has ever experienced. Scales are modified for infants and children to accommodate their level of comprehension.

Treatment

There are many drugs aimed at preventing or treating pain. Nonopioid **analgesics**, narcotic analgesics, **anticonvulsant drugs**, and **tricyclic antidepressants** work by blocking the production, release, or uptake of neurotransmitters. Drugs from different classes may be combined to handle certain types of pain.

Nonopioid analgesics include common over-the-counter medications such as **aspirin**, **acetaminophen** (Tylenol), and ibuprofen (Advil). These are most often

used for minor pain, but there are some prescription-strength medications in this class.

Narcotic analgesics are only available with a doctor's prescription and are used for more severe pain, such as cancer pain. These drugs include codeine, morphine, and **methadone**. **Addiction** to these painkillers is not as common as once thought. Many people who genuinely need these drugs for pain control typically do not become addicted. However, narcotic use is usually limited to patients thought to have a short life span (such as people with terminal cancer) or patients whose pain is only expected to last for a short time (such as people recovering from surgery). In 2004, the Drug Enforcement Administration (DEA) issued new guidelines to help physicians prescribe **narcotics** appropriately without fear of being arrested for prescribing the drugs beyond the scope of their medical practice. DEA is trying to work with physicians to ensure that those who need to drugs receive them but to ensure opioids are not abused.

Anticonvulsants, as well as **antidepressant drugs**, were initially developed to treat seizures and depression, respectively. However, it was discovered that these drugs also have pain-killing applications. Furthermore, since in cases of chronic or extreme pain, it is not unusual for an individual to suffer some degree of depression; antidepressants may serve a dual role. Commonly prescribed anticonvulsants for pain include phenytoin, carbamazepine, and clonazepam. Tricyclic antidepressants include doxepin, amitriptyline, and imipramine.

Intractable (unrelenting) pain may be treated by injections directly into or near the nerve that is transmitting the pain signal. These root blocks may also be useful in determining the site of pain generation. As the underlying mechanisms of abnormal pain are uncovered, other pain medications are being developed.

Drugs are not always effective in controlling pain. Surgical methods are used as a last resort if drugs and local anesthetics fail. The least destructive surgical procedure involves implanting a device that emits electrical signals. These signals disrupt the nerve and prevent it from transmitting the pain message. However, this method may not completely control pain and is not used frequently. Other surgical techniques involve destroying or severing the nerve, but the use of this technique is limited by side effects, including unpleasant **numbness**.

Two effective **pain management** treatments that have been used for generations are heat and cold. Both are used to treat acute and chronic pain. Ice is

generally used to treat inflammation, especially acute injuries to knees and other joints. Treatment usually lasts three to five days. Often it is used as part of the RICE regimen: rest, ice, compression, and elevation. Heat therapy is generally used for increasing tensile strength, increasing blood flow to the injured area, and helping muscles and tendons to relax. Sometimes ice is used in the early stages of an acute injury and then heat for the remainder of treatment. In recent years, scientists have identified heat and cold receptors in the body. This has allowed the development of medications, including patches, creams, and gels, that directly target these receptors, increasing the effectiveness of heat and cold treatments.

Alternative treatment

Both physical and psychological aspects of pain can be dealt with through alternative treatment. Some of the most popular treatment options include **acupressure** and **acupuncture**, massage, **chiropractic**, and relaxation techniques such as **yoga**, hypnosis, and **meditation**. Herbal therapies are gaining increased recognition as viable options; for example, capsaicin, the component that makes cayenne peppers spicy, is used in ointments to relieve the joint pain associated with arthritis. Contrast **hydrotherapy** can also be very beneficial for pain relief.

Lifestyles can be changed to incorporate a healthier diet and regular **exercise**. Regular exercise, aside from relieving **stress**, has been shown to increase endorphins, painkillers naturally produced in the body.

Prognosis

Successful pain treatment is highly dependent on successful resolution of the pain's cause. Acute pain will stop when an injury heals or when an underlying problem is treated successfully. Chronic pain and abnormal pain are more difficult to treat, and it may take longer to find a successful resolution. Some pain is intractable and will require extreme measures for relief.

Prevention

Pain is generally preventable only to the degree that the cause of the pain is preventable. For example, improved surgical procedures, such as those done through a thin tube called a laparoscope, minimize post-operative pain. Anesthesia techniques for surgeries also continuously improve. Some disease and injuries are often unavoidable. However, pain from some surgeries and other medical procedures and

KEY TERMS

Acute pain—Pain in response to injury or another stimulus that resolves when the injury heals or the stimulus is removed.

Chronic pain—Pain that lasts beyond the term of an injury or painful stimulus. Can also refer to cancer pain, pain from a chronic or degenerative disease, and pain from an unidentified cause.

Neuron—A nerve cell.

Neurotransmitters—Chemicals within the nervous system that transmit information from or between nerve cells.

Nociceptor—A neuron that is capable of sensing pain.

Referred pain—Pain felt at a site different from the location of the injured or diseased part of the body. Referred pain is due to the fact that nerve signals from several areas of the body may "feed" the same nerve pathway leading to the spinal cord and brain.

Stimulus—A factor capable of eliciting a response in a nerve.

continuing pain are preventable through drug treatments and alternative therapies.

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ORGANIZATIONS

American Chronic Pain Association, PO Box 850, Rocklin, CA, 95677, (916) 632-3208, (800) 533-3231, APA@pacbell.net, <http://www.theacpa.org>.

American Pain Society, 4700 W. Lake Ave., Glenview, IL, 60025, (847) 375-4715, (866) 574-2654, info@ampainsoc.org, <http://www.ampainsoc.org/>.

Canadian Pain Society, 1143 Wentworth Street West, Suite 202, Oshawa, Canada, Ontario, L1J 8P7, (905) 404-9545, (905) 404-3727, <http://www.canadianpainsociety.ca>.

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Pain management

Definition

Pain itself is defined by the International Association for the Study of Pain (IASP) as “an unpleasant sensory and emotional experience associated with actual or potential tissue damage or described in terms of such damage.” Thus, pain management encompasses all interventions used to understand and ease pain, and if possible to alleviate the cause of the pain.

Purpose

Pain serves to alert a person to potential or actual damage to the body. The definition of damage is quite broad: pain can arise from injury as well as disease. After the message is received and interpreted, further pain can be counterproductive. Pain can have a negative impact on a person’s quality of life and impede recovery from illness or injury, thus contributing to escalating health care costs. Unrelieved pain can become a syndrome in its own right and cause a downward spiral in a person’s health and emotional outlook. Managing pain properly facilitates recovery, prevents additional health complications, and improves an individual’s quality of life.

Yet the experiencing of pain is a completely unique occurrence for each person, a complex combination of several factors other than the pain itself. It is influenced by:

- Ethnic and cultural values. In some cultures, tolerating pain is related to showing strength and endurance. In others, pain is considered punishment for misdeeds.
- Age. Many people have been taught that grownups never cry. On the other hand, in some cultures, the

elderly are allowed to complain freely about pain and discomfort.

- Anxiety and stress. This factor is related to being in a strange or unfamiliar place such as a hospital, and the fear of the unknown consequences of the pain and the condition causing it, which can all combined to make pain feel more severe. For patients being treated for pain, knowing the duration of activity of an analgesic leads to anxiety about the return of pain when the drug wears off. This anxiety can make the pain more severe. In addition, patients who interpret their pain as meaning that their disease is recurring or getting worse often experience pain as more severe.
- Fatigue and depression. It is known that pain in itself can actually cause emotional depression. Fatigue from lack of sleep or the illness itself also contributes to depressed feelings.

Precautions

The perception of pain is an individual experience. Healthcare providers play an important role in understanding their patients’ pain. All too often, both physicians and nurses have been found to incorrectly assess the severity of pain. A study reported in the *Journal of Advanced Nursing* evaluated nurses’ perceptions of a select group of white American and Mexican-American women patients’ pain following gallbladder surgery. Objective assessments of each patient’s pain showed little difference between the perceived severities for each group. Yet, the nurses involved in the study consistently rated all patients’ pain as less than the patients reported, and with equal consistency, believed that better-educated women born in the United States were suffering more than less-educated Mexican-American women. Nurses from a northern European background were more apt to minimize the severity of pain than nurses from eastern and southern Europe or Africa. The study indicated how healthcare staff, and especially nursing staff, need to be aware of how their own background and experience contributes to how they perceive a person’s pain.

Some patient populations are particularly susceptible to inadequate pain management. These include **cancer** patients; children; trauma victims receiving treatment in hospital emergency departments; and the elderly in nursing homes.

Description

Before considering pain management, a review of pain definitions and mechanisms may be useful. Pain is the means by which the peripheral nervous system

KEY TERMS

Acute—Referring to pain in response to injury or other stimulus that resolves when the injury heals or the stimulus is removed.

Central nervous system (CNS)—The part of the nervous system that includes the brain and the spinal cord.

Chronic—Referring to pain that endures beyond the term of an injury or painful stimulus. Can also refer to cancer pain, pain from a chronic or degenerative disease, and pain from an unidentified cause.

Iatrogenic—Resulting from the activity of the physician.

Neuropathy—Nerve damage.

Neurotransmitter—Chemicals within the nervous system that transmit information from or between nerve cells.

Nociceptor—A nerve cell that is capable of sensing pain and transmitting a pain signal.

Nonpharmacological—Referring to therapy that does not involve drugs.

Parasympathetic nervous system—That part of the autonomic nervous system consisting of nerves that arise from the cranial and sacral regions and function in opposition to the sympathetic nervous system.

Peripheral nervous system (PNS)—Nerves that are outside of the brain and spinal cord.

Pharmacological—Referring to therapy that relies on drugs.

Stimulus—A factor capable of eliciting a response in a nerve.

Sympathetic nervous system—That portion of the autonomic nervous system consisting of nerves that originate in the thoracic and lumbar spinal cord and function in opposition to the parasympathetic nervous system.

(PNS) warns the central nervous system (CNS) of injury or potential injury to the body. The CNS comprises the brain and spinal cord, and the PNS is composed of the nerves that stem from and lead into the CNS. PNS includes all nerves throughout the body, except the brain and spinal cord. Pain is sometimes categorized by its site of origin, either cutaneous (originating in the skin, or subcutaneous tissue, such as a shaving nick or paper cut), deep somatic pain (arising

from bone, ligaments and tendons, nerves, or veins and arteries), or visceral (appearing as a result of stimulation of pain receptor nerves around such organs as the brain, lungs, or stomach and intestines).

A pain message is transmitted to the CNS by special PNS nerve cells called nociceptors, which are distributed throughout the body and respond to different stimuli depending on their location. For example, nociceptors that extend from the skin are stimulated by such sensations as pressure, temperature, and chemical changes.

When a nociceptor is stimulated, neurotransmitters are released within the cell. Neurotransmitters are chemicals found within the nervous system that facilitate nerve cell communication. The nociceptor transmits its signal to nerve cells within the spinal cord, which conveys the pain message to the thalamus, a specific region in the brain.

Once the brain has received and processed the pain message and coordinated an appropriate response, pain has served its purpose. The body uses natural painkillers called endorphins to derail further pain messages from the same source. However, these natural painkillers may not adequately dampen a continuing pain message. Also, depending on how the brain has processed the pain information, certain hormones such as prostaglandins may be released. These hormones enhance the pain message and play a role in immune system responses to injury, such as inflammation. Certain neurotransmitters, especially substance P and calcitonin gene-related peptide, actively enhance the pain message at the injury site and within the spinal cord.

Pain is generally divided into two additional categories: acute and chronic. Nociceptive pain, or the pain that is transmitted by nociceptors, is typically called acute pain. This kind of pain is associated with injury, headaches, disease, and many other conditions. Response to acute pain is made by the sympathetic nervous system (the nerves responsible for the fight-or-flight response of the body). It normally resolves once the condition that precipitated it is resolved.

There are some disorders that produce pain that does not resolve following the disorder. Even after healing or a cure has been achieved, the brain continues to perceive pain. In this situation, the pain may be considered chronic. Chronic pain is within the province of the parasympathetic nervous system, and the changeover occurs as the body attempts to adapt to the pain. The time limit used to define chronic pain typically ranges from three to six months, although some healthcare professionals prefer a more flexible

definition and consider chronic pain as pain that endures beyond a normal healing time. The pain associated with cancer, persistent and degenerative conditions, and neuropathy, or nerve damage, is included in the chronic category. Also, unremitting pain that lacks an identifiable physical cause such as the majority of cases of **low back pain** may be considered chronic. The underlying biochemistry of chronic pain appears to be different from that of acute nociceptive pain.

It has been hypothesized that uninterrupted and unrelenting pain can induce changes in the spinal cord. In the past, severing a nerve's connection to the CNS has treated intractable pain. However, the lack of any sensory information being relayed by that nerve can cause pain transmission in the spinal cord to go into overdrive, as evidenced by the phantom limb pain experienced by amputees. Evidence is accumulating that unrelenting pain or the complete lack of nerve signals increases the number of pain receptors in the spinal cord. Nerve cells in the spinal cord may also begin secreting pain-amplifying neurotransmitters independent of actual pain signals from the body. Immune chemicals, primarily cytokines, may play a prominent role in such changes.

Managing pain

Considering the different causes and types of pain, as well as its nature and intensity, management usually requires a multidisciplinary approach. The elements of this approach include treating the underlying cause of pain, pharmacological and non-pharmacological therapies, and some invasive (surgical) procedures.

Treating the cause of pain underlies the basic strategy of pain management. Injuries are repaired, diseases are diagnosed, and certain encounters with pain can be anticipated and treated prophylactically (by prevention). However, there are no guarantees of immediate relief from pain. Recovery can be impeded by pain and quality of life can be damaged. Therefore, pharmacological and other therapies have developed over time to address these aspects of disease and injury.

PHARMACOLOGICAL OPTIONS. General guidelines developed by the World Health Organization (WHO) have been developed for pain management. These guidelines operate upon the three-step ladder approach, including:

- Mild pain is alleviated with acetaminophen or a non-steroidal anti-inflammatory drug (NSAID). NSAIDs and acetaminophen are available as over-the-counter (OTC) and prescription medications, and are frequently the initial pharmacological

treatment for pain. These drugs can also be used as adjuncts to the other drug therapies that might require a doctor's prescription. NSAIDs include aspirin, ibuprofen (Motrin, Advil, Nuprin), naproxen sodium (Aleve), and ketoprofen (Orudis KT). These drugs are used to treat pain from inflammation and work by blocking production of pain-enhancing neurotransmitters. Acetaminophen is also effective against pain, but its ability to reduce inflammation is limited. NSAIDs and acetaminophen are effective for most forms of acute (sharp, but of a short duration) pain.

- Mild to moderate pain is eased with a milder opioid medication, plus acetaminophen or NSAIDs. Opioids include both drugs derived from the opium poppy, such as morphine and codeine, and synthetic drugs based on the structure of opium. This drug class includes drugs such as oxycodone, methadone, and meperidine (Demerol). They provide pain relief by binding to specific opioid receptors in the brain and spinal cord. One drawback of opioids, however, is that they frequently cause constipation because they slow down the rhythmic muscular contractions of the intestines that push food along during the process of digestion.
- Moderate to severe pain is treated with stronger opioid drugs, plus acetaminophen or NSAIDs. Morphine is sometimes referred to as the gold standard of palliative care as it is not expensive; can be given by starting with smaller doses and gradually increased; and is highly effective over a long period of time. It can also be given by a number of different routes, including by mouth, rectally, or by injection.

Although **antidepressant drugs** were developed to treat depression, it has been discovered that they are also effective in combating chronic headaches, cancer pain, and pain associated with nerve damage. Antidepressants that have been shown to have analgesic (pain-reducing) properties include amitriptyline (Elavil), trazodone (Desyrel), and imipramine (Tofranil). **Anticonvulsant drugs** share a similar background with antidepressants. Developed to treat **epilepsy**, anticonvulsants were found to relieve pain as well. Drugs such as phenytoin (Dilantin) and carbamazepine (Tegretol) are prescribed to treat the pain associated with nerve damage.

In some cases, chronic pain caused by complications of diabetes or cancer can be eased by administering local anesthetics. The most commonly used are mexiletine (Mexitil) and a lidocaine patch.

Corticosteroids are another class of drugs commonly given to manage chronic pain caused by

arthritis or other diseases affecting the muscles and joints; they may also be given to control **nausea**. Dexamethasone (Decadron) and prednisone are the most commonly used corticosteroids in pain management. They work by reducing inflammation and suppressing the immune system.

Close monitoring of the effects of pain medications is required in order to assure that adequate amounts of medication are given to produce the desired pain relief. When a person is comfortable with a certain dosage of medication, oncologists typically convert to a long-acting version of that medication. Transdermal fentanyl patches (Duragesic) are a common example of a long-acting opioid drug often used for cancer pain management. A patch containing the drug is applied to the skin and continues to deliver the drug to the person for an average of three days. Pumps are also available that provide an opioid medication upon demand when the person is experiencing pain. By pressing a button, they can release a set dose of medication into an intravenous solution or an implanted catheter. Another mode of administration involves implanted catheters that deliver pain medication directly to the spinal cord. Because these pumps offer the patient some degree of control over the amount of analgesic administered, the system, commonly called patient-controlled analgesia (PCA), reduces the level of **anxiety** about availability of pain medication. Delivering drugs in this way can reduce side effects and increase the effectiveness of the drug. Research is underway to develop toxic substances that act selectively on nerve cells that carry pain messages to the brain. These substances would kill the selected cells and stop transmission of the pain message.

NONPHARMACOLOGICAL OPTIONS. Pain treatment options that do not use drugs are often used as adjuncts to, rather than replacements for, drug therapy. One of the benefits of nondrug therapies is that an individual can take a more active role in pain management. Such relaxation techniques as **yoga** and **meditation** are used to focus the brain elsewhere than on the pain, decrease muscle tension, and reduce **stress**. Tension and stress can also be reduced through **biofeedback**, in which an individual consciously attempts to modify skin temperature, muscle tension, blood pressure, and heart rate.

Hypnosis is another nonpharmacological option for pain relief. Although doctors do not yet fully understand how hypnosis works, it is used successfully in some patients to manage pain related to **childbirth**, oral surgery, burn treatment, and other procedures that require the patient to remain conscious.

Participating in normal activities and exercising can also help control pain levels. Through **physical therapy**, an individual learns beneficial exercises for reducing stress, strengthening muscles, and staying fit. Regular **exercise** has been linked to production of endorphins, the body's natural painkillers.

Acupuncture involves the insertion of small needles into the skin at key points. **Acupressure** uses these same key points, but involves applying pressure rather than inserting needles. Both of these methods may work by prompting the body to release endorphins. Applying heat or being massaged are very relaxing and help reduce stress. Transcutaneous **electrical nerve stimulation** (TENS) applies a small electric current to certain parts of nerves, potentially interrupting pain signals and inducing release of endorphins. To be effective, use of TENS should be medically supervised.

INVASIVE PROCEDURES. There are three types of invasive procedures that may be used to manage or treat pain: anatomic, augmentative, and ablative. These procedures involve surgery, and certain guidelines should be followed before carrying out a procedure with permanent effects. First, the cause of the pain must be clearly identified. Next, surgery should be done only if noninvasive procedures are ineffective. Third, any psychological issues should be addressed. Finally, there should be a reasonable expectation of success.

Anatomic procedures involve correcting the injury or removing the cause of pain. Relatively common anatomic procedures are decompression surgeries such as repairing a **herniated disk** in the lower back or relieving the nerve compression related to **carpal tunnel syndrome**. Another anatomic procedure is neurolysis, also called a nerve block, which involves destroying a portion of a peripheral nerve.

Augmentative procedures include electrical stimulation or direct application of drugs to the nerves that are transmitting the pain signals. Electrical stimulation works on the same principle as TENS. In this procedure, instead of applying the current across the skin, electrodes are implanted to stimulate peripheral nerves or nerves in the spinal cord. Augmentative procedures also include implanted drug-delivery systems. In these systems, catheters are implanted in the spine to allow direct delivery of drugs to the CNS.

Ablative procedures are characterized by severing a nerve and disconnecting it from the CNS. However, this method may not address potential alterations within the spinal cord. These changes perpetuate

pain messages and do not cease, even when the connection between the sensory nerve and the CNS is severed. With growing understanding of neuropathic pain and development of less invasive procedures, ablative procedures are used less frequently. However, they do have applications in select cases of **peripheral neuropathy**, cancer pain, and other disorders.

Preparation

Prior to beginning management, the patient's pain should be thoroughly evaluated, including a psychosocial as well as a physical assessment. Pain scales or questionnaires can be administered by a member of the healthcare team, although there is no single questionnaire that is universally accepted. Some questionnaires are verbal, while others use pictures or drawings to help the patient describe the pain. Some questionnaires are filled out by the patient, while others may be given to relatives or friends to complete. It is often necessary to ask other family members to complete a pain questionnaire if the patient is cognitively impaired.

In spite of their limitations, questionnaires and self-report forms do allow healthcare workers to better understand the pain being suffered by the patient. Evaluation also includes physical examinations and diagnostic tests to determine the underlying physical causes of the pain. Some evaluations require assessments from several viewpoints, including neurology, psychiatry and psychology, and physical therapy. If the pain is caused by a medical procedure, management consists of anticipating the type and intensity of associated pain and managing it preemptively.

Nurses or physicians often take what is called a pain history. This history will help to provide important information that can help health care providers to better manage the patient's pain. A typical pain history includes the following questions:

- Where is the pain located?
- On a scale of 1 to 10, with 1 indicating the least pain, how would the person rate the pain being experienced?
- What does the pain feel like?
- When did (or does) the pain start?
- How long has the person had it?
- Is the person sometimes free of pain?
- Is the pain constant, or is it episodic?
- Does the person know of anything that triggers the pain or makes it worse?

- Does the person have other symptoms (nausea, dizziness, blurred vision, etc.) during or after the pain?
- What pain medications or other measures has the person found to help in easing the pain?
- How does the pain affect the person's ability to carry on normal activities?
- What does it mean to the person that he or she is experiencing pain?

Aftercare

An assessment by nursing staff as well as other healthcare providers should be made to determine the effectiveness of the pain management interventions employed. There are objective, measurable signs and symptoms of pain that can be looked for. The goal of good pain management is the absence of these signs. Signs of acute pain include:

- rise in pulse and blood pressure
- more rapid breathing
- perspiring profusely, clammy skin
- taut muscles
- more tense appearance, fast speech, very alert
- unusually pale skin
- dilated pupils of the eye

Signs of chronic pain include:

- lower pulse and blood pressure
- changeable breathing pattern
- warm, dry skin
- nausea and vomiting
- slow or monotone speech
- inability or difficulty in getting out of bed and performing activities of daily living (ADLs)
- constricted pupils of the eye

When these signs are absent and the patient appears to be comfortable, healthcare providers can consider their interventions to have been successful. It is also important to document interventions used, and which ones were successful.

Risks

Owing to toxicity over the long term, some drugs can only be used for acute pain or as adjuncts in chronic pain management. NSAIDs have the well-known side effect of causing gastrointestinal bleeding, and long-term use of **acetaminophen** has been linked to kidney and liver damage. Other drugs, especially **narcotics**, have such serious side effects as **constipation**, drowsiness, and nausea. Serious side effects can

also accompany pharmacological therapies; mood swings, confusion, bone thinning, cataract formation, increased blood pressure, and other problems may discourage or prevent use of some **analgesics**.

Nonpharmacological therapies carry little or no risks. However, individuals recovering from serious illness or injury should consult with the health care providers or physical therapists before making use of adjunct therapies. Invasive procedures carry risks similar to other surgical procedures, such as infection, reaction to anesthesia, and iatrogenic (injury as a result of treatment) injury.

A traditional concern about narcotics use has been the risk of promoting **addiction**. As narcotic use continues over time, the body becomes accustomed to the drug and adjusts normal functions to accommodate to its presence. Therefore, to elicit the same level of action, it is necessary to increase dosage over time. As dosage increases, an individual may become physically dependent on narcotic drugs.

However, physical dependence is different from psychological addiction. Physical dependence is characterized by discomfort if drug administration suddenly stops, while psychological addiction is characterized by an overpowering craving for the drug for reasons other than pain relief. Psychological addiction is a very real and necessary concern in some instances, but it should not interfere with a genuine need for narcotic pain relief. However, caution must be taken with people who have a history of addictive behavior.

Normal results

Effective application of pain management techniques reduces or eliminates acute or chronic pain. This treatment can improve an individual's quality of life and aid in recovery from injury and disease.

Resources

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American Pain Society, 4700 West Lake Avenue, Glenview, IL, 60025, (847) 375-4715, (866) 574-2654, info@ampainsoc.org, <http://www.ampainsoc.org>.

International Association for the Study of Pain (IASP), 111 Queen Anne Avenue North, Suite 501, Seattle, WA, 98109-4955, (206) 283-0311, (206)283-9403, <http://www.iasp-pain.org>.

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Pain relievers see **Analgesics**

Painful menstruation see **Dysmenorrhea**

Palliative cancer therapy see **Cancer therapy, palliative**

Palliative care

Definition

Palliative care is a multidisciplinary approach to relieving physical, psychological, emotional, social, and spiritual suffering in patients with chronic or terminal illnesses. Palliative care for children includes providing comfort and support for the entire family, regardless of whether the child is in home or hospice care.

Purpose

The purpose of palliative care is to improve the quality of life for patients and their families. The primary goal is to relieve **pain** and other physical symptoms—such as **nausea** or fatigue—caused by diseases, conditions, and treatments. Palliative care also attempts to minimize side effects from medical interventions. In addition, palliative care aims to relieve psychological symptoms, such as **anxiety** or depression, and to provide comfort and support for patients and their families. Pediatric palliative care helps children lead lives that are as normal and happy as possible under the circumstances.

Demographics

It has been estimated that, worldwide, seven million children and their families could benefit from palliative care. In the United States, approximately 27,000 children die annually from illness or other medical conditions. Although the number of pediatric palliative care programs is growing, the number of parents who are solely responsible for overseeing their children's **pain management** and recovery is also increasing, as most children are sent home from the hospital within hours of surgery. Approximately 84% of pediatric surgeries in the United States are performed on an outpatient basis.

Although some children receive treatment and services from adult palliative care programs, of the approximately 2,500 home-, freestanding-, and hospital-based hospice programs in the United States, only about 10% are pediatric. It has been estimated that hospice care is available to less than 1% of American children in need of it.

Description

Children living with serious chronic illnesses—including certain cancers, HIV/AIDS, **cystic fibrosis**, heart disease, lung disease, or kidney failure—can suffer from significant physical symptoms, pain, and

emotional distress. Children undergoing medical treatments can also have physical, psychological, and emotional symptoms that require palliative care. In addition to pain, symptoms such as nausea, weakness, bowel and bladder problems, **constipation**, breathing difficulties, **fatigue**, and mental confusion are treated through palliative care. Although palliative care usually intensifies toward the end of a terminal illness, children may receive palliative care at any stage of—and throughout—any illness.

Palliative care for **cancer** may include **chemotherapy**, **radiation therapy**, or surgery. Although these treatments may not cure cancer that has metastasized or spread, they can diminish the cancer, improve or eliminate symptoms, at least for a period of time, and extend life.

Pediatric illness can have a devastating impact on families. Because many serious childhood illnesses are quite rare, parents and children are often forced to travel to specialized treatment centers. Jobs and support systems can be lost and financial resources depleted. Parents may be at risk for **alcoholism** or drug **abuse**. The sick child's siblings may experience lack of attention. Therefore, pediatric palliative care uses a holistic approach to address the physical, cultural, psychological, emotional, spiritual, and practical needs of the entire family.

Pediatric palliative care differs from adult palliative care for a variety of reasons:

- Serious illnesses in children are relatively rare in developed countries and the course of disease in children is often unpredictable.
- There are often large uncertainties in the prognoses and survival times for life-threatening illnesses in children.
- Children are very resilient and sometimes recover after being close to death.
- Even children with conditions that ultimately prove fatal often have periods of relative health during which they lead comparatively normal lives.
- Pediatricians are strongly committed to their patients and usually want to remain in charge of their care plans.
- Most parents both want and need to be involved in—and have at least some control over—their child's care. Pediatric palliative care encourages their participation.
- Parents are usually not willing to choose between treatments aimed at disease management or a cure and those aimed at making the child comfortable.

- Childhood diseases are usually treated very aggressively with the goal of extending life at any cost. For these reasons, parents often choose “heroic” medical treatments that have only a small chance of success

There are other significant differences between pediatric and adult palliative care:

- Children have unique developmental and psychosocial needs.
- Children are strongly connected to their families and their schools.
- Children may lack verbal skills for expressing their feelings and needs.
- Identifying sources of pain and the drugs and therapies that can effectively manage the pain can be difficult with young children.
- Children are usually dependent on their parents for medical decision-making. However, children as young as seven to ten can often articulate informed opinions about their medical care. Adolescents expect to participate in medical decision-making. Members of a palliative care team may help children and teens to understand their condition and to participate in discussions of treatment options.
- Siblings and grandparents may complicate medical decision-making and other family issues.
- Pediatric palliative care focuses on the entire family rather than on the patient alone.
- Because the initial diagnosis is often the most emotionally difficult time for families, pediatric palliative care usually begins at the time of diagnosis.
- Pediatric palliative care may be much more integrated with medical treatment than adult palliative care, and it can be an important component of any remission or cure.

Pediatric palliative care is being increasingly integrated into treatment protocols and therapies. A palliative care team may include:

- physicians
- specialists
- nurses
- therapists
- social workers
- child-life specialists
- chaplains

Pediatric palliative care is more likely to take place in the home than in a hospital or hospice for a variety of reasons:

- Although hospice care always includes palliative care, it is generally defined as end-of-life care for

patients with a six-month prognosis who will receive no further lifesaving treatments. This definition is often not applicable to children.

- Most hospices do not have pediatric-specific services.
- There is little or no financial reimbursement for pediatric hospice or home care.
- Many families prefer to have their child die at home and many pediatric palliative care programs include end-of-life planning.
- Studies have found that the overall quality of care is generally better in the home than in a hospital or hospice.

Origins

Pediatric palliative care is a relatively new medical subspecialty. The first pediatric palliative home care program in the United States was established in Connecticut in 1974. Since the 1980s a variety of home- and hospice-based pediatric palliative care programs have been established across the United States and around the world. In 1984, Dr. Burton Grebin opened St. Mary's Hospital for Children as a comprehensive palliative care center in Bayside, New York. Grebin sought to normalize the children's environment: St. Mary's has its own New York City Public School; the children wear street clothes and play and socialize; families are encouraged to visit and spend the night. The St. Mary's program includes counseling for parents and siblings.

Benefits

Palliative care can:

- ensure that discomfort and pain are managed adequately
- help normalize life for seriously ill children and their families
- encourage parental participation and ease emotional stress and grief
- promote informed medical decision-making
- promote coordinated medical care and services
- lead to better medical outcomes
- reduce hospital and emergency-room visits, thereby making more efficient use of healthcare spending
- provide families with improved access to resources and services, such as therapy and respite care
- provide children with the most peaceful end of life as possible

Precautions

New medical technologies have complicated treatment decisions for parents. Newborns are often subjected to stressful and painful tests and procedures, even when their condition or illness is incompatible with life. Overwhelmed parents are often unaware of the long-term consequences of the medical decisions that they are asked to make, especially when they are dealing with multiple specialists. In addition, language and cultural barriers can exacerbate communication problems between parents, physicians, and palliative care providers.

It can be very difficult for young children to understand why they are undergoing painful procedures for diagnosis and treatment and many parents want to protect their child from a serious prognosis. However, studies have found that even very young children can understand the seriousness of their disease and that open communication between children and their parents and care providers can help alleviate their fears and maintain hope.

Infants and children are often unable to communicate symptoms and specific sources of discomfort and pain. A major component of pediatric palliative care is the identification and treatment of such symptoms. Prescribing pain medications for children is a specialized skill, because dosages are based on body weight and because children's bodies metabolize medications differently than adults. Furthermore, children often cannot swallow pills. Palliative caregivers must work closely with pharmacists to develop syrups, dissolvable capsules, or skin patches. It is helpful for children to take a medication before leaving the hospital to ensure that they can swallow it. Parents can also request that their pharmacists add their child's favorite flavor to medications.

Parents are often reluctant to administer pain medications, either because they fear that the child will become addicted to **narcotics** or because they are waiting for the child to cry or complain of severe pain. However, pain can interfere with a child's recovery by triggering the body's **stress** response. Prescriptions for painkillers should be filled immediately, so that the medication is available in the middle of the night. Parents must also learn to recognize signs that their child is in pain. These may include:

- fussiness
- refusing to eat or drink
- becoming quiet or withdrawn
- sleep disturbances
- mood or behavior changes

KEY TERMS

Children's hospice—A holistic philosophy that addresses the physical, emotional, social, and spiritual needs of children with life-threatening illnesses, as well as the needs of their families.

Chronic—An illness or condition of long duration, frequent recurrence, or slow progression.

Hospice—A facility or program that provides for the physical and emotional needs of the terminally ill in a caring environment.

Opiate—A drug containing or derived from opium—such as codeine, morphine, and heroin—that alleviates pain and induces sleep.

Palliative treatment—Treatments—such as chemotherapy, radiation therapy, or surgery—that ease the symptoms of a disease without curing it.

Respite care—Temporary care of a patient to provide parents or other caregivers with a period of physical, mental, and emotional rest.

Although medical associations, including the American Academy of Pediatrics, have endorsed pediatric palliative care, it is not universally supported. Some physicians do not believe it is medically necessary and many hospitals claim they cannot afford it. Palliative care is poorly reimbursed by Medicaid and it has been more difficult to demonstrate cost savings from reduced emergency care and hospital stays with pediatric palliative care than with adult programs.

Preparation

Palliative care for children usually begins with the diagnosis of a chronic or life-threatening illness. Preparation may include:

- educating family members about the child's disease or condition
- informing family members about treatment and care options
- advising on various aspects of home care, including administering medication and recognizing symptoms that require immediate attention
- arranging home support services, possibly including transportation, shopping, and meal preparation
- arranging respite care
- helping the family plan for financial strains caused by the child's illness
- helping the family develop a support network

Aftercare

Palliative aftercare may include:

- helping families prepare for a child's death
- helping with funeral and other arrangements
- grief and bereavement counseling and support groups

Risks

Physicians who are inexperienced in treating pain in terminally ill children may be reluctant to prescribe opiates, including morphine. Even when prescribed, parents may withhold medication because of fear of side effects or **addiction**. Parents may consider hastening a suffering child's **death** because they are unaware of the legal options for pain relief, including sedating a child into unconsciousness.

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Children's Hospice International, 1101 King Street, Suite 360, Alexandria, VA, 22314, (703) 684-0330, (800) 2-4-CHILD, info@chionline.org, <http://www.chionline.org>.

Department of Pain Medicine & Palliative Care, Beth Israel Medical Center, First Avenue at 16th Street, New York, NY, 10003, (877) 620-9999, (212) 844-1503, stoppain@chpnet.org, <http://www.stoppain.org>.

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International Association for Hospice and Palliative Care, 5535 Memorial Dr. Suite F, PMB 509, Houston, TX, 77007, (936) 321-9846, (866) 374-2472, (713) 880-2948, <http://www.hospicecare.com>.

National Hospice & Palliative Care Organization, 1731 King Street, Suite 100, Alexandria, VA, 22314, (703) 837-1500, (703) 837-1233, nhpco@AEA-nhpco.org, <http://www.nhpco.org>.

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Palpitations

Definition

The term palpitation refers to a sensation in which a person is aware of an irregular, hard, or rapid heartbeat.

Description

Palpitations mean that the heart is not behaving normally. It can appear to skip beats, beat rapidly, beat irregularly, or thump in the chest. Although palpitations are very common and often harmless, they can be frightening to a person, who is usually unaware of his or her heartbeat.

Palpitations can also be a sign of serious heart trouble. Palpitations that are caused by certain types of abnormal heart rhythms (**arrhythmias**) can be serious, and even fatal if left untreated. Recognizable arrhythmias are present in a small number of patients who have palpitations. Immediate medical attention should be sought for palpitations that feel like a very fast series of heartbeats, last more than two or three minutes, and are unrelated to strenuous physical activity, obvious fright, or anger. Medical attention should also be sought if palpitations are accompanied by chest **pain**, **dizziness**, **shortness of breath**, or an overall feeling of weakness.

Most people have experienced a skipped or missed heartbeat, which is really an early beat and not a skipped beat at all. After a premature heartbeat, the heart rests for an instant then beats with extra force, making a person feel as if the heart has skipped a beat. This type of palpitation is nothing to worry about unless it occurs frequently. Severe palpitations feel like a thudding or fluttering sensation in the chest. After chest pain, palpitations are the most common reason that people are referred for cardiology evaluation.

Causes and symptoms

Palpitations can be caused by **anxiety**, arrhythmias, **caffeine**, certain medications, **cocaine** and amphetamines, emotional **stress**, overeating, panic, somatization, and vigorous **exercise**. There may be no other symptoms. But, anxiety, dizziness, shortness of breath, and chest pain may be signs of more severe arrhythmias.

Diagnosis

Palpitations are diagnosed through a medical history, a **physical examination**, an electrocardiogram

(ECG), and screening for psychiatric disorders. It is often difficult to distinguish palpitations from **panic disorder**, a common problem in which a person experiences frequent and unexplained “fight-or-flight” responses, which is the body’s natural physical reaction to extreme danger or physical exertion, but without the obvious external stimulus.

To accurately diagnose palpitations, one of the irregular heartbeats must be “captured” on an EKG, which shows the heart’s activity. Electrodes covered with a type of gel that conducts electrical impulses are placed on the patient’s chest, arms, and legs. These electrodes send impulses of the heart’s activity to a recorder, which traces them on paper. This **electrocardiography** test takes about 10 minutes and is performed in a physician’s office or hospital. Because palpitations are unlikely to occur during a standard EKG, **Holter monitoring** is often performed. In this procedure, the patient wears a small, portable tape recorder that is attached to a belt or shoulder strap and connected to electrode disks on his or her chest. The Holter monitor records the heart’s rhythm during normal activities. Some medical centers are now using event recorders that the patient can carry for weeks or months. When palpitations occur, the patient presses a button on the device, which captures the information about the palpitations for physician evaluation. Later the recording can be transmitted over the telephone line for analysis.

Treatment

Most palpitations require no treatment. Persistent palpitations can be treated with small doses of a beta blocker. **Beta blockers** are drugs that tend to lower blood pressure. They slow the heart rate and decrease the force with which the heart pumps. If the cause of the palpitations is determined to be an arrhythmia, medical or surgical treatment may be indicated, although surgery is rarely needed.

Alternative treatment

Alternative treatments for palpitations should be used only as a complement to traditional medicine. Alternative treatments include: **aromatherapy**, Chinese herbs, herbal therapies, **homeopathic medicine**, exercise, mind/body medicine, and diet and **nutrition**. In aromatherapy, adding citrus oils to bath water may help with minor palpitations. Some Chinese herbs can also help, but others can worsen arrhythmias, so a qualified herbalist should be consulted. Herbal therapies such as hawthorn (*Crataegus laevigata*) and motherwort (*Leonurus cardiaca*) can help with palpitations.

KEY TERMS

Arrhythmia—Any variation from the normal heartbeat. Some arrhythmias are harmless, while others, such as ventricular tachycardia, ventricular fibrillation, and ventricular standstill, can be fatal.

Somatization—Anxiety converted into physical symptoms. Somatization is a sign of panic disorder.

Homeopathic remedies such as *Lachesis*, *Digitalis*, and *Aconite* (*Aconitum napellus*) may be used to control palpitations but should be taken only when prescribed by a homeopathic physician. Mind/body medicine such as **meditation** and **yoga** can help the person relax, eliminating or reducing palpitations caused by anxiety or stress. Reducing or eliminating tea, cola, coffee, and chocolate, and consuming adequate amounts of the **minerals calcium**, magnesium, and potassium can help reduce or eliminate palpitations. Alternative treatments should be used with care, as the benefits of many such treatments have not been confirmed by scientific research.

Prognosis

Most palpitations are harmless, but some can be a sign of heart trouble, which could be fatal if left untreated.

Prevention

Palpitations not caused by arrhythmias can be prevented by reducing or eliminating anxiety and emotional stress, and reducing or eliminating consumption of tea, cola, coffee, and chocolate. Exercise can also help, but a treadmill **stress test** performed by a physician should be considered first to make sure that exercise is safe.

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American Institute of Stress, 124 Park Ave., Yonkers, NY, 10703, (914) 963–1200, <http://www.stress.org>.

Center for Disease Control (CDC). Division for Heart Disease and Stroke Prevention, 4770 Buford Hwy NE, Atlanta, GA, 30341–3717, (770) 488–2424, www.cdc.gov/cholesterol/faqs.htm.

European Society of Cardiology, The European Heart House, 2035 Route des Colles, B.P. 179–Les Templiers, Sophia–Antipolis, France, 06903, 33 4 9294 7600, 33 4 9294 7601, <http://www.escardio.org>.

Heart Foundation, 80 William St., Level 3, SydneyNSW, Australia, 2011, 02 9219 2444, 300 36 27 87, <http://www.heartfoundation.org.au>.

National Heart, Lung, and Blood Institute, PO Box 30105, Bethesda, MD, 20824–0105, (301) 592–8573, (204) 629–3246, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov>.

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Panax quinquefolius see **Ginseng**

Pancreas removal see **Pancreatectomy**

Pancreas transplantation

Definition

Pancreas transplantation is a surgical procedure in which a diseased pancreas is replaced with a healthy pancreas that has been obtained from an immunologically compatible cadaver or living donor.

Purpose

The pancreas is an organ that secretes insulin, a peptide hormone that regulates glucose (blood sugar) metabolism. Patients with type 1 diabetes have experienced partial or complete damage to the insulin-producing beta cells of the pancreas. Consequently, they are unable to generate sufficient insulin to control blood glucose levels. Long-term uncontrolled high blood glucose levels can cause damage to every system of the body, so type 1 patients must inject insulin to do the work of the beta cells. Pancreas transplantation allows the body to once again make and secrete its own

KEY TERMS

Cadaver organ—A pancreas, kidney, or other organ from a brain-dead organ donor.

Duodenum—The section of the small intestine immediately after the stomach.

insulin, and establishes insulin independence for these individuals.

Demographics

It is estimated that about 1.4 million people in the United States have type 1 **diabetes mellitus** (also called insulin-dependant diabetes or juvenile diabetes). Among these individuals, the best candidates for pancreas transplantation are typically:

- between the ages of 20 and 40
- those who have extreme difficulty regulating their glucose levels with insulin therapy (a condition called brittle diabetes)
- those who have few secondary complications of diabetes
- those who are in good cardiovascular health

Pancreas-only transplants account for about 10% of the 1200 pancreas transplants performed each year in the United States as of 2010. More common is the combined kidney-pancreas transplant, or simultaneous pancreas-kidney transplantation (SPK), which is performed in about 75% of patients. The remaining 15% of patients receive a PAK, or pancreas after kidney transplant, according to the United Network for Organ Sharing (UNOS). There are about 100 medical centers in the United States that perform pancreas transplants as of early 2010.

Description

Once a donor pancreas is located and **tissue typing** deems it compatible, the patient is contacted and prepared for surgery. Blood tests, a **chest x ray**, and an electrocardiogram (ECG) are performed and an intravenous (IV) line is started for fluid and medication administration. Once the transplant procedure is ready to start, **general anesthesia** is administered.

The surgeon makes an incision under the ribs and locates the pancreas and duodenum. The pancreas and duodenum (part of the small intestine) are removed. The new pancreas and duodenum are then connected to the patient's duodenum, and the blood vessels are sutured together to restore blood flow to the new

pancreas. The patient's original pancreas is left in place.

Replacing the duodenum allows the pancreas to drain into the gastrointestinal system. The transplant can also be done creating bladder drainage. Bladder drainage makes it easier to monitor organ rejection because pancreatic secretions can be measured in the patient's urine. Once the new pancreas is in place, the abdomen and skin are sutured closed. This surgery is often done at the same time as kidney **transplant surgery**.

Diagnosis/Preparation

After the patient and doctor have decided on a pancreas transplant, a complete immunological study is performed to match the patient to a donor. An extensive medical history and **physical examination** is performed, including radiological exams, blood and urine tests, and psychological evaluation. Once the patient is approved for transplant, he or she will be placed on the United Network for Organ Sharing (UNOS) Organ Center waiting list. The timing of surgery depends on the availability of a donated living or cadaver organ.

Aftercare

Patients receiving a pancreas transplantation are monitored closely for organ rejection. The average hospital stay is three weeks, and it takes about six months to recover from surgery. Patients will take **immunosuppressant drugs** for the rest of their lives.

Risks

Diabetes and poor kidney function greatly increase the risk of complications from anesthesia during surgery. Organ rejection, excessive bleeding, and infection are other major risks associated with this surgery.

The reason that simultaneous kidney-pancreas transplants and pancreas-after-kidney transplants are performed more frequently than pancreas-only transplants is the relative risk of immunosuppressant drugs in people with diabetes. People with type 1 diabetes are already at risk for autoimmune problems, are more prone to infections, and have a complicated medical history that makes suppressing the immune system unadvisable.

On the other hand, diabetes is also the number one cause of **chronic kidney failure**, or end-stage renal disease (ESRD), which makes this group more likely to eventually require a kidney transplant for survival.

In those patients with diabetes who will receive or are already receiving immunosuppressive treatment for a life-saving kidney transplant, a pancreas transplant can return their ability to self-produce insulin.

Patients with type 1 diabetes considering pancreas transplantation alone must weigh the risks and benefits of the procedure and decide with their doctors whether life-long treatment with immunosuppressive drugs is preferable to life-long insulin dependence.

Normal results

In a successful transplant, the pancreas begins producing insulin, bringing the regulation of glucose back under control. Natural availability of insulin prevents the development of additional complications associated with diabetes, including kidney damage, vision loss, and nerve damage. Many patients report an improved quality of life.

Morbidity and mortality rates

According to the Mayo Clinic, as of 2009, the transplanted pancreas is still functioning after one year in about 87 percent of people who receive a simultaneous pancreas-kidney transplant. After five years, the rate drops to 72 percent. In about 77 percent of people who receive a pancreas-after-kidney transplant, the transplanted pancreas is still functioning after one year; five years after transplant, the rate drops to 59 percent. In about 85 percent of people who receive a pancreas-only transplant, the transplanted pancreas is still functioning after one year. That rate is about 52 percent after five years.

Alternatives

Innovations in islet cell transplants, a procedure that involves transplanting a culture of the insulin-producing islet cells of a healthy pancreas to a patient with type 1 diabetes, have increased the frequency of this procedure. The Edmonton Protocol, a type of islet cell transplant developed in 1999 by Dr. James Shapiro at the University of Alberta (Canada), uses a unique immunosuppressant drug regimen that has dramatically improved success rates of the islet transplant procedure. As of early 2010, the Edmonton Protocol is still considered investigational in the United States, and a number of clinical trials are ongoing. One center that is recruiting patients for clinical trials of islet transplantation is the Schulze Diabetes Institute of the University of Minnesota.

Researchers in Japan and elsewhere are also investigating stem cells as a possible source of insulin-

National transplant waiting list by organ type (June 2010)

Organ needed	Persons waiting
Kidney	85,296
Liver	16,031
Heart	3,141
Kidney/Pancreas	2,199
Lung	1,802
Pancreas	1,450
Intestine	242
Heart/Lung	79

SOURCE: U.S. Department of Health and Human Services, Organ Procurement and Transplantation Network. Available online at: <http://optn.transplant.hrsa.gov/data/default.asp> (accessed June 8, 2010).

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

producing cells available for transplantation. This research is still in its early stages as of 2010.

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ORGANIZATIONS

American Diabetes Association, 1701 North Beauregard Street, Alexandria, VA, 22311, (800) 342-2383, <http://www.diabetes.org>.

Schulze Diabetes Institute, 420 Delaware Street S.E., Minneapolis, MN, 55455, (612) 626-3016, <http://www.surg.umn.edu/diabinst/home.html>.

United Network for Organ Sharing (UNOS), 700 North 4th St., Richmond, VA, 23219, (888) 894-6361, <http://www.transplantliving.org>.

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Pancreatectomy

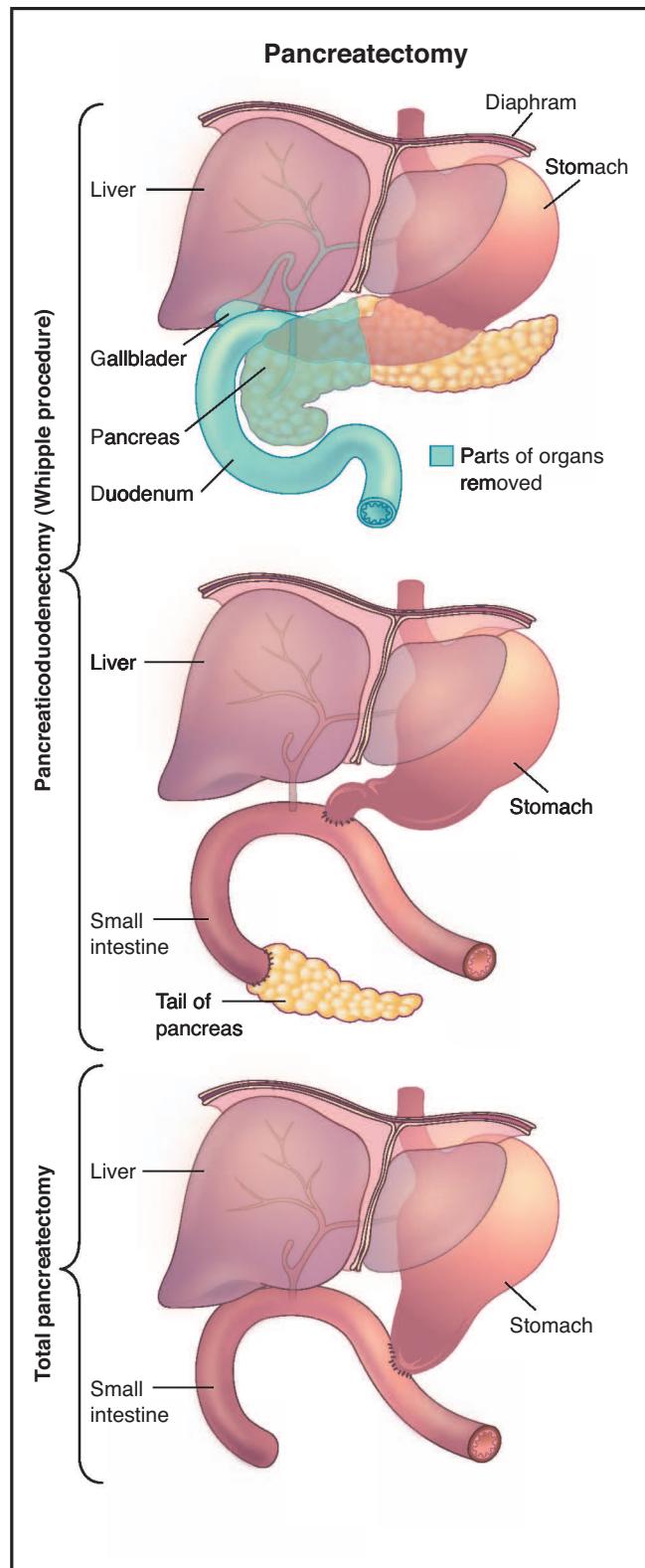
Definition

A pancreatectomy is the surgical removal of the pancreas. A pancreatectomy may be total, in which case the entire organ is removed, usually along with the spleen, gallbladder, common bile duct, and portions of the small intestine and stomach. A pancreatectomy may also be distal, meaning that only the body and tail of the pancreas are removed, leaving the head of the organ attached. When the duodenum is removed along with all or part of the pancreas, the procedure is called a pancreaticoduodenectomy, which surgeons sometimes refer to as Whipple's procedure. Pancreaticoduodenectomies are increasingly used to treat a variety of malignant and benign diseases of the pancreas. This procedure often involves removal of the regional lymph nodes as well.

Purpose

A pancreatectomy is the most effective treatment for **cancer** of the pancreas, an abdominal organ that secretes digestive enzymes, insulin, and other hormones. The thickest part of the pancreas near the duodenum (a part of the small intestine) is called the head, the middle part is called the body, and the thinnest part adjacent to the spleen is called the tail.

While surgical removal of tumors in the pancreas is the preferred treatment, it is only possible in the 10–15% of patients who are diagnosed early enough for a potential cure. Patients who are considered suitable for surgery usually have small tumors in the head of the pancreas (close to the duodenum, or first part of the small intestine), have **jaundice** as their initial



Two types of pancreatectomies: pancreaticoduodenectomy (top, also known as the Whipple procedure) and total pancreatectomy (bottom). (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

KEY TERMS

Chemotherapy—A cancer treatment that uses synthetic drugs to destroy the tumor either by inhibiting the growth of the cancerous cells or by killing the cancer cells.

Computed tomography (CT) scan—An imaging technique that creates a series of pictures of areas inside the body, taken from different angles. The pictures are created by a computer linked to an x-ray machine.

Endoscopic retrograde cholangiopancreatography (ERCP)—A procedure to x-ray the ducts (tubes) that carry bile from the liver to the gallbladder and from the gallbladder to the small intestine.

Laparoscopy—In this procedure, a laparoscope (a thin, lighted tube) is inserted through an incision in the abdominal wall to determine if the cancer is within the pancreas only or has spread to nearby tissues and if it can be removed by surgery later. Tissue samples may be removed for biopsy.

Magnetic resonance imaging (MRI)—A procedure in which a magnet linked to a computer is used to create detailed pictures of areas inside the body.

Pancreas—A large gland located on the back wall of the abdomen, extending from the duodenum (first part of the small intestine) to the spleen. The pancreas produces enzymes essential for digestion, and

the hormones insulin and glucagon, which play a role in diabetes.

Pancreaticoduodenectomy—Removal of all or part of the pancreas along with the duodenum. Also known as “Whipple’s procedure” or “Whipple’s operation.”

Pancreatitis—Inflammation of the pancreas, either acute (sudden and episodic) or chronic, usually caused by excessive alcohol intake or gallbladder disease.

Positron emission tomography (PET) scan—An imaging system that creates a picture showing the location of tumor cells in the body. A substance called radionuclide dye is injected into a vein, and the PET scanner rotates around the body to create the picture. Malignant tumor cells show up brighter in the picture because they are more active and take up more dye than normal cells.

Radiation therapy—A treatment using high energy radiation from x-ray machines, cobalt, radium, or other sources.

Ultrasonogram—A procedure where high-frequency sound waves that cannot be heard by human ears are bounced off internal organs and tissues. These sound waves produce a pattern of echoes which are then used by the computer to create sonograms, or pictures of areas inside the body.

symptom, and have no evidence of metastatic disease (spread of cancer to other sites). The stage of the cancer will determine whether the pancreatectomy to be performed should be total or distal.

A partial pancreatectomy may be indicated when the pancreas has been severely injured by trauma, especially injury to the body and tail of the pancreas. While such surgery removes normal pancreatic tissue as well, the long-term consequences of this surgery are minimal, with virtually no effects on the production of insulin, digestive enzymes, and other hormones.

Chronic **pancreatitis** is another condition for which a pancreatectomy is occasionally performed. Chronic pancreatitis—or continuing inflammation of the pancreas that results in permanent damage to this organ—can develop from long-standing, recurring episodes of acute (periodic) pancreatitis. This painful condition usually results from alcohol **abuse** or the presence of **gallstones**. In most patients with the alcohol-induced disease, the pancreas is widely involved, therefore, surgical correction is almost impossible.

Description

A pancreatectomy can be performed through an open surgery technique, in which case one large incision is made, or it can be performed laparoscopically, in which case the surgeon makes four small incisions to insert tube-like surgical instruments. The abdomen is filled with gas, usually carbon dioxide, to help the surgeon view the abdominal cavity. A camera is inserted through one of the tubes and displays images on a monitor in the operating room. Other instruments are placed through the additional tubes. The laparoscopic approach allows the surgeon to work inside the patient’s abdomen without making a large incision.

If the pancreatectomy is partial, the surgeon clamps and cuts the blood vessels, and the pancreas is stapled and divided for removal. If the disease affects the splenic artery or vein, the spleen is also removed.

If the pancreatectomy is total, the surgeon removes the entire pancreas and attached organs. He

or she starts by dividing and detaching the end of the stomach. This part of the stomach leads to the small intestine, where the pancreas and bile duct both attach. In the next step, he removes the pancreas along with the connected section of the small intestine. The common bile duct and the gallbladder are also removed. To reconnect the intestinal tract, the stomach and the bile duct are then connected to the small intestine.

During a pancreatectomy procedure, several tubes are also inserted for postoperative care. To prevent tissue fluid from accumulating in the operated site, a temporary drain leading out of the body is inserted, as well as a **gastrostomy** or g-tube leading out of the stomach in order to help prevent **nausea and vomiting**. A jejunostomy or j-tube may also be inserted into the small intestine as a pathway for supplementary feeding.

Diagnosis/Preparation

Patients with symptoms of a pancreatic disorder undergo a number of tests before surgery is even considered. These can include ultrasonography, x-ray examinations, **computed tomography scans** (CT scan), and **endoscopic retrograde cholangiopancreatography** (ERCP), a specialized imaging technique to visualize the ducts that carry bile from the liver to the gallbladder. Tests may also include **angiography**, another imaging technique used to visualize the arteries feeding the pancreas, and needle aspiration cytology, in which cells are drawn from areas suspected to contain cancer. Such tests are required to establish a correct diagnosis for the pancreatic disorder and in the planning the surgery.

Since many patients with pancreatic cancer are undernourished, appropriate nutritional support, sometimes by **tube feedings**, may be required prior to surgery.

Some patients with pancreatic cancer deemed suitable for a pancreatectomy will also undergo **chemotherapy** and/or **radiation therapy**. This treatment is aimed at shrinking the tumor, which will improve the chances for successful surgical removal. Sometimes, patients who are not initially considered surgical candidates may respond so well to chemoradiation that surgical treatment becomes possible. Radiation therapy may also be applied during the surgery (intraoperatively) to improve the patient's chances of survival, but this treatment is not yet in routine use. Some studies have shown that intraoperative radiation therapy extends survival by several months.

Patients undergoing **distal pancreatectomy** that involves removal of the spleen may receive preoperative medication to decrease the risk of infection.

Aftercare

Pancreatectomy is major surgery. Therefore, extended hospitalization is usually required with an average hospital stay of two to three weeks.

Some pancreatic cancer patients may also receive combined chemotherapy and radiation therapy after surgery. This additional treatment has been clearly shown to enhance survival rates.

After surgery, patients experience **pain** in the abdomen and are prescribed pain medication. Follow-up exams are required to monitor the patient's recovery and remove implanted tubes.

A total pancreatectomy leads to a condition called pancreatic insufficiency, because food can no longer be normally processed with the enzymes normally produced by the pancreas. Insulin secretion is likewise no longer possible. These conditions are treated with pancreatic enzyme replacement therapy, which supplies digestive enzymes; and with insulin injections. In some case, distal pancreatectomies may also lead to pancreatic insufficiency, depending on the patient's general health condition before surgery and on the extent of pancreatic tissue removal.

Risks

There is a fairly high risk of complications associated with any pancreatectomy procedure. A recent Johns Hopkins study documented complications in 41% of cases. The most devastating complication is postoperative bleeding, which increases the mortality risk to 20–50%. In cases of postoperative bleeding, the patient may be returned to surgery to find the source of hemorrhage, or may undergo other procedures to stop the bleeding.

One of the most common complications from a pancreaticoduodenectomy is delayed gastric emptying, a condition in which food and liquids are slow to leave the stomach. This complication occurred in 19% of patients in the Johns Hopkins study. To manage this problem, many surgeons insert feeding tubes at the original operation site, through which nutrients can be fed directly into the patient's intestines. This procedure, called enteral **nutrition**, maintains the patient's nutrition if the stomach is slow to recover normal function. Certain medications, called promotility agents, can help move the nutritional contents through the gastrointestinal tract.

The other most common complication is pancreatic anastomotic leak. This is a leak in the connection that the surgeon makes between the remainder of the pancreas and the other structures in the abdomen.

Most surgeons handle the potential for this problem by checking the connection during surgery.

Normal results

After a total pancreatectomy, the body loses the ability to secrete insulin, enzymes, and other substances; therefore, the patient has to take supplements for the rest of his or her life.

Patients usually resume normal activities within a month after surgery, although they are asked to avoid heavy lifting for six to eight weeks and not to drive as long as they take narcotic medication.

When a pancreatectomy is performed for chronic pancreatitis, the majority of patients obtain some relief from pain. Some studies report that one-half to three-quarters of patients become free of pain.

Morbidity and mortality rates

The mortality rate for pancreatectomy has decreased in recent years to 5–10%, depending on the extent of the surgery and the experience of the surgeon. A study of 650 patients at Johns Hopkins Medical Institution, Baltimore, found that only nine patients, or 1.4%, died from complications related to surgery.

Unfortunately, pancreatic cancer is the most lethal form of gastrointestinal malignancy. However, for a highly selective group of patients, a pancreatectomy offers a chance for cure, especially when performed by experienced surgeons. The overall five-year survival rate for patients who undergo pancreatectomy for pancreatic cancer is about 10%; patients who undergo pancreaticoduodenectomy have a 4–5% survival at five years. The risk for tumor recurrence is thought to be unaffected by whether the patient undergoes a total pancreatectomy or a pancreaticoduodenectomy, but is increased when the tumor is larger than 1.2 in (3 cm) and the cancer has spread to the lymph nodes or surrounding tissue.

Alternatives

Depending on the medical condition, a **pancreas transplantation** may be considered as an alternative for some patients.

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ORGANIZATIONS

American College of Gastroenterology, P.O. Box 342260, Bethesda, MD, 20827-2260, (301) 263-9000, <http://www.acg.gi.org>.

American Gastroenterological Association (AGA), 4930 Del Ray Avenue, Bethesda, MD, 20814, (301) 654-2055, (301) 654-5920, member@gastro.org, <http://www.gastro.org>.

National Cancer Institute, NCI Public Inquiries Office, 6116 Executive Boulevard, Bethesda, MD, 20892-8322, (800) 422-6237, <http://www.cancer.gov>.

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Pancreatic cancer, endocrine

Definition

Endocrine pancreatic **cancer** is a disease in which cancerous cells originate within the tissues of the pancreas that produce hormones.

Description

The pancreas is a 6–8 in (15–20 cm) long, slipper-shaped gland located in the abdomen. It lies behind the stomach, within a loop formed by the small intestine. Other nearby organs include the gallbladder, spleen, and liver. The pancreas has a wide end (head), a narrow end (tail), and a middle section (body). A healthy pancreas is important for normal food digestion and plays a critical role in the body's metabolic processes. The pancreas has two main functions, each performed by distinct types of tissue. The exocrine tissue secretes fluids into the other organs of the digestive system, while the endocrine tissue secretes substances that are circulated in the bloodstream. The exocrine pancreas makes up the vast majority of the gland; it produces pancreatic juices containing enzymes that help break down proteins and fatty food. The endocrine tissue of the pancreas makes up only 2% of the gland's total mass. It consists of small patches of cells that produce hormones (like insulin) that control how the body stores and uses nutrients. These patches are called islets (islands)

of Langerhans or islet cells and are interspersed evenly throughout the pancreas. Each islet contains approximately 1,000 endocrine cells and a dense network of capillaries (tiny blood vessels), which allows immediate entry of hormones into the circulatory system.

Pancreatic tumors are classified as either exocrine or endocrine tumors depending on which type of tissue they arise from within the gland. Endocrine tumors of the pancreas are very rare, accounting for only 5% of all pancreatic cancers. The majority of endocrine pancreatic tumors are functional adenocarcinomas that overproduce a specific hormone. There are several types of islet cells and each produces its own hormone or peptide (small protein molecule). Functional endocrine tumors are named after the hormone they secrete. Insulinoma is the most common tumor of the endocrine pancreas. Patients with this disease usually develop **hypoglycemia** due to increased insulin production that leads to abnormally low blood sugar levels. **Gastrinoma**, a disease in which gastrin (hormone that stimulates stomach acid production) is overproduced, causes multiple ulcers in the upper gastrointestinal (GI) tract. Gastrinoma was first described in patients with a rare form of severe peptic ulcer disease known as Zollinger-Ellison syndrome (ZES). The less common glucagonoma causes mild diabetes due to excess glucagon (hormone that stimulates glucose production) secretion. Other rare islet cell tumors include vipoma (vasoactive intestinal peptide) and somatostatinoma. Nonfunctional pancreatic endocrine tumors are not associated with an excess production of any hormone and can be difficult to distinguish from **exocrine pancreatic cancer**. Cancers of the endocrine pancreas are relatively slow-growing compared to the more common ductal adenocarcinomas of the exocrine pancreas.

Between one and four cases of insulinoma occur per million people per year, and 90% of these tumors are benign. They occur mostly between the ages of 50 and 60 and affect men and women equally. Less than three cases of gastrinoma per million people are diagnosed each year, but it is the most common functional islet cell tumor in patients with multiple endocrine tumors, a condition known as multiple endocrine neoplasia (MEN) syndrome. Vipoma and glucagonoma are even rarer and they occur more frequently in women. Somatostatinoma is exceedingly uncommon, and less than 100 cases have been reported worldwide. Nonfunctional islet cell cancers account for approximately one-third of all cancers of the endocrine pancreas, and the majority of these are malignant.

Causes and symptoms

There are no known causes of islet cell cancer, but a small percentage of cases occur due to hereditary syndromes such as MEN. This is a condition that frequently causes more than one tumor in several endocrine glands, such as the parathyroid and pituitary, in addition to the islet cells of the pancreas. Twenty-five percent of gastrinomas and less than 10% of insulinomas occur in MEN patients. Von Hippel-Lindau (VHL) syndrome is another genetic disorder that causes multiple tumors, and 10–15% of VHL patients will develop islet cell cancer.

Symptoms vary among the different islet cell cancer types. Insulinoma causes repeated episodes of hypoglycemia, sweating, and **tremors**, while patients with gastrinoma have inflammation of the esophagus, epigastric **pain**, multiple ulcers, and possibly **diarrhea**. Symptoms of glucagonoma include a distinctive skin rash, inflammation of the stomach, glucose intolerance, weight loss, weakness, and anemia (less common). Patients with vipoma have episodes of profuse, watery diarrhea, even after **fasting**. Somatostatinoma causes mild diabetes, diarrhea/steatorrhea (fatty stools), weight loss, and gallbladder disease. Nonfunctional endocrine tumors frequently produce the same symptoms as cancer of the exocrine pancreas such as abdominal pain, **jaundice**, and weight loss.

Diagnosis

A thorough physical exam is usually performed when a patient presents with the above symptoms, however, functional endocrine tumors of the pancreas tend to be small and are not detected by palpating the abdomen. Once other illnesses such as infection are ruled out, the doctor will order a series of blood and urine tests. The functional endocrine tumors can be identified through increased levels of hormone in the bloodstream.

Functional endocrine tumors can occur in multiple sites in the pancreas and are often small (less than 1 cm), making them difficult to diagnose. Nonfunctional tumors tend to be larger, which makes them difficult to distinguish from tumors of the exocrine pancreas. Methods such as computed tomography (CT) scan and **magnetic resonance imaging** (MRI) are used to take pictures of the internal organs and allow the doctor to determine whether a tumor is present. Somatostatin receptor scintigraphy (trade name OctreoScan) is an imaging system used to localize endocrine tumors, especially gastrinomas and somatostatinomas. Endoscopic ultrasound (EUS) is

a more sensitive technique that may be used if a CT scan fails to detect a tumor. Endocrine tumors usually have many blood vessels, so **angiography** may be useful in the doctor's assessment and staging of the tumor. Surgical exploration is sometimes necessary in order to locate very small tumors that occur in multiple sites. These techniques also help the doctor evaluate how far the tumor has spread. A biopsy can be taken to confirm diagnosis, but more often, doctors look at the size and local invasion of the tumor in order to plan a treatment strategy.

Treatment

Staging

The staging system for islet cell cancer is still evolving, but the tumors typically fall into three categories: cancers that arise in one location within the pancreas, cancers that arise in several locations within the pancreas, and cancers that have spread to nearby lymph nodes or to other organs in the body.

Surgery is the only curative method for islet cell cancers, and studies have shown that an aggressive surgical approach can improve survival and alleviate symptoms of the disease. As with most forms of cancer, the earlier it is diagnosed, the greater the chance for survival. With the exception of insulinoma, the majority of islet cell tumors are malignant at the time of diagnosis, and more than half are metastatic. However, surgery and **chemotherapy** have been shown to improve the outcome of patients even if they have metastatic disease. Surgery may include partial or total removal of the pancreas, and in patients with gastrinoma, the stomach may be removed as well. Streptozotocin, doxorubicin, and 5-fluorouracil (5-FU) are chemotherapeutic agents commonly used in the treatment of islet cell cancer. Patients may experience **nausea and vomiting** as well as kidney toxicity from streptozotocin, and bone marrow suppression from doxorubicin. Hormone therapy is used to relieve the symptoms of functional tumors by inhibiting excess hormone production. Other techniques may be used to block blood flow to the liver in an attempt to kill the cancer cells that have spread there. Abdominal pain, **nausea, vomiting** and **fever** may result from this type of treatment. Radiation has little if any role in the treatment of islet cell cancer.

Prognosis

Islet cell cancers overall have a more favorable prognosis than cancers of the exocrine pancreas, and the median survival from diagnosis is three and a half

KEY TERMS

Adenocarcinoma—A malignant tumor that arises within the tissues of a gland and retains its glandular structure.

Angiography—Diagnostic technique used to study blood vessels in a tumor.

Biopsy—Removal and microscopic examination of cells to determine whether they are cancerous.

Chemotherapy—Drug treatment administered to kill cancerous cells.

Endocrine—Refers to glands that secrete hormones circulated in the bloodstream.

Endoscopic ultrasonography (EUS)—Diagnostic imaging technique where an ultrasound probe is inserted down a patient's throat to determine if a tumor is present.

Gastrinoma—Tumor that arises from the gastrin-producing cells in the pancreas.

Insulinoma—Tumor that arises from the insulin-producing cells in the pancreas.

Islets of Langerhans—Clusters of cells in the pancreas that make up the endocrine tissue.

years. This is mainly due to their slow-growing nature. Insulinomas have a five-year survival rate of 80% and gastrinomas have 65%. When malignant, islet cell cancers do not generally respond well to chemotherapy, and the treatment is mainly palliative. Most patients with metastasis do not survive five years. Islet cell cancer tends to spread to the surrounding lymph nodes, stomach, small intestine, and liver.

Prevention

There are no known risk factors associated with sporadic islet cell cancer. Therefore, it is not clear how to prevent its occurrence. Individuals with MEN syndrome or VHL, however, have a genetic predisposition to developing islet cell cancer and should be screened regularly in an effort to catch the disease early.

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ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd., NE, Atlanta, GA 30329, (404) 320-3333, (800) ACS-2345. <http://www.cancer.org>.

Cancer Research and Prevention Foundation, 1600 Duke Street, Suite 500, Alexandria, VA 22314, (703) 836-4412, (800) 227-2732, info@preventcancer.org, <http://www.preventcancer.org>.

National Cancer Institute, 9000 Rockville Pike, Bldg. 31, Rm. 10A16, Bethesda, MD, 20892, (800) 422-6237, <http://www.nci.nih.gov>.

National Familial Pancreas Tumor Registry, The Johns Hopkins Hospital, 600 North Wolfe St, Baltimore, MD, 21287-6417, (410) 377-7450

National Organization for Rare Disorders, 100 Route 37, PO Box 8923, New Fairfield, CT, 06812, (203) 746-6518, <http://www.nord-rdb.com/~orphan>.

National Pancreas Foundation, 101 Federal Street, Suite 1900, Boston, MA 02110, (617) 578-0382, (866) 726-2737, (617) 578-0383. <http://www.pancreasfoundation.org>.

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Pancreatic cancer, exocrine

Definition

Exocrine pancreatic **cancer** is a disease in which cancerous cells originate within the tissues of the pancreas that produce digestive juices.

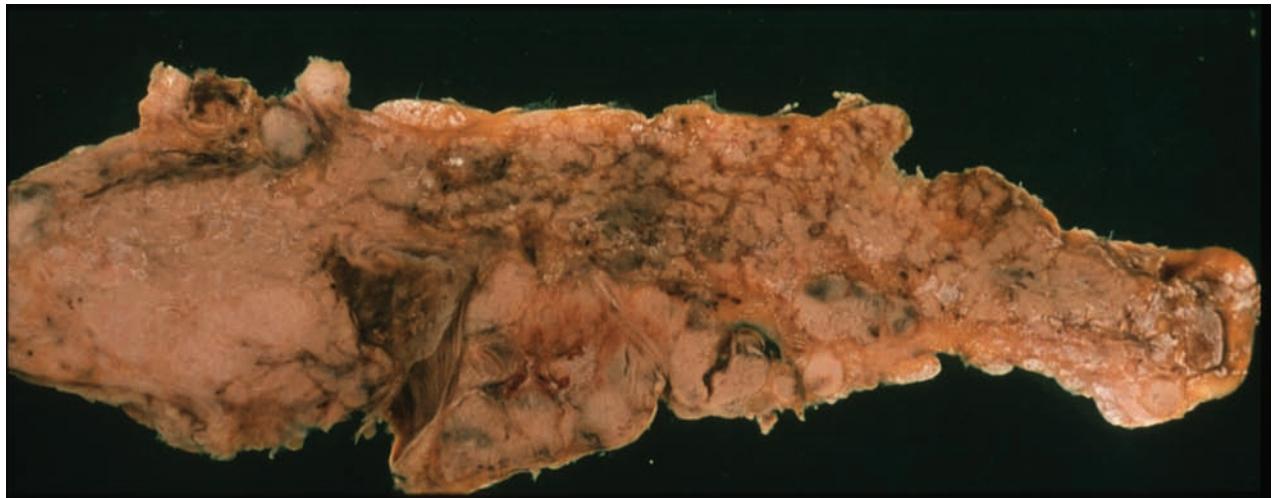
Description

The pancreas is a 6–8 in (15–20 cm) long, slipper-shaped gland located in the abdomen. It lies behind the stomach, within a loop formed by the small intestine. Other nearby organs include the gallbladder, spleen, and liver. The pancreas has a wide end (head), a narrow end (tail), and a middle section (body). A healthy pancreas is important for normal food digestion and also plays a critical role in the body's metabolic processes. The pancreas has two main functions, and each are performed by distinct types of tissue. The exocrine tissue makes up the vast

majority of the gland and secretes fluids into the other organs of the digestive system. The endocrine tissue secretes hormones (like insulin) that are circulated in the bloodstream, and these substances control how the body stores and uses nutrients. The exocrine tissue of the pancreas, comprised mostly of acinar cells and ductal cells, produces pancreatic (digestive) juices. These juices contain several enzymes that help break down proteins and fatty foods. The exocrine pancreas forms an intricate system of channels or ducts, which are tubular structures that carry pancreatic juices to the small intestine where they are used for digestion.

Pancreatic tumors are classified as either exocrine or endocrine tumors depending on which type of tissue they arise from within the gland. Ninety-five percent of pancreatic cancers occur in the tissues of the exocrine pancreas. Ductal adenocarcinomas arise in the cells that line the ducts of the exocrine pancreas and account for 80% to 90% of all tumors of the pancreas. Unless specified, nearly all reports on pancreatic cancer refer to ductal adenocarcinomas. Less common types of pancreatic exocrine tumors include acinar cell carcinoma, cystic tumors that are typically benign but may become cancerous, and papillary tumors that grow within the pancreatic ducts. Pancreatoblastoma is a very rare disease that primarily affects young children. Two-thirds of pancreatic tumors occur in the head of the pancreas, and tumor growth in this area can lead to the obstruction of the nearby common bile duct that empties bile fluid into the small intestine. When bile cannot be passed into the intestine, patients may develop yellowing of the skin and eyes (**jaundice**) due to the buildup of bilirubin (a component of bile) in the bloodstream. Tumor blockage of bile or pancreatic ducts may also cause digestive problems since these fluids contain critical enzymes in the digestive process. Depending on their size, pancreatic tumors may cause abdominal **pain** by pressing on the surrounding nerves. Because of its location deep within the abdomen, pancreatic cancer often remains undetected until it has spread to other organs such as the liver or lung. Pancreatic cancer tends to rapidly spread to other organs, even when the primary (original) tumor is relatively small.

Though pancreatic cancer accounts for only 3% of all cancers, it is the fifth most frequent cause of cancer deaths. In 2010, an estimated 43,140 new cases of pancreatic cancer were diagnosed in the United States. Pancreatic cancer is primarily a disease associated with advanced age, with 80% of cases occurring between the ages of 60 and 80. Men are almost twice as likely to develop this disease than women. Countries



Carcinoma of the head of the pancreas. Tumors appear as gritty, gray, hard nodules, invading the adjacent gland. (Biophoto Associates/Science Source/Photo Researchers, Inc.)

with the highest frequencies of pancreatic cancer include the United States, New Zealand, Western European nations, and Scandinavia. The lowest occurrences of the disease are reported in India, Kuwait and Singapore. African Americans have the highest incidence of pancreatic cancer of any ethnic group worldwide. Whether this difference is due to diet or environmental factors remains unclear.

Causes and symptoms

Although the exact cause for pancreatic cancer is not known, several risk factors have been shown to increase susceptibility to this particular cancer, the greatest of which is cigarette **smoking**. Approximately one-third of pancreatic cancer cases occur among smokers. People who have diabetes develop pancreatic cancer twice as often as non-diabetics. Numerous studies suggest that a family history of pancreatic cancer is another strong risk factor for developing the disease, particularly if two or more relatives in the immediate family have the disease. Other risk factors include chronic (long-term) inflammation of the pancreas (**pancreatitis**), **diets** high in fat, and occupational exposure to certain chemicals such as petroleum.

Pancreatic cancer often does not produce symptoms until it reaches an advanced stage. Patients may then present with the following signs and symptoms:

- upper abdominal and/or back pain
- jaundice
- weight loss

- loss of appetite
- diarrhea
- weakness
- nausea

These symptoms may also be caused by other illnesses; therefore, it is important to consult a doctor for an accurate diagnosis.

Diagnosis

Pancreatic cancer is difficult to diagnose, especially in the absence of symptoms, and there is no current screening method for early detection. The most sophisticated techniques available often do not detect very small tumors that are localized (have not begun to spread). At advanced stages where patients show symptoms, a number of tests may be performed to confirm diagnosis and to assess the stage of the disease. Approximately half of all pancreatic cancers are metastatic (have spread to other sites) at the time of diagnosis.

The first step in diagnosing pancreatic cancer is a thorough medical history and complete **physical examination**. The abdomen will be palpated to check for fluid accumulation, lumps, or masses. If there are signs of jaundice, blood tests will be performed to rule out the possibility of liver diseases such as hepatitis. Urine and stool tests may be performed as well.

Non-invasive imaging tools such as computed tomography (CT) scans and **magnetic resonance imaging** (MRI) can be used to produce detailed pictures of the internal organs. CT is the tool most often

used to diagnose pancreatic cancer, as it allows the doctor to determine if the tumor can be removed by surgery or not. It is also useful in staging a tumor by showing the extent to which the tumor has spread. During a CT scan, patients receive an intravenous injection of a contrast dye so the organs can be visualized more clearly. MRI may be performed instead of CT if a patient has an allergy to the CT contrast dye. In some cases where the tumor is impinging on blood vessels or nearby ducts, MRI may be used to generate an image of the pancreatic ducts.

If the doctor suspects pancreatic cancer and no visible masses are seen with a CT scan, a patient may undergo a combination of invasive tests to confirm the presence of a pancreatic tumor. Endoscopic ultrasound (EUS) involves the use of an ultrasound probe at the end of a long, flexible tube that is passed down the patient's throat and into the stomach. This instrument can detect a tumor mass through high frequency sound waves and echoes. EUS can be accompanied by fine needle aspiration (FNA), where a long needle, guided by the ultrasound, is inserted into the tumor mass in order to take a biopsy sample. **Endoscopic retrograde cholangiopancreatography** (ERCP) is a technique often used in patients with severe jaundice because it enables the doctor to relieve blockage of the pancreatic ducts. The doctor, guided by **endoscopy** and x rays, inserts a small metal or plastic stent into the duct to keep it open. During ERCP, a biopsy can be done by collecting cells from the pancreas with a small brush. The cells are then examined under the microscope by a pathologist, who determines the presence of any cancerous cells.

In some cases, a biopsy may be performed during a type of surgery called **laparoscopy**, which is done under **general anesthesia**. Doctors insert a small camera and instruments into the abdomen after a minor incision is made. Tissue samples are removed for examination under the microscope. This procedure allows a doctor to determine the extent to which the disease has spread and decide if the tumor can be removed by further surgery.

An **angiography** is a type of test that studies the blood vessels in and around the pancreas. This test may be done before surgery so that the doctor can determine the extent to which the tumor invades and interacts with the blood vessels within the pancreas. The test requires **local anesthesia** and a catheter is inserted into the patient's upper thigh. A dye is then injected into blood vessels that lead into the pancreas, and x rays are taken.

In April 2001, doctors at major cancer research institutions such as Memorial Sloan-Kettering Cancer Center in New York were investigating CT angiography, an imaging technique that is less invasive than angiography alone. CT angiography is similar to a standard CT scan, but allows doctors to take a series of pictures of the blood vessels that support tumor growth. A dye is injected as in a CT scan (but at rapid intervals) and no catheter or **sedation** is required. A computer generates 3D images from the pictures that are taken, and the information is gathered by the surgical team who will develop an appropriate strategy if the patient's disease can be operated on.

Treatment

Staging

After cancer of the pancreas has been diagnosed, doctors typically use a TNM staging system to classify the tumor based on its size and the degree to which it has spread to other areas in the body. T indicates the size and local advancement of the primary tumor. Since cancers often invade the lymphatic system before spreading to other organs, regional lymph node involvement (N) is an important factor in staging. M indicates whether the tumor has metastasized (spread) to distant organs. In stage I, the tumor is localized to the pancreas and has not spread to surrounding lymph nodes or other organs. Stage II pancreatic cancer has spread to nearby organs such as the small intestine or bile duct, but not the surrounding lymph nodes. Stage III indicates lymph node involvement, whether the cancer has spread to nearby organs or not. Stage IVA pancreatic cancer has spread to organs near the pancreas such as the stomach, spleen, or colon. Stage IVB is a cancer that has spread to distant sites (liver, lung). If pancreatic cancer has been treated with success and then appears again in the pancreas or in other organs, it is referred to as recurrent disease.

Treatment of pancreatic cancer will depend on several factors, including the stage of the disease and the patient's age and overall health status. A combination of therapies is often employed in the treatment of this disease to improve the patient's chances for survival. Surgery is used whenever possible and is the only means by which cancer of the pancreas can be cured. However, less than 15% of pancreatic tumors can be removed by surgery. By the time the disease is diagnosed (usually at stage III), therapies such as radiation and **chemotherapy** or both are used in addition to surgery to relieve a patient's symptoms and

enhance quality of life. For patients with metastatic disease, chemotherapy and radiation are used mainly as palliative (pain alleviating) treatments.

Surgery

Three types of surgery are used in the treatment of pancreatic cancer, depending on what section of the pancreas the tumor is located in. A Whipple procedure removes the head of the pancreas, part of the small intestine and some of the surrounding tissues. This procedure is most common since the majority of pancreatic cancers occur in the head of the organ. A total **pancreatectomy** removes the entire pancreas and the organs around it. **Distal pancreatectomy** removes only the body and tail of the pancreas. Chemotherapy and radiation may precede surgery (neoadjuvant therapy) or follow surgery (adjuvant therapy). Surgery is also used to relieve symptoms of pancreatic cancer by draining fluids or bypassing obstructions. Side effects from surgery can include pain, weakness, **fatigue**, and digestive problems. Some patients may develop diabetes or malabsorption as a result of partial or total removal of the pancreas.

Radiation therapy

Radiation therapy is sometimes used to shrink a tumor before surgery or to remove remaining cancer cells after surgery. Radiation may also be used to relieve pain or digestive problems caused by the tumor if it cannot be removed by surgery. External radiation therapy refers to radiation applied externally to the abdomen using a beam of high-energy x rays. High-dose intraoperative radiation therapy is sometimes used during surgery on tumors that have spread to nearby organs. Internal radiation therapy refers to the use of small radioactive seeds implanted in the tumor tissue. The seeds emit radiation over a period of time to kill tumor cells. Radiation treatment may cause side effects such as fatigue, tender or itchy skin, **nausea**, **vomiting**, and digestive problems.

Chemotherapy

Chemotherapeutic agents are powerful drugs that are used to kill cancer cells. They are classified according to the mechanism by which they induce cancer cell **death**. Multiple agents are often used to increase the chances of tumor cell death. Gemcitabine is the standard drug used to treat pancreatic cancers and can be used alone or in combination with other drugs, such as 5-flourouracil (5-FU). Other drugs are being tested in combination with gemcitabine in several ongoing clinical trials, specifically irinotecan (CPT-11) and oxaliplatin. Chemotherapy may be administered orally or

intravenously in a series of doses over several weeks. During treatment, patients may experience fatigue, nausea, **vomiting**, hair loss, and mouth sores, depending on which drugs are used.

Biological treatments

Numerous vaccine treatments are being developed in an effort to stimulate the body's immune system into attacking cancer cells. This is also referred to as immunotherapy. Another type of biological treatment involves using a targeted monoclonal antibody to inhibit the growth of cancer cells. The antibody is thought to bind to and neutralize a protein that contributes to the growth of the cancer cells. Investigational treatments such as these may be considered by patients with metastatic disease who would like to participate in a clinical trial. Biological treatments typically cause flu-like symptoms (chills, **fever**, loss of appetite) during the treatment period.

Alternative treatment

Acupuncture or **hypnotherapy** may be used in addition to standard therapies to help relieve the pain associated with pancreatic cancer. Because of the poor prognosis associated with pancreatic cancer, some patients may try special diets with vitamin supplements, certain **exercise** programs, or unconventional treatments not yet approved by the FDA. Patients should always inform their doctors of any alternative treatments they are using as they could interfere with standard therapies. The National Cancer Institute (NCI) is funding phase III clinical trials of a controversial treatment for pancreatic cancer that involves the use of supplemental pancreatic enzymes (to digest cancerous cells) and coffee **enemas** (to stimulate the liver to detoxify the cancer). These theories remain unproven and the study is widely criticized in the medical community. It remains to be seen whether this method of treatment has any advantage over the standard chemotherapeutic regimen in prolonging patient survival or improving quality of life.

Prognosis

Unfortunately, cancer of the pancreas is often fatal, and median survival from diagnosis is less than six months, while the five-year survival rate is 4%. This is mainly due to the lack of screening methods available for early detection of the disease. Yet, even when localized tumors can be removed by surgery, patient survival after five years is only 10% to 15%. These statistics demonstrate the aggressive nature of most pancreatic cancers and their tendency to recur. Pancreatic cancers tend to be resistant to radiation

KEY TERMS

Acinar cell carcinoma—A malignant tumor arising from the acinar cells of the pancreas.

Angiography—Diagnostic technique used to study blood vessels in a tumor.

Biopsy—Removal and microscopic examination of cells to determine whether they are cancerous.

Cancer vaccines—A treatment that uses the patient's immune system to attack cancer cells.

Chemotherapy—Drug treatment administered to kill cancerous cells.

Ductal adenocarcinoma—A malignant tumor arising from the duct cells within a gland.

Endoscopic retrograde cholangiopancreatography (ERCP)—Diagnostic technique used to obtain a biopsy. Also a surgical method of relieving biliary obstruction caused by a tumor.

Endoscopic ultrasonography (EUS)—Diagnostic imaging technique in which an ultrasound probe

is inserted down a patient's throat to determine if a tumor is present.

Exocrine—Refers to glands which secrete their products through a duct.

Laparoscopic surgery—Minimally invasive surgery in which a camera and surgical instruments are inserted through a small incision.

Pancreatectomy—Partial or total surgical removal of the pancreas.

Radiation therapy—Use of radioisotopes to kill tumor cells. Applied externally through a beam of x rays, intraoperatively (during surgery), or deposited internally by implanting radioactive seeds in tumor tissue.

Whipple procedure—Surgical removal of the head of the pancreas, part of the small intestine, and some surrounding tissue.

and chemotherapy and these modes of treatment are mainly used to relieve pain and tumor burden.

Prevention

Although the exact cause of pancreatic cancer is not known, there are certain risk factors that may increase a person's chances of developing the disease. Quitting smoking will certainly reduce the risk for pancreatic cancer and many other cancers. The American Cancer Society recommends a diet rich in fruits, vegetables, and dietary fiber in order to reduce the risk of pancreatic cancer. According to the NCI, workers who are exposed to petroleum and other chemicals may be at greater risk for developing the disease and should follow their employer's safety precautions. People with a family history of pancreatic cancer are at greater risk than the general population, as a small percentage of pancreatic cancers are considered hereditary.

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ORGANIZATIONS

CancerNet, National Cancer Institute, 9000 Rockville Pike, Bldg. 31, Rm.10A16, Bethesda, MD, 20892, (800) 422-6237, <http://www.cancer.org>.

Hirshberg Foundation for Pancreatic Cancer Research, 375 Homewood Rd, Los Angeles, CA, 90049, (310) 472-6310, <http://www.pancreatic.org>.

National Pancreas Foundation, P.O. Box 935, Wexford, PA, 15090-0935, <http://www.pancreasfoundation.org>.

Pancreatic Cancer Action Network, P.O. Box 1010, Torrance, CA, 90505, (877) 272-6226, <http://www.pancan.org>.

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Pancreatitis

Definition

Pancreatitis is an inflammation of the pancreas, an organ that is important in digestion. Pancreatitis can be acute (beginning suddenly, usually with the patient recovering fully) or chronic (progressing slowly with continued, permanent injury to the pancreas).

Demographics

The incidence of acute pancreatitis is approximately 40 cases per year per 100,000 adults, with the United States and Finland being the two most predominant countries of its occurrence. Mild forms of pancreatitis account for approximately 80% of the cases, with a mortality rate as low as 1%, while severe pancreatitis (approximately 20%) has as high as a 30% rate of mortality. African Americans have the highest incidence (approximately 20.5%), Caucasians approximately 5.5%, and Native Americans, approximately 4% per 100,000 population. The most common age for pancreatitis to occur is between 35–64 years of age.

Description

The pancreas is located in the midline of the back of the abdomen, closely associated with the liver, stomach, and duodenum (the first part of the small intestine). The pancreas is considered a gland. A gland is an organ whose primary function is to produce chemicals that pass either into the main blood circulation (called an endocrine function), or pass into another organ (called an exocrine function). The pancreas is unusual because it has both endocrine and exocrine functions. Its endocrine function produces three hormones. Two of these hormones, insulin and glucagon, are central to the processing of sugars in the diet (carbohydrate metabolism or breakdown). The third hormone produced by the endocrine cells of the pancreas affects gastrointestinal functioning. This hormone is called vasoactive intestinal polypeptide (VIP). The pancreas's exocrine function produces a variety of digestive enzymes (trypsin, chymotrypsin, lipase, and amylase, among others). These enzymes are passed into the duodenum through a channel called the pancreatic duct. In the duodenum, the enzymes begin the process of breaking down a variety of food components, including, proteins, fats, and starches.

Acute pancreatitis occurs when the pancreas suddenly becomes inflamed but improves. Patients recover

fully from the disease, and in almost 90% of cases the symptoms disappear within about a week after treatment. The pancreas returns to its normal architecture and functioning after healing from the illness. After an attack of acute pancreatitis, tissue and cells of the pancreas return to normal. With chronic pancreatitis, damage to the pancreas occurs slowly over time. Symptoms may be persistent or sporadic, but the condition does not disappear and the pancreas is permanently impaired. Pancreatic tissue is damaged, and the tissue and cells function poorly.

Causes and symptoms

There are a number of causes of acute pancreatitis. The most common, however, are gallbladder disease and **alcoholism**. These two diseases are responsible for more than 80% of all hospitalizations for acute pancreatitis. Other factors in the development of pancreatitis include:

- certain drugs
- infections
- structural problems of the pancreatic duct and bile ducts (channels leading from the gallbladder to the duodenum)
- injury to the abdomen resulting in injury to the pancreas (including injuries occurring during surgery)
- abnormally high levels of circulating fats in the bloodstream
- malfunction of the parathyroid gland, with high blood levels of calcium
- complications from kidney transplants
- a hereditary tendency toward pancreatitis.

Pancreatitis caused by drugs accounts for about 5% of all cases. Some drugs that are definitely related to pancreatitis include:

- Azathioprine, 6-mercaptopurine (Imuran)
- Dideoxyinosine (Videx)
- Estrogens (birth control pills)
- Furosemide (Lasix)
- Pentamidine (NebuPent)
- Sulfonamides (Urobak, Azulfidine)
- Tetracycline
- Thiazide diuretics (Diuril, Enduron)
- Valproic acid (Depakote).

Some drugs that are probably related to pancreatitis include:

- Acetaminophen (Tylenol)
- Angiotensin-converting enzyme (ACE) inhibitors (Capoten, Vasotec)

- Erythromycin
- Methyldopa (Aldomet)
- Metronidazole (Flagyl, Protostat)
- Nitrofurantoin (Furadantin, Furan)
- Nonsteroidal anti-inflammatory drugs (NSAIDs) (Aleve, Naprosyn, Motrin)
- Salicylates (aspirin).

All of these causes of pancreatitis seem to have a similar mechanism in common. Under normal circumstances, many of the extremely potent enzymes produced by the pancreas are not active until they are passed into the duodenum, where contact with certain other chemicals allow them to function. In pancreatitis, something allows these enzymes to become prematurely activated, so that they actually begin their digestive functions within the pancreas. The pancreas, in essence, begins digesting itself. A cycle of inflammation begins, including swelling and loss of function. Digestion of the blood vessels in the pancreas results in bleeding. Other active pancreatic chemicals cause blood vessels to become leaky, and fluid begins leaking out of the normal circulation into the abdominal cavity. The activated enzymes also gain access to the bloodstream through leaky, eroded blood vessels, and begin circulating throughout the body.

Pain is a major symptom in pancreatitis. The pain is usually quite intense and steady, located in the upper right hand corner of the abdomen, and often described as “boring.” This pain is also often felt all the way through to the patient’s back. The patient’s breathing may become quite shallow because deeper breathing tends to cause more pain. Relief of pain by sitting up and bending forward is characteristic of pancreatic pain. **Nausea and vomiting**, and abdominal swelling are all common as well. A patient will often have a slight **fever**, with an increased heart rate and low blood pressure.

Classic signs of **shock** may appear in more severely ill patients. Shock is a very serious syndrome that occurs when the volume (quantity) of fluid in the blood is very low. In shock, a patient’s arms and legs become extremely cold, the blood pressure drops dangerously low, the heart rate is quite fast, and the patient may begin to experience changes in mental status.

In very severe cases of pancreatitis (called necrotizing pancreatitis), the pancreatic tissue begins to die, and bleeding increases. Due to the bleeding into the abdomen, two distinctive signs may be noted in patients with necrotizing pancreatitis. Turner’s sign is a reddish-purple or greenish-brown color to the

flank area (the area between the ribs and the hip bone). Cullen’s sign is a bluish color around the navel.

Some of the complications of pancreatitis are due to shock. When shock occurs, all of the body’s major organs are deprived of blood (and, therefore, oxygen), resulting in damage. Kidney, respiratory, and **heart failure** are serious risks of shock. The pancreatic enzymes that have begun circulating throughout the body (as well as various poisons created by the abnormal digestion of the pancreas by those enzymes) have severe effects on the major body systems. Any number of complications can occur, including damage to the heart, lungs, kidneys, lining of the gastrointestinal tract, liver, eyes, bones, and skin. As the pancreatic enzymes work on blood vessels surrounding the pancreas, and even blood vessels located at a distance, the risk of **blood clots** increases. These blood clots complicate the situation by blocking blood flow in the vessels. When blood flow is blocked, the supply of oxygen is decreased to various organs and the organ can be damaged.

The pancreas may develop additional problems, even after the pancreatitis decreases. When the entire organ becomes swollen and suffers extensive cell **death** (pancreatic necrosis), the pancreas becomes extremely susceptible to serious infection. A local collection of pus (called a pancreatic **abscess**) may develop several weeks after the illness subsides, and may result in increased fever and a return of pain. Another late complication of pancreatitis, occurring several weeks after the illness begins, is called a pancreatic pseudocyst. This occurs when dead pancreatic tissue, blood, white blood cells, enzymes, and fluid leaked from the circulatory system accumulate. In an attempt to enclose and organize this abnormal accumulation, a kind of wall forms from the dead tissue and the growing scar tissue in the area. Pseudocysts cause additional abdominal pain by putting pressure on and displacing pancreatic tissue (resulting in more pancreatic damage). Pseudocysts also press on other nearby structures in the gastrointestinal tract, causing more disruption of function. Pseudocysts are life-threatening when they become infected (abscess) and when they rupture. Simple rupture of a pseudocyst causes death 14% of the time. Rupture complicated by bleeding causes death 60% of the time.

As the pancreatic tissue is increasingly destroyed in chronic pancreatitis, many digestive functions become disturbed. The quantity of hormones and enzymes normally produced by the pancreas begins to seriously decrease. Decreases in the production of enzymes result in the inability to appropriately digest food. Fat digestion, in particular, is impaired. A patient’s stools become greasy as fats are passed out

of the body. The inability to digest and use proteins results in smaller muscles (wasting) and weakness. The inability to digest and use the nutrients in food leads to **malnutrition**, and a generally weakened condition. As the disease progresses, permanent injury to the pancreas can lead to diabetes.

Diagnosis

Diagnosis of pancreatitis can be made very early in the disease by noting high levels of pancreatic enzymes circulating in the blood (amylase and lipase). Later in the disease, and in chronic pancreatitis, these enzyme levels will no longer be elevated. Because of this fact, and because increased amylase and lipase can also occur in other diseases, the discovery of such elevations are helpful but not mandatory in the diagnosis of pancreatitis. Other abnormalities in the blood may also point to pancreatitis, including increased white blood cells (occurring with inflammation and/or infection), changes due to **dehydration** from fluid loss, and abnormalities in the blood concentration of **calcium**, magnesium, **sodium**, potassium, bicarbonate, and sugars.

X rays or ultrasound examination of the abdomen may reveal **gallstones**, perhaps responsible for blocking the pancreatic duct. The gastrointestinal tract will show signs of inactivity (**ileus**) due to the presence of pancreatitis. Chest x rays may reveal abnormalities due to air trapping from shallow breathing, or due to lung complications from the circulating pancreatic enzyme irritants. **Computed tomography scans** (CT scans) of the abdomen may reveal the inflammation and fluid accumulation of pancreatitis, and may also be useful when complications like an abscess or a pseudocyst are suspected.

In the case of chronic pancreatitis, a number of blood tests will reveal the loss of pancreatic function that occurs over time. Blood sugar (glucose) levels will rise, eventually reaching the levels present in diabetes. The levels of various pancreatic enzymes will fall, as the organ is increasingly destroyed and replaced by non-functioning scar tissue. Calcification of the pancreas can also be seen on x rays. **Endoscopic retrograde cholangiopancreatography** (ERCP) may be used to diagnose chronic pancreatitis in severe cases. In this procedure, the doctor uses a medical instrument fitted with a fiber-optic camera to inspect the pancreas. A magnified image of the area is shown on a television screen viewed by the doctor. Many endoscopes also allow the doctor to retrieve a small sample (biopsy) of pancreatic tissue to examine under a microscope. A

contrast product may also be used for radiographic examination of the area.

Treatment

Treatment of pancreatitis involves quickly and sufficiently replacing lost fluids by giving the patient new fluids through a needle inserted in a vein (intravenous or IV fluids). These IV solutions need to contain appropriate amounts of salts, sugars, and sometimes even proteins, in order to correct the patient's disturbances in blood chemistry. Pain is treated with a variety of medications. In order to decrease pancreatic function (and decrease the discharge of more potentially harmful enzymes into the bloodstream), the patient is not allowed to eat. A thin, flexible tube (nasogastric tube) may be inserted through the patient's nose and down into his or her stomach. The nasogastric tube can empty the stomach of fluid and air, which may accumulate due to the inactivity of the gastrointestinal tract. Oxygen may need to be administered by nasal prongs or by a mask.

The patient will need careful monitoring in order to identify complications that may develop. Infections (often occurring in cases of necrotizing pancreatitis, abscesses, and pseudocysts) will require **antibiotics** through the IV. Severe necrotizing pancreatitis may require surgery to remove part of the dying pancreas. A pancreatic abscess can be drained by a needle inserted through the abdomen and into the collection of pus (percutaneous needle aspiration). If this is not sufficient, an abscess may also require surgical removal. Pancreatic pseudocysts may shrink on their own (in 25–40% of cases) or may continue to expand, requiring needle aspiration or surgery. When diagnostic exams reveal the presence of gallstones, surgery may be necessary for their removal. When a patient is extremely ill from pancreatitis, however, such surgery may need to be delayed until any infection is treated, and the patient's condition stabilizes.

Because chronic pancreatitis often includes repeated flares of acute pancreatitis, the same kinds of basic treatment are necessary. Patients cannot take solids or fluids by mouth. They receive IV replacement fluids, receive pain medication, and are monitored for complications. Treatment of chronic pancreatitis caused by alcohol consumption requires that the patient stop drinking alcohol entirely. As chronic pancreatitis continues and insulin levels drop, a patient may require insulin injections in order to be able to process sugars in his or her diet. Pancreatic enzymes can be replaced with oral medicines, and patients

sometimes have to take as many as eight pills with each meal. As the pancreas is progressively destroyed, some patients stop feeling the abdominal pain that was initially so severe. Others continue to have constant abdominal pain, and may even require a surgical procedure for relief. Drugs can be used to reduce the pain, but when **narcotics** are used for pain relief there is danger of the patient becoming addicted.

Prognosis

A number of systems have been developed to help determine the prognosis of an individual with pancreatitis. A very basic evaluation of a patient will allow some prediction to be made based on the presence of dying pancreatic tissue (necrosis) and bleeding. When necrosis and bleeding are present, as many as 50% of patients may die.

More elaborate systems have been created to help determine the prognosis of patients with pancreatitis. The most commonly used system identifies 11 different signs (Ranson's signs) that can be used to determine the severity of the disease. The first five categories are evaluated when the patient is admitted to the hospital:

- age over 55 years
- blood sugar level over 200 mg/dL
- serum lactic dehydrogenase over 350 IU/L (increased with increased breakdown of blood, as would occur with internal bleeding, and with heart or liver damage)
- AST over 250 mu (a measure of liver function, as well as a gauge of damage to the heart, muscle, brain, and kidney)
- white blood count over 16,000 u L

The next six of Ranson's signs are reviewed 48 hours after admission to the hospital. These are:

- greater than 10% decrease in hematocrit (a measure of red blood cell volume)
- increase in BUN greater than 5 mg/dL (blood urea nitrogen, an indicator of kidney function)
- blood calcium less than 8 mg/dL
- PaO₂ less than 60 mm Hg (a measure of oxygen in the blood)
- base deficit greater than 4 mEq/L (a measure of change in the normal acidity of the blood)
- fluid sequestration greater than 6 L (an estimation of the quantity of fluid that has leaked out of the blood circulation and into other body spaces)

Once a doctor determines how many of Ranson's signs are present and gives the patient a score, the doctor can better predict the risk of death. The more signs

KEY TERMS

Abscess—A pocket of infection; pus.

Acute—Of short and sharp course. Illnesses that are acute appear quickly and can be serious or life-threatening. The illness ends and the patient usually recovers fully.

Chronic—Of long duration and slow progression. Illnesses that are chronic develop slowly over time, and do not end. Symptoms may be continual or intermittent, but the patient usually has the condition for life.

Diabetes—A disease characterized by an inability to process sugars in the diet, due to a decrease in or total absence of insulin production. May require injections of insulin before meals to aid in the metabolism of sugars.

Duodenum—The first section of the small intestine that receives partly digested material from the stomach.

Endocrine—A system of organs that produces chemicals that go into the bloodstream to reach other organs whose functioning they affect.

Enzyme—A chemical that speeds up or makes a particular chemical reaction more efficient. In the digestive system, enzymes are involved in breaking down large food molecules into smaller molecules that can be processed and utilized by the body.

Exocrine—A system of organs that produces chemicals that go through a duct (or tube) to reach other organs whose functioning they affect.

Gland—Collections of tissue that produce chemicals needed for chemical reactions elsewhere in the body.

Hormone—A chemical produced in one part of the body that travels to another part of the body in order to exert an effect.

present, the greater the chance of fatal complications. A patient with less than three positive Ranson's signs has a 95% survival rate. A patient with three to four positive Ranson's signs has an 80–85% survival rate.

The results of a CT scan can also be used to predict the severity of pancreatitis. Slight swelling of the pancreas indicates mild illness. Significant swelling, especially with evidence of destruction of the pancreas and/or fluid build-up in the abdominal cavity, indicates more severe illness. With severe illness, there is a worse prognosis.

Prevention

Alcoholism is essentially the only preventable cause of pancreatitis. Patients with chronic pancreatitis must stop drinking alcohol entirely. The drugs that cause or may cause pancreatitis should also be avoided.

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ORGANIZATIONS

- National Digestive Diseases Information Clearinghouse, 2 Information Way, Bethesda, MD, 20892-3570, (800) 891-5389, <http://www.niddk.nih.gov/health/digest/nddc.htm>.
 National Institutes of Health (NIH), 9000 Rockville Pike, Bethesda, MD, 20892, 301-496-4000, <http://www.nih.gov/index.html>.
 U.S. National Library of Medicine, 8600 Rockville Pike, Bethesda, MD, 20894, <http://www.nlm.nih.gov/medlineplus/medlineplus.html>.

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Panic attack see **Panic disorder**

Panic disorder

Definition

A panic disorder is a psychological state characterized by acute (rapid onset) feelings, which engulf a person with a deep sense of destruction, **death** and imminent doom. The main feature of panic disorder (PD) is a history of previous panic attacks (PA). The PA symptoms are pronounced and the affected person will gasp for air, have increased breathing (hyperventilate), feel dizzy (light headed), and develop a loss of

sensation (paresthesia). Most patients will run outside and symptoms like increased breathing will slow and the PA symptoms will subside. Most PA last three to ten minutes. It is rare for PA to extend in duration over 30 minutes.

Description

The essential characteristics of panic disorder , consist of specific and common criteria. The affected person usually has recurrent and unexpected panic attacks (the active presentation of panic disorder). The PA is characterized by a discrete, rapid onset feeling of intense fear or discomfort. Affected persons have several somatic (referring to physical signs) or cognitive (thinking) symptoms. Affected persons usually react in a manner that indicates impending doom. They commonly exhibit signs of a sweating, racing heart beat, chest **pain, shortness of breath**, and the perception of feeling smothered. The panic attack (PA) is usually followed by one month (or more) of one or more of the following thought processes:

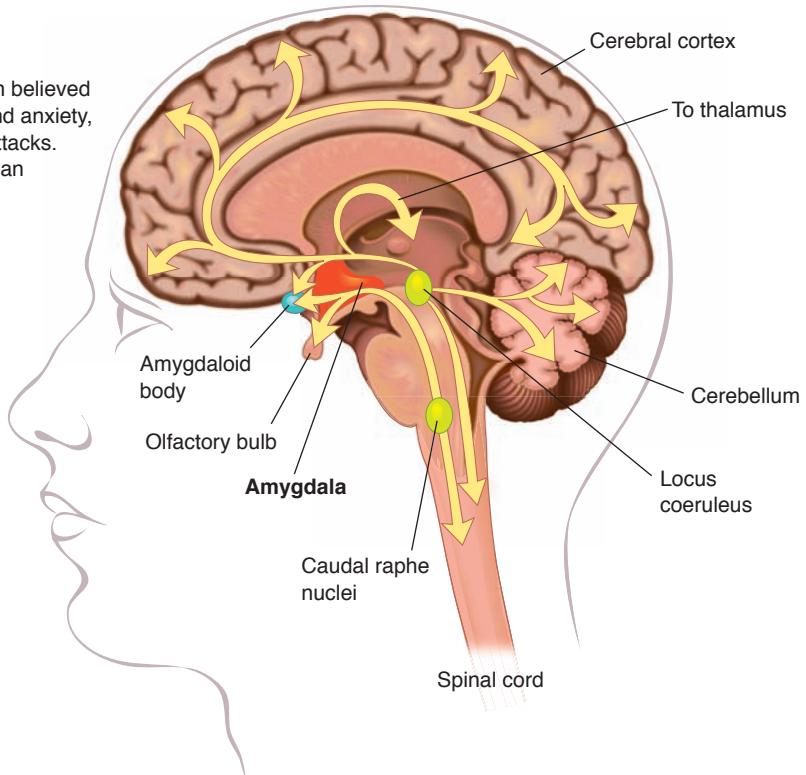
- Persistent concern or preoccupation about having future attacks
- Worry about the possible consequences, complications, or behavioral changes associated with attacks (e.g. losing control, going crazy, or having a serious medical condition like a heart attack).

Genetic profile

Panic disorder definitely runs in families and twin studies suggest that about 20% of patients who have the criteria for diagnosis have first-degree relatives with the disorder. In families with no history of affected first-degree relatives the prevalence decreases to 4%. The ratio between monozygotic twins (identical) to dizygotic (non-identical) twins is 5:1 for PD. Recent evidence suggests that there is a genetic mutation in the SLC6A4 gene. This gene is related to a brain chemical called serotonin, a chemical in the brain, which is known to effect mood. If the transport of serotonin is imbalanced then certain parts of the brain may not receive the correct stimulus causing alterations in mood. Some studies have demonstrated that there is no positive family history in about 50% of patients diagnosed with PD. Other possible causes of PD include social learning and autonomic responsiveness (the attack will affect the body and hypersensitizes nerve cells in the brain). Another gene possibly associated with panic disorder is the COMT gene that provides instructions for making an enzyme called catechol-O-methyltransferase. Mutations in this gene have been associated with other disorders that affect

Panic disorder

The amygdala is the area of the brain believed to regulate emotions, such as fear and anxiety, both of which are involved in panic attacks. Panic disorder may also be linked to an imbalance of serotonin, a chemical that helps control mood.



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thought and emotion, with studies suggesting that these conditions may be due to inefficient processing of information in the prefrontal cortex.

Demographics

PD usually begins during the affected persons late teens or in the twenties, and is uncommon after age 35 and unusual after age 45 years. Global studies suggest that the lifetime prevalence of PD is between 1.5% and 3.5%. In the United States, the National Institute of Mental Health (NIMH) estimates that panic disorder affects about 6 million American adults and is twice as common in women as men.

Agoraphobia (anxiety) state about being in situations or places that might make escape embarrassing or difficult is seen in approximately one-third to one-half of persons who meet the criteria for PD diagnosis. Other reports indicate that about 95% of persons affected with agoraphobia also have a previous history

or current diagnosis of PD. In some cultures PA is believed to be associated with magic or witchcraft. Additional causes of PA may include intentional suppression of one's freedoms or public life.

Signs and symptoms

Criteria for panic attack:

1. Cardiac palpitations (pounding, racing or accelerated heart rate).
2. Sweating.
3. Shaking (trembling).
4. Breathing difficulties, including shortness of breath or perceptions of being smothered.
5. Feeling of choking.
6. Chest discomfort or pain.
7. Feeling light-headed (faint, dizzy or unsteady).
8. Stomach discomfort or nausea.

9. Affected individuals may lose contact with reality during the attack.
10. A feeling of being detached and out of contact with oneself.
11. Fear of losing control of oneself (going “crazy”).
12. Fear of dying.
13. Tingling or numbness sensations.

Criteria for panic disorder:

1. Recurrent and unexpected PA.
2. Worry about the consequences, implications, or behavioral changes associated with PA (perceptions of going “crazy,” losing control of actions, or suffering from a life threatening condition, such as a heart attack).
3. PA is not caused by or associated with a medical condition.
4. PA is not associated with another mental disorder, such as phobia (an exaggerated fear to something like spiders or heights). Exposure to a specific phobia situation or object can promote a PA.

Criteria for agoraphobia:

1. The essential feature of agoraphobia is anxiety about being in situations or places that make escape embarrassing or difficult. These fears usually involve characteristic clusters of situations that include being on a bridge, being in a crowd, standing in line in a department store, or traveling in a train, bus, or automobile. Elevators are another common cause promoting the occurrence of PA. These situations, which lead to the PA, are often difficult or embarrassing to abruptly flee from.
2. Avoidance of the affected person’s fear, which usually limits travel away from home, causing impaired functioning.

Criteria for PD without agoraphobia:

Recurrent unexpected PA; at least one attack followed by one month or more of one or more of the following symptoms:

- Persistent concern about having future attacks
- Worry about consequences associated with attacks
- A change in behavioral patterns related to the attacks (e.g., the affected person avoids travel).
- Absence of agoraphobia
- PA are not due to a medical condition

KEY TERMS

Palpitation—An irregular heartbeat.

Phobia—An exaggerated fear.

- PA not associated with another mental disorder (e.g., phobias).

Criteria for panic disorder with agoraphobia:

1. Criteria 1, 2, and 5 for PD without agoraphobia must be present.
2. The presence of agoraphobia.

Diagnosis

There are no specific laboratory findings associated with diagnosing PD. However, evidence suggests that some affected persons may have low levels of carbon dioxide and an important ion in the human body called bicarbonate (helps in regulating blood from becoming too acidic or alkaline). These chemical changes may hypersensitize (making cells excessively sensitive) nerve cells, which can increase the activity of other structures throughout the body, such as sweat glands (sweating) and the heart (racing, accelerated or pounding rate). Additionally, lactic acid (a chemical made in the body from sugar) plays a role in nerve cell hypersensitivity. The diagnosis of PD can be made accurately if the specific symptoms and criteria are established.

Neuroimaging studies indicate that the arteries (vessels that deliver oxygen rich blood to cells and tissues) are constricted (smaller diameter) as a result of increased breathing rates during a PA.

The consulting clinician must exclude other possible causes of panic attacks such as intoxication with stimulant drugs (**cocaine**, **caffeine**, amphetamines [speed]). Withdrawal from alcohol and **barbiturates** can also induce panic-like behaviors. Additionally, the consulting therapist should obtain a comprehensive medical history and examination to determine if the PA is caused by a medical condition frequently observed in hormonal diseases (overactive thyroid), tumors that secrete chemicals causing a person to have pronounced “hyper” changes (racing heartbeat, sweating, shaking). Other causes include a possible cardiac (heart) disease such as an irregularly beating heart.

Treatment and management

Moderate to severe PD is characterized by frequent PA ranging from five to seven times a week or with significant disability associated with anxiety between episodes. In addition to **cognitive-behavioral therapy** an affected person will usually require medications. There are three classes of medications commonly prescribed for PD patients.

Tricyclic antidepressants

Tricyclic antidepressants are a class of medications used to treat depression and other closely related mental disorders. Individuals affected with PD are usually given imipramine, which has been shown in some studies to be effective in approximately 70% of cases. Medications in this category usually have a prolonged lag time until a positive response is observed. This is primarily due to adverse side effects, which prevent rapid increases of dosage and also because they act on specific chemical imbalances in the brain, which take time to stabilize.

The first choice of medication treatment for PD is tricyclics (imipramine, desipramine and nortriptyline). These medications require careful dosing and monitoring. The actual blood level (therapeutic level necessary to make improvements) may vary in special populations who have the disorder. Elderly patients may require a smaller dose, due to decrease in metabolism (in this context metabolism refers to the breakdown of large chemicals to smaller ones for usage) and kidney function, which are part of **aging**. Some patients may develop gastrointestinal (stomach) side effects, which may interfere with absorption from the gut, thereby decreasing beneficial blood levels. Furthermore, patients who receive tricyclics may develop **dry mouth** and low blood pressure. The heart may be adversely affected (altered rate and rhythm) especially in patients with preexisting diseases, causing direct damage or strain in the heart. Affected persons receiving tricyclics also commonly experience changes in sexual functioning, including loss of desire and ejaculation. Adverse (negative) side effects usually decrease patient compliance (the person stops taking medications to avoid side effects). Recently, a new group of tricyclics was made available. These tricyclics (fluoxetine, sertraline, paroxetine and fluvoxamine) act on specific areas in the brain to correct potential chemical imbalances.

Monoamine oxidase inhibitors (MAOIs)

A second line category of medications used to treat PD are the monoamine oxidase (a chemical that assists in storing certain chemicals in nerve cells)

inhibitors (MAOI). MAOI will stop the action of MAO, thereby decreasing the amount of certain chemicals in the brain that may influence PAs. This group of medications is effective in approximately 75–80% of cases, especially for refractory (not active) depression. Affected individuals using MAOI must avoid specific foods to prevent a hypertensive crisis (when the blood pressure rapidly increases). These foods include cheeses (except cream cheese, cottage cheese, and fresh yogurt); liver of all types; meat and yeast extracts; fermented or aged meats (such as salami and bologna); broad and Chinese bean pods; all types of alcohol-containing products; soy sauce; shrimp and shrimp paste; and sauerkraut. Although MAOI are effective medications for treatment of PD, they are underutilized due to strict dietary limitations.

Benzodiazepines

Benzodiazepines are another class of medications used to treat PD. They include medications such as diazepam (Valium), lorazepam, and clonazepam. They have been reported to be effective in 70–90% of patients with PD. However, the effective dose is approximately two to three times higher for PD than milder forms of simple anxiety (these medications are usually indicated for mild anxiety). This increased dosing in patients with PD is undesirable since there is risk of physical dependence and withdrawal (commonly exhibited when the medication is rapidly tapered down or stopped). However, they are indicated when PD affected patients respond poorly to tricyclics or have a fear of taking MAOIs due to dietary restrictions and problems associated with eating the wrong foods accidentally.

Long term management

Reassuring the PD patient that anticipated panic attacks are unlikely while taking medication is essential for long-term maintenance. Cognitive-behavioral therapy is also important for long-term treatment. Weaning off medications must be done slowly since patients develop a sense of security that they will not have an attack while actively dosing.

Clinical trials

Clinical trials on panic disorder are currently sponsored by the National Institutes of Health (NIH) and other agencies. As of 2009, NIH was reporting 98 on-going and completed studies.

Examples include:

- The evaluation of the effectiveness of psychodynamic psychotherapy in treating adults with panic disorder. (NCT00128388)

- A study to identify genes that increase the risk of developing panic disorder. (NCT00083265)
- A study to examine brain and noradrenaline function in panic disorder. (NCT00103987)
- The evaluation of the relative effectiveness of three psychotherapies in treating people with a panic disorder. (NCT00353470)

Clinical trial information is constantly updated by NIH and the most recent information on panic disorder trials can be found at: <http://clinicaltrials.gov/ct2/results?term=panic+disorder>.

Prognosis

The course of PD and agoraphobia varies considerably over time. Some cases may experience spontaneous remissions (the disorder is present but it is not active). The course can be so variable that an affected person may go on for years without a PA, then have several attacks, and then enter a second phase of remission, which may last for years. In some cases a decrease in PA may be closely related to a decrease and avoidance of anxiety-associated situations, which promote agoraphobia. Agoraphobia itself may become chronic (long term or permanent) with or without PA. In general, approximately 50–60% will recover substantially five to 20 years after the initial attack. Approximately 20% will still have long term impairment, which will stay the same or slightly worsen. Generally, the earlier treatment is sought, the better the outcome. The course in children and adolescents is chronic (long term), usually lasting about three years. Generally, PD shows the highest risk of developing new psychological disorders during follow up visits. If PA is treated early, anticipatory anxiety and phobia may be more manageable and responsive to treatment.

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ORGANIZATIONS

- American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209, (888) 357-7924, <http://www.psych.org>.
- American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (800) 374-2721, (202) 336-5568, <http://www.apa.org>.
- Mental Health America, 2000 N. Beauregard Street, 6th Floor, Alexandria, VA, 22311, (800) 969-6642, (703) 684-5968, <http://www.nmha.org>.

National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Bethesda, MD, 20892-9663, (866) 615-6464, <http://www.nimh.nih.gov>.

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Pap test

Definition

The Pap test is a procedure in which a physician scrapes cells from the cervix or vagina to check for **cervical cancer**, **vaginal cancer**, or abnormal changes that could lead to cancer. It often is called a Pap smear.

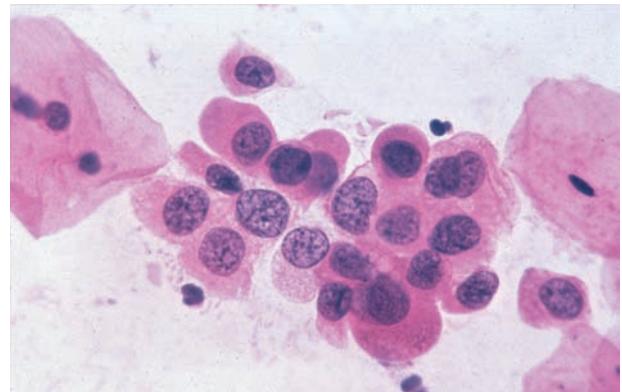
Purpose

The Pap test is used to detect abnormal growth of cervical cells at an early stage so that treatment can be started when the condition is easiest to treat. This microscopic analysis of cells can detect cervical cancer, precancerous changes, inflammation (vaginitis), infections, and some **sexually transmitted diseases** (STDs). The Pap test can occasionally detect endometrial (uterine) cancer or **ovarian cancer**, although it was not designed for this purpose.

Women should begin to have Pap tests at the age of 21 or within three years of becoming sexually active, whichever comes first. Young people are more likely to have multiple sex partners, which increases their risk of certain diseases that can cause cancer, such as human papillomavirus (HPV).

The American Cancer Society (ACS) updated its guidelines concerning Pap test frequency in late 2002. In brief, women should continue screening every year with regular Pap tests until age 30, every two years if using the liquid-based Pap test. Once a woman age 30 and older has had three normal results in a row, she may get screened every two to three years. A doctor may suggest more frequent screening if a woman has certain risk factors for cervical cancer. Women who have had total hysterectomies including the removal of the cervix do not need Pap tests unless the **hysterectomy** resulted from cervical cancer. Those over age 70 who have had three normal results generally do not need to continue having Pap tests under the new guidelines.

Women with certain risk factors may have yearly tests. Those at highest risk for cervical cancer are women who started having sex before age 18, those with many sex partners (especially if they did not use



These malignant cells were taken from a woman's cervix during a Pap test. (Parviz M. Pour/Photo Researchers, Inc.)

condoms, which protect against STDs), those who have had STDs such as **genital herpes** or **genital warts**, and those who smoke. Women older than 40 may have the test yearly, if experiencing bleeding after **menopause**. Women who have had a positive test result in the past may need screening every six months. Women who have had cervical cancer or precancer should have regular Pap smears.

Other women also benefit from the Pap test. Women over age 65 account for 25% of all cases of cervical cancer and 41% of deaths from this disease. Women over age 65 who have never had a Pap smear benefit the most from the test. Some women have the cervix left in place after hysterectomy and will continue to receive regular Pap tests. Finally, a pregnant woman should have a Pap test as part of her first prenatal examination.

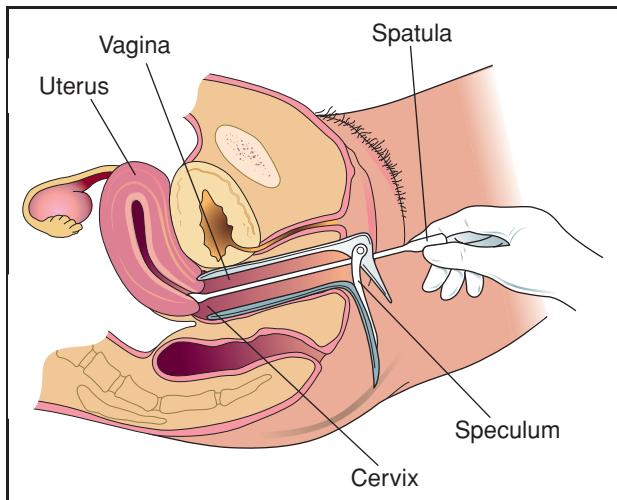
The Pap smear is a screening test. It identifies women who are at increased risk of cervical dysplasia (abnormal cells) or cervical cancer. Only an examination of the cervix with a special lighted instrument (**colposcopy**) and samples of cervical tissue (biopsies) can actually diagnose these problems.

Precautions

The Pap test is usually not done during the menstrual period because of the presence of blood cells. The best time is in the middle of the menstrual cycle.

Description

The Pap test is an extremely cost-effective and beneficial exam. Cervical cancer used to be a leading cause of cancer deaths in American women, but widespread use of this diagnostic procedure reduced the **death** rate from this disease by 74% between 1955 and



The Pap test is a procedure used to detect abnormal growth of cervical cells which may be a precursor to cancer of the cervix. It is administered by a physician who inserts a speculum into the vagina to open and separate the vaginal walls. A spatula is then inserted to scrape cells from the cervix. These cells are transferred onto glass slides for laboratory analysis. The Pap test may also identify vaginitis, some sexually transmitted diseases, and cancers of the uterus and ovaries. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

1992. A 2003 study reported that the test reduces rates of invasive cervical cancer by as much as 94%. In 2003, the FDA approved a new screening test that combines DNA testing for the HPV type that causes the most cases of cervical cancer with the standard Pap test, increasing its screening value.

The Pap test, sometimes called a cervical smear, is the microscopic examination of cells scraped from both the outer cervix and the cervical canal. (The cervix is the opening between the vagina and the uterus, or womb.) It is called the "Pap" test after its developer, Dr. George N. Papanicolaou. This simple procedure is performed during a gynecologic examination and is usually covered by insurance. For those with coverage, Medicare will pay for one screening Pap smear every three years.

During the pelvic examination, an instrument called a speculum is inserted into the vagina to open it. The doctor then uses a tiny brush, or a cotton-tipped swab and a small spatula to wipe loose cells off the cervix and to scrape them from the inside of the cervix. The cells are transferred or "smeared" onto glass slides, the slides are treated to stabilize the cells, and the slides are sent to a laboratory for microscopic examination. The entire procedure is usually painless and takes five to 10 minutes at most.

The newer method called liquid-based cytology, or the liquid-based Pap test, involves spreading the cells more evenly on a slide after removing them from the sample. The liquid-based method prevents cells from drying out and becoming distorted. Studies show that liquid-based testing slightly improves cancer detection and greatly improves detection of pre-cancers, but it costs more than the traditional Pap test. Trade names in 2003 for liquid-based Pap smears were ThinPrep and AutoCytex.

Preparation

The Pap test may show abnormal results when a woman is healthy or normal results in women with cervical abnormalities as much as 25% of the time. It may even miss up to 5% of cervical cancers. Some simple preparations may help to ensure that the results are reliable. Among the measures that may help increase test reliability are:

- avoiding sexual intercourse for two days before the test
- not using douches for two or three days before the test
- avoiding tampons, vaginal creams, or birth control foams or jellies for two to three days before the test
- scheduling the Pap smear when not menstruating.

However, most women are not routinely advised to make any special preparations for a Pap test.

If possible, women may want to ensure that their test is performed by an experienced gynecologist, physician, or provider and sent to a reputable laboratory. The physician should be confident in the accuracy of the chosen lab.

Before the exam, the physician will take a complete sexual history to determine a woman's risk status for cervical cancer. Questions may include date and results of the last Pap test, any history of abnormal Pap tests, date of last menstrual period and any irregularity, use of hormones and birth control, family history of gynecologic disorders, and any vaginal symptoms. These topics are relevant to the interpretation of the Pap test, especially if any abnormalities are detected. Immediately before the Pap test, the woman should empty her bladder to avoid discomfort during the procedure.

Aftercare

Harmless cervical bleeding is possible immediately after the test; a woman may need to use a sanitary napkin. She should also be sure to comply with her doctor's orders for follow-up visits.

Risks

No appreciable health risks are associated with the Pap test. However, abnormal results (whether valid or due to technical error) can cause significant **anxiety**. Women may wish to have their sample double-checked, either by the same laboratory or by the new technique of computer-assisted rescreening. The Food and Drug Administration (FDA) has approved the use of AutoPap and PAPNET to doublecheck samples that have been examined by technologists. AutoPap may also be used to perform initial screening of slides, which are then checked by a technologist. Any abnormal Pap test should be followed by colposcopy, not by double checking the Pap test.

Normal results

Normal (negative) results from the laboratory exam mean that no atypical, dysplastic, or cancer cells were detected, and the cervix is normal.

Abnormal results

Terminology

Abnormal cells found on the Pap test may be described using two different grading systems. Although this can be confusing, the systems are quite similar. The Bethesda system is based on the term “squamous intraepithelial lesion” (SIL). Precancerous cells are classified as atypical squamous cells of undetermined significance, low-grade SIL, or high-grade SIL. Low-grade SIL includes mild dysplasia (abnormal cell growth) and abnormalities caused by HPV; high-grade SIL includes moderate or severe dysplasia and carcinoma in situ (cancer that has not spread beyond the cervix).

Another term that may be used is “cervical intraepithelial neoplasia” (CIN). In this classification system, mild dysplasia is called CIN I, moderate is CIN II, and severe dysplasia or carcinoma in situ is CIN III.

Regardless of terminology, it is important to remember that an abnormal (positive) result does not necessarily indicate cancer. Results may be falsely abnormal after infection or irritation of the cervix. Up to 40% of mild dysplasia reverts to normal tissue without treatment, and only 1% of mild abnormalities ever develop into cancer.

Changes of unknown cause

ASCUS or LSIL cells are found in 5%–10% of all Pap tests. The most common abnormality is atypical

squamous cells of undetermined significance, which are found in 4% of all Pap tests. Sometimes these results are described further as either reactive or pre-cancerous. Reactive changes suggest that the cervical cells are responding to inflammation, such as from a yeast infection. These women may be treated for infection and then undergo repeat Pap testing in three to six months. If those results are negative, no further treatment is necessary. This category may also include atypical “glandular” cells, which could imply a more severe type of cancer and requires repeat testing and further evaluation.

Dysplasia

The next most common finding (in about 25 of every 1,000 tests) is low-grade SIL, which includes mild dysplasia or CIN I and changes caused by HPV. Unlike cancer cells, these cells do not invade normal tissues. Women are most susceptible to cervical dysplasia between the ages of 25 and 35. Typically, dysplasia causes no symptoms, although women may experience abnormal vaginal bleeding. Because dysplasia is precancerous, it should be treated if it is moderate or severe.

Treatment of dysplasia depends on the degree of abnormality. In women with no other risk factors for cervical cancer, mild precancerous changes may be simply observed over time with repeat testing, perhaps every four to six months. This strategy works only if women are diligent about keeping later appointments. Premalignant cells may remain that way without causing cancer for five to ten years, and may never become malignant.

In women with positive results or risk factors, the gynecologist must perform colposcopy and biopsy. A colposcope is an instrument that looks like binoculars, with a light and a magnifier, used to view the cervix. Biopsy, or removal of a small piece of abnormal cervical or vaginal tissue for analysis, is usually done at the same time.

High-grade SIL (found in three of every 50 Pap tests) includes moderate to severe dysplasia or carcinoma in situ (CIN II or III). After confirmation by colposcopy and biopsy, it must be removed or destroyed to prevent further growth. Several outpatient techniques are available: conization (removal of a cone-shaped piece of tissue), **laser surgery**, **cryotherapy** (freezing), or the “loop electrosurgical excision procedure.” Cure rates are nearly 100% after prompt and appropriate treatment of carcinoma in situ. Of course, frequent checkups are then necessary.

KEY TERMS

Carcinoma in situ—Malignant cells that are present only in the outer layer of the cervix.

Cervical intraepithelial neoplasia (CIN)—A term used to categorize degrees of dysplasia arising in the epithelium, or outer layer, of the cervix.

Dysplasia—Abnormal changes in cells.

Human papillomavirus (HPV)—The most common STD in the United States. Various types of HPV are known to cause cancer.

Neoplasia—Abnormal growth of cells, which may lead to a neoplasm, or tumor.

Squamous intraepithelial lesion (SIL)—A term used to categorize the severity of abnormal changes arising in the squamous, or outermost, layer of the cervix.

Cancer

HPV, the most common STD in the United States, may be responsible for many cervical cancers. Cancer may be manifested by unusual vaginal bleeding or discharge, bowel and bladder problems, and pain. Women are at greatest risk of developing cervical cancer between the ages of 30 and 40 and between the ages of 50 and 60. Most new cancers are diagnosed in women between 50 and 55. Although the likelihood of developing this disease begins to level off for Caucasian women at the age of 45, it increases steadily for African Americans for another 40 years. Biopsy is indicated when any abnormal growth is found on the cervix, even if the Pap test is negative.

Doctors have traditionally used **radiation therapy** and surgery to treat cervical cancer that has spread within the cervix or throughout the pelvis. In severe cases, postoperative radiation is administered to kill any remaining cancer cells, and **chemotherapy** may be used if cancer has spread to other organs. Recent studies have shown that giving chemotherapy and radiation at the same time improves a patient's chance of survival. The National Cancer Institute has urged physicians to strongly consider using both chemotherapy and radiation to treat patients with invasive cervical cancer. The survival rate at five years after treatment of early invasive cancer is 91%; rates are below 70% for more severe invasive cancer. That is why prevention, risk reduction, and frequent Pap tests are the best defense for a woman's gynecologic health.

The Pap test is a procedure used to detect abnormal growth of cervical cells which may be a precursor to cancer of the cervix. It is administered by a physician who inserts a speculum into the vagina to open and separate the vaginal walls. A spatula is then inserted to scrape cells from the cervix. These cells are transferred onto glass slides for laboratory analysis. The Pap test may also identify vaginitis, some sexually transmitted diseases, and cancers of the uterus and ovaries.

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American Cancer Society, 1599 Clifton Road NE, Atlanta, GA, 30329-4251, (800) 227-2345, <http://www.cancer.org>.

American College of Obstetricians and Gynecologists, 409 Twelfth Street SW, P.O. Box 96920, Washington, DC, 20090-6920, (202) 638-5577, <http://www.acog.org>.

National Cancer Institute, Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD, 20892-2580, (800) 422-6237, <http://www.nci.nih.gov>.

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Papanicolaou test see **Pap test**

Papilledema

Definition

Papilledema is a swelling of the optic nerve, at the point where this nerve joins the eye, that is caused by an increase in fluid pressure within the skull (intracranial pressure). Swelling of the optic nerve due to other causes such as infection or inflammatory disease is not called papilledema.

Description

The optic nerve is the nerve that transmits signals from the eye to the brain. Papilledema is a swelling

of this nerve where it meets the eye (the optic disc) caused by an increase in intracranial pressure. Almost all cases of papilledema are bilateral (affect both eyes). Papilledema can be observed in people of any age, but is relatively uncommon in infants because the bones of the skull are not fully fused together at this age.

Causes and symptoms

Papilledema is caused by an increase in the pressure of the fluid (cerebrospinal fluid) that is present between the brain and the skull, inside the head. This increase in intracranial pressure may be caused by any of a variety of conditions within the skull, brain, or spinal cord. The most common causes of papilledema are:

- tumor of the brain, spinal cord, skull, spinal column, or optic nerve
- abscess (the accumulation of pus within a confined space)
- craniosynostosis (an abnormal closure of the bones of the skull)
- hemorrhage (bleeding)
- hydrocephalus (an accumulation of cerebrospinal fluid within the skull)
- intracranial infection (any infection within the skull such as meningitis and encephalitis)
- head injury

The symptoms of papilledema include:

- headaches, which are usually worse upon awakening and exacerbated by coughing, holding the breath, or other maneuvers that tend to increase intracranial pressure.
- nausea and vomiting.
- changes in vision, such as temporary and transient blurring, graying, flickering, or double vision

Diagnosis

A diagnosis of papilledema is achieved by visual examination of the eye with an ophthalmoscope. This instrument shines light through the pupil of the eye and illuminates the retina while the clinician looks through it. Eye drops to dilate the pupils are used to insure a thorough examination.

Treatment

Treatment of papilledema is generally aimed at the treatment of the underlying disorder that is causing papilledema.

KEY TERMS

Craniosynostosis—A premature closure of one or more of the joints (fissures) between the bones of the skull, which causes an abnormally shaped skull.

Hydrocephalus—The accumulation of cerebrospinal fluid within the skull.

Ophthalmoscope—A medical instrument which shines a light through the pupil of the patient's eye and illuminates the retina (back) of the eye, allowing a visual examination of the interior of the eye.

Diuretic drugs combined with a weight reduction program may be useful in cases of papilledema that are caused by an abnormally high production of cerebrospinal fluid.

Corticosteroids have been shown to be effective in relieving the symptoms in some patients with papilledema caused by inflammatory disorders.

Alternative treatment

Alternative treatments for conditions that cause the occurrence of papilledema include **acupuncture**, **aromatherapy**, **hydrotherapy**, massage, and herbal remedies.

Prognosis

With prompt medical care to treat the underlying cause of papilledema, a person affected with papilledema will not have permanent damage to his or her eye-sight. However, prolonged papilledema can result in permanent damage to the optic nerve which could lead to blindness.

Prevention

Preventing papilledema is only possible if the underlying condition causing the papilledema can be found. Treatment of this underlying condition may prevent recurrences of papilledema.

Resources

BOOKS

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ORGANIZATIONS

National Eye Institute, 31 Center Drive MSC 2510, Bethesda, MD, (301) 496-5248, <http://www.nei.nih.gov>.

Paul A. Johnson, Ed.M.

Papillomavirus infection see **Genital warts**

Papule see **Skin lesions**

Paracentesis

Definition

Paracentesis is a procedure during which fluid from the abdomen is removed through a needle.

Purpose

There are two reasons to take fluid out of the abdomen. One is to analyze it. The other is to relieve pressure.

Liquid that accumulates in the abdomen is called **ascites**. Ascites seeps out of organs for several reasons related either to disease in the organ or fluid pressures that are changing.

Liver disease

All the blood flowing through the intestines passes through the liver on its way back to the heart. When progressive disease such as alcohol damage or hepatitis destroys enough liver tissue, the scarring that results shrinks the liver and constricts the blood flow. Such scarring of the liver is called **cirrhosis**. Pressure builds up in the intestinal circulation, slowing flow and pushing fluid into the tissues. Slowly the fluid accumulates in areas with the lowest pressure and greatest capacity. The free space around abdominal organs receives most of it. This space is called the peritoneal space because it is enclosed by a thin membrane called the peritoneum. The peritoneum wraps around nearly every organ in the abdomen, providing many folds and spaces for the fluid to gather.

Infections

Peritonitis is an infection of the peritoneum. Infection changes the dynamics of body fluids, causing them to seep into tissues and spaces. Peritonitis can

develop in several ways. Many abdominal organs contain germs that do not belong elsewhere in the body. If they spill their contents into the peritoneum, infection is the result. The gall bladder, the stomach, any part of the intestine, and especially the appendix all cause peritonitis when they leak or rupture. **Tuberculosis** can infect many organs in the body; it is not confined to the lungs. Tuberculous peritonitis causes ascites.

Other inflammations

Peritoneal fluid is not just produced by infections. The pancreas can cause a massive sterile peritonitis when it leaks its digestive enzymes into the abdomen.

Cancer

Any **cancer** that begins in or spreads to the abdomen can leak fluid. One particular tumor of the ovary that leaks fluid, the resulting presentation of the disease, is Meigs' syndrome.

Kidney disease

Since the kidneys are intimately involved with the body's fluid balance, diseases of the kidney often cause excessive fluid to accumulate. Nephrosis and **nephrotic syndrome** are the general terms for diseases that cause the kidneys to retain water and provoke its movement into body tissues and spaces.

Heart failure

The ultimate source of fluid pressure in the body is the heart, which generates blood pressure. All other pressures in the body are related to blood pressure. As the heart starts to fail, blood backs up, waiting to be pumped. This increases back pressure upstream, particularly below the heart where gravity is also pulling blood away from the heart. The extra fluid from **heart failure** is first noticed in the feet and ankles, where gravitational effects are most potent. In the abdomen, the liver swells first, then it and other abdominal organs start to leak.

Pleural fluid

The other major body cavity is the chest. The tissue in the chest corresponding to the peritoneum is called the pleura, and the space contained within the pleura, between the ribs and the lungs, is called the pleural space. Fluid is often found in both cavities, and fluid from one cavity can find its way into the other.

Fluid that accumulates in the abdomen creates abnormal pressures on organs in the abdomen. Digestion is hindered; blood flow is slowed. Pressure upward on the chest compromises breathing. The

KEY TERMS

Ectopic pregnancy—A pregnancy occurring outside the womb that often ruptures and requires surgical removal.

kidneys function poorly in the presence of such external pressures and may even fail with tense, massive ascites.

Description

During paracentesis, special needles puncture the abdominal wall, being careful not to hit internal organs. If fluid is needed only for analysis, just a bit is removed. If pressure relief is an additional goal, many quarts may be removed. Rapid removal of large amounts of fluid can cause blood pressure to drop suddenly. For this reason, the physician will often leave a tube in place so that fluid can be removed slowly, giving the circulation time to adapt.

A related procedure called culpocelesis removes ascitic fluid from the very bottom of the abdominal cavity through the back of the vagina. This is used mostly to diagnose female genital disorders like **ectopic pregnancy** that bleed or exude fluid into the peritoneal space.

Fluid is sent to the laboratory for testing, where cancer and blood cells can be detected, infections identified, and chemical analysis can direct further investigations.

Aftercare

An adhesive bandage and perhaps a single stitch close the hole. Nothing more is required.

Risks

Risks are negligible. It is remotely possible that an organ could be punctured and bleed or that an infection could be introduced.

Normal results

A diagnosis of the cause and/or relief from accumulated fluid pressure are the expected results.

Abnormal results

Fluid will continue to accumulate until the cause is corrected. Repeat procedures may be needed.

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J. Ricker Polsdorfer, MD

Paracoccidioidomycosis see **South American blastomycosis**

Paragonamiasis see **Fluke infections**

Paralysis

Definition

Paralysis is defined as complete loss of strength in an affected limb or muscle group.

Description

The chain of nerve cells that runs from the brain through the spinal cord out to the muscle is called the motor pathway. Normal muscle function requires intact connections all along this motor pathway. Damage at any point reduces the brain's ability to control the muscle's movements. This reduced efficiency causes weakness, also called paresis. Complete loss of communication prevents any willed movement at all. This lack of control is called paralysis. Certain inherited abnormalities in muscle cause **periodic paralysis**, in which the weakness comes and goes.

The line between weakness and paralysis is not absolute. A condition causing weakness may progress to paralysis. On the other hand, strength may be restored to a paralyzed limb. Nerve regeneration or regrowth is one way in which strength can return to a paralyzed muscle. Paralysis almost always causes a change in muscle tone. Paralyzed muscle may be flaccid, flabby, and without appreciable tone, or it may be spastic, tight, and with abnormally high tone that increases when the muscle is moved.

Paralysis may affect an individual muscle, but it usually affects an entire body region. The distribution of weakness is an important clue to the location of the nerve damage that is causing the paralysis. Words describing the distribution of paralysis use the suffix “-plegia,” from the Greek word for “stroke.” The types of paralysis are classified by region:

- monoplegia, affecting only one limb
- diplegia, affecting the same body region on both sides of the body (both arms, for example, or both sides of the face)
- hemiplegia, affecting one side of the body
- paraplegia, affecting both legs and the trunk
- quadriplegia, affecting all four limbs and the trunk

Causes and symptoms

Causes

The nerve damage that causes paralysis may be in the brain or spinal cord (the central nervous system) or it may be in the nerves outside the spinal cord (the peripheral nervous system). The most common causes of damage to the brain are:

- stroke
- tumor
- trauma (caused by a fall or a blow)
- Multiple sclerosis (a disease that destroys the protective sheath covering nerve cells)
- cerebral palsy (a condition caused by a defect or injury to the brain that occurs at or shortly after birth)
- metabolic disorder (a disorder that interferes with the body's ability to maintain itself)

Damage to the spinal cord is most often caused by trauma, such as a fall or a car crash. Other conditions that may damage nerves within or immediately adjacent to the spine include:

- tumor
- herniated disk (also called a ruptured or slipped disk)
- spondylosis (a disease that causes stiffness in the joints of the spine)
- rheumatoid arthritis of the spine
- neurodegenerative disease (a disease that damages nerve cells)
- multiple sclerosis

Damage to peripheral nerves may be caused by:

- trauma
- compression or entrapment (such as carpal tunnel syndrome)
- Guillain-Barré syndrome (a disease of the nerves that sometimes follows fever caused by a viral infection or immunization)
- chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) (a condition that causes pain and swelling in the protective sheath covering nerve cells)

- radiation
- inherited demyelinating disease (a condition that destroys the protective sheath around the nerve cell)
- toxins or poisons

Symptoms

The distribution of paralysis offers important clues to the site of nerve damage. Hemiplegia is almost always caused by brain damage on the side opposite the paralysis, often from a **stroke**. Paraplegia occurs after injury to the lower spinal cord, and quadriplegia occurs after damage to the upper spinal cord at the level of the shoulders or higher (the nerves controlling the arms leave the spine at that level). Diplegia usually indicates brain damage, most often from **cerebral palsy**. Monoplegia may be caused by isolated damage to either the central or the peripheral nervous system. Weakness or paralysis that occurs only in the arms and legs may indicate demyelinating disease. Fluctuating symptoms in different parts of the body may be caused by **multiple sclerosis**.

Sudden paralysis is most often caused by injury or stroke. Spreading paralysis may indicate degenerative disease, inflammatory disease such as **Guillain-Barré syndrome** or CIDP, metabolic disorders, or inherited demyelinating disease.

Other symptoms often accompany paralysis from any cause. These symptoms may include **numbness and tingling**, **pain**, changes in vision, difficulties with speech, or problems with balance. **Spinal cord injury** often causes loss of function in the bladder, bowel, and sexual organs. High spinal cord injuries may cause difficulties in breathing.

Diagnosis

Careful attention should be paid to any events in the patient's history that might reveal the cause of the paralysis. The examiner should look for incidents such as falls or other traumas, exposure to toxins, recent infections or surgery, unexplained **headache**, preexisting metabolic disease, and family history of weakness or other neurologic conditions. A neurologic examination tests strength, reflexes, and sensation in the affected area and normal areas.

Imaging studies, including **computed tomography scans** (CT scans), **magnetic resonance imaging** (MRI) scans, or **myelography** may reveal the site of the injury. **Electromyography** and nerve conduction velocity tests are performed to test the function of the muscles and peripheral nerves.

KEY TERMS

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures.

Electromyography—A test that uses electrodes to record the electrical activity of muscle. The information gathered is used to diagnose neuromuscular disorders.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct images of internal structures.

Myelin—The insulation covering nerve cells. Demyelinating disease causes a breakdown of myelin.

Myelography—An x-ray process that uses a dye or contrast medium injected into the space around the spine.

Nerve conduction velocity test—A test that measures the time it takes a nerve impulse to travel a specific distance over the nerve after electronic stimulation.

Treatment

The only treatment for paralysis is to treat its underlying cause. The loss of function caused by long-term paralysis can be treated through a comprehensive **rehabilitation** program. Rehabilitation includes:

- Physical therapy. The physical therapist focuses on mobility. Physical therapy helps develop strategies to compensate for paralysis by using those muscles that still have normal function, helps maintain and build any strength and control that remain in the affected muscles, and helps maintain range of motion in the affected limbs to prevent muscles from shortening (contracture) and becoming deformed. If nerve regrowth is expected, physical therapy is used to retrain affected limbs during recovery. A physical therapist also suggests adaptive equipment such as braces, canes, or wheelchairs.
- Occupational therapy. The occupational therapist focuses on daily activities such as eating and bathing. Occupational therapy develops special tools and techniques that permit self-care and suggests ways to modify the home and workplace so that a patient with an impairment may live a normal life.

- Other specialties. The nature of the impairment may mean that the patient needs the services of a respiratory therapist, vocational rehabilitation counselor, social worker, speech-language pathologist, nutritionist, special education teacher, recreation therapist, or clinical psychologist.

Prognosis

The likelihood of recovery from paralysis depends on what is causing it and how much damage has been done to the nervous system.

Prevention

Prevention of paralysis depends on prevention of the underlying causes. Risk of stroke can be reduced by controlling high blood pressure and cholesterol levels. Seatbelts, air bags, and helmets reduce the risk of injury from motor vehicle accidents and falls. Good prenatal care can help prevent premature birth, which is a common cause of cerebral palsy.

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Richard Robinson

Paralysis agitans see **Parkinson's disease**

Paralytic shellfish poisoning see **Fish and shellfish poisoning**

Paranoia

Definition

Paranoia is an unfounded or exaggerated distrust of others, sometimes reaching delusional proportions. Paranoid individuals constantly suspect the motives of those around them, and believe that certain individuals, or people in general, are “out to get them.”

Description

Paranoid perceptions and behavior may appear as features of a number of mental illnesses, including depression and **dementia**, but are most prominent in three types of psychological disorders: paranoid **schizophrenia**, delusional disorder (persecutory type), and paranoid personality disorder (PPD).

Individuals with paranoid schizophrenia and persecutory delusional disorder experience what is known as persecutory **delusions**: an irrational, yet unshakable, belief that someone is plotting against them. Persecutory delusions in paranoid schizophrenia are bizarre, sometimes grandiose, and often accompanied by auditory **hallucinations**. Delusions experienced by individuals with delusional disorder are more plausible than those experienced by paranoid schizophrenics; not bizarre, though still unjustified. Individuals with delusional disorder may seem offbeat or quirky rather than mentally ill, and, as such, may never seek treatment.

Persons with paranoid personality disorder tend to be self-centered, self-important, defensive, and emotionally distant. Their paranoia manifests itself in constant suspicions rather than full-blown delusions. The disorder often impedes social and personal relationships and career advancement. Some individuals with PPD are described as “litigious,” as they are constantly initiating frivolous law suits. PPD is more common in men than in women, and typically begins in early adulthood.

Causes and symptoms

The exact cause of paranoia is unknown. Potential causal factors may be genetics, neurological abnormalities, changes in brain chemistry, and **stress**. Paranoia is also a possible side effect of drug use and **abuse** (for example, alcohol, **marijuana**, amphetamines, **cocaine**, PCP). Acute, or short term, paranoia may occur in some individuals overwhelmed by stress.

The *Diagnostic and Statistical Manual of Mental Disorders*, fourth edition (*DSM-IV*), the diagnostic standard for mental health professionals in the United States, lists the following symptoms for paranoid personality disorder:

- suspicious; unfounded suspicions; believes others are plotting against him/her
- preoccupied with unsupported doubts about friends or associates
- reluctant to confide in others due to a fear that information may be used against him/her
- reads negative meanings into innocuous remarks

KEY TERMS

Persecutory delusion—A fixed, false, and inflexible belief that others are engaging in a plot or plan to harm an individual.

- bears grudges
- perceives attacks on his/her reputation that are not clear to others, and is quick to counterattack
- maintains unfounded suspicions regarding the fidelity of a spouse or significant other

Diagnosis

Patients with paranoid symptoms should undergo a thorough **physical examination** and patient history to rule out possible organic causes (such as dementia) or environmental causes (such as extreme stress). If a psychological cause is suspected, a psychologist will conduct an interview with the patient and may administer one of several clinical inventories, or tests, to evaluate mental status.

Treatment

Paranoia that is symptomatic of paranoid schizophrenia, delusional disorder, or paranoid personality disorder should be treated by a psychologist and/or psychiatrist. Antipsychotic medication such as thioridazine (Mellaril), haloperidol (Haldol), chlorpromazine (Thorazine), clozapine (Clozaril), or risperidone (Risperdal) may be prescribed, and cognitive therapy or **psychotherapy** may be employed to help the patient cope with their paranoia and/or persecutory delusions. Antipsychotic medication, however, is of uncertain benefit to individuals with paranoid personality disorder and may pose long-term risks.

If an underlying condition, such as depression or drug abuse, is found to be triggering the paranoia, an appropriate course of medication and/or psychosocial therapy is employed to treat the primary disorder.

Prognosis

Because of the inherent mistrust felt by paranoid individuals, they often must be coerced into entering treatment. As unwilling participants, their recovery may be hampered by efforts to sabotage treatment (for example, not taking medication or not being forthcoming with a therapist), a lack of insight into their condition, or the belief that the therapist is plotting against them. Albeit with restricted lifestyles,

some patients with PPD or persecutory delusional disorder continue to function in society without treatment.

ORGANIZATIONS

American Psychiatric Association (APA), 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209, (888) 357-7924, apa@psych.org, <http://www.psych.org>.

American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (202) 336-5500, (800) 374-2721, <http://www.apa.org/>.

National Alliance for the Mentally Ill (NAMI), 3803 N. Fairfax Dr., Ste. 100, Arlington, VA, 22203, 703 524-7600, (703) 524-9094, (800) 950-6264, <http://www.nami.org>.

National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Bethesda, MD 20892, 301-443-4513, 1-866-615-6464, 301-443-8431 (TTY), 1-866-415-8051 (TTY toll-free), 301-443-4279 (Fax), nimhinfo@nih.gov, <http://www.nimh.nih.gov/index.shtml>.

Paula Anne Ford-Martin

Parapharyngeal abscess see **Abscess**

Paraphilias see **Sexual perversions**

Paraplegia see **Paralysis**

Parasomnia see **Sleep disorders**

Parathyroid gland removal see
Parathyroidectomy

Differential diagnosis of hyperparathyroidism

PTH is also useful in the differential diagnosis of overactive parathyroid glands (**hyperparathyroidism**). Primary hyperparathyroidism is most often caused by a benign tumor in one or more of the parathyroid glands. It is rarely caused by parathyroid **cancer**. Patients with this condition have high PTH and calcium levels.

Secondary hyperparathyroidism is often seen in patients with chronic renal failure (CRF). The kidneys fail to excrete sufficient phosphate, and the parathyroid gland secretes PTH in an effort to lower calcium levels to balance the calcium-phosphate ratio. Because of the constant stimulation of the parathyroid, CRF patients have high PTH and normal or slightly low calcium levels.

Tertiary hyperparathyroidism occurs when CRF causes a severe imbalance in the calcium-phosphate ratio, leading to very high PTH production that results in hypercalcemia. Patients with this condition have high PTH and high calcium levels.

Specific PTH assays

PTH is broken down in the body into three different molecular forms: the intact PTH molecule and several smaller fragments which include an amino acid or N-terminal, a midregion or midmolecule, and a carboxyl or C-terminal. Two tests are currently used to measure intact PTH and its terminal fragments. While both tests are used to diagnose hyper or **hypoparathyroidism**, each test also has specific applications as well. The C-terminal PTH assay is used to diagnose the ongoing disturbances in PTH metabolism that occur with secondary and tertiary hyperparathyroidism. The assay for intact PTH and the N-terminal fragment, which are both measured at the same time, is more accurate in detecting sudden changes in the PTH level. For this reason, the N-terminal PTH assay is used to monitor a patient's response to therapy.

Parathyroid hormone test

Definition

The parathyroid hormone (PTH) test is a blood test performed to determine the serum levels of a hormone secreted by the parathyroid gland in response to low blood **calcium** levels. PTH works together with vitamin D to maintain healthy bones. The parathyroid glands are small paired glands located near the thyroid gland at the base of the neck.

Purpose

The PTH level is measured to evaluate the level of blood calcium. It is routinely monitored in patients with a kidney disorder called chronic renal failure (CRF). Because PTH is one of the major factors affecting calcium metabolism, the PTH test helps to distinguish nonparathyroid from parathyroid causes of too much calcium in the blood (**hypercalcemia**).

Precautions

Drug interactions

Some prescription drugs affect the results of PTH tests. Drugs that *increase* PTH levels include phosphates, anticonvulsants, **steroids**, isoniazid, lithium, and rifampin. Drugs that *decrease* PTH include cimetidine and propranolol.

Timing

PTH levels are subject to daily variation, ranging from a peak around 2:00 a.m. to a low point around

KEY TERMS

Assay—An analysis of the chemical composition or strength of a substance.

Hypercalcemia—Abnormally high levels of blood calcium.

Hyperparathyroidism—Overactivity of the parathyroid glands. Symptoms include generalized aches and pains, depression, and abdominal pain.

Hypoparathyroidism—Insufficient production of parathyroid hormone, which results in low levels of blood calcium.

2:00 p.m. Specimens are usually drawn at 8:00 a.m. The laboratory should be notified if the patient works a night shift so that this difference in biological rhythm can be taken into account.

Other serum level tests

Due to the relationship between PTH and calcium, calcium levels should be tested at the same time as PTH. Most laboratories have established reference values to indicate what PTH level is normal for a particular calcium level. In addition, the effects of PTH on kidney function and bone strength indicate that serum calcium, phosphorus, and creatinine levels should be measured together with PTH. The **creatinine test** measures kidney function and aids in the diagnosis of parathyroid dysfunction.

Description

The PTH test is performed on a sample of the patient's blood, withdrawn from a vein into a vacuum tube. The procedure, which is called a venipuncture, takes about five minutes.

Preparation

The patient should have nothing to eat or drink from midnight of the day of the test.

Risks

Risks for this test are minimal, but may include slight bleeding from the puncture site, a small bruise or swelling in the area, or **fainting** or feeling lightheaded.

Normal results

Reference ranges for PTH tests vary somewhat depending on the laboratory, and must be interpreted

in association with calcium results. The following ranges are typical:

- Intact PTH: 10–65 pg/mL
- PTH N-terminal (includes intact PTH): 8–24 pg/mL
- PTH C-terminal (includes C-terminal, intact PTH, and midmolecule): 50–330 pg/mL

Abnormal results

When measured with serum calcium levels, abnormally *high* PTH values may indicate primary, secondary, or tertiary hyperparathyroidism, chronic renal failure, **malabsorption syndrome**, and **vitamin D deficiency**. Abnormally *low* PTH levels may indicate hypoparathyroidism, hypocalcemia, and certain malignancies.

Resources

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Janis O. Flores

Parathyroid scan

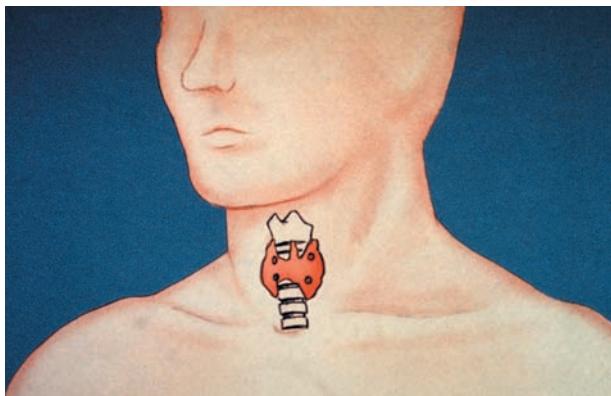
Definition

A parathyroid scan is sometimes called a parathyroid localization scan or parathyroid scintigraphy. This scan uses radioactive pharmaceuticals that are readily taken up by cells in the parathyroid glands to obtain an image of the glands and any abnormally active areas within them.

Purpose

The parathyroid glands, embedded in the thyroid gland in the neck, but separate from the thyroid in function, control **calcium** metabolism in the body. The parathyroid glands produce parathyroid hormone (PTH). PTH regulates the level of calcium in the blood.

Calcium is critical to cellular metabolism, as well as being the main component of bones. If too much PTH is secreted, the bones release calcium into the bloodstream. Over time, the bones become brittle and more likely to break. A person with levels of calcium in the blood that are too high feels tired, run down, irritable, and has difficulty sleeping. Additional signs of too much calcium in the blood are **nausea and vomiting**, frequent urination, **kidney stones** and bone **pain**. A parathyroid scan is administered when the



The parathyroid glands, embedded in the thyroid gland in the neck but separate from the thyroid gland in function, control calcium metabolism in the body by producing parathyroid hormone, or PTH. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

parathyroid appears to be overactive and a tumor is suspected.

Precautions

Parathyroid scans are not recommended for pregnant women because of the potential harm to the developing fetus. People who have had another recent nuclear medicine procedure or an intravenous contrast test may need to wait until the earlier radioactive markers have been eliminated from their system in order to obtain accurate results from the parathyroid scan.

Description

A parathyroid scan is a non-invasive procedure that uses two radiopharmaceuticals (drugs with a radioactive marker) to obtain an image of highly active areas of the parathyroid glands. The test can be done in two ways.

Immediate scan

If the test is to be performed immediately, the patient lies down on an imaging table with his head and neck extended and immobilized. The patient is injected with the first radiopharmaceutical. After waiting 20 minutes, the patient is positioned under the camera for imaging. Each image takes five minutes. It is essential that the patient remain still during imaging.

After the first image, the patient is injected with a second radiopharmaceutical, and imaging continues for another 25 minutes. Total time for the test is about one hour: injection 10 minutes, waiting period 20 minutes, and imaging 30 minutes.

KEY TERMS

Cyst—An abnormal sac containing fluid or semi-solid material.

Goiter—Chronic enlargement of the thyroid gland.

Neoplasm—An uncontrolled growth of new tissue.

Another way to do this test is as follows. After the first images are acquired, the patient returns two hours later for additional images. Time for this procedure totals about three hours: injection 10 minutes, waiting period two hours and 20 minutes, and imaging 30 minutes.

Delayed scan

In a delayed parathyroid scan, the patient is asked to swallow capsules containing the first radiopharmaceutical. The patient returns after a four hour waiting period, and the initial image is made. Then the patient is injected with the second radiopharmaceutical. Imaging continues for another 25 minutes. The total time is about four hours and 40 minutes: waiting period four hours, injection 10 minutes, and imaging 30 minutes.

Preparation

No special preparations are necessary for this test. It is not necessary to fast or maintain a special diet. The patient should wear comfortable clothing and no metal jewelry around the neck.

Aftercare

The patient should not feel any adverse effects of the test and can resume normal activities immediately.

Risks

The only risk associated with this test is to the fetus of a pregnant woman.

Normal results

Normal results will show no unusual activity in the parathyroid glands.

Abnormal results

A concentration of radioactive materials in the parathyroid gland beyond background levels suggests excessive activity and the presence of a tumor. False positive results sometimes result from the presence of multinodular goiter, neoplasm, or cysts. False positive

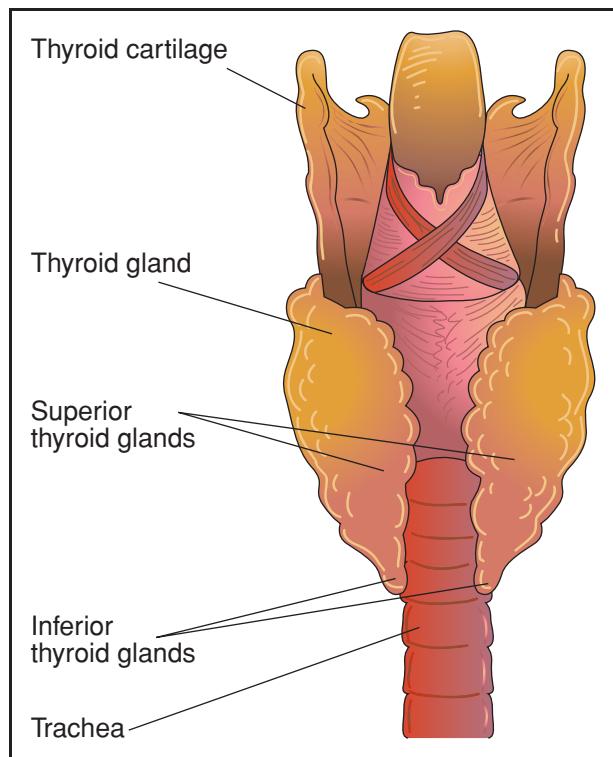
tests are tests that interpret the results as abnormal when this is not true.

Resources

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Tish Davidson, A.M.



Parathyroidectomy

Definition

Parathyroidectomy is the removal of one or more of the parathyroid glands. The parathyroid glands are usually four in number, although the exact number may vary from three to seven. They are located in the neck in front of the Adam's apple and are closely linked to the thyroid gland. The parathyroid glands regulate the balance of **calcium** in the body.

Purpose

Parathyroidectomy is usually done to treat **hyperparathyroidism** (abnormal over-functioning of the parathyroid glands).

Precautions

Parathyroidectomy should only be done when other non-operative methods have failed to control the patient's hyperparathyroidism.

Description

Parathyroidectomy is an operation done most commonly by a general surgeon, or occasionally by an otolaryngologist, in the operating room of a hospital. The operation begins when the anesthesiologist anesthetizes or puts the patient to sleep. The surgeon makes an incision in the front of the neck where a tight-fitting necklace would rest. All of the parathyroid glands are identified. The surgeon then identifies the gland or glands with the disease and confirms the diagnosis by sending a piece of the gland(s) to the pathology department for immediate microscopic examination. The glands are then removed and the incision is closed and a dressing is placed over the incision.

Parathyroidectomy refers to the surgical removal of one or more of the parathyroid glands due to hyperparathyroidism (an abnormal over-functioning of the parathyroid glands). It is usually done after other non-operative methods have failed to control or correct this condition. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Patients generally stay overnight in the hospital after completion of the operation and may remain for one or two additional days. These procedures are reimbursed by insurance companies. Surgeon's fees typically range from \$1,000–\$2,000. Anesthesiologists charge for their services based on the medical status of the patient and the length of the operative procedure. Hospitals charge for use of the operating suite, equipment, lab and diagnostic tests, and medications.

Preparation

Prior to the operation, the diagnosis of hyperparathyroidism should be confirmed using lab tests. Occasionally, physicians order **computed tomography scans** (CT scans), ultrasound exams, and/or **magnetic resonance imaging** (MRI) tests to determine the total number of parathyroid glands and their location prior to the procedure.

KEY TERMS

Anesthesiologist—A physician who specializes in anesthetizing patients for operations.

Ectopic parathyroid tissue—A condition where the thyroid tissue is located in an abnormal place.

Hyperparathyroidism—Abnormal over-functioning of the parathyroid glands.

Hypoparathyroidism—Abnormal under-functioning of the parathyroid glands.

Otolaryngologist—A surgeon who treats people with abnormalities in the head and neck regions of the body.

Aftercare

The incision should be watched for signs of infection. In general, no specific wound care is required.

The level of calcium in the body should be monitored during the first 48 hours after the operation by obtaining frequent blood samples for laboratory analysis.

Risks

The major risk of parathyroidectomy is injury to the recurrent laryngeal nerve (a nerve that lies very near the parathyroid glands and serves the larynx or voice box). If this nerve is injured, the voice may become hoarse or weak.

Occasionally, too much parathyroid tissue is removed, and the patient may develop **hypoparathyroidism** (under-functioning of the parathyroid glands). If this occurs, the patient will require daily calcium supplements.

Sometimes not all of the parathyroid glands are found in the initial operation. A fifth or sixth gland may be located in an aberrant location such as the chest (ectopic parathroid). If this occurs, the patient's hyperparathyroidism may not be corrected, and a second procedure may be required to find the other gland(s).

Hematoma formation (collection of blood under the incision) is a possible complication of any operative procedure. However, in procedures that involve the neck it is of particular concern, because a rapidly enlarging hematoma can obstruct the airway.

Infection of the surgical incision may occur, as with any operative procedure, but this is not common.

Normal results

Most patients require only two or three days of hospitalization to recover from the operation. They usually can resume most of their normal activities within one to two weeks.

Resources

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Mary Jeanne Krob, MD, FACS

Paratyphoid fever

Definition

Paratyphoid fever, which is sometimes called *Salmonella paratyphi* infection, is a serious contagious disease caused by a gram-negative bacterium. It is also grouped together with **typhoid fever** under the name enteric fever.

Description

Enteric fever is increasingly rare in the United States. Of the 500 cases reported in an average year, about 60% are infections acquired during travel in Mexico, India, or South America.

Paratyphoid fever has three stages: an early stage marked by high fever; a toxic stage with abdominal pain and intestinal symptoms, and a long period of recovery from fever (defervescence). In adults, these three phases may cover a period of four to six weeks; in children, they are shorter and may cover 10 days to two weeks. During the toxic stage there is a 1–10% chance of intestinal perforation or hemorrhage.

Causes and symptoms

Paratyphoid fever is caused by any of three strains of *Salmonella paratyphi*: *S. paratyphi A*; *S. schottmuelleri* (also called *S. paratyphi C*); or *S. hirschfeldii* (also called *S. paratyphi B*). It can be transmitted from animals or animal products to humans or from person to person. The incubation

period is one to two weeks but is often shorter in children. Symptom onset may be gradual in adults but is often sudden in children.

Paratyphoid fever is marked by high fever, **headache**, loss of appetite, **vomiting**, and **constipation** or **diarrhea**. The patient typically develops an enlarged spleen. About 30% of patients have rose spots on the front of the chest during the first week of illness. The rose spots develop into small hemorrhages that may be hard to see in African or Native Americans.

Patients with intestinal complications have symptoms resembling those of **appendicitis**: intense cramping pain with soreness in the right lower quadrant of the abdomen.

Diagnosis

The diagnosis is usually made on the basis of a history of recent travel and culturing the paratyphoid organism. Because the disease is unusual in the United States, the doctor may not consider paratyphoid in the diagnosis unless the patient has the classic symptoms of an enlarged spleen and rose spots. The doctor will need to rule out other diseases with high fevers, including **typhus**, **brucellosis**, **tularemia** (rabbit fever), psittacosis (parrot fever), mononucleosis, and **Kawasaki syndrome**. *S. paratyphi* is easily cultured from samples of blood, stool, urine, or bone marrow.

Treatment

Medications

Paratyphoid fever is treated with **antibiotics** over a two- to three-week period with trimethoprim-sulfamethoxazole (Bactrim, Septra); amoxicillin (Amoxil, Novamoxin); and ampicillin (Ampcill). Third-generation **cephalosporins** (ceftriaxone [Rocephin], cefotaxime [Claforan], or cefixime [Suprax]) or chloramphenicol (Chloromycetin) may be given if the specific strain is resistant to other antibiotics.

Surgery

Patients with intestinal perforation or hemorrhage may need surgery if the infection cannot be controlled by antibiotics.

Supportive care

Patients with paratyphoid fever need careful monitoring for signs of complications as well as bed rest and nutritional support. Patients with severe infections may require fluid replacement or blood transfusions.

KEY TERMS

Defervescence—Return to normal body temperature after high fever.

Enteric fever—A term that is sometimes used for either typhoid or paratyphoid fever.

Rose spots—Small slightly raised reddish pimples that are a distinguishing feature of typhoid or paratyphoid infection.

Prognosis

Most patients with paratyphoid fever recover completely, although intestinal complications can result in **death**. With early treatment, the mortality rate is less than 1%.

Prevention

Immunization

Vaccination against paratyphoid fever is not necessary within the United States but is recommended for travel to countries with high rates of enteric fever.

Hygienic measures

Travelers in countries with high rates of paratyphoid fever should be careful to wash hands before eating and to avoid meat, egg, or poultry dishes unless they have been thoroughly cooked.

Resources

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Rebecca J. Frey, PhD

Paresthesias see **Numbness and tingling**

Parkinson's disease

Definition

Parkinson's disease (PD) is a progressive degenerative brain disorder marked by **tremors**, rigidity, slow movements (bradykinesia), and posture instability. It occurs when cells in one of the movement-control

centers of the brain begin to die for unknown reasons. PD was first described by British physician James Parkinson in the early 1817.

Demographics

About 1% of people over age 60 develop PD with an approximate prevalence of 120 cases per 100,000 population. The likelihood of developing PD increases with age with an estimated 15% of those ages 65–74, and almost 30% of those ages 75–84 showing symptoms. Because PD is difficult to diagnose accurately, these numbers are only estimates. PD is about 1.5 times more common in men than in women. Average age of onset is 60 years; the disease is uncommon in people under age 40.

Description

Usually beginning in a person's late fifties or early sixties, Parkinson's disease causes a progressive decline in movement control, affecting the ability to control initiation, speed, and smoothness of motion. Many cases of PD are sporadic. This means that there is a spontaneous and permanent change in nucleotide sequences (the building blocks of genes). Sporadic mutations also involve unknown environmental factors in combination with genetic defects. The abnormal gene (mutated gene) will form an altered end product or protein. This will cause abnormalities in specific areas in the body where the protein is used. Some evidence suggests that there is also a genetic component that predisposes some people to develop the disease when exposed to certain (as yet undiscovered) environmental factors. Recent research has linked PD with a gene that codes for a protein called alpha-synuclein. Further research is attempting to fully understand the relationship with this protein and nerve cell degeneration.

Risk factors

Age is the greatest risk factor for developing PD. Gender also is a risk factor, as men are more likely to be diagnosed with the disease. Family history can increase risk; people with a first-degree relative (parent, sibling, child) with PD have double the chance of developing the disease compared to people without PD in the immediate family.

Smoking tobacco has consistently been shown to protect against the development of PD as has **caffeine** consumption. However, other health risks of smoking far outweigh the potential protective effect.

Causes and symptoms

The immediate cause of PD is degeneration of brain cells in the area known as the substantia nigra, one of the movement control centers of the brain. Damage to this area leads to the cluster of symptoms known as "parkinsonism." In PD, degenerating brain cells contain Lewy bodies which are not found in healthy brain cells and which help to identify the disease. The cell **death** leading to parkinsonism may be caused by a number of conditions, including infection, trauma, and **poisoning**. Some drugs given for **psychosis**, such as haloperidol (Haldol) or chlorpromazine (Thorazine, Largactil), may cause parkinsonism. When no cause for nigral cell degeneration can be found, the disorder is called idiopathic parkinsonism, or Parkinson's disease. Parkinsonism may be seen in other degenerative conditions, known as the "parkinsonism plus" syndromes, such as **progressive supranuclear palsy**.

The substantia nigra, or "black substance," is one of the principal movement control centers in the brain. By releasing the neurotransmitter dopamine, it helps to refine movement patterns throughout the body. The dopamine released by nerve cells of substantia nigra stimulates another brain region, the corpus striatum. Without enough dopamine, the corpus striatum cannot control its targets, and so on down the line. Ultimately, the movement patterns of walking, writing, reaching for objects, and other basic programs cannot operate properly, and the symptoms of parkinsonism are the result.

Some known toxins can cause parkinsonism, most notoriously a chemical called MPTP, found as an impurity in some illegal drugs. Parkinsonian symptoms appear within hours of ingestion and are permanent. MPTP may exert its effects through generation of toxic molecular fragments called free radicals, and reducing free radicals has been a target of several experimental treatments for PD using **antioxidants**.

It is possible that early exposure to some as-yet-unidentified environmental toxin or virus leads to undetected nigral cell death, and PD then manifests as normal age-related decline brings the number of functioning nigral cells below the threshold needed for normal movement. It is also possible that, for genetic reasons, some people are simply born with fewer cells in their substantia nigra than others, and they develop PD as a consequence of normal decline.

KEY TERMS

AADC inhibitors—Drugs that block the amino acid decarboxylase; one type of enzyme that breaks down dopamine. Also called DC inhibitors, they include carbidopa and benserazide.

Akinesia—A loss of the ability to move; freezing in place.

Antioxidant—A molecule that prevents oxidation. In the body antioxidants attach to other molecules called free radicals and prevent the free radicals from causing damage to cell walls, DNA, and other parts of the cell.

Bradykinesia—Extremely slow movement.

COMT inhibitors—Drugs that block catechol-o-methyl transferase, an enzyme that breaks down dopamine. COMT inhibitors include entacapone and tolcapone.

Dopamine—A neurochemical made in the brain that is involved in many brain activities, including movement and emotion.

Dyskinesia—Impaired ability to make voluntary movements.

Free radical—A molecule with an unpaired electron that has a strong tendency to react with other molecules in DNA (genetic material), proteins, and lipids (fats), resulting in damage to cells. Free radicals are neutralized by antioxidants.

Idiopathic—Of unknown origin; without a known cause.

MAO-B inhibitors—Inhibitors of the enzyme monoamine oxidase B. MAO-B helps break down dopamine; inhibiting it prolongs the action of dopamine in the brain. Selegiline is an MAO-B inhibitor.

Orthostatic hypotension—A sudden decrease in blood pressure upon sitting up or standing. May be a side effect of several types of drugs.

Substantia nigra—One of the movement control centers of the brain.

Symptoms

The identifying symptoms of PD include:

- Tremors, usually beginning in the hands, often occurring on one side before the other. The classic tremor of PD is called a “pill-rolling tremor,” because the movement resembles rolling a pill between the thumb and forefinger. This tremor occurs at a frequency of about three per second.
- Slow movements (bradykinesia) occur, which may involve slowing down or stopping in the middle of familiar tasks such as walking, eating, or shaving. This may include freezing in place during movements (akinesia).
- Muscle rigidity or stiffness, occurring with jerky movements replacing smooth motion.
- Postural instability or balance difficulty occurs. This may lead to a rapid, shuffling gait (festination) to prevent falling.
- In most cases, there is a “masked face,” with little facial expression and decreased eye-blinking.

In addition, a wide range of other symptoms may often be seen, some beginning earlier than others:

- depression (reported in about half of all individuals with PD)
- speech changes, including rapid speech without inflection changes

- problems with sleep, including restlessness and nightmares
- emotional changes, including fear, irritability, and insecurity
- incontinence
- constipation
- handwriting changes, with letters becoming smaller across the page (micrographia)
- progressive problems with intellectual function (dementia)

Diagnosis

The diagnosis of Parkinson's disease involves a careful medical history and a **neurological exam** to look for characteristic symptoms. There are no definitive tests for PD, although a variety of lab tests may be done to rule out other causes of symptoms, especially if only some of the identifying symptoms are present. Tests for other causes of parkinsonism may include brain scans, blood tests, **lumbar puncture**, and x rays.

Treatment

There is no cure for Parkinson's disease. Treatment can be complicated and is based on the individual's age, level of impairment, cognitive function and response to treatment.

Exercise, nutrition, and physical therapy

Regular, moderate **exercise** has been shown to improve motor function without an increase in medication for a person with PD. Exercise helps maintain range of motion in stiff muscles, improve circulation, and stimulate appetite. An exercise program designed by a physical therapist has the best chance of meeting the specific needs of the person with PD. A physical therapist may also suggest strategies for balance compensation and techniques to stimulate movement during slowdowns or freezes.

Good **nutrition** is important to maintenance of general health. A person with PD may lose some interest in food, especially if depressed, and may have **nausea** from the disease or from medications, especially those known as dopamine agonists. Slow movements may make it difficult to eat quickly, and delayed gastric emptying may lead to a feeling of fullness without having eaten much. Increasing fiber in the diet can improve **constipation**, soft foods can reduce the amount of needed chewing, and a prokinetic drug can increase the movement of food through the digestive system.

People with PD may need to limit the amount of protein in their **diets**. The main drug used to treat PD, L-dopa, is an amino acid, and is absorbed by the digestive system by the same transporters that pick up other amino acids broken down from proteins in the diet. Limiting protein, under the direction of the physician or a nutritionist, can improve the absorption of L-dopa.

No evidence indicates that vitamin or mineral supplements can have any effect on the disease other than in the improvement of the patient's general health. No antioxidants used to date have shown promise as a treatment except for selegiline, an MAO-B inhibitor that is discussed below. A large, carefully controlled study of vitamin E demonstrated that it could not halt disease progression. However, in a preliminary study, the antioxidant co-enzyme Q10 appeared to slow the progression of PD. Co-enzyme Q10 remains under investigation.

Drugs

The pharmacological treatment of Parkinson's disease is complex. While there are a large number of drugs that can be effective, their effectiveness varies with the patient, disease progression, and the length of time the drug has been used. Dose-related side effects may preclude using the most effective dose, or require the introduction of a new drug to counteract them. Response to drug therapy is monitored and drugs may be adjusted in an attempt to find a treatment regimen

that provides the most benefits with the fewest side effects. Research is ongoing in an effort to find drugs to treat PD. Individuals should consult their doctor about advances in drug therapy and clinical trials underway to test new PD drugs. There are six classes of drugs currently used to treat PD.

DRUGS THAT REPLACE DOPAMINE. One drug that helps replace dopamine, levodopa (L-dopa), is the single most effective treatment for the symptoms of PD. L-dopa is a derivative of dopamine, and is converted into dopamine by the brain. It may be started when symptoms begin, or when they become serious enough to interfere with work or daily living.

L-dopa therapy usually remains effective for five years or longer. Following this, many patients develop motor fluctuations, including peak-dose "dyskinesias" (abnormal movements such as tics, twisting, or restlessness), rapid loss of response after dosing (known as the "on-off" phenomenon), and unpredictable drug response. Higher doses may be tried, but often lead to an increase in dyskinesias. In addition, side effects of L-dopa include **nausea and vomiting**, and low blood pressure upon standing (**orthostatic hypotension**), which can cause **dizziness**. These effects may lessen after several weeks of therapy.

ENZYME INHIBITORS. Dopamine is broken down by several enzyme systems in the brain and elsewhere in the body, and blocking these enzymes is a key strategy to prolonging the effect of dopamine. The two most commonly prescribed forms of L-dopa contain a drug to inhibit the amino acid decarboxylase (an AADC inhibitor), one type of enzyme that breaks down dopamine. These combination drugs are Sine-met and Parcopa (L-dopa plus carbidopa) and Madopar (L-dopa plus benzaseride). Controlled-release formulations also aid in prolonging the effective interval of an L-dopa dose.

The enzyme monoamine oxidase B (MAO-B) inhibitor selegiline(Eldepryl) may be given as add-on therapy for L-dopa. Selegiline appears to have a neuroprotective effect, sparing nigral cells from damage by free radicals. Because of this, and the fact that it has few side effects, it is frequently prescribed early in the disease before L-dopa is begun. Rasagiline (Azilect) is a second-generation MAO-B inhibitor with fewer potential side effects than selegiline. Entacapone (Comtan) and tolcapone (Tasmar), two inhibitors of another enzyme system called catechol-o-methyl transferase (COMT) are also available to treat PD symptoms with fewer motor fluctuations and decreased daily L-dopa requirements.

CHOLINESTERASE INHIBITORS. The cholinesterase inhibitor Exelon (rivastigmine) both as a tablet and a transdermal patch is used to treat **dementia** in mild to moderate PD.

DOPAMINE AGONISTS. Dopamine works by stimulating receptors on the surface of corpus striatum cells. Drugs that also stimulate these cells are called dopamine agonists, or DAs. DAs may be used before L-dopa therapy, or added on to avoid requirements for higher L-dopa doses late in the disease. DAs available in the United States as of 2009 include Apomorphine (Apokyn), a short-acting DA, bromocriptine (Parlodel), ropinirole (Requip), and pramipexole (Mirapex). In 2007, the U.S. Food and Drug Administration (FDA) approved cabergoline (Dostinex) for treatment of PD. Other dopamine agonists in use elsewhere include lisuride (Dopergine) and apomorphine. Side effects of all the DAs are similar to those of dopamine, plus confusion and **hallucinations** at higher doses. In 2007, the drug pergolide (Permax) was withdrawn from sale in the United States and elsewhere after studies showed it increased the risk of serious heart valve damage.

ANTICHOLINERGIC DRUGS. Anticholinergics maintain dopamine balance as levels decrease. However, the side effects of anticholinergics (**dry mouth**, constipation, confusion, and blurred vision) are usually too severe in older patients or in patients with dementia. In addition, anticholinergics rarely work for very long. They are often prescribed for younger patients who have predominant shaking. Trihexyphenidyl (Artane) is the drug most commonly prescribed.

DRUGS WHOSE MODE OF ACTION IS UNCERTAIN. Amantadine (Symmetrel) is sometimes used as an early therapy before L-dopa is begun, and as an add-on later in the disease. Its anti-Parkinsonian effects are mild, and are not seen in many patients. Clozapine (Clozaril) is effective especially against psychiatric symptoms of late PD, including psychosis and hallucinations.

Surgery

Two surgical procedures are used for treatment of PD that cannot be controlled adequately with drug therapy. In PD, a brain structure called the globus pallidus (GPi) receives excess stimulation from the corpus striatum. In a pallidotomy, the GPi is destroyed by heat, delivered by long thin needles inserted under anesthesia. Electrical stimulation of the GPi is another way to reduce its action. In this procedure, fine electrodes are inserted to deliver the stimulation, which may be adjusted or turned

off as the response dictates. Other regions of the brain may also be stimulated by electrodes inserted elsewhere. In most patients, these procedures lead to significant improvement for some motor symptoms, including peak-dose dyskinesias. This allows the patient to receive more L-dopa, since these dyskinesias are usually what causes an upper limit on the L-dopa dose.

A third procedure, transplant of fetal nigral cells, is still highly experimental. Its benefits to date have been modest, although improvements in technique and patient selection are likely to change that. Also, **gene therapy** is showing promise as a future treatment for PD. In one trial by Cornell University scientists involving 12 patients with PD, all had their symptoms improved by at least 25% for up to a year after gene therapy. Further research is being conducted.

Alternative treatment

Currently, the best treatments for PD involve the use of conventional drugs such as levodopa. Alternative therapies, including **acupuncture**, massage, and **yoga**, can help relieve some symptoms of the disease and loosen tight muscles. Alternative practitioners have also applied herbal and dietary therapies, including amino acid supplementation, antioxidant (**vitamins A, C, E, selenium, and zinc**) therapy, B vitamin supplementation, and **calcium** and magnesium supplementation, to the treatment of PD. Anyone using these therapies in conjunction with conventional drugs should check with their doctor to avoid the possibility of adverse interactions. For example, vitamin B₆ (either as a supplement or from foods such as whole grains, bananas, beef, fish, liver, and potatoes) can interfere with the action of L-dopa when the drug is taken without carbidopa.

Prognosis

Despite medical treatment, the symptoms of Parkinson's disease worsen over time, and become less responsive to drug therapy. Late-stage psychiatric symptoms are often the most troubling, including difficulty sleeping, nightmares, intellectual impairment (dementia), hallucinations, and loss of contact with reality (psychosis).

Prevention

There is no known way to prevent Parkinson's disease.

Resources

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- Robinson, Richard, ed. "Parkinson's Disease." WeMove.org. December 7, 2008. <http://www.wemove.org/par/> (accessed December 20, 2010).

ORGANIZATIONS

- American Parkinson Disease Association, 135 Parkinson Ave., Staten Island, NY, 10305, (718) 981-8001, (800) 223-2732, (718) 981-4399, adpa@adpaparkinson.org, <http://www.apdparkinson.org>.
- National Institute of Neurological Disorders and Stroke (NINDS), P.O. Box 5801, Bethesda, MD, 20828, (301) 496-5751. TTY: (301) 468-5981, (800) 352-9424, <http://www.ninds.nih.gov>.
- National Parkinson Foundation, 1501 N.W. 9th Avenue/ Bob Hope Road, Miami, FL, 33136-1494, (305) 243-6666, (800) 327-4545, (305) 243-5595, contact@parkinson.org, <http://www.parkinson.org>.
- Parkinson's Disease Foundation, 1359 Broadway, Suite 1509, New York, NY, 10018, (212) 923-4700, (800) 457-6676, (212) 923-4778, info@pdf.org, <http://www.pdf.org>.
- The Parkinson's Institute and Clinical Center, 672 Almanor Ave, Sunnyvale, CA, 94068, (408) 734-2800, (800) 655-2273, info2@thepi.org, <http://www.thepi.org>.

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Parkinsonism see **Parkinson's disease**

Parotid gland removal see **Parotidectomy**

Parotid gland scan see **Salivary gland scan**

Parotidectomy

Definition

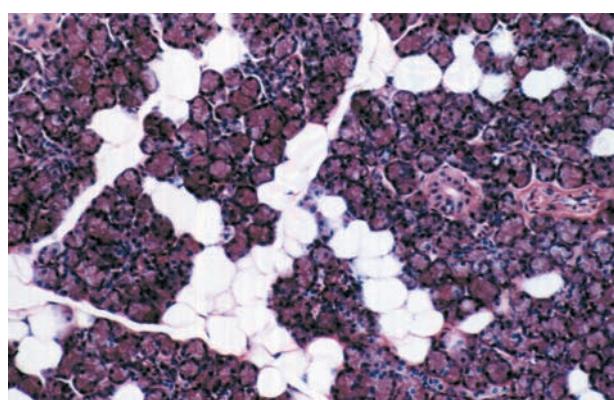
Parotidectomy is the removal of the parotid gland, a salivary gland near the ear.

Purpose

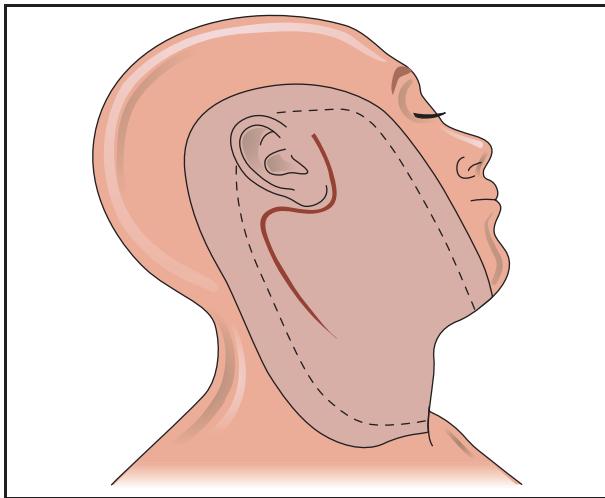
The main purpose of parotidectomy is to remove cancerous tumors in the parotid gland. A number of tumors can develop in the parotid gland. Many of these are tumors that have spread from other areas of the body, entering the parotid gland by way of the lymphatic system. Among the tumors seen in the parotid gland are lymphoma, melanoma, and squamous cell carcinoma.

Description

The parotid gland is the largest of the salivary glands. There are two parotid glands, one on each side of the face. They lie just in front of the ears and a duct runs from each to the inside of the cheek. Each parotid gland has several lobes. Surgery is recommended as part of the treatment for all cancers in the parotid gland. Superficial or localized parotidectomy is recommended by some authorities, unless a lipoma or Warthin's tumor is present. One of the advantages to this approach is that nerves to facial muscles are left intact. Many facial nerves run through the same area as the parotid gland and can be damaged during more complete parotidectomies. Most authorities recommend total parotidectomy, especially if **cancer** is



A micrograph of a normal human parotid gland. One of the salivary glands, the parotid consists of acini arranged in lobes. This image shows a junction between several lobes; the clear spaces represent the interlobular connective tissue. The masses of secretory cells produce a watery secretion which is passed to the intralobular. (Custom Medical Stock Photo, Inc. Reproduced by permission.)



Parotidectomy is a surgical procedure performed to remove cancerous tumors in the parotid gland, a salivary gland near the ear. Among the tumors seen in the parotid gland are lymphoma, melanoma, and squamous cell carcinoma. The illustration above shows the facial incision sites for this procedure. (Illustration by Electronic Illustrators Group.)

Reproduced by permission of Gale, a part of Cengage Learning.)

found in both the superficial and deep lobes of the parotid gland. If the tumor has spread to involve the facial nerve, the operation is expanded to include parts of bone behind the ear (mastoid) to remove as much tumor as possible. Some authorities recommend post-surgery radiation as follow-up treatment for cancer.

Aftercare

After surgery, the patient will remain in the hospital for one to three days. The site of incision will be watched closely for signs of infection and heavy bleeding (hemorrhage). The incision site should be kept clean and dry until it is completely healed. The patient should not wash their hair until the stitches have been removed. If the patient has difficulty smiling, winking, or drinking fluids, the physician should be contacted immediately. These are signs of facial nerve damage.

Risks

There are a number of complications that follow parotidectomy. Facial nerve **paralysis** after minor surgery should be minimal. During surgery, it is possible to repair cut nerves. After major surgery, a graft is attempted to restore nerve function to facial muscles. Salivary fistulas can occur when saliva collects in the incision site or drains through the incision. Reoccurrence of cancer is the single most important consideration for patients who have undergone parotidectomy. Long term survival rates are largely dependent on the tumor types

KEY TERMS

Fistula—An abnormal opening or duct through tissue that results from injury, disease, or other trauma.

Salivary gland—Three pairs of glands that secrete into the mouth and aid digestion.

and the stage of tumor development at the time of the operation.

Other risks include **blood clots** (hematoma) and infection. The most common long-term complication of parotidectomy is redness and sweating in the cheek, known as Frey's syndrome. Rarely, paralysis may extend throughout all the branches of the facial nervous system.

Resources

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Mary K. Fyke

Parotitis, epidemic see **Mumps**

Paroxetine see **Selective serotonin reuptake inhibitors**

Paroxysmal atrial tachycardia

Definition

A period of very rapid and regular heart beats that begins and ends abruptly. The heart rate is usually between 160 and 200 beats per minute. This condition is also known as paroxysmal supraventricular tachycardia.

Description

The term paroxysmal means that the event begins suddenly, without warning and ends abruptly. Atrial tachycardia means that the upper chambers of the heart are beating abnormally fast. Paroxysmal atrial

tachycardia can occur without any heart disease being present. It is usually more annoying than dangerous.

Causes and symptoms

Paroxysmal atrial tachycardia may be caused by several different things. The fast rate may be triggered by a premature atrial beat that sends an impulse along an abnormal electrical path to the ventricles. Other causes stem from **anxiety**, stimulants, overactive thyroid, and in some women, the onset of menstruation.

Though seldom life-threatening, paroxysmal atrial tachycardia produces annoying symptoms which can include lightheadedness, chest **pain**, **palpitations**, anxiety, sweating, and **shortness of breath**.

Diagnosis

Diagnosis is not always easy, because the event is usually over by the time the patient sees a doctor. A careful description of the episode will aid the doctor in his diagnosis. If the rapid heart rate is still occurring, an electrocardiograph (ECG) will show the condition. If the event is over, physicians often recommend a period of ambulatory electrocardiographic monitoring (called **Holter monitoring**) to confirm the diagnosis.

Treatment

The doctor may suggest that during an episode of paroxysmal atrial tachycardia the following practice may help. Briefly hold the nose and mouth closed and breathe out, or by bearing down, as though straining at a bowel movement. The doctor may try to stop the episode by gently massaging an area in the neck called the carotid sinus.

If these conservative measures do not work, an injection of the drug verapamil or adenosine should stop the episode quickly.

In rare cases, the drugs do not work and electrical shock (**cardioversion**) may be necessary, particularly if serious symptoms are also present with the tachycardia.

Prognosis

Paroxysmal atrial tachycardia is not a disease, and is seldom life-threatening. The episodes are usually more unpleasant than they are dangerous, and the prognosis is generally good.

Prevention

Frequent episodes are usually cause for medication. In rare cases, the doctor may recommend a procedure called **catheter ablation**, which will remove

KEY TERMS

Premature atrial beat—A beat that occurs before it would normally be expected.

Supraventricular—A term for an event that occurs in the upper chambers (atria) of the heart.

(or ablate) the precise area of the heart responsible for triggering the fast heart rate.

In a catheter ablation procedure, the doctor will place a special catheter against the area of the heart responsible for the problem. Radio-frequency energy is then passed to the tip of the catheter, so that it heats up and destroys the target area. Catheter ablation is considered a non-surgical technique.

ORGANIZATIONS

American Heart Association National Center, 7272

Greenville Avenue, Dallas, TX, 75231, (800) 242-8721,
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Dorothy Elinor Stonely

Paroxysmal supraventricular tachycardia see
Paroxysmal atrial tachycardia

Parrot fever

Definition

Parrot fever is a rare **infectious disease** that causes **pneumonia** in humans. It is transmitted from pet birds or poultry. The illness is caused by a chlamydia, which is a type of intracellular parasite closely related to bacteria. Parrot fever is also called chlamydiosis, psittacosis, or ornithosis.

Description

Parrot fever, which is referred to as avian psittacosis when it infects birds, is caused by *Chlamydia psittaci*. Pet birds in the parrot family, including parrots, parakeets, macaws, and cockatiels, are the most common carriers of the infection. Other birds that may also spread *Chlamydia psittaci* include pigeons, doves, mynah birds, and turkeys. Birds that are carrying the organism may appear healthy, but can shed it in their feces. The symptoms of avian psittacosis include inactivity, loss of appetite and ruffled feathers,

diarrhea, runny eyes and nasal discharge, and green or yellow-green urine. Sick birds can be treated with **antibiotics** by a veterinarian.

Chlamydia psittaci is usually spread from birds to humans through exposure to infected bird feces during cage cleaning or by handling infected birds. In humans, parrot fever ranges in severity from minor flu-like symptoms to severe and life-threatening pneumonia.

Causes and symptoms

Parrot fever is usually transmitted by inhaling dust from dried bird droppings or by handling infected birds. Humans can also spread the disease by person-to-person contact, but that is very rare. The symptoms usually develop within five to 14 days of exposure and include fever, **headache**, chills, loss of appetite, **cough**, and tiredness. In the most severe cases of parrot fever, the patient develops pneumonia. People who work in pet shops or who keep pet birds are the most likely individuals to become infected.

Diagnosis

Only 100–200 cases of parrot fever are reported each year in the United States. It is possible, however, that the illness is more common since it is easily confused with other types of **influenza** or pneumonia. Doctors are most likely to consider a diagnosis of parrot fever if the patient has a recent history of exposure to birds. The diagnosis can be confirmed by blood tests for antibodies, usually complement fixation or immunofluorescence tests. The organism is difficult to culture. A **chest x ray** may also be used to diagnose the pneumonia caused by *Chlamydia psittaci*.

Treatment

Psittacosis is treated with an antibiotic, usually tetracycline (Achromycin, Sumycin); doxycycline (Doxo, Vibramycin); or erythromycin (Eryc, Ilotycin). Oral medication is typically prescribed for at least 10–14 days. Severely ill patients may be given intravenous antibiotics for the first few days of therapy.

Prognosis

The prognosis for recovery is excellent; with antibiotic treatment, more than 99% of patients with parrot fever will recover. Severe infections, however, may be fatal to the elderly, untreated persons, and persons with weak immune systems.

KEY TERMS

Avian chlamydiosis—An illness in pet birds and poultry caused by *Chlamydia psittaci*. It is also known as parrot fever in birds.

Chlamydia psittaci—An organism related to bacteria that infects some types of birds and can be transmitted to humans to cause parrot fever.

Chlamydiosis, psittacosis, or ornithosis—Other names for parrot fever in humans.

Prevention

There is no vaccine that is effective against parrot fever. Birds that are imported into the country as pets should be quarantined to ensure that they are not infected before they can be sold. Health authorities recommend that breeders and importers feed imported birds a special blend of feed mixed with antibiotics for 45 days to ensure that any *Chlamydia psittaci* organisms are destroyed. In addition, bird cages and food and water bowls should be cleaned daily.

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Centers for Disease Control and Prevention, 1600 Clifton Rd., Atlanta, GA, 30333, (404) 639-3311, (800) 311-3435, <http://www.cdc.gov>.

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Partial birth abortion see **Abortion, partial birth**

Partial thromboplastin time

Definition

The partial thromboplastin time (PTT) test is a blood test that is done to investigate bleeding disorders and to monitor patients taking an anticoagulant drug (heparin).

KEY TERMS

Activated partial thromboplastin time—Partial thromboplastin time test that uses activators to shorten the clotting time, making it more useful for heparin monitoring.

Clotting factors—Substances in the blood that act in sequence to stop bleeding by forming a clot.

Coagulation—The process of blood clotting.

Coagulation cascade—The sequence of biochemical activities, involving clotting factors, that stop bleeding by forming a clot.

Common pathway—The pathway that results from the merging of the extrinsic and intrinsic pathways.

The common pathway includes the final steps before a clot is formed.

Extrinsic pathway—One of three pathways in the coagulation cascade.

Heparin—A medication that prevents blood clots.

Intrinsic pathway—One of three pathways in the coagulation cascade.

Partial thromboplastin time—A test that checks the clotting factors of the intrinsic pathway.

Plasma—The fluid part of blood, as distinguished from blood cells.

Purpose

Diagnosis

Blood clotting (coagulation) depends on the action of substances in the blood called clotting factors. Measuring the partial thromboplastin time helps to assess which specific clotting factors may be missing or defective.

Monitoring

Certain surgical procedures and diseases cause **blood clots** to form within blood vessels. Heparin is used to treat these clots. The PTT test can be used to monitor the effect of heparin on a patient's coagulation system.

Precautions

Certain medications besides heparin can affect the results of the PPT test. These include **antihistamines**, vitamin C (ascorbic acid), **aspirin**, and chlorpromazine (Thorazine).

Description

When a body tissue is injured and begins to bleed, it starts a sequence of clotting factor activities called the coagulation cascade, which leads to the formation of a blood clot. The cascade has three pathways: extrinsic, intrinsic, and common. Many of the thirteen known clotting factors in human blood are shared by both pathways; several are found in only one. The PTT test evaluates the factors found in the intrinsic and common pathways. It is usually done in combination with other tests, such as the prothrombin test, which evaluate the factors of the extrinsic pathway.

The combination of tests narrows the list of possible missing or defective factors.

Heparin prevents clotting by blocking certain factors in the intrinsic pathway. The PTT test allows a doctor to check that there is enough heparin in the blood to prevent clotting, but not so much as to cause bleeding. The test is done before the first dose of heparin or whenever the dosage level is changed; and again when the heparin has reached a constant level in the blood. The PTT test is repeated at scheduled intervals.

The PTT test uses blood to which a chemical has been added to prevent clotting before the test begins. About 5 mL of blood are drawn from a vein in the patient's inner elbow region. Collection of the sample takes only a few minutes. The blood is spun in a centrifuge, which separates the pale yellow liquid part of blood (plasma) from the cells. **Calcium** and activating substances are added to the plasma to start the intrinsic pathway of the coagulation cascade. The partial thromboplastin time is the time it takes for a clot to form, measured in seconds.

The test can be done without activators, but they are usually added to shorten the clotting time, making the test more useful for monitoring heparin levels. When activators are used, the test is called activated partial thromboplastin time or APTT.

Test results can be obtained in less than one hour. The test is usually covered by insurance.

Preparation

The doctor should check to see if the patient is taking any of the medications that may influence the test results. If the patient is on heparin therapy, the blood sample is drawn one hour before the next dose of heparin.

Aftercare

Aftercare includes routine care of the puncture site. In addition, patients on heparin therapy must be watched for signs of spontaneous bleeding. The patient should not be left alone until the doctor or nurse is sure that bleeding has stopped. Patients should also be advised to watch for bleeding gums, bruising easily, and other signs of clotting problems; to avoid activities that might cause minor cuts or **bruises**; and to avoid using aspirin.

Risks

The patient may develop a bruise or swelling around the puncture site, which can be treated with moist warm compresses. People with coagulation problems may bleed for a longer period than normal.

Normal results

Normal results vary based on the method and activators used. Normal APTT results are usually between 25–40 seconds; PTT results are between 60–70 seconds. APTT results for a patient on heparin should be 1.5–2.5 times normal values. An APTT longer than 100 seconds indicates spontaneous bleeding.

Abnormal results

Increased levels in a person with a bleeding disorder indicate a clotting factor may be missing or defective. Further tests are done to identify the factor involved. **Liver disease** decreases production of factors, increasing the PTT.

Low levels in a patient on heparin indicate too little heparin is in the blood to prevent clots. High levels indicate too much heparin is present, placing the person at risk of excessive bleeding.

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Paruresis

Definition

The inability to urinate in the presence of others.

Description

Paruresis, also known as shy or bashful bladder, is the inability or difficulty to urinate in the presence of other people, when under time pressure, or on vehicles such as trains or airplanes. Urination is normal when those constraints or factors are absent, typically when in the bathroom at home. Research suggests up to 17 million Americans, 3.25 million Canadians, and 51 million Europeans suffer from the social **anxiety** disorder. Paruresis ranges in intensity from mild, in which the person can urinate in public facilities under certain circumstances, to severe, in which the person can only urinate when alone at home. The condition almost exclusively affects males although it can occur in females.

Paruresis can be socially disabling and can often completely take over a person's life. Examples include avoiding travel, social functions, and sports arenas. Just as serious are the psychological consequences, such as depression, and anxiety. Job choices and career decisions are often adversely affected. People with the condition often avoid jobs where there is mandatory drug testing done by the supervised collection of a urine sample.

Causes and symptoms

Paruretics (people who suffer from paruresis) commonly refer to three triggers that influence them when in public restrooms. For the typical paruretic, these triggers must be removed, or the person must try another toilet, for urination to occur on a particular occasion. First, the condition occurs much more frequently when strangers are present in the restroom as opposed to friends or relatives. Second, proximity plays a role in the problem. Proximity for the paruretic is both physical, involving the relative closeness of others in or near the restroom, and psychological, involving the need for privacy. The most frequent complaint about physical stimuli in public facilities is the absence of suitable partitions and doors on urinals or stalls. Third, temporary psychological states, especially anxiety, anger, and fear can interfere with urination.

Diagnosis

The condition is diagnosed on the basis of the sufferer's account of their symptoms. In severe cases,

sufferers can waste considerable time waiting for everyone else to leave the toilet before they can urinate, and might totally avoid urinating in public toilets. The condition is usually self-diagnosed when any or all of the three main triggers of paruresis are present and the condition is chronic.

Treatment

The most well documented current treatment is based upon **cognitive-behavioral therapy**, of which the aim is to reorganize the “abnormal” emotional schemes arising from the anxiety generating elements that trigger this problem. This can be done individually in a self-help situation, in a support group, or through **psychotherapy** with a psychologist or psychiatrist.

Therapy includes three separate but linked components:

- Cognitive—An attempt to modify the abnormal thoughts and ideas around the object of anxiety, such as the thought, “When I use a toilet, everybody looks at me and wonders what I’m doing.”
- Behavioral—Step by step desensitization by very gradual exposure to the feared situation, the aim being to achieve a series of small successes, and thus reassure the subconscious mind that it is “safe” to urinate in a situation that previously led to panic and failure. This can be thought of as relearning urination in a social situation.
- Relaxation—Learning techniques that facilitate relaxation, both mental and physical, such as sphincter relaxation exercises.

Drug treatments, usually with medications used to treat benign prostate hyperplasia (BPH), an enlargement of the prostate gland, such as *terazosin* (Hytrin), *tamsulosin* (Flomax), and *alfuzosin* (Uroxatral) are the subject of much debate and usually produce poor results.

Alternative treatment

One possible alternative medicine treatment is **saw palmetto**, used to treat urinary problems in men with BPH, an enlargement of the prostate gland. BPH results in a swelling of the prostate gland that obstructs the urethra. This causes painful urination, reduced urine flow, difficulty starting or stopping the flow, dribbling after urination, and more frequent nighttime urination. A typical dose is 320 mg per day of standardized extract. It may take up to four weeks of use before beneficial effects are seen.

KEY TERMS

Benign prostate hyperplasia (BPH)—Enlargement of the prostate gland.

Psychotherapy—The treatment of mental disorders by psychological methods, usually by a psychiatrist or psychologist.

Sphincter—A circular band of muscle that surrounds an opening or passage in the body and narrows or closes the opening by contracting.

Urethra—The tube in humans that carries urine from the bladder out of the body.

Prognosis

Most people who suffer from the condition never seek help or treatment. Many never even discuss the problem with anyone. But anecdotal evidence suggests that those who do seek help have a good success rate at overcoming their fear or anxiety over time, sometimes a year or longer.

Prevention

There is no known way to prevent a person from developing paruresis. Anecdotal evidence suggests it often does not occur until around the age of **puberty**. One suggestion for prevention is to condition children from an early age to urinate in public restrooms.

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Parvovirus B19 infection see **Fifth disease**

Pasteurellosis see **Animal bite infections**

Patau syndrome

Definition

Patau syndrome, also called trisomy 13, is a congenital (present at birth) disorder associated with the presence of an extra copy of chromosome 13. The extra chromosome 13 causes numerous physical and mental abnormalities, especially heart defects. Patau syndrome is named for Dr. Klaus Patau, who reported the syndrome and its association with trisomy in 1960. It is sometimes called Bartholin-Patau syndrome, named in part for Thomas Bartholin, a French physician who described an infant with the syndrome in 1656.

Description

Children normally inherit 23 chromosomes from each parent, for a total of 46 chromosomes. A typical human being has 46 chromosomes: 22 pairs of non-sex linked chromosomes and one pair of sex-linked chromosomes, that determine that child's sex. Sometimes a child may end up with more than 46 chromosomes because of problems with the father's sperm or the mother's egg; or, because of mutations that occurred after the sperm and the egg fused to form the embryo (conception).

Normally, there are two copies of each of the 23 chromosomes: one from each parent. A condition called trisomy occurs when three, instead of two, copies of a chromosome are present in a developing human embryo. An extra copy of a particular chromosome can come either from the egg or sperm, or because of mutations that occur after conception.

The best-known trisomy-related disorder is **Down syndrome** (trisomy 21), in which the developing embryo has an extra copy of chromosome 21. Patau syndrome is trisomy 13, in which the developing embryo has three copies of chromosome 13.

An extra copy of chromosome 13 is not the only cause of Patau syndrome. Other changes in chromosome 13, such as mispositioning (translocation), can also result in the characteristics classified as Patau syndrome. In these cases, an error occurs that causes a portion of chromosome 13 to be exchanged for a portion of another chromosome. There is no production of extra chromosomes, but a portion of each affected chromosome is "misplaced" (translocated) to another chromosome.

Patau syndrome causes serious physical and mental abnormalities including: heart defects; incomplete brain development; such unusual facial features as a

sloping forehead, a smaller than average head (microcephaly), small or missing eyes, low-set ears, and **cleft palate** or hare lip; extra fingers and toes (**polydactyly**); abnormal genitalia; spinal defects; seizures; gastrointestinal hernias, particularly at the navel (omphalocele); and **mental retardation**. Due to the severity of these conditions, fewer than 20% of those affected with Patau syndrome survive beyond infancy. Most infants with the syndrome die within the first three months of life; the average life expectancy of the survivors is about 10 years.

Genetic profile

When an extra copy (trisomy) of a chromosome is made, it may either be a total trisomy (in which an extra copy of the entire chromosome is made), or partial trisomy (in which only one part of the chromosome is made an extra time).

In most cases of trisomy, errors in chromosome duplication occur at conception because of problems with the egg or the sperm that are coming together to produce an offspring. In these cases, every cell in the body of the offspring has an extra copy of the affected chromosome. However, errors in chromosome duplication may also occur during the rapid cell division that takes place immediately after conception. In these cases, only some cells of the body have the extra chromosome error. The condition in which only some of the cells in the body have the extra chromosome is called mosaicism.

Seventy-five to 80 percent of the cases of Patau syndrome are caused by a trisomy of chromosome 13. Some of these cases are the result of a total trisomy, while others are the result of a partial trisomy. Partial trisomy generally causes less severe physical symptoms than full trisomy. Ten percent of these cases are of the mosaic type, in which only some of the body's cells have the extra chromosome. The physical symptoms of the mosaic form of Patau syndrome depends on the number and type of cells that carry the trisomy.

Most cases of trisomy are not passed on from one generation to the next. Usually they result from a malfunction in the cell division (mitosis) that occurs after conception. At least 75% of the cases of Patau syndrome are caused by errors in chromosome replication that occur after conception. The remaining 25% are caused by the inheritance of translocations of chromosome 13 with other chromosomes within the parental chromosomes. In these cases, a portion of another chromosome switches places with a portion of chromosome 13. This leads to errors in the genes on

both chromosome 13 and the chromosome from which the translocated portion originated.

Patau syndrome occurs in approximately one in 8,000–12,000 live births in the United States. In many cases, spontaneous abortion (**miscarriage**) occurs and the fetus does not survive to term. In other cases, the affected individual is stillborn. As appears to be the case in all trisomies, the risks of Patau syndrome seem to increase with the mother's age, particularly if she is over 30 when pregnant. Male and female children are equally affected, and the syndrome occurs in all races and ethnic groups. Females with Patau syndrome, however, have a better chance of surviving past infancy than males.

Causes and symptoms

The severity and symptoms of Patau syndrome vary with the type of chromosomal anomaly, from extremely serious conditions to nearly normal appearance and functioning.

Full trisomy 13, which is present in the majority of the cases, results in the most severe and numerous internal and external abnormalities. Commonly, the forebrain fails to divide into lobes or hemispheres (holoprosencephaly) and the entire head is unusually small (microcephaly). The spinal cord may protrude through a defect in the vertebrae of the spinal column (myelomeningocele). Children who survive infancy have profound mental retardation and may experience seizures. In a few rare cases Patau syndrome may coexist with Klinefelter's syndrome or other chromosomal abnormalities.

Incomplete development of the optic (sight) and olfactory (smell) nerves often accompany the brain defects described above. The eyes may be unusually small (**microphthalmia**) or one eye may be absent (**anophthalmia**). The eyes are sometimes set close together (hypotelorism) or even fused into a single structure. Incomplete development of any structures in the eye (coloboma) or failure of the retina to develop properly (retinal dysplasia) will also produce vision problems. Patau syndrome affected individuals may be born either partially or totally deaf and many are subject to recurring ear infections.

The facial features of many Patau syndrome-affected individuals appear flattened. The ears are generally malformed and lowset. Frequently, a child with trisomy 13 has a **cleft lip**, a cleft palate, or both. Other physical characteristics include loose folds of skin at the back of the neck, extra fingers or toes (polydactyly), permanently flexed (closed) fingers (camptodactyly), noticeably prominent heels, "rocker-bottom" foot,"

and missing ribs. Genital malformations are common in individuals affected with Patau syndrome and include undescended testicles (cryptorchidism), an abnormally developed scrotum, and ambiguous genitalia in males, or an abnormally formed uterus (bicornuate uterus) in females.

In nearly all cases, Patau syndrome affected infants have respiratory difficulties and heart defects, including atrial and ventricular septal defects (holes between chambers of the heart); malformed ducts that cause abnormal direction of blood flow (**patent ductus arteriosus**); holes in the valves of the lungs and the heart (pulmonary and aortic valves); and misplacement of the heart in the right, rather than the left, side of the chest (dextrocardia). The kidneys and gastrointestinal system may also be affected with cysts similar to those seen in **polycystic kidney disease**. These defects are frequently severe and life-threatening.

Partial trisomy of the distal segment of chromosome 13 results in generally less severe, but still serious, symptoms and a distinctive facial appearance including a short upturned nose, a longer than usual area between the nose and upper lip (philtrum), bushy eyebrows, and tumors made up of blood capillaries on the forehead (frontal capillary hemangioma). Partial trisomy of the proximal segment of chromosome 13 is much less likely to be fatal and has been associated with a variety of facial features including a large nose, a short upper lip, and a receding jaw. Both forms of partial trisomy also result in severe mental retardation.

Beyond one month of age, other symptoms that are seen in individuals with Patau syndrome are: feeding difficulties and **constipation**, reflux disease, slow growth rates, curvature of the spine (**scoliosis**), irritability, sensitivity to sunlight, low muscle tone, high blood pressure, sinus infections, urinary tract infections, and ear and eye infections.

Diagnosis

Patau syndrome is detectable during **pregnancy** through the use of ultrasound imaging, **amniocentesis**, and **chorionic villus sampling** (CVS). At birth, the newborn's numerous malformations indicate a possible chromosomal abnormality. Trisomy 13 is confirmed by examining the infant's chromosomal pattern through karyotyping or another procedure. Karyotyping involves the separation and isolation of the chromosomes present in cells taken from an individual. These cells are generally extracted from cells found in a blood sample. The 22 non-sex linked chromosomes are identified by size, from largest to smallest, as chromosomes 1 through 22. The sex-determining chromosomes are also

KEY TERMS

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman’s abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Chorionic villus sampling (CVS)—A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother’s vagina or abdominal wall and a sample of cells is collected from around the fetus. These cells are then tested for chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the body consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Changes in either the total number of chromosomes or their shape and size (structure) may lead to physical or mental abnormalities.

identified. The diagnosis of Patau syndrome is confirmed by the presence of three, rather than the normal two, copies of the thirteenth largest chromosome.

A newer method of diagnosing trisomies that has the advantages of speed and lower cost is the quantitative fluorescent PCR (QF-PCR) assay. QF-PCR testing allows a doctor to determine the presence of a chromosomal abnormality within 24 hours with a very high degree of accuracy.

Treatment

Some infants born with Patau syndrome have severe and incurable **birth defects**. However, children with better prognoses require medical treatment to correct structural abnormalities and associated complications. For feeding problems, special formulas, positions, and techniques may be used. Tube feeding or the placement of a gastric tube (**gastrostomy**) may be required. Structural abnormalities such as cleft lip and cleft palate can be corrected through surgery. Special **diets**, **hearing aids**, and vision aids can be used to mitigate the symptoms of Patau syndrome. **Physical therapy**, **speech therapy**, and other types of developmental therapy will help the child reach his or her potential.

Karyotyping—A laboratory procedure in which chromosomes are separated from cells, stained, and arranged so that their structure can be studied under the microscope.

Mosaicism—A genetic condition resulting from a mutation, crossing over, or nondisjunction of chromosomes during cell division, causing a variation in the number of chromosomes in the cells.

Translocation—The transfer of one part of a chromosome to another chromosome during cell division. A balanced translocation occurs when pieces from two different chromosomes exchange places without loss or gain of any chromosome material. An unbalanced translocation involves the unequal loss or gain of genetic information between two chromosomes.

Trisomy—The condition of having three identical chromosomes instead of the normal two in a cell.

Ultrasound—An imaging technique that uses sound waves to help visualize internal structures in the body.

Since the translocation form of Patau syndrome is genetically transmitted, **genetic counseling** for the parents should be part of the management of the disease.

Prognosis

Approximately 45% of trisomy 13 babies die within their first month of life; up to 70% in the first six months; and over 70% by one year of age. Survival to adulthood is very rare. Only one adult is known to have survived to age 33.

Most survivors have profound mental and physical disabilities; however, the capacity for learning in children with Patau syndrome varies from case to case. Older children may be able to walk with or without a walker. They may also be able to understand words and phrases, follow simple commands, use a few words or signs, and recognize and interact with others.

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ORGANIZATIONS

- National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.
- Rainbows Down Under—A Trisomy 18 and Trisomy 13 Resource. SOFT Australia., 198 Oak Road, Kirrawee, Australia, NSW 2232, 029521-6031, SOFTAus@optushome.com.au, <http://members.optushome.com.au>.
- Support Organization for Trisomy 18, 13, and Related Disorders (SOFT), 2982 South Union Street, Rochester, NY, 14624, (585) 594-4621, (800) 716-7638, barbsoft@rochester.rr.com, <http://www.trisomy.org>.

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Patent ductus arteriosus

Definition

Patent ductus arteriosus (PDA) is a heart defect that occurs when the ductus arteriosus (the temporary fetal blood vessel that connects the aorta and the pulmonary artery) does not close at birth.

Description

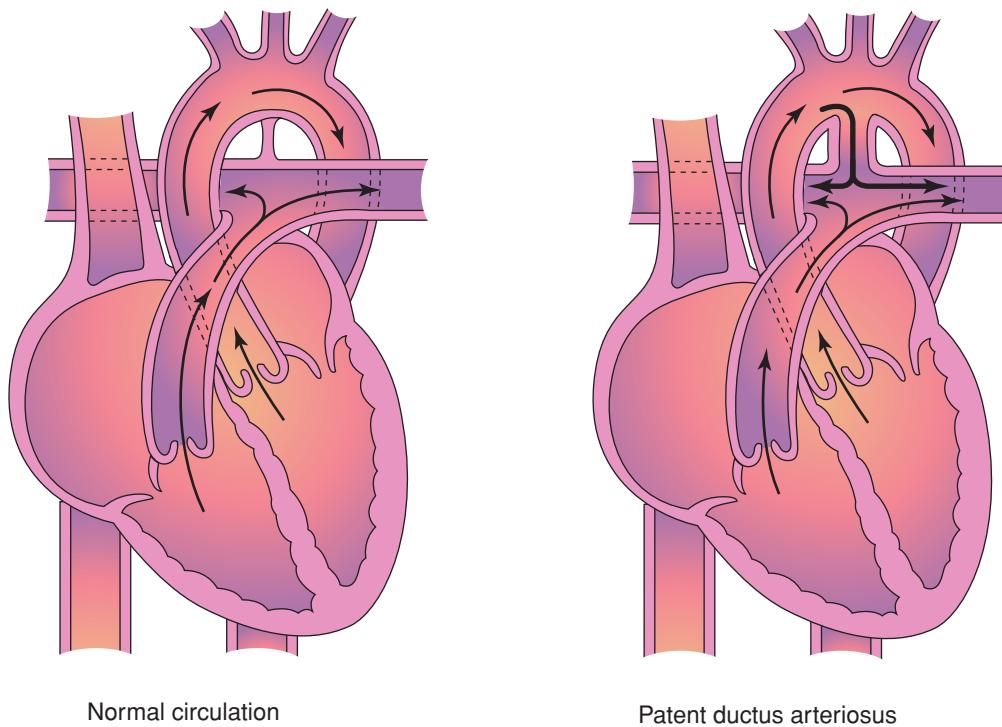
The ductus arteriosus is a temporary fetal blood vessel that connects the aorta and the pulmonary artery before birth. The ductus arteriosus should be

present and open before birth while the fetus is developing in the uterus. Since oxygen and nutrients are received from the placenta and the umbilical cord instead of the lungs, the ductus arteriosus acts as a "short cut" that allows blood to bypass the deflated lungs and go straight out to the body. After birth, when the lungs are needed to add oxygen to the blood, the ductus arteriosus normally closes. The closure of the ductus arteriosus ensures that blood goes to the lungs to pick up oxygen before going out to the body. Closure of the ductus arteriosus usually occurs at birth as levels of certain chemicals, called prostaglandins, change and the lungs fill with air. If the ductus arteriosus closes correctly, the blood pumped from the heart goes to the lungs, back into the heart, and then out to the body through the aorta. The blood returning from the lungs and moving out of the aorta carries oxygen to the cells of the body.

In some infants, the ductus arteriosus remains open (or patent) and the resulting heart defect is known as patent ductus arteriosus. In most cases, a small PDA does not result in physical symptoms. If the PDA is larger, health complications may occur.

In an average individual's body, the power of blood being pumped by the heart and other forces leads to a certain level of pressure between the heart and lungs. The pressure between the heart and lungs of an individual affected by PDA causes some of the oxygenated blood that should go out to the body (through the aorta) to return back through the PDA into the pulmonary artery. The pulmonary artery takes the blood immediately back to the lungs. The recycling of the already oxygenated blood forces the heart to work harder as it tries to supply enough oxygenated blood to the body. In this case, usually the left side of the heart grows larger as it works harder and must contain all of the extra blood moving back into the heart. This is known as a left-to-right or aortic-pulmonary shunt.

As noted, the size of the PDA determines how much harder the heart has to work and how much bigger the heart becomes. If the PDA is large, the bottom left side of the heart is forced to pump twice as much blood because it must supply enough blood to recycle back to the lungs and move out to the body. As the heart responds to the increased demands for more oxygenated blood by pumping harder, the pulmonary artery has to change in size and shape in order to adapt to the increased amount and force of the blood. In some cases, the increase in size and shape changes the pressure in the pulmonary artery and lungs. If the pressure in the lungs is higher than that of the heart



Patent ductus arteriosus (PDA) is the failure of the ductus arteriosus to close after birth, allowing blood to inappropriately flow from the aorta into the pulmonary artery. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

and body, blood returning to the heart will take the short cut back into the aorta from the pulmonary artery through the PDA instead of going to the lungs. This backward flowing of blood does not carry much oxygen. If blood without much oxygen is being delivered to the body, the legs and toes will turn blue or cyanotic. This is called a shunt reversal.

When a PDA results in a large amount of blood being cycled in the wrong order, either through a left-to-right shunt or shunt reversal, the overworked, enlarged heart may stop working (congestive **heart failure**) and the lungs can become filled with too much fluid (**pulmonary edema**). At this time, there is also an increased risk for a bacterial infection that can inflame the lining of the heart (**endocarditis**). These three complications are very serious.

PDA is a very common heart defect. Though an exact incidence of PDA is difficult to determine, one review in 1990 found that approximately 8% of live births were found to be affected by PDA. PDA can occur in full-term infants, but it seen most frequently in preterm infants, infants born at a high altitude, and babies whose mothers were affected by the German

measles (rubella) during **pregnancy**. PDA is two to three times more common in females than males. PDA occurs in individuals of every ethnic origin and does not occur more frequently in any one country or ethnic population.

Causes and symptoms

PDA can be a result of an environmental exposure before birth, inheriting a specific changed or mutated gene or genes, a symptom of a genetic syndrome, or be caused by a combination of genetic and environmental factors (multifactorial).

Environmental exposures that can increase the chance for a baby to be affected by PDA include fetal exposure to rubella before birth, preterm delivery, and birth at a high altitude location.

PDA can be an inherited condition running in families as isolated PDA or part of a genetic syndrome. In either case, there are specific gene changes or mutations which lead to a defect in the elastic tissue forming the walls of the ductus arteriosus. The genes causing isolated PDA have not been identified, but it is

KEY TERMS

Aorta—The main artery located above the heart which pumps oxygenated blood out into the body. Many congenital heart defects affect the aorta.

Catheterization—The process of inserting a hollow tube into a body cavity or blood vessel.

Ductus arteriosus—The temporary channel or blood vessel between the aorta and pulmonary artery in the fetus.

Echocardiograph—A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

Electrocardiogram (ECG, EKG)—A test used to measure electrical impulses coming from the heart

in order to gain information about its structure or function.

Endocarditis—A dangerous infection of the heart valves caused by certain bacteria.

Oxygenated blood—Blood carrying oxygen through the body.

Pulmonary artery—An artery that carries blood from the heart to the lungs.

Pulmonary edema—A problem caused when fluid backs up into the veins of the lungs. Increased pressure in these veins forces fluid out of the vein and into the air spaces (alveoli). This interferes with the exchange of oxygen and carbon dioxide in the alveoli.

known that PDA can be inherited through a family in an autosomal dominant pattern or an autosomal recessive pattern.

Every person has approximately 30,000 genes, which tell our bodies how to grow and develop correctly. Each gene is present in pairs since one is inherited from our mother, and one is inherited from our father. In an autosomal dominant condition, only one specific changed or mutated copy of the gene for PDA is necessary for a person to have PDA. If a parent has an autosomal dominant form of PDA, there is a 50% chance for each child to have the same or similar condition.

PDA can also be inherited in an autosomal recessive manner. A recessive condition occurs when a child receives two changed or mutated copies of the gene for a particular condition, such as PDA (one copy from each parent). Individuals with a single changed or mutated copy of a gene for a recessive condition, are known as “carriers,” and have no health problems related to the condition. In fact, each of us carries between five and 10 genes for harmful, recessive conditions. However, when two people who each carry a changed or mutated copy of the same gene for a recessive condition meet, there is a chance, with each pregnancy, for the child to inherit the two changed or mutated copies from each parent. In this case, the child would have PDA. For two known carriers, there is a 25% risk with each child to have a child with PDA, a 50% chance to have a child who is a carrier, and a 25% chance to have a child who is neither affected nor a carrier.

Most cases of PDA occur as the result of multifactorial inheritance which is caused by the combination of genetic factors and environmental factors. The

combined factors lead to isolated defects in the elastic tissue forming the walls of the ductus arteriosus. Family studies can provide different recurrence risks depending on the family member affected by multifactorial PDA. If an individual is affected by isolated, multifactorial PDA, they have a 2–4% chance of having a child affected by PDA. If a couple has one child with isolated, multifactorial PDA, there is a 3% chance that another of their children could be affected by PDA. If a couple has two children affected by isolated, multifactorial PDA, there is a 10–25% chance that they could have another child affected by PDA.

Unless a specific pattern of inheritance, preterm delivery, or known exposure is found through the examination of a detailed pregnancy and family history, the multifactorial family studies are used to estimate the possible risk of recurrence of PDA in a family.

The main sign of PDA is a constant heart murmur that sounds like the hum of a refrigerator or other machinery. This murmur is usually heard by the doctor using a stethoscope. Otherwise, there are no specific symptoms of PDA, unless the ductus arteriosus size is large. Children and adults with a large ductus arteriosus can show difficulty in breathing during moderate physical **exercise**, an enlarged heart, and failure to gain weight. In some cases, heart failure and pulmonary congestion can indicate a PDA.

Diagnosis

Diagnosis is most often made by detecting the characteristic “machinery” heart murmur heard by a

doctor through a stethoscope. Tests such as a **chest x-ray**, echocardiograph, and ECG are used to support the initial diagnosis. Other indications of PDA include failure to gain weight, frequent chest infections, heavy breathing during mild physical exertion, congestive heart failure, and pulmonary **edema**. Prenatal ultrasounds are unable to detect PDA because the heart defect does not occur until the time of birth.

Treatment

The treatment and management of PDA depends upon the size of the PDA and symptoms being experienced by the affected individual. In some cases, a PDA can correct itself in the first months of life. In preterm infants experiencing symptoms, the first step in correcting a PDA is treatment through medications such as indomethacin. In preterm infants whose PDA is not closed through medication, full term infants affected by PDA, and adults, surgery is an option for closing the ductus arteriosus. In 2000 and 2001, researchers developed and reviewed alternatives to surgical closure such as interventional **cardiac catheterization** and video-assisted thorascopic surgical repair. A cardiologist can help individuals determine the best method for treatment based on their physical symptoms and medical history.

Prognosis

Adults and children can survive with a small opening remaining in the ductus arteriosus. Treatment, including surgery, of a larger PDA is usually successful and frequently occurs without complications. Proper treatment allows children and adults to lead normal lives.

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American Society of Cataract and Refractive Surgery, 2112 North Wilkins Road, Swanton, OH, 43558, (419) 825-5575, (419) 825-2880, CHASER@compuserve.com, <http://www.csun.edu>.

Kids With Heart, National Association for Children’s Heart Disorders, Inc., P.O. Box 12504, Green Bay, WI, 54307-2504, (920) 498-0058, (800) 538-5390, michelle@kidswithheart.org, <http://www.kidswithheart.org/>.

Dawn A. Jacob

PCV see **Hematocrit**

Pediculosis see **Lice infestation**

Pedophilia see **Sexual perversions**

Pellagra

Definition

Pellagra is a disorder brought on by a deficiency of the nutrient called niacin or nicotinic acid, one of the B-complex **vitamins**.

Description

Nicotinic acid plays a crucial role in the cellular process called respiration. Respiration is the process by which nutrients (specifically sugar, or glucose) and oxygen are taken in, chemical reactions take place, energy is produced and stored, and carbon dioxide and wastes are given off. This process is absolutely central to basic cell functioning, and thus the functioning of the body as a whole.

Niacin is a B vitamin found in such foods as yeast, liver, meat, fish, whole-grain cereals and breads, and legumes. Niacin can also be produced within the body from the essential amino acid called tryptophan. Dietary requirements for niacin depend on the age, gender, size, and activity level of the individual. Niacin requirements range from 5 mg in infants up to 20 mg in certain adults.

Causes and symptoms

Pellagra can be either primary or secondary. Primary pellagra results when the diet is extremely deficient in niacin-rich foods. A classic example occurs in geographic locations where Indian corn (maize) is the dietary staple. Maize does contain niacin, but in a form which cannot be absorbed from the intestine (except when it has been treated with alkali, as happens in the preparation of tortillas). People who rely on maize as their major food source often develop pellagra. Pellagra can also occur when a hospitalized patient, unable to eat for a very prolonged period of time, is given fluids devoid of vitamins through a needle in the vein (intravenous or IV fluids).

Secondary pellagra occurs when adequate quantities of niacin are present in the diet, but other diseases or conditions interfere with its absorption and/or processing. This is seen in various diseases that cause prolonged **diarrhea**, with **cirrhosis** of the liver and **alcoholism**, with long-term use of the anti-tuberculosis drug called isoniazid, in patients with malignant carcinoid tumor, and in patients suffering from **Hartnup disease** (an inherited disorder which results in disordered absorption of amino acids from the intestine and kidney).

Pellagra causes a variety of symptoms affecting the skin; mucous membranes (moist linings of the mouth, organs, etc.); central nervous system (including the brain and nerves); and the gastrointestinal system. The classic collection of symptoms includes redness and swelling of the mouth and tongue, diarrhea, skin rash, and abnormal mental functioning, including **memory loss**. While early patients may simply have a light skin rash, over time the skin becomes increasingly thickened, pigmented, and may slough off in places. Areas of the skin may become prone to bacterial infection. The mouth and tongue, and sometimes the vagina, become increasingly thick, swollen, and red. Abdominal **pain** and bloating occur, with **nausea and vomiting**, and bloody diarrhea to follow. Initial mental changes appear as inability to sleep (**insomnia**), **fatigue**, and a sense of disconnectedness (apathy). These mental changes progress to memory loss, confusion, depression, and **hallucinations** (in which the individual sees sights or hears sounds that do not really exist). The most severe states include stiffness of the arms and legs, with resistance to attempts to move the limbs; variations in level of consciousness; and the development of involuntary sucking and grasping motions. This collection of symptoms is called “encephalopathic syndrome.”

Diagnosis

Diagnosis is purely based on the patient’s collection of symptoms, together with information regarding the patient’s diet. When this information points to niacin deficiency, replacement is started, and the diagnosis is then partly made by evaluating the patient’s response to increased amounts of niacin. There are no chemical tests available to definitively diagnose pellagra.

Treatment

Treatment of pellagra usually involves supplementing the individual’s diet with a form of niacin called niacinamide (niacin itself in pure supplementation form causes a number of unpleasant side effects, including sensations of **itching**, burning, and flushing). The niacinamide can be given by mouth (orally) or by injection

KEY TERMS

Niacinamide—A form of niacin, which is usually used as a dietary supplement for people with insufficient niacin.

Respiration—Respiration is the process by which nutrients (specifically sugar, or glucose) and oxygen are taken in to a cell; chemical reactions take place; energy is produced and stored; and carbon dioxide and wastes are given off.

(when diarrhea would interfere with its absorption). The usual oral dosage is 300–500 mg each day; the usual dosage of an injection is 100–250 mg, administered two to three times each day. When pellagra has progressed to the point of the encephalopathic syndrome, a patient will require 1,000 mg of niacinamide orally, and 100–250 mg of niacinamide by injection. Once the symptoms of pellagra have subsided, a maintenance dose of niacin can be calculated, along with attempting (where possible) to make appropriate changes in the diet. Because many B-complex vitamin deficiencies occur simultaneously, patients will usually require the administration of other B-complex vitamins as well.

Prognosis

Untreated pellagra will continue progressing over the course of several years, and is ultimately fatal. Often, **death** is due to complications from infections, massive **malnutrition** brought on by continuous diarrhea, blood loss due to bleeding from the gastrointestinal tract, or severe encephalopathic syndrome.

Prevention

Prevention of pellagra is completely possible; what is required is either a diet adequate in niacin-rich foods, or appropriate supplementation. However, in many geographic locations in the world such foods are unavailable to the general population, and pellagra becomes an unavoidable complication of poverty.

ORGANIZATIONS

American Dietetic Association, 120 S. Riverside Plaza, Suite 2000, Chicago, IL, 60606-6995, (312) 899-0040, (800) 877-1600, <http://www.eatright.org/>.

Rosalyn Carson-DeWitt, MD

Pelvic endoscopy see **Laparoscopy**

Pelvic exam

Definition

A pelvic examination is a routine procedure used to assess the well being of the female patient's lower genito-urinary tract. This is done as part of a usual health screening and prevention tool, and is an element of the total health care for the female patient.

Purpose

Pelvic exams are useful as a screening tool for **sexually transmitted diseases** such as **gonorrhea**, **chlamydia**, **genital warts**, **herpes**, and **syphilis**. In addition, exams detect some forms of **cancer** that may affect the genitalia. By analyzing the cervical region with a Pap-nicolaou or Pap smear, clinicians are able to look for signs of **cervical cancer**. The American Cancer Society and the American College of Obstetricians and Gynecologists recommend pelvic exams with Pap tests for women starting at age 18. It is also recommended that exams start earlier if the teenager requests oral **contraception**. Pap smears should continue once yearly for three years and at the physician's discretion following this time. Various groups differ in opinions on when to discontinue screening for cervical cancer, however, the United States Preventative Services Task Force recommends screening continue until age 65 if the patient has not had previous abnormal results. Women who have undergone a total **hysterectomy** for reasons other than cervical cancer do not need to be screened.

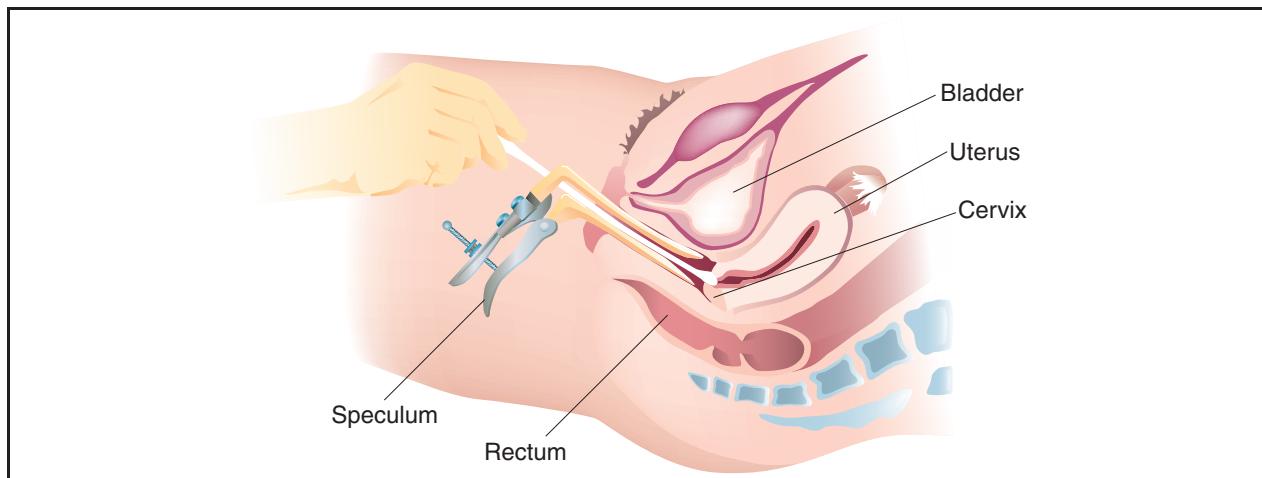
Precautions

Pelvic examinations are safe procedures, thus no precautions are necessary.

Description

The first part of the examination involves visual inspection and palpation of the external genitalia. The examiner will note the characteristics of the labia majora, labia minora, clitoris, urethral orifice, and the Skene's and Bartholin's glands. In addition, the perineum and anus will be checked. The clinician will be examining these areas for any indication of swelling, inflammation, abnormal discharge, polyps, abnormal odor, or other lesions.

The next part involves examining the internal genitalia. The examiner will first insert a gloved finger into the vagina in order to palpate the cervix. Next an instrument called a speculum is inserted. This device is made of plastic or metal and used to open the vaginal cavity in order for the examiner to be able to view the vaginal walls and cervix. Any lesions, bleeding, or abnormal discharge can be visualized with the speculum in place. If indicated, a Pap smear will then be performed. With the speculum still in place, the examiner gently scrapes the patient's cervix with a wooden or plastic spatula as well as a cylindrical-type brush. The spatula collects cells from the outer surface of the cervix, while the brush is used to collect cells from the inner-cervix. The collected cells are then spread on a glass slide, sprayed with a fixative, and sent to a laboratory for analysis. The examiner may then insert a cotton or Dacron swab



During a pelvic exam, cells from the cervix are scraped on a spatula and are tested for abnormalities. ((Illustration by Argosy, Inc. Reproduced by permission of Gale, a part of Cengage Learning.)

KEY TERMS

Bacterial vaginitis—This is the term for inflammation of the vagina due to a bacterial infection.

Bartholin's glands—These glands are embedded in the vestibule of the vagina and function to maintain moisture.

Cervical dysplasia—Dysplasia is the abnormal growth of the epithelial cells. This is what a Pap smear will detect in the cervix.

Colposcopy—This procedure is done when a Pap smear reveals abnormal results. With an endoscope placed through the vagina and into the cervix, a physician can determine exactly where lesions of the cervix are.

Hematoma—Hematomas are masses of blood (or clotted blood) that accumulate in tissues and may result from trauma.

Myoma—These are benign (non-malignant) tumors of the uterus.

Papanicolaou or Pap smear—This is a screening test for cancer of the cervix. Cells are scraped from the cervix, smeared on a glass plate, and sent to a laboratory to examine for any abnormal cells or dysplasia. This test may also detect other cells seen in certain vaginal infections.

Skene's glands—These are the glands of the female urethra.

Speculum—A speculum is an instrument that is used during the internal genitalia examination. It can be made of plastic or metal and is used to open up the vaginal cavity in order for the examiner to view the walls of the vagina and the cervix.

Urethral meatus—This is the external opening of the urethra.

into the cervix. This will be held in place for 10–30 seconds and when withdrawn spread on a plastic plate or into a tube containing a reagent for the specimen. This procedure may be repeated again with the anus. Such swab tests are used to check for gonorrhea and chlamydia, or bacterial vaginitis, which is a bacterial infection resulting in inflammation of the vagina.

Following the Pap smear is the bimanual examination during which the examiner will place an index and middle finger into the vagina to first examine the vaginal walls for any irregularities or tenderness. The cervix will then be palpated in order to note its shape, consistency, mobility, and any tenderness. The examiner will then place his or her other hand on the abdomen and gently push down while pushing the cervix up. This is done to assess the size and shape of the uterus, and also to note any tenderness or abnormal lesions. During this time, the ovaries are also checked for any masses, or tenderness.

The last part of the pelvic exam is the rectovaginal examination. This allows the clinician to better examine the pelvic organs and structures. The examiner will place their index finger into the vagina and a lubricated, gloved middle finger against the anus. During this part, the patient may feel an urgency to have a bowel movement. However, this is a natural feeling and a bowel movement will not occur. The patient will then be asked to strain down in order for the anal sphincter to relax. As

relaxation occurs the examiner will insert the middle finger into the rectum, enabling the position and shape of the uterus to be better assessed. In addition, any masses or tenderness can be evaluated at this point. The anal canal and rectum can also be examined for any polyps, or other lesions at this time. After the rectovaginal exam, the patient will be allowed clean off any excess lubricant and get dressed. The examiner will then discuss the procedure and any findings with the patient.

Preparation

Pelvic exams require the patient to void prior to starting, as a full bladder can add to discomfort and make palpation difficult for the examiner. Even though some tests cannot be done on a menstruating patient, an examination can still be performed. Any tampons should be removed prior to the exam. Douching is not recommended before an exam due to the hazard of washing away cells that are needed for examination. If a Pap smear is to be done, the patients should also refrain from sexual intercourse or using vaginal suppositories for 24 to 48 hours prior to the exam. The patient will be asked to undress and put on a gown. The examiner will instruct the patient to lie on the examination table on her back and may assist her in putting her feet in stirrups. The buttocks are then slid to the edge of the table in order for a full view of the area to be examined.

Aftercare

Even with the invasiveness of this procedure, the patient should be able to immediately resume normal daily activities.

Risks

Other than minor discomfort, there are no risks associated with a routine pelvic examination.

Normal results

No significant findings by the examiner indicate a normal pelvic examination. The external and internal genitalia will be free of any lesions or abnormal discharge. The Pap smear will not reveal cervical dysplasia or abnormal tissue development, and there will not be any abnormal masses or tenderness upon palpation.

Abnormal results

The examiner may discover abnormal lesions during the course of the exam that may require additional tests. Ulcerations, bumps, sores, blisters, or vesicles on the external genitalia may be signs of a sexually transmitted disease. Some of the sexually transmitted diseases that may cause lesions to the external genitalia include venereal **warts**, syphilis, and **genital herpes**. Gonorrhea or chlamydia may also cause inflammation to the urethral meatus or the external opening of the urethra. These, in addition to bacterial infections, can also cause inflammation of the Skene's glands, Bartholin's glands, and vulva. Infections may result in an irritating discharge. Discharge may also be noted in yeast infections. Other abnormal findings of the external genitalia include carcinomas, vulvar tumors, or hematomas. Hematomas are masses of accumulated blood that appears as a bluish swelling of the labium that may occur following trauma to this area. Examination of the internal genitalia may reveal similar findings in regards to sexually transmitted diseases and carcinomas. Cervical abnormalities can also be found and may include lacerations, infections, ulcers, cysts, and polyps. All of these will require further evaluation in order to determine the underlying cause.

Since Pap smears screen for cervical cancer, abnormal results require special attention. Due to the incidence of false-positives or false-negatives, the test may be repeated or the physician may choose to have the patient undergo a **colposcopy**. This procedure uses an endoscope and will examine the vagina and cervix

in more depth. This will identify 100% of lesions present. A biopsy may then be taken of the lesion in order to determine the exact type of abnormality. Several new techniques are now available that improve the accuracy of the Pap smear including automated analysis machines. Bimanual and rectovaginal exams may reveal abnormalities of the uterus or other pelvic structures. One commonly encountered finding is a myoma, which is a benign uterine tumor. In addition, the uterus may be positioned abnormally by being angled too far forward or backward. **Ovarian cysts** and tumors, as well as some disorders of the fallopian tubes, can be findings of these two exams.

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Laith Farid Gulli, M.D.
Robert Ramirez, B.S.

Pelvic fracture

Definition

A pelvic fracture is a break in one or more bones of the pelvis.

Description

The pelvis is a butterfly-shaped group of bones located at the base of the spine. The pelvis consists of the pubis, ilium, and ischium bones (among others) held together by tough ligaments. With a cavity in its

center, the pelvis forms one major ring and two smaller rings of bone that support and protect internal organs such as the bladder, intestines, and rectum. In women, the pelvis also surrounds the uterus and vagina. The pelvis is wider and has a larger cavity in females than in males because it must accommodate **childbirth**.

Fractures of the pelvis are uncommon, accounting for only 0.3–6% of all fractures. Pelvic rings often break in more than one place. Pelvic fractures range widely in severity. Disruption of the major ring is usually a severe injury, while disruption of a minor ring is often not serious. A mild fracture (for example, one that occurs due to the impact of jogging) may heal in several weeks without surgery. However, a serious pelvic fracture can be a life-threatening event requiring emergency medical care and lengthy **rehabilitation**. The latter type of injury may involve damage to nearby internal organs.

Pelvic fractures are classified as stable or unstable, and as open or closed. A stable fracture is one in which the pelvis remains stable and involves one break-point in the pelvic ring with minimal hemorrhage. An unstable fracture is one in which the pelvis is unstable with two or more break-points in the pelvic ring with moderate to severe hemorrhage. All types of pelvic fractures are further divided into “open” or “closed,” depending on whether open skin **wounds** in the lower abdomen are present, or not present.

Causes and symptoms

Most pelvic fractures occur during high-speed accidents (such as car or motorcycle crashes) or falls from great heights. The greater the force, the greater the opportunity for a severe fracture. Pelvic fractures can also occur spontaneously or after minor falls in people with bone-weakening diseases such as **osteoporosis**. Less commonly, pelvic fractures may occur during athletic activities such as football, hockey, skiing, and long-distance running.

The primary symptom of a pelvic fracture is **pain** in the groin, hip, or lower back, which may worsen when walking or moving the legs. Other symptoms may include abdominal pain; numbness/tingling in the groin or legs; bleeding from the vagina, urethra (urine tube), or rectum; difficulty urinating; and difficulty walking or standing. A stress fracture that occurs while jogging may cause pain in the thigh or buttock.

Diagnosis

A pelvic fracture is typically diagnosed by an emergency physician looking for bone tenderness, limitations of movement, difficulty walking, and any loss

of nerve function in the lower part of the body. In addition, the physician looks for signs of injury to nearby organs of the intestinal or genitourinary systems. This search may include checking the rectum, vagina, and urethra for signs of bleeding. The physician will order a plain x ray of the pelvis; this test will usually detect the presence of a fracture. Blood and urine tests may also be done. A computed tomography (CT) scan will be performed in complicated cases. Depending on the severity of the fracture, other imaging procedures may be required as well, such as contrasting studies involving the injection of a radioactive dye. The pictures can be used to evaluate organs and structures in the pelvic area, such as the urethra, bladder, and blood vessels.

Treatment

In the case of a potentially serious pelvic fracture (such as that occurring after an accident or high fall), emergency assistance should be summoned. The person with the injury should be covered with a blanket or jacket (to maintain body heat), and should not be moved by non-trained personnel, especially if there is severe pain or signs of possible nerve injury.

Treatment depends on the severity of the injury. In the case of a minor fracture, treatment may consist of bed rest and over-the-counter (OTC) or prescription pain killers. **Physical therapy**, the use of crutches, and surgery may also be recommended. Healing can take anywhere from a few weeks to several months.

Severe injuries to the pelvis (such as those involving more than one break) can be life threatening, resulting in **shock**, extensive internal bleeding, and damage to internal organs. In these situations, the immediate goal is to control bleeding and stabilize the injured person’s condition. Resuscitation procedures may be required, as well as large amounts of intravenous fluids and blood transfusions if internal bleeding is present. These injuries often require extensive surgery as well as lengthy rehabilitation.

Alternative treatment

To speed up the healing process, some practitioners of alternative medicine recommend **magnetic field therapy**, hydrogen peroxide therapy, **calcium**, vitamin D, vitamin B complex, and zinc.

Prognosis

The prognosis for minor pelvic fractures is excellent, with most people gaining full mobility in a matter of weeks or months. Severe pelvic fractures can be fatal due to internal bleeding or damage to

KEY TERMS

Computed tomography (CT) scan—An imaging procedure that produces a series of thin x-ray slices of internal body organs or structures.

Fracture—A break in a bone.

Orthopedist—A doctor who specializes in disorders of the musculoskeletal system.

Osteoporosis—A decrease in the amount of bone mass, leading to fractures.

Shock—A condition of profound physiological disturbance characterized by failure of the circulatory system to maintain adequate blood supply to vital organs.

Stress fracture—A crack in a bone (usually the result of overuse).

nearby organs, or result in chronic pain and physical disabilities.

Prevention

People with bone-weakening conditions such as osteoporosis or **cancer**, or tendencies to fall are more vulnerable to bone fractures. They should follow their treatment regimens and make use of canes and other walking aids as well as safety devices in the home (bars, non-skidding mats) and avoid climbing up, even on a small stool.

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ORGANIZATIONS

American Academy of Orthopaedic Surgeons, 6300 North River Rd., Rosemont, IL, 60018–4262, (800) 346–AAOS, <http://www.aaos.org>.

American College of Surgeons, 633 North Saint Claire St., Chicago, IL, 60611, (312) 202–5000, <http://www.facs.org>.

American Pain Society, 4700 W. Lake Ave., Glenview, IL, 60025, (847) 375–4715, <http://www.ampainsoc.org>.

American Society for Bone and Mineral Research, 2025 M St., NW, Suite 800, Washington, DC, 20036–3309, (202) 367–1161, <http://www.asbmr.org>.

Greg Annussek
Laura Jean Cataldo, RN, Ed.D.

Pelvic gynecologic sonogram see **Pelvic ultrasound**

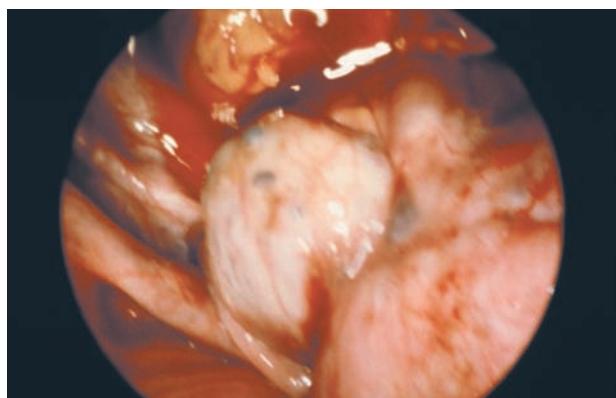
Pelvic inflammatory disease

Definition

Pelvic inflammatory disease (PID) is a term used to describe any infection in the lower female reproductive tract that spreads to the upper female reproductive tract. The lower female genital tract consists of the vagina and the cervix. The upper female genital tract consists of the body of the uterus, the fallopian or uterine tubes, and the ovaries.

Demographics

PID is the most common and the most serious consequence of infection with **sexually transmitted diseases (STD)** in women. Over one million cases of PID are diagnosed annually in the United States, and it is the most common cause for hospitalization of reproductive-age women. Sexually active women aged 15–25 are at highest risk for developing PID. The disease can also occur, although less frequently, in women having monogamous sexual relationships. The most serious consequences of PID are increased risk of **infertility** and **ectopic pregnancy**.



Laparoscopic view of pelvic inflammatory disease. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

To understand PID, it is helpful to understand the basics of inflammation. Inflammation is the body's response to disease-causing (pathogenic) microorganisms. The affected body part may swell due to accumulation of fluid in the tissue or may become reddened due to an excessive accumulation of blood. A discharge (pus) may be produced that consists of white blood cells and dead tissue. Following inflammation, scar tissue may form by the proliferation of scar-forming cells and is called fibrosis. **Adhesions** of fibrous tissue form and cause organs or parts of organs to stick together.

PID may be used synonymously with the following terms:

- salpingitis (inflammation of the fallopian tubes)
- endometritis (inflammation of the inside lining of the body of the uterus)
- tubo-ovarian abscesses (abscesses in the tubes and ovaries)
- pelvic peritonitis (inflammation inside of the abdominal cavity surrounding the female reproductive organs)

Causes and symptoms

The two major causes of STDs are the organisms *Neisseria gonorrhoeae* and *Chlamydia trachomatis*. The main symptom of *N. gonorrhoeae* infection (**gonorrhea**) is a vaginal discharge of mucus and pus. Sometimes bacteria from the colon normally in the vaginal cavity may travel upward to infect the upper female genital organs, facilitated by the infection with gonorrhea. Infections with *C. trachomatis* and other non-gonococcal organisms are more likely to have mild or no symptoms.

Normally, the cervix produces mucus, which acts as a barrier to prevent disease-causing microorganisms (called pathogens) from entering the uterus and moving upward to the tubes and ovaries. This barrier may be breached in two ways. A sexually transmitted pathogen, usually a single organism, invades the lining cells, alters them, and gains entry. Another way for organisms to gain entry happens when trauma or alteration to the cervix occurs. **Childbirth**, spontaneous or induced abortion, or use of an intrauterine contraceptive device (**IUD**) are all conditions that may alter or weaken the normal lining cells, making them susceptible to infection, usually by several organisms. During menstruation, the

cervix widens and may allow pathogens entry into the uterine cavity.

Recent evidence suggests that **bacterial vaginosis** (BV), a bacterial infection of the vagina, may be associated with PID. BV results from the alteration of the balance of normal organisms in the vagina, by douching, for example. While the balance is altered, conditions are formed that favor the overgrowth of anaerobic bacteria, which thrive in the absence of free oxygen. A copious discharge is usually present. Should some trauma occur in the presence of anaerobic bacteria, such as menses, abortion, intercourse, or childbirth, these organisms may gain entrance to the upper genital organs.

The most common symptom of PID is pelvic **pain**. However, many women with PID have symptoms so mild that they may be unaware that an infection exists.

In acute salpingitis, a common form of PID, swelling of the fallopian tubes may cause tenderness on **physical examination**. **Fever** may be present. Abscesses may develop in the tubes, ovaries, or in the surrounding pelvic cavity. Infectious discharge may leak into the peritoneal cavity and cause **peritonitis**, or abscesses may rupture causing a life-threatening surgical emergency.

Chronic salpingitis may follow an acute attack. Subsequent to inflammation, scarring and resulting adhesions may result in chronic pain and irregular menses. Due to blockage of the tubes by scar tissue, women with chronic salpingitis are at high risk of having an ectopic **pregnancy**. The fertilized ovum is unable to travel down the fallopian tube to the uterus and implants itself in the tube, on the ovary, or in the peritoneal cavity. This condition can also be a life-threatening surgical emergency.

IUD

IUD usage has been strongly associated with the development of PID. Bacteria may be introduced to the uterine cavity while the IUD is being inserted or may travel up the tail of the IUD from the cervix into the uterus. Uterine tissue in association with the IUD shows areas of inflammation that may increase its susceptibility to pathogens.

Susceptibility to STDs

Susceptibility to STDs involves many factors, some of which are not known. The ability of the organism to produce disease and the circumstances that place the organism in the right place at a time

KEY TERMS

Adhesion—The joining or sticking together of parts of an organ that are not normally joined together.

C-reactive protein (CRP)—A protein present in blood serum in various abnormal states, like inflammation.

Ectopic—Located away from normal position; ectopic pregnancy results in the attachment and growth of the fertilized egg outside of the uterus, a life-threatening condition.

Endometriosis—The presence and growth of functioning endometrial tissue in places other than the uterus; often results in severe pain and infertility.

Erythrocyte sedimentation rate (ESR)—The rate at which red blood cells settle out in a tube of unclotted blood, expressed in millimeters per hour; elevated sedimentation rates indicate the presence of inflammation.

Fibrosis—The formation of fibrous, or scar, tissue that may follow inflammation and destruction of normal tissue.

Hysterectomy—Surgical removal of the uterus.

Laparoscope—A thin flexible tube with a light on the end that is used to examine the inside of the abdomen; the tube is inserted into the abdomen by way of a small incision just below the navel.

when a trauma or alteration to the lining cells has occurred are factors. The individual's own immune response also helps to determine whether infection occurs.

Risk factors

A number of factors affect the risk of developing PID. They include:

- Age—The incidence of PID is very high in younger women and decreases as a woman ages.
- Race—The incidence of PID is 8–10 times higher in nonwhites than in whites.
- Socioeconomic status—The higher incidence of PID in women of lower socioeconomic status is due in part to a woman's lack of education and awareness of health and disease and her accessibility to medical care.
- Contraception—Induced abortion, use of an IUD, non-use of barrier contraceptives such as condoms, and frequent douching are all associated with a higher risk of developing PID.
- Lifestyle—High risk behaviors, such as drug and alcohol abuse, early age of first intercourse, number of sexual partners, and smoking all are associated with a higher risk of developing PID.
- Types of sexual practices—Intercourse during menses and frequent intercourse may offer more opportunities for the admission of pathogenic organisms to the inside of the uterus.
- Disease—Sixty to 75% of cases of PID are associated with STDs. A prior episode of PID increases the chances of developing subsequent infections.

Diagnosis

Examination

If PID is suspected, a physician will take a complete medical history and perform an internal pelvic examination. Other diseases that may cause pelvic pain, such as **appendicitis** and **endometriosis**, must be ruled out. If pelvic examination reveals tenderness or pain in that region, or tenderness on movement of the cervix, these are good physical signs that PID is present.

Tests

Specific diagnosis of PID is difficult to make because the upper pelvic organs are hard to reach for samplings. The physician may take samples directly from the cervix to identify the organisms that may be responsible for infection. Two blood tests may help to establish the existence of an inflammatory process. A positive **C-reactive protein (CRP)** and an elevated **erythrocyte sedimentation rate (ESR)** indicate the presence of inflammation. The physician may take fluid from the cavity surrounding the ovaries called the *cul de sac*; this fluid may be examined directly for bacteria or may be used for culture.

Procedures

Diagnosis of PID may also be done by performing a surgical **laparoscopy**, which allows the doctor to view the pelvic organs. Equipment for this procedure includes a camera, and a narrow scope that has a light on the end of it for visual purposes. The procedure enables the doctor to take photos and also to take fluid or tissue specimens to send to the lab for further evaluation.

Treatment

Traditional

The goals of treatment are to reduce or eliminate the clinical symptoms and abnormal physical findings, to get rid of the microorganisms, and to prevent long term consequences such as infertility and the possibility of ectopic pregnancy.

Drugs

If acute salpingitis is suspected, treatment with **antibiotics** should begin immediately. Early intervention is crucial to keep the fallopian tubes undamaged. The patient is usually treated with at least two broad spectrum antibiotics that can kill both *N. gonorrhoeae* and *C. trachomatis* plus other types of bacteria that may have the potential to cause infection. Hospitalization may be required to ensure compliance. Treatment for chronic PID may involve **hysterectomy**, which may be helpful in some cases.

If a woman is diagnosed with PID, she should see that her sexual partner is also treated to prevent the possibility of reinfection.

Alternative

Alternative therapy should be complementary to antibiotic therapy. For pain relief, an experienced practitioner may apply castor oil packs, or use **acupressure** or **acupuncture**. Some herbs, such as *Echinacea* (*Echinacea* spp.) and *calendula* (*Calendula officinalis*) are believed to have antimicrobial activity and may be taken to augment the action of prescribed antibiotics. General tonic herbs, as well as good **nutrition** and rest, are important in recovery and strengthening after an episode of PID. Blue cohosh (*Caulophyllum thalictroides*) and false unicorn root (*Chamaelirium luteum*) are recommended as tonics for the general well-being of the female genital tract.

Prognosis

PID can be cured if the initial infection is treated immediately. If infection is not recognized, as frequently happens, the process of tissue destruction and scarring that result from inflammation of the tubes results in irreversible changes in the tube structure that cannot be restored to normal. Subsequent bouts of PID increase a woman's risks manyfold. Thirty to forty percent of cases of female infertility are due to acute salpingitis.

With modern antibiotic therapy, **death** from PID is almost nonexistent. In rare instances, death may occur from the rupture of tubo-ovarian abscesses

and the resulting infection in the abdominal cavity. One recent study has linked infertility, a consequence of PID, with a higher risk of **ovarian cancer**.

Prevention

The prevention of PID is a direct result of the prevention and prompt recognition and treatment of STDs or of any suspected infection involving the female genital tract. The main symptom of infection is an abnormal discharge. To distinguish an abnormal discharge from the mild fluctuations of normal discharge associated with the menstrual cycle takes vigilance and self-awareness. Sexually active women must be able to detect symptoms of lower genital tract disease. Ideally, these women will be able to have a frank dialogue regarding their sexual history, risks for PID, and treatment options with their physicians. Also, these women should have open discussions with their sexual partners regarding disclosure of significant symptoms of possible infection.

Lifestyle changes should be geared toward preventing the transfer of organisms when the body's delicate lining cells are unprotected or compromised. Barrier contraceptives, such as **condoms**, diaphragms, and cervical caps should be used. Women in monogamous relationships should use barrier contraceptives during menses and take their physician's advice regarding intercourse following abortion, childbirth, or biopsy procedures.

Resources

BOOKS

- Ford, Melissa. *Navigating the Land of If: Understanding Infertility and Exploring Your Options*. Berkeley, CA: Seal Press, 2009.
- Marr, Lisa. *Sexually Transmitted Diseases: A Physician Tells You What You Need to Know*. 2nd ed. Baltimore: The Johns Hopkins University Press, 2007.
- Wilson, Michael R. *Pelvic Inflammatory Disease*. New York: Rosen Publishing Group, 2009.

ORGANIZATIONS

- American Society for Reproductive Medicine, 1209 Montgomery Highway, Birmingham, AL, 35216-2809, (205) 978-5000, <http://www.asrm.com>.
- International Center for Infertility Information Dissemination, P.O. Box 6836, Arlington, VA, 22206, (703) 379-9178, <http://www.asrm.org>.
- National Institutes of Health (NIH), 9000 Rockville Pike, Bethesda, MD, 20892, (301) 496-4000, <http://www.nih.gov>.
- National Women's Health Network, 514 10th Street NW, Suite 400, Washington, DC, 20004, (202) 628-7814, <http://www.nwhn.org>.

RESOLVE, 8405 Greensboro Drive, Suite 800, McLean, VA, 22102-5120, (703) 556-7172, <http://www.resolve.org>.

U.S. National Library of Medicine, 8600 Rockville Pike, Bethesda, MD, 20894, <http://www.nlm.nih.gov/medlineplus/medlineplus.html>.

Karen J. Wells
Laura Jean Cataldo, RN, EdD

Pelvic relaxation

Definition

Pelvic relaxation is a weakening of the supportive muscles and ligaments of the pelvic floor. This condition, which affects women and is usually caused by **childbirth**, **aging**, and problems with support, causes the pelvic floor to sag and press into the wall of the vagina.

Demographics

Pelvic relaxation is a common condition in adult females and may affect one in three women, especially those who have had children.

Description

The pelvic floor normally holds the uterus and the bladder in position above the vagina. When the pelvic floor becomes stretched and damaged, these organs can sag into the vagina, sometimes bulging out through the vaginal opening. A sagging uterus is referred to as a uterine prolapse, pelvic floor **hernia**, or pudendal hernia. A sagging bladder is referred to as a bladder prolapse, or cystocele. Other organs, such as the rectum and intestine, can also sag into the vagina as a result of a weakened pelvic floor.

Causes and symptoms

Childbirth increases the risk of pelvic relaxation. Other causes include **constipation**, a chronic **cough**, **obesity**, and heavy lifting. Some women develop the condition after **menopause**, when the body loses the estrogen that helps maintain muscle tone. Mild pelvic relaxation may cause no symptoms. More severe pelvic relaxation can cause the following symptoms:

- an aching sensation in the vagina, lower abdomen, groin or lower back
- heaviness or pressure in the vaginal area, as if something is about to “fall out” of the vagina

KEY TERMS

Cystocele—Bulging of the bladder into the vagina.

Cystourethrocele—Bulging of the bladder neck into the vagina.

Enterocèle—Bulging of the intestine into the upper part of the vagina.

Kegel exercises—Pelvic muscle exercises that strengthen bladder and bowel control.

Pessary—A device inserted into the vagina to support sagging organs.

Rectocele—Bulging of the rectum into the vaginal wall.

Uterine prolapse—Bulging of the uterus into the vagina.

Vaginal prolapse—Bulging of the top of the vagina into the lower vagina or outside the opening of the vagina.

- bladder control problems that worsen with heavy lifting, coughing, or sneezing
- frequent urinary tract infections
- difficulty having a bowel movement

Diagnosis

A thorough **pelvic exam** can help diagnose pelvic relaxation, as can tests of bladder function.

Treatment

Exercises called Kegel exercises can strengthen pelvic floor muscles and lessen the symptoms of pelvic relaxation. These exercises involve squeezing the muscles that stop the flow of urine. The pelvic floor can also be strengthened by estrogen supplements. Physicians sometimes prescribe the insertion of a supportive ring-shaped device called a pessary into the vagina, to prevent the uterus and bladder from pressing into the vagina. Sometimes surgery is recommended to repair a sagging bladder or uterus, and sometimes surgical removal of the uterus (**hysterectomy**) is recommended. Patients are often advised to adhere to a high-fiber diet to reduce the strain of bowel movements, maintain a moderate weight, and avoid activities that strain the pelvic floor. They are sometimes prescribed medications to help control urination and prevent leakage.

Prognosis

Mild cases of pelvic relaxation can sometimes be reversed through Kegel exercises, while severe cases usually do not respond to **exercise** or estrogen therapy, but usually require pessary support or surgery.

Prevention

To limit **stress** on the pelvic support system, women are advised to maintain a normal body weight, limit heavy lifting, and avoid unnecessary straining to have bowel movements.

Resources

BOOKS

- Nelson, Miriam, and Jennifer Ackerman. *The Strong Woman's Guide to Total Health*. New York, NY: Rodale Books, 2010.
- Rosenfeld, Jo Ann, editor. *Handbook of Women's Health*, 2nd ed. New York, NY: Cambridge University Press, 2009.
- Thacker, Holly. *The Cleveland Clinic Guide to Menopause*. New York, NY: Kaplan Publishing, 2009.

ORGANIZATIONS

- American College of Obstetricians and Gynecologists (ACOG), 409 12th St., S.W., PO Box 96920, Washington, DC, 20090–6920, <http://www.acog.org>.
- National Association For Continence (NAFC), PO Box 1019, Charleston, SC, 29402–1019, (800) BLADDER, <http://www.nafc.org>.
- National Kidney and Urologic Diseases Information Clearinghouse (NIDDK), 3 Information Way, Bethesda, MD, 20892, (800) 891–5390, <http://kidney.niddk.nih.gov>.

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Pelvic ultrasound

Definition

Pelvic ultrasound is a procedure where harmless, high-frequency sound waves are projected into the abdomen. These waves reflect off of the internal structures and create shadowy black and white pictures on a display screen.

Purpose

Ultrasound is performed routinely during **pregnancy**. Early in the pregnancy (at about seven weeks), it might be used to determine the size of the

uterus or the fetus, to detect multiple or **ectopic pregnancy**, to confirm that the fetus is alive (or viable), or to confirm the due date. Toward the middle of the pregnancy (at about 16–20 weeks), ultrasound may be used to confirm fetal growth, to reveal defects in the anatomy of the fetus, and to check the placenta. Toward the end of pregnancy, it may be used to evaluate fetal size, position, growth, or to check the placenta. Doctors may use ultrasound during diagnostic procedures like **amniocentesis** and **chorionic villus sampling**. Both of these tests use long needles inserted through the mother's abdomen into the uterus or placenta to gather cells. Ultrasound can also be used in men or women to examine other internal organs, such as the liver, gallbladder, kidney, and heart. The procedure can be useful in detecting cysts, tumors, and **cancer** of the uterus, ovaries, and breasts.

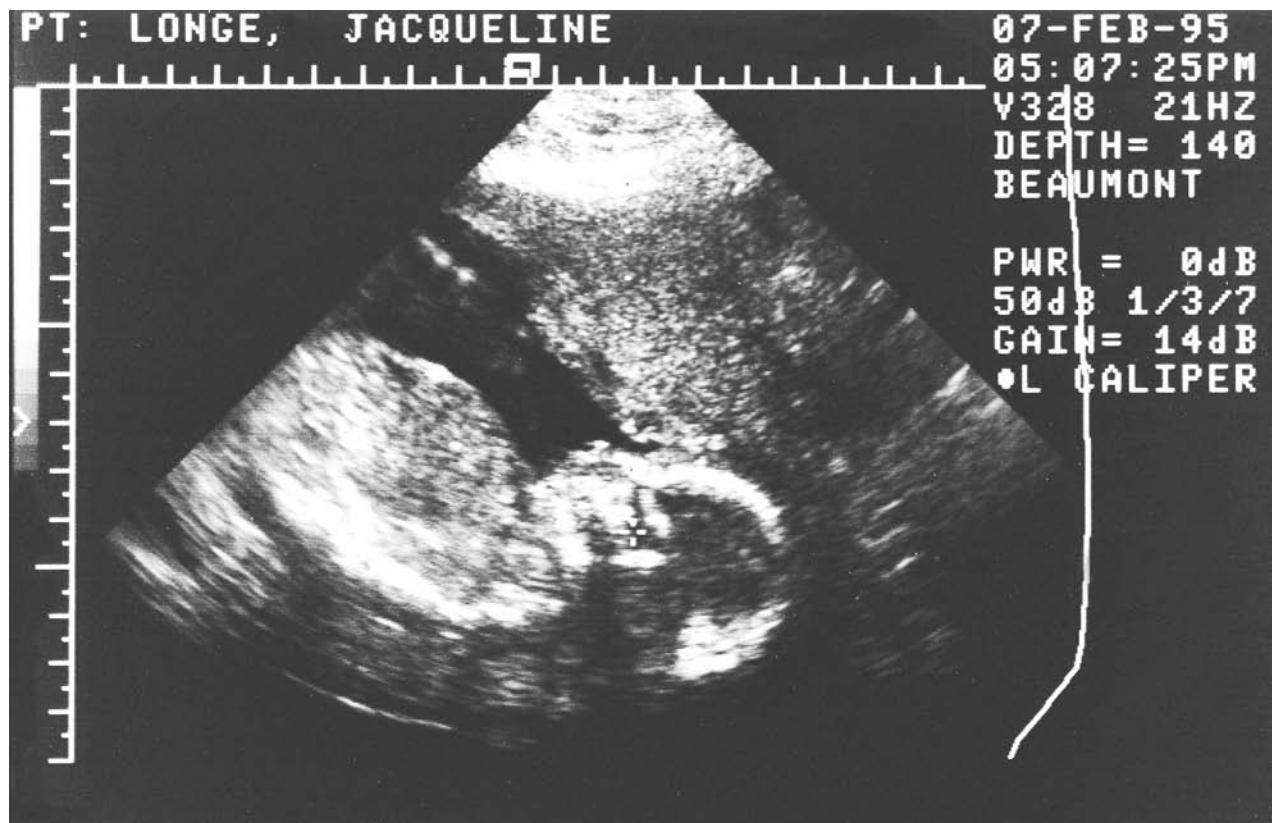
Precautions

There are no special precautions recommended before an ultrasound examination. Unlike x rays, ultrasound does not produce any harmful radiation and does not pose a risk to the mother or the fetus. While many woman have an ultrasound as part of their prenatal care, there may be no medical need to perform the procedure.

Description

Ultrasound examinations can be done in a doctor's office, clinic, or hospital setting. Typically, the pregnant woman will lie on an examination table with her abdomen exposed. Gel or oil is applied to the area. The doctor or technician will move a hand-held scanner (called a transducer) over the abdomen. The transducer emits high-frequency sound waves (usually in the range of 3.5–7.0 megahertz) into the abdomen. The waves are reflected back to the transducer and the wave patterns are shown as an image on a display screen. An ultrasound scan reveals the shapes, densities, and even movements of organs and tissues. Although the pictures transmitted by an ultrasound scan appear gray and grainy, a trained technician can identify the fetus within the uterus, monitor its heartbeat, and sometimes determine its sex. Using computerized tools, the technician can measure various structures shown on the screen. For example, the length of the upper thigh bone (femur) or the distance between the two sides of the skull can indicate the age of the fetus.

Ultrasound technology has been used safely in medical settings for over 30 years, and several significant



An ultrasound image of Anabelle Ashlyn Longe at 20 weeks. (Courtesy of Jacqueline Longe.)

improvements have been made to the procedure. A specially designed transducer probe can be placed in the vagina to provide better ultrasound images. This transvaginal or endovaginal scan is particularly useful in early pregnancy or in cases where ectopic pregnancy is suspected. Doppler ultrasound uses enhanced sound waves to monitor subtle events, like the flow of fetal blood through the heart and arteries. Color imaging is a recent addition to ultrasound technology. With this process, color can be assigned to the various shades of gray for better visualization of subtle tissue details. A new technology under development is three-dimensional ultrasound, which has the potential for detecting even very subtle fetal defects.

Preparation

Before undergoing a pelvic ultrasound, a woman may be asked to drink several glasses of water and to avoid urinating for about one hour before the examination. When the bladder is full, the uterus and fetus are easier to see. A lubricating gel or mineral oil may be applied to the area to make moving the transducer easier.

Aftercare

The lubricating jelly or oil applied to the abdomen is wiped off at the end of the procedure. After an ultrasound examination, a patient can immediately resume normal activities.

Risks

There are no known risks, to either the mother or the fetus, associated with the use of ultrasound.

Normal results

The reliability of ultrasound readings depends on the skill of the technician or doctor performing the scan. Patients should be aware that fetal abnormalities cannot be detected with 100% accuracy using ultrasound. A normal ultrasound result does not necessarily guarantee that the fetus will be normal.

Abnormal results

Ultrasound examinations in obstetrics may detect abnormalities or defects in the fetus. This information may reveal that the fetus cannot survive on its own

KEY TERMS

Amniocentesis—A procedure where a needle is inserted through the pregnant mother's abdomen and into the uterus to draw off some of the amniotic fluid surrounding the fetus.

Chorionic villus sampling—A procedure where a needle is inserted into the placenta to draw off some of the placenta's inner wall cells surrounding the fetus.

Ectopic pregnancy—A pregnancy where the fertilized egg becomes implanted somewhere other than in the uterus. A tubal pregnancy is when the fertilized egg implants in the fallopian tube.

Fetus—A term for an unborn baby, usually from the end of week eight to the moment of birth.

Placenta—The organ that allows interchange between the fetus and the mother. Blood from the fetus and the mother do not directly mix, but the thin placental membrane allows the fetus to absorb nutrients and oxygen from the mother. Waste products from the fetus can exit through the placenta.

Ultrasonography—Another term for ultrasound.

after birth or that it will require extensive treatment or care. Some surgical procedures can be performed to correct defects while the fetus is still in the uterus. Parents faced with information regarding possible **birth defects** may require counseling to consider their choice to either continue or end the pregnancy.

The diagnostic use of ultrasound may reveal the presence of cysts, tumors, or cancer in internal organs.

ORGANIZATIONS

American Institute of Ultrasound in Medicine, 14750 Sweitzer Lane, Suite 100, Laurel, MD, 20707-5906, (301) 498-4100, (301) 498-4450, <http://www.aium.org>.

Altha Roberts Edgren

Penicillin V see **Penicillins**

Penicillins

Definition

Penicillins are antibiotic medicines that kill bacteria and prevent their growth and spread.

Purpose

Penicillins are used to treat infections in the middle ears, sinuses, mouth, throat and lungs, and urinary system. They are also used as part of a multi-drug treatment program for peptic ulcers caused by the *Helicobacter pylori* bacterium.

These drugs are also used to prevent infections in people who are susceptible to recurrent heart or **kidney disease** prior to having dental and other procedures.

Penicillins will *not* cure or prevent colds, flu, and other infections caused by viruses.

Description

Examples of penicillins include penicillin V (Pen-Vee K, V-cillin K), cloxicillin, amoxicillin (Amoxil, Polymox, Trimox), and carbenicillin. Penicillin can be combined with clavulanic acid (Augmentin) to broaden the scope of its effectiveness.

Recommended dosage

Available only by prescription, penicillins come as capsules, tablets (regular and chewable), liquids and injectables.

Antibiotics should always be taken as directed for as long as they are prescribed. Do not stop taking them when symptoms improve.

Precautions

A full ten days of treatment must be taken when treating beta-streptococcal infections to prevent the secondary development of **rheumatic fever** or kidney disease (**glomerulonephritis**).

When using these drugs to treat **sexually transmitted diseases** (STD), follow up blood tests for syphilis should be done monthly for four months to assure that this disease, requiring longer term treatment, is not present.

Penicillins may change the results of the urine test for glucose.

ALLERGIES. People who have hay **fever**, **asthma**, **eczema**, or other **allergies** (or who have had such allergies in the past) may be more likely to be allergic to the penicillins.

Anyone who has had unusual reactions to penicillins or **cephalosporins** in the past should let his or her physician know before taking the drugs again. Physicians should also know about allergies to foods, dyes, preservatives, or other substances.

Special conditions

DIABETES. Penicillins may cause false positive results on urine sugar tests.

PHENYLKETONURIA. Augmentin chewable tablets contain phenylalanine. People with **phenylketonuria** (PKU) should consult their physician before taking this medicine.

Side effects

The most common adverse effects from penicillins are abdominal **pain, nausea**, upset stomach, mild **diarrhea** or skin rash with **itching**.

Rarely, there may be soreness of the mouth or tongue or severe diarrhea.

Severe allergic reactions rarely occur, but require immediate medical attention. Those symptoms include:

- difficulty breathing
- fever
- lightheadedness or faintness
- joint pain
- hives, itching, or red, scaly skin
- swelling or puffiness in the face, mouth or throat

Interactions

Tetracycline antibiotics reduce the effectiveness of penicillins.

Some penicillins reduce the effectiveness of birth control pills. Secondary methods of preventing **pregnancy** are advised while taking them.

Penicillins may reduce the blood pressure effects of beta-blocker drugs, like atenolol, used to treat high blood pressure.

Penicillins may increase the blood levels of methotrexate.

Allopurinol (Zyloprim) may increase the blood levels of penicillins.

James Waun, MD, RPh

Penile cancer

Definition

Penile **cancer** is the growth of malignant cells on the external skin and in the tissues of the penis.

Description

Penile cancer is a disease in which cancerous cells appear on the penis. If left untreated, this cancer can grow and spread from the penis to the lymph nodes in the groin and eventually to other parts of the body.

Demographics

Penile cancer is a rare form of cancer that develops in about one out of 100,000 men per year in the United States. Penile cancer is more common in other parts of the world, particularly Africa and Asia. In Uganda, penile cancer is the most common form of cancer for men.

Causes and symptoms

The cause of penile cancer is unknown. The most common symptoms of penile cancer are:

- a tender spot, an open sore, or a wart-like lump on the penis
- unusual liquid discharges from the penis
- pain or bleeding in the genital area

Diagnosis

In order to diagnose penile cancer, the doctor examines the patient's penis for lumps or other abnormalities. A tissue sample, or biopsy, may be ordered to distinguish cancerous cells from **syphilis** and penile **warts**. If the results confirm a diagnosis of cancer, additional tests are done to determine whether the disease has spread to other parts of the body.

Treatment

In Stage I penile cancer, malignant cells are found only on the surface of the head (glans) and on the foreskin of the penis. If the cancer is limited to the foreskin, treatment may involve wide local excision and **circumcision**. Wide local excision is a form of surgery that removes only cancer cells and a small amount of normal tissue adjacent to them. Circumcision is removal of the foreskin.

If the Stage I cancer is only on the glans, treatment may involve the use of a fluorouracil cream (Adrucil, Efdex), and/or microsurgery. Microsurgery removes cancerous tissue and the smallest possible amount of normal tissue. During microsurgery, the doctor uses a special instrument that provides a comprehensive view of the area where cancer cells are located and makes it possible to determine that all malignant cells have been removed.

In Stage II, the penile cancer has spread to the surface of the glans, tissues beneath the surface, and the shaft of the penis. The treatment recommended may be

amputation of all or part of the penis (total or partial penectomy). If the disease is diagnosed early enough, surgeons are often able to preserve enough of the organ for urination and sexual activity. Treatment may also include microsurgery and external **radiation therapy**, in which a machine provides radiation to the affected area. **Laser surgery** is an experimental treatment for Stage II cancers. Laser surgery uses an intense precisely focused beam of light to dissolve or burn away cancer cells.

In Stage III, malignant cells have spread to lymph nodes in the groin, where they cause swelling. The recommended treatment may include amputation of the penis and removal of the lymph nodes on both sides. Radiation therapy may also be suggested. More advanced disease requires systemic treatments using drugs (**chemotherapy**). In chemotherapy, medicines are administered intravenously or taken by mouth. These drugs enter the bloodstream and kill cancer cells that have spread to any part of the body.

In Stage IV, the disease has spread throughout the penis and lymph nodes in the groin, or has traveled to other parts of the body. Treatments are similar to that for Stage III cancer.

Recurrent penile cancer is disease that recurs in the penis or develops in another part of the body after treatment has eradicated the original cancer cells.

Cure rates are high for cancers diagnosed in Stage I or II, but much lower for Stages III and IV, by which time cancer cells have spread to the lymph nodes.

Alternative treatment

In addition to the treatments previously described, biological therapy is another treatment that is currently being studied. Biological therapy is a type of treatment that is sometimes called biological response modifier (BRM) therapy. It uses natural or artificial substances to boost, focus, or reinforce the body's disease-fighting resources.

Prevention

Conditions which increase a person's chance of getting penile cancer include:

- infection with genital warts (human papillomavirus, or HPV)
- a skin disease called psoriasis
- a condition called phimosis, in which the foreskin becomes difficult to retract
- other conditions that result in repeated irritation of the penis.
- a history of smoking.

KEY TERMS

Circumcision—Surgical removal of the foreskin of the penis. It is usually performed shortly after birth.

Fluorouracil—A cell-killing (cytotoxic) medication that can be applied in cream form to treat cancer of the penis.

There appears to be a connection between development of the disease and lack of personal hygiene. Failure to regularly and thoroughly cleanse the part of the penis covered by the foreskin increases the risk of developing the disease. Penile cancer is also more common in uncircumcised men.

Resources

OTHER

"What is penile cancer?" American Cancer Society. July 20, 2010. <http://www.cancer.org/Cancer/PenileCancer/DetailedGuide/penile-cancer-what-is-penile-cancer> (accessed December 20, 2010).

ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329, (800) 227-2345, <http://www.cancer.org>.
Cancer Group Institute, 17620 9th Ave. NE, Miami, FL, 33162, (305) 493-1980, <http://www.cancergroup.com>.

Maureen Haggerty
Paul A. Johnson, Ed.M.

Penile implant surgery see **Penile prostheses**

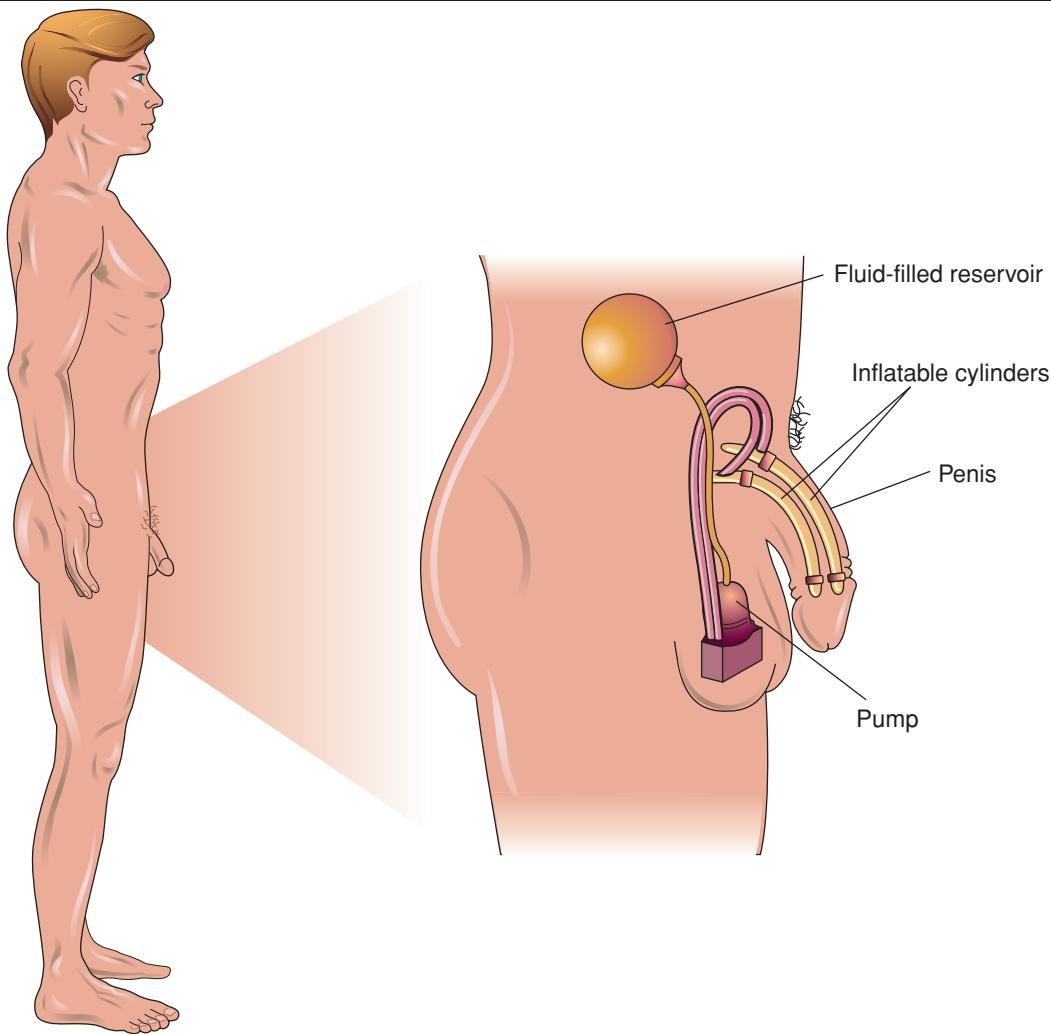
Penile prostheses

Definition

Penile prostheses are semirigid or inflatable devices that are implanted into penises to alleviate **impotence**.

Purpose

The penis is composed of one channel for urine and semen and three compartments with tough, fibrous walls containing "erectile tissue." With appropriate stimulation, the blood vessels that lead out of these compartments constrict, trapping blood. Blood pressure fills and hardens the compartments producing an erection of sufficient firmness to perform sexual intercourse. Additional stimulation leads to ejaculation, where semen is pumped out of the urethra. When this



The inflatable implant is a common penile prosthesis. This device connects through a tube to a flexible fluid reservoir and a pump. The pump is shaped like a testicle and inserted in the scrotum. When the pump is squeezed, the fluid is forced into the inflatable cylinders implanted inside the penis, producing an erection. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

system fails, impotence (failure to create and maintain an erection) occurs.

Impotence can be caused by a number of conditions, including diabetes, **spinal cord injury**, prolonged drug **abuse**, and removal of a prostate gland. If the medical condition is irreversible, a penile prosthesis may be considered. Patients whose impotence is caused by psychological problems are not recommended for implant surgery.

Description

Penile implant surgery is conducted on patients who have exhausted all other areas of treatment. The semirigid device consists of two rods that are easier and less

expensive to implant than the inflatable cylinders. Once implanted, the semirigid device needs no follow-up adjustments, however it produces a penis which constantly remains semi-erect. The inflatable cylinders produce a more natural effect. The patient is able to simulate an erection by using a pump located in the scrotum.

With the patient asleep under **general anesthesia**, the device is inserted into the erectile tissue of the penis through an incision in the fibrous wall. In order to implant the pump for the inflatable implant, incisions are made in the abdomen and the perineum (area between the anus and the genitals). A fluid reservoir is inserted into the groin and the pump is placed in the scrotum. The cylinders, reservoir, and pump are

KEY TERMS

General anesthesia—Deep sleep induced by a combination of medicines that allows surgery to be performed.

Genital—Sexual organ.

Perineum—Area between the anus and genitals.

Scrotum—The external pouch containing the male reproductive glands (testes) and part of the spermatic cord.

connected by tubes and tested before the incisions are closed.

Preparation

Surgery always requires an adequately informed patient, both as to risks and benefits. In this case, the sexual partner should also be involved in the discussion. Prior to surgery, antibacterial cleansing occurs and the surrounding areas are shaved.

Aftercare

To minimize swelling, ice packs are applied to the penis for the first 24 hours following surgery. The incision sites are cleansed daily to prevent infection. Pain relievers may be taken.

Risks

With any implant, there is a slightly greater risk of infection. The implant may irritate the penis and cause continuous pain. The inflatable prosthesis may need follow-up surgery to repair leaks in the reservoir or to reconnect the tubing.

Resources

BOOKS

Tanagho, Emil A., Jack W McAninch, and Donald Ridge-way Smith. *Smith's General Urology*. New York: McGraw-Hill Medical, 2008.

J. Ricker Polsdorfer, MD

Pentoxifylline see **Blood-viscosity reducing drugs**

Peptic ulcer disease see **Heliobacteriosis**

Percutaneous renal biopsy see **Kidney biopsy**

Percutaneous transhepatic cholangiography

Definition

Percutaneous transhepatic cholangiography (PTHC) is an x-ray test used to identify obstructions either in the liver or bile ducts that slow or stop the flow of bile from the liver to the digestive system.

Purpose

Because the liver and bile ducts are not normally seen on x rays, the doctor injects the liver with a special dye that will show up on the resulting picture. This dye distributes evenly to fill the whole liver drainage system. If the dye does not distribute evenly, this is indicative of a blockage, which may be caused by a gallstone or a tumor in the liver, bile ducts, or pancreas.

Precautions

Patients should report allergic reactions to:

- anesthetics
- dyes used in medical tests
- iodine
- shellfish

PTHC should not be performed on anyone who has **cholangitis** (inflammation of the bile duct), massive **ascites**, a severe allergy to iodine, or a serious uncorrectable or uncontrollable bleeding disorder. Patients who have diabetes should inform their doctor.

Description

PTHC is performed in a hospital, doctor's office, or outpatient surgical or x-ray facility. The patient lies on a movable x-ray table and is given a local anesthetic. The patient will be told to hold his or her breath, and a doctor, nurse, or laboratory technician will inject a special dye into the liver as the patient exhales.

The patient may feel a twinge when the needle penetrates the liver, a pressure or fullness, or brief discomfort in the upper right side of the back. Hands and feet may become numb during the 30–60 minute procedure.

The x-ray table will be rotated several times during the test, and the patient helped to assume a variety of positions. A special x-ray machine called a fluoroscope will track the dye's movement through the bile

ducts and show whether the fluid is moving freely or if its passage is obstructed.

PTHC costs about \$1,600. The test may have to be repeated if the patient moves while x rays are being taken.

Preparation

An intravenous antibiotic may be given every 4–6 hours during the 24 hours before the test. The patient will be told to fast overnight. Having an empty stomach is a safety measure in case of complications, such as bleeding, that might require emergency repair surgery. Medications such as **aspirin**, or non-steroidal anti-inflammatory drugs that thin the blood, should be stopped three–seven days prior to taking the PRHC test. Patients may also be given a sedative a few minutes before the test begins.

Aftercare

A nurse will monitor the patient's vital signs and watch for:

- itching
- flushing
- nausea and vomiting
- sweating
- excessive flow of saliva
- possible serious allergic reactions to contrast dye

The patient should stay in bed for at least six hours after the test, lying on the right side to prevent bleeding from the injection site. The patient may resume normal eating habits and gradually resume normal activities. The doctor should be informed right away if **pain** develops in the right abdomen or shoulder or in case of **fever**, **dizziness**, or a change in stool color to black or red.

Risks

Septicemia (blood poisoning) and bile **peritonitis** (a potentially fatal infection or inflammation of the membrane covering the walls of the abdomen) are rare but serious complications of this procedure. Dye occasionally leaks from the liver into the abdomen, and there is a slight risk of bleeding or infection.

Normal results

Normal x rays show dye evenly distributed throughout the bile ducts. **Obesity**, gas, and failure to fast can affect test results.

KEY TERMS

Ascites—Abnormal accumulation of fluid in the abdomen.

Bile ducts—Tubes that carry bile, a thick yellowish-green fluid that is made by the liver, stored in the gallbladder, and helps the body digest fats.

Cholangitis—Inflammation of the bile duct.

Fluoroscope—An x-ray machine that projects images of organs.

Granulomatous disease—Characterized by growth of tiny blood vessels and connective tissue.

Jaundice—Disease that causes bile to accumulate in the blood, causing the skin and whites of the eyes to turn yellow. Obstructive jaundice is caused by blockage of bile ducts, while non-obstructive jaundice is caused by disease or infection of the liver.

Abnormal results

Enlargement of bile ducts may indicate:

- obstructive or non-obstructive jaundice
- cholelithiasis (gallstones)
- hepatitis (inflammation of the liver)
- cirrhosis (chronic liver disease)
- granulomatous disease
- pancreatic cancer
- bile duct or gallbladder cancers

Resources

BOOKS

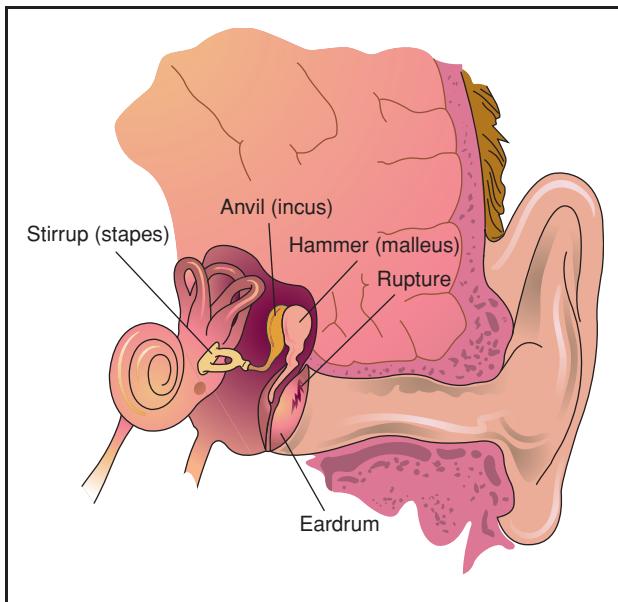
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Maureen Haggerty

Perforated eardrum

Definition

A perforated eardrum exists when there is a hole or rupture in the eardrum, the thin membrane that separates the outer ear canal from the middle ear. A perforated eardrum may cause temporary **hearing loss** and occasional discharge.



A perforated eardrum is caused by a hole or rupture in the eardrum, the thin membrane that separates the outer ear canal from the middle ear. It may result in temporary hearing loss and occasional discharge. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Demographics

Perforated eardrum can affect individuals of all ages. However, it is most commonly associated with individuals who have frequent colds and upper respiratory ailments (especially children). Young children (up to two years of age) are more susceptible to perforated eardrum due to the frequency of middle ear infections they experience.

Description

The eardrum (tympanic membrane) is a thin wall that separates the outer ear from the middle ear, vibrating when sound waves strike the membrane. The middle ear is connected to the nose by the Eustachian tube.

In addition to conducting sound, the eardrum also protects the middle ear from bacteria. When it is perforated, bacteria can more easily get into this part of the ear, causing ear infections.

In general, the larger the hole in the eardrum, the greater the temporary loss of hearing. The location of the perforation also affects the degree of hearing loss. Severe hearing loss may follow a skull fracture that disrupts the bones in the middle ear. Eardrum perforation caused by a loud noise may result in ringing in the ear (**tinnitus**), in addition to a temporary hearing

loss. Over time, this hearing loss improves and the ringing usually fades in a few days.

Causes and symptoms

The eardrum can become damaged by a direct injury. It is possible to perforate the eardrum:

- with a cotton-tipped swab or another foreign object
- by hitting the ear with an open hand
- after a skull fracture
- after a loud explosion or other loud noise

In addition, an ear infection can rupture the eardrum as pressure within the middle ear rises when fluid builds up. If the eardrum is punctured by pressure from an ear infection, there may be infected or bloody drainage from the ear.

Rarely, a small hole may remain in the eardrum after a pressure-equalizing tube falls out or is removed by a doctor.

Symptoms include an earache or **pain** in the ear, which may be severe, or a sudden decrease in ear pain, followed by ear drainage of clear, bloody, or pus-filled fluid, hearing loss, or ear noise/buzzing.

Diagnosis

A doctor can diagnose a perforated eardrum by direct inspection with an otoscope. Hearing tests may reveal a hearing loss.

Treatment

A perforated eardrum usually heals by itself within two months. **Antibiotics** may be given to prevent infection or to treat an existing ear infection. Painkillers can relieve any ear pain.

Sometimes, a paper patch is placed over the eardrum until the membrane heals. Three or four patches may be needed before the perforation closes completely. If the eardrum does not heal on its own, surgical repair (tympanoplasty) may be necessary.

The ear should be kept clean and dry while the eardrum heals. Patients should insert cotton balls into the ear when showering or shampooing to block any water from getting into the ear. Pain in the ear may be eased by applying warm compresses.

Prognosis

While a perforated eardrum may be uncomfortable, it usually heals on its own. Any hearing loss that accompanies the perforation is usually temporary.

KEY TERMS

Eustachian tube—The air duct that connects the area behind the nose to the middle ear.

Otoscope—An instrument used to examine the ear, to inspect the outer ear canal and the eardrum, and to detect diseases in the middle ear.

Prevention

A perforated eardrum can be prevented by avoiding insertion of any object into the ear to clean it. If a foreign object becomes lodged in the ear, only a doctor should try to remove it.

Promptly treating all ear infections is another way to guard against a ruptured eardrum.

Resources

BOOKS

Plack, Christopher. *Oxford Handbook of Auditory Science: Hearing*. New York, NY: Oxford University Press, 2010.

ORGANIZATIONS

American Academy of Otolaryngology—Head and Neck Surgery, Inc., One Prince St., Alexandria, VA, 22314–3357, (703) 836–4444, <http://www.entnet.org>.

American Hearing Research Foundation (AHRF), 55 E. Washington St., Suite 2022, Chicago, IL, 60602, (312) 726–9670, <http://www.american-hearing.org>.

American Speech-Language–Hearing Association (ASHA), 10801 Rockville Pike, Rockville, MD, 20852, (800) 638–8255, <http://www.asha.org>.

Better Hearing Institute (BHI), 515 King St., Suite 420, Alexandria, VA, 22314, (703) 684–3391, <http://www.betterhearing.org>.

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Perforated septum

Definition

A perforated septum is a hole in the nasal septum, the vertical plane of tissue that separates the nostrils.

Description

The nasal septum is a thin structure in the middle of the nose. In front, it is cartilage, further back it is

bone. On either side, it is covered with mucus membranes. The cartilage depends upon the blood vessels in the mucus membranes on either side for its **nutrition**. If that blood supply is shut off, the cartilage dies, producing a hole or perforation.

Causes and symptoms

There are several causes of a perforated septum.

- Wearing ornaments in the nose. To hang an ornament from the middle of the nose requires that the tissue directly in front of the septal cartilage be pierced or perforated.
- Sniffing cocaine. Cocaine is a potent vasoconstrictor, which means that it causes small blood vessels to close. It is used in nose surgery because it shrinks mucus membranes, permitting better visualization and access into the nose. Used continuously, tissues are deprived of blood and die. The nasal septum is the most vulnerable to this effect of sniffing cocaine.
- Getting the septum cauterized. Nosebleeds usually come from the front part of the nasal septum, which is rich in blood vessels. Uncontrolled repeated bleeding from these vessels may require cautery-burning the vessels with electricity or chemicals to close them off. Injudicious cautery of both sides of the septum has in the past led to death of tissue and consequent perforation.
- More and more people are having cosmetic surgery done on their nose. The procedure, called rhinoplasty, occasionally damages the septum's blood supply.
- Contracting certain diseases. Several diseases—typhoid, syphilis, systemic lupus erythematosus, and tuberculosis—can infect this tissue and destroy it.
- Being exposed to harmful vapors. Toxic air pollutant-like acid fumes, phosphorus, and copper vapor—and sometimes even cortisone sprays—can destroy nasal tissue.

Perforation is not serious. It causes irritation, mostly complaints of dryness and crusting. Sometimes air blowing past it whistles. Picking at the crusts can cause bleeding.

Treatment

Surgical repair is not difficult. The surgeon may devise a plastic button that fits exactly into the defect and stays in place like a collar button.

KEY TERMS

Systemic lupus erythematosus—A collagen-vascular disease in the autoimmune category that causes damage to many different parts of the body.

Alternative treatment

Saline nasal sprays may be sufficient to control symptoms and prevent the need for surgery.

Prevention

Nosebleeds from the septum can usually be controlled with pinching. Vaginal estrogen cream has also been used successfully to toughen the blood vessels.

Resources

BOOKS

Wilson, William R., J. B. Nadol, Jr., and Gregory W. Randolph. *Clinical Handbook of Ear, Nose and Throat Disorders*. New York: Informa Healthcare, 2004.

J. Ricker Polsdorfer, MD

Pericardiocentesis

Definition

Pericardiocentesis is the removal by needle of pericardial fluid from the sac surrounding the heart for diagnostic or therapeutic purposes.

Purpose

The pericardium, the sac (or membrane) that surrounds the heart muscle, normally contains a small amount of fluid that cushions and lubricates the heart as the heart expands and contracts. When too much fluid gathers in the pericardial cavity, the space between the pericardium and the outer layers of the heart, a condition known as pericardial effusion occurs. Abnormal amounts of fluid may result from:

- pericarditis (caused by infection, inflammation)
- trauma (producing blood in the pericardial sac)
- surgery or other invasive procedures performed on the heart
- cancer (producing malignant effusions)
- myocardial infarction, congestive heart failure
- renal failure

Possible causes of **pericarditis** include chest trauma, systemic infection (bacterial, viral, or fungal), myocardial infarction (**heart attack**), or **tuberculosis**. When pericarditis is suspected, pericardiocentesis may be advisable in order to obtain a fluid sample for laboratory analysis to identify the underlying cause of the condition.

Pericardiocentesis is also used in emergency situations to remove excessive accumulations of blood or fluid from the pericardial sac, such as with **cardiac tamponade**. When fluid builds up too rapidly or excessively in the pericardial cavity, the resulting compression on the heart impairs the pumping action of the vascular system. Cardiac tamponade is a life-threatening condition that requires immediate treatment.

Precautions

Whenever possible, an echocardiogram (ultrasound test) should be performed to confirm the presence of the pericardial effusion and to guide the pericardiocentesis needle during the procedure. Because of the risk of accidental puncture to major arteries or organs in pericardiocentesis, surgical drainage may be a preferred treatment option for pericardial effusion in non-emergency situations.

Description

The patient's vital signs are monitored throughout the procedure, and an ECG tracing is continuously run. If time allows, **sedation** is administered, the puncture site is cleaned with an antiseptic iodine solution, and a local anesthetic is injected into the skin to numb the area. The patient is instructed to remain still. The physician performing pericardiocentesis will insert a syringe with an attached cardiac needle slowly into the chest wall until the needle tip reaches the pericardial sac. The patient may experience a sensation of pressure as the needle enters the membrane. When the needle is in the correct position, the physician will aspirate, or withdraw, fluid from the pericardial sac.

When the procedure is performed for diagnostic purposes, the fluid will be collected into specimen tubes for laboratory analysis. If the pericardiocentesis is performed to treat a cardiac tamponade or other significant fluid build-up, a pericardial catheter may be attached to the needle to allow for continuous drainage.

After the cardiac needle is removed, pressure is applied to the puncture site for approximately five minutes, and the site is then bandaged.

Preparation

Prior to pericardiocentesis, the test procedure is explained to the patient, along with the risks and possible complications involved, and the patient is asked to sign an informed consent form. If the patient is incapacitated, the same steps are followed with a family member.

No special diet or **fasting** is required for the test. After the patient changes into a hospital gown, an intravenous line is inserted into a vein in the arm. The IV will be used to administer sedation, and any required medications or blood products. Leads for an electrocardiogram (ECG) tracing are attached to the patient's right and left arms and legs, and the fifth lead is attached to the cardiac needle used for the procedure. The patient is instructed to lie flat on the table, with the upper body elevated to a 60-degree angle.

Aftercare

The site of the puncture and any drainage catheter should be checked regularly for signs of infection such as redness and swelling. Blood pressure and pulse are also monitored following the procedure. Patients who experience continued bleeding or abnormal swelling of the puncture site, sudden **dizziness**, difficulty breathing, or chest pains in the days following a pericardiocentesis procedure should seek immediate medical attention.

Risks

Pericardiocentesis is an invasive procedure, and infection of the puncture site or pericardium is always a risk. Possible complications include perforation of a major artery, lung, or liver. The myocardium, the outer muscle layer of the heart, could also be damaged if the cardiac needle is inserted too deeply.

Normal results

Normal pericardial fluid is clear to straw-colored in appearance with no bacteria, blood, **cancer** cells or pathogens. There is typically a minimal amount of the fluid (10–50 mL) in the pericardial cavity.

Abnormal results

A large volume of pericardial fluid (over 50 mL) indicates the presence of pericardial effusion. Laboratory analysis of the fluid can aid in the diagnosis of the cause of pericarditis. The presence of an infectious organism such as *staphylococcus aureus* is a sign of bacterial pericarditis. Excessive protein is present in cases of **systemic lupus erythematosus** or myocardial

KEY TERMS

Cardiac tamponade—Compression and restriction of the heart that occurs when the pericardium fills with blood or fluid. This increase in pressure outside the heart interferes with heart function and can result in shock and/or death.

Catheter—A long, thin, flexible tube used to drain or administer fluids.

Echocardiogram—An imaging test using high-frequency sound waves to obtain pictures of the heart and surrounding tissues.

Electrocardiogram—A cardiac test that measures the electrical activity of the heart.

Myocardium—The middle layer of the heart wall.

Pericardium—A double membranous sac that envelops and protects the heart.

infarction (heart attack). An elevated white blood count may point to a fungal infection. If the patient has a hemorrhage, a cardiac rupture, or cancer, there may be blood in the pericardial fluid.

Resources

BOOKS

Maisch, Bernhard, et al. *Interventional Pericardiology: Pericardiocentesis, Pericardioscopy, Pericardial Biopsy, Balloon Pericardiectomy, and Intrapericardial Therapy*. New York: Springer, 2010.

ORGANIZATIONS

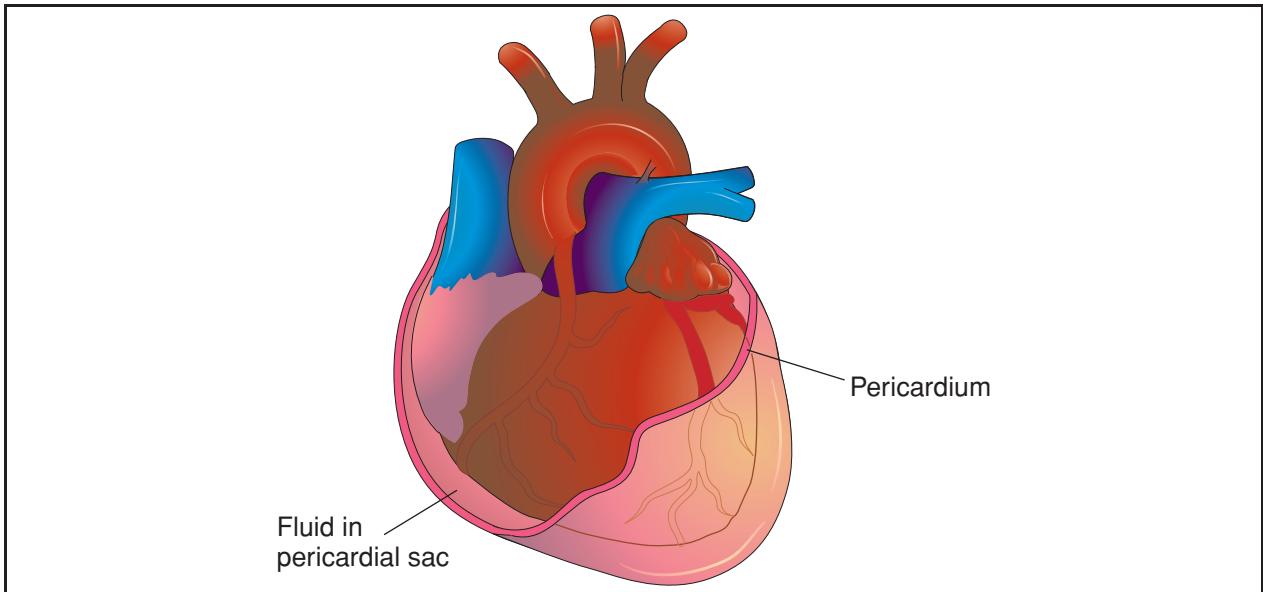
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Pericarditis

Definition

Pericarditis is an inflammation of the two layers of the thin, sac-like membrane that surrounds the heart. This membrane is called the pericardium, so the term pericarditis means inflammation of the pericardium.



Cardiac tamponade occurs when fluid collects in the pericardial sac between the heart and the surrounding pericardium. A medical emergency, cardiac tamponade deprives the body of oxygen and requires immediate treatment. ((Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Description

Pericarditis is fairly common. It affects approximately one in 1,000 people. The most common form is caused by infection with a virus. People in their 20s and 30s who have had a recent upper respiratory infection are most likely to be affected, along with men aged 20–50. One out of every four people who have had pericarditis will get it again, but after two years these relapses are less likely.

Causes and symptoms

The viruses that cause pericarditis include those that cause **influenza**, **polio**, and **rubella** (German measles). In children, the most common viruses that cause pericarditis are the adenovirus and the coxsackievirus (which is most likely to affect children during warmer weather).

Although pericarditis is usually caused by a virus, it also can be caused by an injury to the heart or it can follow a **heart attack**. It may also be caused by certain inflammatory diseases such as **rheumatoid arthritis** or **systemic lupus erythematosus**. Bacteria, fungi, parasites, **tuberculosis**, **cancer** or kidney failure may also affect the pericardium. Sometimes the cause is unknown.

There are several forms of pericarditis, depending on the cause.

Acute pericarditis

This is caused by infection with a virus, bacteria, or fungus—usually in the lungs and upper respiratory tract. This form of the disease causes a sharp, severe **pain** that starts in the region of the breastbone. If the pericarditis is caused by a bacteria, it is called bacterial or purulent pericarditis.

Cardiac tamponade

Sometimes fluid collects between the heart and the pericardium. This is called pericardial effusion, and may lead to a condition called **cardiac tamponade**. When the fluid accumulates, it can squeeze the heart and prevent it from filling with blood. This keeps the rest of the body from getting the necessary supply of oxygen and can cause dangerously low blood pressure. A cardiac tamponade can happen when the chest is injured during surgery, **radiation therapy**, or an accident. Cardiac tamponade is a serious medical emergency and must be treated immediately.

Constrictive pericarditis

When the pericardium is scarred or thickened, the heart has difficulty contracting. This is because the pericardium has shrunken or tightened around the heart, constricting the muscle's heart movement. This usually occurs as a result of tuberculosis, which now is rarely

KEY TERMS

Computed tomography (CT) scan—A CT scan uses x rays to scan the body from many angles. A computer compiles the x rays into a picture of the area being studied. The images are viewed on a monitor and printed-out.

Echocardiogram—An echocardiogram bounces sound waves off the heart to create a picture of its chambers and valves.

Electrocardiogram (ECG)—An ECG is a test to measure electrical activity in the heart.

Heart catheterization—A heart catheterization is used to view the heart's chamber and valves. A tube (catheter) is inserted into an artery, usually in

the groin. A dye is then put into the artery through the tube. The dye makes its way to the heart to create an image of the heart on x-ray film. The image is photographed and stored for further examination.

Pericardiocentesis—Pericardiocentesis is a procedure used to test for viruses, bacteria, and fungus. The physician puts a small tube through the skin, directly into the pericardial sac, and withdraws fluid. The fluid then is tested for viruses, bacteria, and fungus.

Pericardium—The pericardium is the thin, sac-like membrane that surrounds the heart. It has two layers: the serous pericardium and the fibrous pericardium.

found in the United States, except in immigrant, AIDS, and prison populations.

Symptoms of pericarditis

Symptoms likely to be associated with pericarditis include:

- rapid breathing
- breathlessness
- dry cough
- fever and chills
- weakness
- broken blood vessels (hemorrhages) in the mucus membrane of the eyes, the back, the chest, fingers, and toes
- feelings of anxiety
- A sharp or dull pain that starts in the front of the chest under the breastbone and radiates to the left side of the neck, upper abdomen, and left shoulder. The pain is less intense when the patient sits up or leans forward and worsens when lying down; it may worsen with a deep breath, like pleurisy, which may accompany pericarditis

In cardiac tamponade, neck veins may be swollen and blood pressure may be very low.

Diagnosis

The heart of a person with pericarditis is likely to produce a grating sound (friction rub) when heard through a stethoscope. This sound occurs because the roughened pericardium surfaces are rubbing against each other.

The following tests will also help diagnose pericarditis and what is causing it:

- electrocardiograph (ECG) and echocardiogram to distinguish between pericarditis and a heart attack.
- x ray to show the traditional “water bottle” shadow around the heart that is often seen in pericarditis where there is a sufficient fluid build up.
- computed tomography scan (CT scan) of the chest.
- heart catheterization to view the heart's chambers and valves.
- pericardiocentesis to test for viruses, bacteria, fungus, cancer, and tuberculosis.
- blood tests such as LDH and CPK to measure cardiac enzymes and distinguish between a heart attack and pericarditis, as well as a complete blood count (CBC) to look for infection.

Treatment

Since most pericarditis is caused by a virus and will heal naturally, there is no specific, curative treatment. Ordinary **antibiotics** do not work against viruses. Pericarditis that comes from a virus usually clears up in two weeks to three months. Medications may be used to reduce inflammation, however. They include **nonsteroidal anti-inflammatory drugs** (NSAIDs), such as ibuprofen and **aspirin**. **Corticosteroids** are helpful if the pericarditis was caused by a heart attack or systemic lupus erythematosus. **Analgesics** (painkillers such as aspirin or **acetaminophen**) also may be given.

If the pericarditis recurs, removal of all or part of the pericardium (pericardectomy) may be necessary. In the case of constrictive pericarditis, the pericardectomy

may be necessary to remove the stiffened parts of the pericardium that are preventing the heart from beating correctly.

If a cardiac tamponade is present, it may be necessary to drain excess fluid from the pericardium. **Pericardiocentesis**, the same procedure used for testing, will be used to withdraw the fluid.

For most people, home care with rest and medications to relieve pain are sufficient. A warm heating pad or compress also may help relieve pain. Sitting in an upright position and bending forward helps relieve discomfort. A person with pericarditis may also be kept in bed, with the head of the bed elevated to reduce the heart's need to work hard as it pumps blood. Along with painkillers and antibiotics, diuretic drugs ("water pills") to reduce fluids may also be used judiciously.

Prognosis

Prognosis is good. Most people recover within three weeks to several months and do not need any additional treatment.

Prevention

There is no way to prevent pericarditis, but a healthy lifestyle with proper **nutrition** and **exercise** will help keep the body's immune system strong and more likely to fight off invading microorganisms.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

Christine Kuehn Kelly

Perinatal infection

Definition

An infection caused by a bacteria or virus that can be passed from a mother to her baby during **pregnancy** or delivery is called a perinatal infection.

Description

Perinatal infections include bacterial or viral illnesses that can be passed from a mother to her baby

either while the baby is still in the uterus, during the delivery process, or shortly after birth. Maternal infection can, in some cases, cause complications at birth. The mother may or may not experience active symptoms of the infection during the pregnancy. The most serious and most common perinatal infections, and the impact of these diseases on the mother and infant, are discussed below in alphabetical order. It is important to note that men can become infected and can transmit many of these infections to other women. The sexual partners of women who have these infections also should seek medical treatment.

Causes and symptoms

Chlamydia

Chlamydia trachomatis is the most common bacterial sexually transmitted disease in the United States, causing more than 4 million infections each year. The majority of women with chlamydial infection experience no obvious symptoms. The infection affects the reproductive tract and causes **pelvic inflammatory disease**, **infertility**, and **ectopic pregnancy** (the fertilized egg implants somewhere other than in the uterus). This infection can cause premature rupture of the membranes and early labor. It can be passed to the infant during delivery and can cause ophthalmia neonatorum (an eye infection) within the first month of life and **pneumonia** within one to three months of age. Symptoms of **chlamydial pneumonia** are a repetitive **cough** and rapid breathing. **Wheezing** is rare and the infant does not develop a **fever**.

Cytomegalovirus

Cytomegalovirus (CMV) is a common virus in the herpes virus family. It is found in saliva, urine, and other body fluids and can be spread through sexual contact or other more casual forms of physical contact like kissing. In adults, CMV may cause mild symptoms of swollen lymph glands, fever, and **fatigue**. Many people who carry the virus experience no symptoms at all. Infants can become infected with CMV while still in the uterus if the mother becomes infected or develops a recurrence of the infection during pregnancy. Most infants exposed to CMV before birth develop normally and do not show any symptoms. As many as 6,000 infants who were exposed to CMV before birth are born with serious complications each year. CMV interferes with normal fetal development and can cause **mental retardation**, blindness, deafness, or **epilepsy** in these infants.

Genital herpes

Genital herpes, which is usually caused by herpes simplex virus type 2 (HSV-2), is a sexually transmitted disease that causes painful sores on the genitals. Women who have their first outbreak of genital herpes during pregnancy are at high risk of **miscarriage** or delivering a low birth weight baby. The infection can be passed to the infant at the time of delivery if the mother has an active sore. The most serious risk to the infant is the possibility of developing HSV-2 **encephalitis**, an inflammation of the brain, with symptoms of irritability and poor feeding.

Hepatitis B

Hepatitis B is a contagious virus that causes liver damage and is a leading cause of chronic **liver disease** and **cirrhosis**. Approximately 20,000 infants are born each year to mothers who test positive for the hepatitis B virus. These infants are at high risk for developing hepatitis B infection through exposure to their mothers' blood during delivery.

Human immunodeficiency virus (HIV)

Human **immunodeficiency** virus (HIV) is a serious, contagious virus that causes acquired immunodeficiency syndrome (**AIDS**). About one-fourth of pregnant women with HIV pass the infection on to their newborn infants. An infant with HIV usually develops AIDS and dies before the age of two.

Human papillomavirus

Human papillomavirus (HPV) is a sexually transmitted disease that causes **genital warts** and can increase the risk of developing some cancers. HPV appears to be transferred from the mother to the infant during the birth process.

Rubella (German measles)

Rubella is a virus that causes German **measles**, an illness that includes rash, fever, and symptoms of an upper respiratory tract infection. Most people are exposed to rubella during childhood and develop antibodies to the virus so they will never get it again. Rubella infection during early pregnancy can pass through the placenta to the developing infant and cause serious **birth defects** including heart abnormalities, mental retardation, blindness, and deafness.

Streptococcus

Group B streptococcus (GBS) infection is the most common bacterial cause of infection and **death** in newborn infants. Although rates have declined in

the United States since the introduction of **antibiotics** to at-risk women during labor in the 1980s, about 1,600 cases and 80 newborn deaths still occur each year. In women, GBS can cause vaginitis and urinary tract infections. Both infections can cause premature birth and the bacteria can be transferred to the infant in the uterus or during delivery. GBS causes pneumonia, **meningitis**, and other serious infections in infants.

Syphilis

Syphilis is a sexually transmitted bacterial infection that can be transferred from a mother to an infant through the placenta before birth. Up to 50% of infants born to mothers with syphilis will be premature, stillborn, or will die shortly after birth. Infected infants may have severe birth defects. Those infants who survive infancy may develop symptoms of syphilis up to two years later.

Diagnosis

Chlamydia

Chlamydial bacteria can be diagnosed by taking a cotton swab sample of the cervix and vagina during the third trimester of the pregnancy. Chlamydial cell cultures take three to seven days to grow but many laboratories are not equipped to run the tests necessary to confirm the diagnosis.

Cytomegalovirus

Past or recent infection with CMV can be identified by antibody tests and CMV can be grown from body fluids.

Genital herpes

The appearance of a genital sore is enough to suspect an outbreak of genital herpes. The sore can be cultured and tested to confirm that HSV-2 is present.

Hepatitis B

A blood test can be used to screen pregnant women for the hepatitis B surface antigen (HBsAg) in prenatal health programs.

Human immunodeficiency virus (HIV)

HIV can be detected using a blood test and is part of most prenatal screening programs.

Human papillomavirus

HPV causes the growth of **warts** in the genital area. The wart tissue can be removed with a scalpel

and tested to determine what type of HPV virus caused the infection.

Rubella (German measles)

Pregnant women are usually tested for antibodies to rubella, which would indicate that they have been previously exposed to the virus and therefore would not develop infection during pregnancy if exposed.

Streptococcus

GBS can be detected by a vaginal or rectal swab culture, and sometimes from a **urine culture**. Blood tests can be used to confirm GBS infection in infants who exhibit symptoms.

Syphilis

Pregnant women are usually tested for syphilis as part of the prenatal screening.

Treatment

Chlamydia

Pregnant women can be treated during the third trimester with oral erythromycin, for seven–14 days depending on the dose used. Newborn infants can be treated with erythromycin liquid for 10–14 days at a dosage determined by their body weight.

Cytomegalovirus

No drugs or vaccines are currently available for prevention or treatment of CMV.

Genital herpes

The **antiviral drugs** acyclovir or famciclovir can be administered to the mother during pregnancy. Little is known about the risks of these drugs to the fetus, however, the risk of birth defects does not seem to be any higher than for women who do not take these medications. Infants with suspected HSV-2 can be treated with acyclovir. Delivery of the infant by **cesarean section** is recommended if the mother has an active case of genital herpes.

Hepatitis B

Infants born to mothers who test positive to the HBsAg test should be treated with hepatitis B immune globulin at birth to give them immediate protection against developing hepatitis B. These infants, as well as all infants, should also receive a series of three hepatitis B vaccine injections as part of their routine immunizations.

Human immunodeficiency virus (HIV)

Recent studies have shown that prenatal care and HIV testing before delivery are major opportunities to prevent perinatal HIV infection. Pregnant women with HIV should be treated as early in the pregnancy as possible with zidovudine (AZT). Other newer drugs designed to treat HIV/AIDS also may be used during pregnancy with the knowledge that these drugs may have unknown effects on the infant. The risks and benefits of such treatments need to be discussed. Infants born with HIV should receive aggressive drug treatment to prevent development of AIDS.

Human papillomavirus

Genital warts are very difficult to treat and frequently recur even after treatment. They can be removed by **cryotherapy** (freezing), laser or electrocauterization (burning), or surgical excision (cutting) of the warts. Some medications (imiquimod 5% cream, podophyllin, trichloroacetic acid or topical 5-fluorouracil) can be applied to help dissolve genital warts. Cesarean delivery rather than vaginal delivery seems to reduce the risk of transmission of HPV from mothers to infants.

Rubella (German measles)

No treatment is available. Some health care providers may recommend giving the mother an injection of immune globulin (to boost the immune system to fight off the virus) if she is exposed to rubella early in the pregnancy. However, no evidence to support the use of these injections exists. Exposure to rubella early in pregnancy poses a high risk that the infant will have serious birth defects. Termination of the pregnancy may be considered. Women who have not been previously exposed to rubella will usually be vaccinated immediately after the first pregnancy to protect infants of future pregnancies.

Streptococcus

Pregnant women diagnosed with GBS late in the pregnancy should be treated with antibiotics injected intravenously to prevent **premature labor**. In 2003, the Centers for Disease Control and Prevention (CDC) issued revised guidelines for preventing perinatal GBS disease. They began recommending that women not only be tested as soon as they learn of their pregnancy, but again at 35 to 37 weeks gestation. The CDC also recommended updated **prophylaxis** regimens for women with penicillin **allergies**, as well as new guidelines for patients with threatened preterm deliveries and other new recommendations. If transmission of

GBS to the newborn infant already is suspected or if the baby develops symptoms of infection, infants often are treated with antibiotics.

Syphilis

Antibiotic therapy, usually penicillin, given early in the pregnancy can be used to treat the infection and may prevent transmission to the infant.

Prognosis

Chlamydia

Without treatment, the most serious consequences of chlamydial infection are related to complications of premature delivery. Treatment of the mother with antibiotics during the third trimester can prevent premature delivery and the transfer of the infection to the baby. Infants treated with antibiotics for eye infection or pneumonia generally recover.

Cytomegalovirus

The chance for recovery after exposure to CMV is very good for both the mother and the infant. Exposure to CMV can be serious and even life threatening for mothers and infants whose immune systems are compromised, for example those receiving **chemotherapy** or who have HIV/AIDS. Those infants who develop birth defects after CMV exposure may have serious, lifelong complications.

Genital herpes

Once a woman or infant is infected, outbreaks of genital herpes sores can recur at any point during their lifetimes.

Hepatitis B

Infants treated at birth with immune globulin and the series of vaccinations will be protected from development of hepatitis B infection. Infants infected with hepatitis B develop a chronic, mild form of hepatitis and are at increased risk for developing liver disease.

Human immunodeficiency virus (HIV)

Treatment with AZT during pregnancy significantly reduces the chance that the infant will be infected with HIV from the mother.

Human papillomavirus

Once infected with HPV, there is a lifelong risk of developing warts and an increased risk of some cancers.

KEY TERMS

Cesarean section—A surgical procedure in which an incision is made in a woman's abdomen to deliver the infant from the uterus.

Ectopic pregnancy—A condition that ends in miscarriage, in which the fertilized ovum attaches somewhere other than in the uterus (for example in the fallopian tube or abdomen).

Encephalitis—Inflammation or swelling of the brain.

Perinatal—The period of time around the time of pregnancy and delivery.

Pneumonia—An infection and inflammation of the lungs that usually causes shortness of breath, cough, fever, and chest pain.

Rubella (German measles)

Infants exposed to rubella virus in the uterus are at high risk for severe birth defects including heart defects, blindness, and deafness.

Streptococcus

Infection of the urinary tract or genital tract of pregnant women can cause premature birth. Infants infected with GBS can develop serious, life-threatening infections.

Syphilis

Premature birth, birth defects, or the development of serious syphilis symptoms is likely to occur in untreated pregnant women.

Prevention

Use of a barrier method of contraceptive (condom) can prevent transmission of some of the infections. Intravenous drug use and sexual intercourse with infected partners increases the risks of exposure to most of these infections. Pregnant women can be tested for many of the bacterial or viral infections described; however, effective treatment may not be available to protect the infant. New studies show that a woman's nutritional status may contribute to her ability to fight off infections, particularly in cases of **malnutrition**. Proper prenatal care may improve outcomes and prevent some infections.

Resources

PERIODICALS

- Goldenberg, Robert L. "The Plausibility of Micronutrient Deficiency in Relationship to Perinatal Infection." *The Journal of Nutrition* May 2003: 1645S.
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Periodic paralysis

Definition

Periodic **paralysis** (PP) is the name for several rare, inherited muscle disorders marked by temporary weakness, especially following rest, sleep, or **exercise**.

Description

Periodic paralysis disorders are genetic disorders that affect muscle strength. There are two major forms, hypokalemic and hyperkalemic, each caused by defects in different genes.

In hypokalemic PP, the level of potassium in the blood falls in the early stages of a paralytic attack, while in hyperkalemic PP, it rises slightly or is normal. (The root of both words, "kali," refers to potassium.) Hyperkalemic PP is also called potassium-sensitive PP.

Causes and symptoms

Causes

Both forms of PP are caused by inheritance of defective genes. Both genes are dominant, meaning that only one copy of the defective gene is needed for a person to develop the disease. A parent with the gene has a 50% chance of passing it along to each offspring, and the likelihood of passing it on is unaffected by the results of previous pregnancies.

The gene for hypokalemic PP is present equally in both sexes, but leads to noticeable symptoms more often in men than in women. The normal gene is responsible for a muscle protein controlling the flow of **calcium** during muscle contraction.

The gene for hyperkalemic PP affects virtually all who inherit it, with no difference in male-vs.-female expression. The normal gene is responsible for a muscle protein controlling the flow of **sodium** during muscle contraction.

Symptoms

The attacks of weakness in hypokalemic PP usually begin in late childhood or early adolescence and often become less frequent during middle age. The majority of patients develop symptoms before age 16. Since they begin in the school years, the symptoms of hypokalemic PP are often first seen during physical education classes or after-school sports, and may be mistaken for laziness, or lack of interest on the part of the child.

Attacks are most commonly brought on by:

- strenuous exercise followed by a short period of rest
- large meals, especially ones rich in carbohydrates or salt
- emotional stress
- alcohol use
- infection
- pregnancy

The weakness from a particular attack may last from several hours to as long as several days, and may be localized to a particular limb, or might involve the entire body.

The attacks of weakness of hyperkalemic PP usually begin in infancy or early childhood, and may become less severe later in life. As in the hypokalemic form, attacks are brought on by **stress**, **pregnancy**, and exercise followed by rest. In contrast, though, hyperkalemic attacks are not associated with a heavy meal but rather with missing a meal, with high potassium intake, or use of glucocorticoid drugs such as prednisone. (Glucocorticoids are a group of **steroids** that regulate metabolism and affect muscle tone.)

Weakness usually lasts less than three hours, and often persists for only several minutes. The attacks are usually less severe, but more frequent, than those of the hypokalemic form. Weakness usually progresses from the lower limbs to the upper, and may involve the facial muscles as well.

Diagnosis

Diagnosis of either form of PP begins with a careful medical history and a complete physical and **neurological exam**. A family medical history may reveal other affected relatives. Blood and urine tests done at

the onset of an attack show whether there are elevated or depressed levels of potassium. Electrical tests of muscle and a muscle biopsy show characteristic changes.

Challenge tests, to aid in diagnosis, differ for the two forms. In hypokalemic PP, an attack of weakness can be brought on by administration of glucose and insulin, with exercise if necessary. An attack of hyperkalemic PP can be induced with administration of potassium after exercise during **fasting**. These tests are potentially hazardous and require careful monitoring.

Genetic tests are available at some research centers and are usually recommended for patients with a known family history. However, the number of different possible mutations leading to each form is too great to allow a single comprehensive test for either form, thus limiting the usefulness of **genetic testing**.

Treatment

Severe respiratory weakness from hypokalemic PP may require intensive care to ensure adequate ventilation. Potassium chloride may be given by mouth or intravenously to normalize blood levels.

Attacks requiring treatment are much less common in hyperkalemic PP. Glucose and insulin may be prescribed. Eating carbohydrates may also relieve attacks.

Prognosis

Most patients learn to prevent their attacks well enough that no significant deterioration in the quality of life occurs. Strenuous exercise must be avoided, however. Attacks often lessen in severity and frequency during middle age. Frequent or severe attacks increase the likelihood of permanent residual weakness, a risk in both forms of periodic paralysis.

Prevention

There is no way to prevent the occurrence of either disease in a person with the gene for the disease. The likelihood of an attack of either form of PP may be lessened by avoiding the triggers (the events or combinations of circumstances which cause an attack) for each.

Hypokalemic PP attacks may be prevented with use of acetazolamide (or another carbonic anhydrase inhibitor drug) or a diuretic to help retain potassium in the bloodstream. These attacks may also be prevented by avoiding such triggers as salty food, large meals, a high-carbohydrate diet, and strenuous exercise.

KEY TERMS

Gene—A biologic unit of heredity transmitted from parents to offspring.

Attacks of hyperkalemic PP may be prevented with frequent small meals high in carbohydrates, and the avoidance of foods high in potassium such as orange juice or bananas. Acetazolamide or thiazide (a diuretic) may be prescribed.

ORGANIZATIONS

Muscular Dystrophy Association, 3300 East Sunrise Drive, Tucson, AZ, 85718, (800) 572-1717, <http://www.mdausa.org>.

Periodic Paralysis Association, 155 West 68th St., Suite 17, New York, NY, 10023, (407) 339-9499, <http://www.periodicparalysis.org>.

Richard Robinson

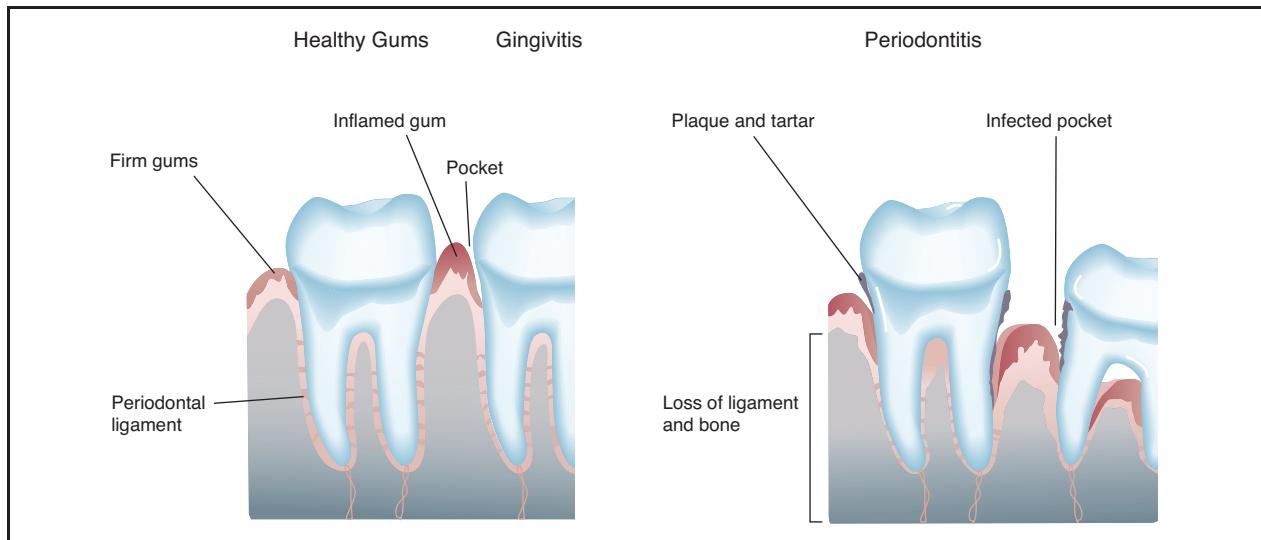
Periodontal disease

Definition

Periodontal diseases are a group of diseases that affect the tissues that support and anchor the teeth. Left untreated, periodontal disease results in the destruction of the gums, alveolar bone (the part of the jaws where the teeth arise), and the outer layer of the tooth root.

Description

Periodontal disease is usually seen as a chronic inflammatory disease. An acute infection of the periodontal tissue may occur, but is not usually reported to the dentist. The tissues that are involved in periodontal diseases are the gums, which include the gingiva, periodontal ligament, cementum, and alveolar bone. The gingiva is a pink-colored mucus membrane that covers parts of the teeth and the alveolar bone. The periodontal ligament is the main part of the gums. The cementum is a calcified structure that covers the lower parts of the teeth. The alveolar bone is a set of ridges from the jaw bones (maxillary and mandible) in which the teeth are embedded. The main area involved in periodontal disease is the gingival sulcus, a pocket between the teeth and the gums. Several distinct forms of periodontal disease



Healthy gums support the teeth. When gingivitis goes untreated, the gums become weak and pockets form around the teeth. Plaque and tartar build up in the pockets, the gum recedes, and periodontitis occurs. ((Illustration by Argosy, Inc. Reproduced by permission of Gale, a part of Cengage Learning.)

are known. These are gingivitis, acute necrotizing ulcerative gingivitis, adult periodontitis, and localized juvenile periodontitis. Although periodontal disease is thought to be widespread, serious cases of periodontitis are not common. Gingivitis is also one of the early signs of leukemia in some children.

Gingivitis

Gingivitis is an inflammation of the outermost soft tissue of the gums. The gingivae become red and inflamed, lose their normal shape, and bleed easily. Gingivitis may remain a chronic disease for years without affecting other periodontal tissues. Chronic gingivitis may lead to a deepening of the gingival sulcus. Acute necrotizing ulcerative gingivitis is mainly seen in young adults. This form of gingivitis is characterized by painful, bleeding gums, and **death** (necrosis) and erosion of gingival tissue between the teeth. It is thought that **stress, malnutrition, fatigue, and poor oral hygiene** are among the causes for acute necrotizing ulcerative gingivitis.

Adult periodontitis

Adult periodontitis is the most serious form of the periodontal diseases. It involves the gingiva, periodontal ligament, and alveolar bone. A deep periodontal pocket forms between the teeth, the cementum, and the gums. Plaque, calculus, and debris from food and other sources collect in the pocket. Without treatment, the periodontal ligament can be destroyed and resorption of the alveolar

bone occurs. This allows the teeth to move more freely and eventually results in the loss of teeth. Most cases of adult periodontitis are chronic, but some cases occur in episodes or periods of tissue destruction.

Localized juvenile periodontitis

Localized juvenile periodontitis is a less common form of periodontal disease and is seen mainly in young people. Primarily, localized juvenile periodontitis affects the molars and incisors. Among the distinctions that separate this form of periodontitis are the low incidence of bacteria in the periodontal pocket, minimal plaque formation, and mild inflammation.

Herpetic gingivostomatitis

Herpes infection of the gums and other parts of the mouth is called herpetic gingivostomatitis and is frequently grouped with periodontal diseases. The infected areas of the gums turn red in color and have whitish herpes lesions. There are two principal differences between this form of periodontal diseases and most other forms. Herpetic gingivostomatitis is caused by a virus, Herpes simplex, not by bacteria, and the viral infection tends to heal by itself in approximately two weeks. Also, herpetic gingivostomatitis is infectious to other people who come in contact with the herpes lesions or saliva that contains virus from the lesion.



An extreme case of juvenile periodontitis. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)



Gingivitis, an inflammation of the gums, is a common periodontal disease. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)

Pericoronitis

Pericoronitis is a condition found in children who are in the process of producing molar teeth. The disease is seen more frequently in the lower molar teeth. As the molar emerges, a flap of gum still covers the tooth. The flap of gum traps bacteria and food, leading to a mild irritation. If the upper molar fully emerges before the lower one, it may bite down on the flap during chewing. This can increase the irritation of the flap and lead to an infection. In bad cases, the infection can spread to the neck and cheeks.

Desquamative gingivitis

Desquamative gingivitis occurs mainly in postmenopausal women. The cause of the disease is not understood. The outer layers of the gums slough off, leaving raw tissue and exposed nerves.

Trench mouth

Trench mouth is an acute, necrotizing (causing tissue death), ulcerating (causing open sores) form of gingivitis. It causes **pain** in the affected gums. **Fever** and fatigue are usually present also. Trench mouth, also known as Vincent's disease, is a complication of mild cases of gingivitis. Frequently, poor oral hygiene is the main cause. Stress, an unbalanced diet, or lack of sleep are frequent cofactors in the development of trench mouth. This form of periodontal disease is more common in people who smoke. The term "trench mouth" was created in World War I, when the disease was common in soldiers who lived in the trenches. Symptoms of trench mouth appear suddenly. The initial symptoms include painful gums and foul breath. Gum tissue between teeth becomes infected and dies, and starts to disappear. Often, what appears to be remaining gum is dead tissue. Usually, the gums bleed easily, especially when chewing. The pain can increase to the point where eating and swallowing become difficult. Inflammation or infection from trench mouth can spread to nearby tissues of the face and neck.

Periodontitis

Periodontitis is a condition in which gingivitis has extended down around the tooth and into the supporting bone structure. Periodontitis is also called pyorrhoea. Plaque and tarter buildup sometimes lead to the formation of large pockets between the gums and teeth. When this happens, anaerobic bacteria grow in the pockets. The pockets eventually extend down around the roots of the teeth where the bacteria cause damage to the bone structure supporting the teeth. The teeth become loose and tooth loss can result. Some medical conditions are associated with an increased likelihood of developing periodontitis. These diseases include diabetes, **Down syndrome**, Crohn's disease, **AIDS**, and any disease that reduces the number of white blood cells in the body for extended periods of time.

Causes and symptoms

Several factors play a role in the development of periodontal disease. The most important are age and oral hygiene. The number and type of bacteria present on the gingival tissues also play a role in the development of periodontal diseases. The presence of certain species of bacteria in large enough numbers in the gingival pocket and related areas correlates with the development of periodontal disease. Also, removal of the bacteria correlates with reduction or elimination of

disease. In most cases of periodontal disease, the bacteria remain in the periodontal pocket and do not invade surrounding tissue.

The mechanisms by which bacteria in the periodontal pocket cause tissue destruction in the surrounding region are not fully understood. Several bacterial products that diffuse through tissue are thought to play a role in disease formation. Bacterial endotoxin is a toxin produced by some bacteria that can kill cells. Studies show that the amount of endotoxin present correlates with the severity of periodontal disease. Other bacterial products include proteolytic enzymes, molecules that digest protein found in cells, thereby causing cell destruction. The immune response has also been implicated in tissue destruction. As part of the normal immune response, white blood cells enter regions of inflammation to destroy bacteria. In the process of destroying bacteria, periodontal tissue is also destroyed.

Gingivitis usually results from inadequate oral hygiene. Proper brushing of the teeth and flossing decreases plaque buildup. The bacteria responsible for causing gingivitis reside in the plaque. Plaque is a sticky film that is largely made from bacteria. Tartar is plaque that has hardened. Plaque can turn into tartar in as little as three days if not brushed off. Tartar is difficult to remove by brushing. Gingivitis can be aggravated by hormones, and sometimes becomes temporarily worse during **pregnancy**, **puberty**, and when the patient is taking birth control pills. Interestingly, some drugs used to treat other conditions can cause an overgrowth of the gingival tissue that can result in gingivitis because plaque builds up more easily. Drugs associated with this condition are phenytoin, used to treat seizures; cyclosporin, given to organ transplant patients to reduce the likelihood of organ rejection; and **calcium** blockers, used to treat several different heart conditions. **Scurvy**, a vitamin C deficiency, and **pellagra**, a niacin deficiency, can also lead to bleeding gums and gingivitis.

The initial symptoms of periodontitis are bleeding and inflamed gums, and **bad breath**. Periodontitis follows cases of gingivitis, which may not be severe enough to cause a patient to seek dental help. Although the symptoms of periodontitis are also seen in other forms of periodontal diseases, the key characteristic in periodontitis is a large pocket that forms between the teeth and gums. Another characteristic of periodontitis is that pain usually does not develop until late in the disease, when a tooth loosens or an **abscess** forms.

KEY TERMS

Anaerobic bacteria—Microorganisms that grow in the absence of oxygen.

Inflammation—A painful redness and swelling of an area of tissue in response to infection or injury.

Diagnosis

Diagnosis is made by observation of infected gums. Usually, a dentist is the person to diagnose and characterize the various types of periodontal disease. In cases such as acute herpetic gingivostomatitis, there are characteristic herpetic lesions. Many of the periodontal diseases are distinguished based on the severity of the infection and the number and type of tissues involved.

Diagnosis of periodontitis includes measuring the size of the pockets formed between the gums and teeth. Normal gingival pockets are shallow. If periodontal disease is severe, jaw bone loss will be detected in x-ray images of the teeth. If too much bone is lost, the teeth become loose and can change position. This will also be seen in x-ray images.

Treatment

Tartar can only be removed by professional dental treatment. Following treatment, periodontal tissues usually heal quickly. Gingivitis caused by vitamin deficiencies is treated by administering the needed vitamin. There are no useful drugs to treat herpetic gingivostomatitis. Because of the pain associated with the herpes lesions, patients may not brush their teeth while the lesions are present. Herpes lesions heal by themselves without treatment. After the herpetic lesions have disappeared, the gums usually return to normal if good oral hygiene is resumed. Pericoronitis is treated by removing debris under the flap of gum covering the molar. This operation is usually performed by a dentist. Surgery is used to remove molars that are not likely to form properly.

Treatment for trench mouth starts with a complete cleaning of the teeth, removal of all plaque, tartar, and dead tissue on the gums. For the first few days after cleaning, the patient uses hydrogen peroxide mouth washes instead of brushing. After cleaning, the gum tissue will be very raw and rinsing minimizes damage to the gums that might be caused by the toothbrush. For the first few days, the patient should visit the dentist daily for checkups and then every second or third day for the next two weeks. Occasionally, antibiotic treatment is used to supplement dental cleaning of the teeth.

and gums. Surgery may be needed if the damage to the gums is extensive and they do not heal properly.

Treatment of periodontitis requires professional dental care. The pockets around the teeth must be cleaned, and all tartar and plaque removed. In periodontitis, tartar and plaque can extend far down the tooth root. Normal dental hygiene, brushing and flossing, cannot reach deep enough to be effective in treating periodontitis. In cases where pockets are very deep (more than 0.25 in [0.64 cm] deep), surgery is required to clean the pocket. This is performed in a dental office. Sections of gum that are not likely to reattach to the teeth may be removed to promote healing by healthy sections of gum. Abscesses are treated with a combination of **antibiotics** and surgery. The antibiotics may be delivered directly to the infected gum and bone tissues to ensure that high concentrations of the antibiotic reach the infected area. Abscess infections, especially of bone, are difficult to treat and require long term antibiotic treatments to prevent a reoccurrence of infection.

Prognosis

Periodontal diseases can be easily treated. The gums usually heal and resume their normal shape and function. In cases where they do not, prostheses or surgery can restore most of the support for proper functioning of the teeth.

Prevention

Most forms of periodontal disease can be prevented with good dental hygiene. Daily use of a toothbrush and flossing is sufficient to prevent most cases of periodontal disease. Tartar control toothpastes help prevent tartar formation, but do not remove tartar once it has formed.

Resources

BOOKS

Mandell, Gerald L., et al. *Mandell, Douglas, and Bennett's Principles and Practice of Infectious Diseases*.

John T. Lohr, PhD

Periodontitis see **Periodontal disease**

Periorbital cellulitis see **Orbital and periorbital cellulitis**

Peripheral arterial disease see **Peripheral vascular disease**

Peripheral neuritis see **Peripheral neuropathy**

Peripheral neuropathy

Definition

The term peripheral neuropathy encompasses a wide range of disorders in which the nerves outside of the brain and spinal cord—peripheral nerves—have been damaged. Peripheral neuropathy may also be referred to as peripheral neuritis, or if many nerves are involved, the terms polyneuropathy or polyneuritis may be used.

Description

Peripheral neuropathy is a widespread disorder, and there are many underlying causes. Some of these causes are common, such as diabetes, and others are extremely rare, such as acrylamide **poisoning** and certain inherited disorders. The most common worldwide cause of peripheral neuropathy is **leprosy**. Leprosy is caused by the bacterium *Mycobacterium leprae*, which attacks the peripheral nerves of affected people. According to statistics gathered by the World Health Organization, an estimated 1.15 million people have leprosy worldwide.

Leprosy is extremely rare in the United States, where diabetes is the most commonly known cause of peripheral neuropathy. It has been estimated that more than 17 million people in the United States and Europe have diabetes-related polyneuropathy. Many neuropathies are idiopathic, meaning that no known cause can be found. The most common of the inherited peripheral neuropathies in the United States is Charcot-Marie-Tooth disease, which affects approximately 125,000 persons.

Another of the better known peripheral neuropathies is **Guillain-Barré syndrome**, which arises from complications associated with viral illnesses, such as cytomegalovirus, **Epstein-Barr virus**, and human **immunodeficiency** virus (HIV), or bacterial infection, including *Campylobacter jejuni* and **Lyme disease**. The worldwide incidence rate is approximately 1.7 cases per 100,000 people annually. Other well-known causes of peripheral neuropathies include chronic **alcoholism**, infection of the varicella-zoster virus, **botulism**, and poliomyelitis. Peripheral neuropathy may develop as a primary symptom, or it may be due to another disease. For example, peripheral neuropathy is only one symptom of diseases such as amyloid neuropathy, certain cancers, or inherited neurologic disorders. Such diseases may affect the peripheral nervous system (PNS) and the central nervous system (CNS), as well as other body tissues.

To understand peripheral neuropathy and its underlying causes, it may be helpful to review the structures and arrangement of the PNS.

Nerve cells and nerves

Nerve cells are the basic building block of the nervous system. In the PNS, nerve cells can be threadlike—their width is microscopic, but their length can be measured in feet. The long, spidery extensions of nerve cells are called axons. When a nerve cell is stimulated, by touch or pain, for example, the message is carried along the axon, and neurotransmitters are released within the cell. Neurotransmitters are chemicals within the nervous system that direct nerve cell communication.

Certain nerve cell axons, such as the ones in the PNS, are covered with a substance called myelin. The myelin sheath may be compared to the plastic coating on electrical wires—it is there both to protect the cells and to prevent interference with the signals being transmitted. Protection is also given by Schwann cells, special cells within the nervous system that wrap around both myelinated and unmyelinated axons. The effect is similar to beads threaded on a necklace.

Nerve cell axons leading to the same areas of the body may be bundled together into nerves. Continuing the comparison to electrical wires, nerves may be compared to an electrical cord—the individual components are coated in their own sheaths and then encased together inside a larger protective covering.

Peripheral nervous system

The nervous system is classified into two parts: the CNS and the PNS. The CNS is made up of the brain and the spinal cord, and the PNS is composed of the nerves that lead to or branch off from the CNS.

The peripheral nerves handle a diverse array of functions in the body. This diversity is reflected in the major divisions of the PNS—the afferent and the efferent divisions. The afferent division is in charge of sending sensory information from the body to the CNS. When afferent nerve cell endings, called receptors, are stimulated, they release neurotransmitters. These neurotransmitters relay a signal to the brain, which interprets it and reacts by releasing other neurotransmitters.

Some of the neurotransmitters released by the brain are directed at the efferent division of the PNS. The efferent nerves control voluntary movements, such as moving the arms and legs, and involuntary movements, such as making the heart pump blood. The nerves controlling voluntary movements are

called motor nerves, and the nerves controlling involuntary actions are referred to as autonomic nerves. The afferent and efferent divisions continually interact with each other. For example, if a person were to touch a hot stove, the receptors in the skin would transmit a message of heat and pain through the sensory nerves to the brain. The message would be processed in the brain and a reaction, such as pulling back the hand, would be transmitted via a motor nerve.

Neuropathy

NERVE DAMAGE. When an individual has a peripheral neuropathy, nerves of the PNS have been damaged. Nerve damage can arise from a number of causes, such as disease, physical injury, poisoning, or malnutrition. These agents may affect either afferent or efferent nerves. Depending on the cause of damage, the nerve cell axon, its protective myelin sheath, or both may be injured or destroyed.

CLASSIFICATION. There are hundreds of peripheral neuropathies. Reflecting the scope of PNS activity, symptoms may involve sensory, motor, or autonomic functions. To aid in diagnosis and treatment, the symptoms are classified into principal neuropathic syndromes based on the type of affected nerves and how long symptoms have been developing. Acute development refers to symptoms that have appeared within days, and subacute refers to those that have evolved over a number of weeks. Early chronic symptoms are those that take months to a few years to develop, and late chronic symptoms have been present for several years.

The classification system is composed of six principal neuropathic syndromes, which are subdivided into more specific categories. By narrowing down the possible diagnoses in this way, specific medical tests can be used more efficiently and effectively. The six syndromes and a few associated causes are listed below:

- Acute motor paralysis, accompanied by variable problems with sensory and autonomic functions. Neuropathies associated with this syndrome are mainly accompanied by motor nerve problems, but the sensory and autonomic nerves may also be involved. Associated disorders include Guillain-Barré syndrome, diphtheritic polyneuropathy, and porphyritic neuropathy.
- Subacute sensorimotor paralysis. The term sensorimotor refers to neuropathies that are mainly characterized by sensory symptoms, but also have a minor component of motor nerve problems. Poisoning with heavy metals (e.g., lead, mercury, and arsenic),

chemicals, or drugs are linked to this syndrome. Diabetes, Lyme disease, and malnutrition are also possible causes.

- Chronic sensorimotor paralysis. Physical symptoms may resemble those in the above syndrome, but the time scale of symptom development is extended. This syndrome encompasses neuropathies arising from cancers, diabetes, leprosy, inherited neurologic and metabolic disorders, and hypothyroidism.
- Neuropathy associated with mitochondrial diseases. Mitochondria are organelles—structures within cells—responsible for handling a cell's energy requirements. If the mitochondria are damaged or destroyed, the cell's energy requirements are not met and it can die.
- Recurrent or relapsing polyneuropathy. This syndrome covers neuropathies that affect several nerves and may come and go, such as Guillain-Barré syndrome, porphyria, and chronic inflammatory demyelinating polyneuropathy.
- Mononeuropathy or plexopathy. Nerve damage associated with this syndrome is limited to a single nerve or a few closely associated nerves. Neuropathies related to physical injury to the nerve, such as carpal tunnel syndrome and sciatica, are included in this syndrome.

Causes and symptoms

Typical symptoms of neuropathy are related to the type of affected nerve. If a sensory nerve is damaged, common symptoms include **numbness**, **tingling** in the area, a prickling sensation, or pain. Pain associated with neuropathy can be quite intense and may be described as cutting, stabbing, crushing, or burning. In some cases, a nonpainful stimulus may be perceived as excruciating or pain may be felt even in the absence of a stimulus. Damage to a motor nerve is usually indicated by weakness in the affected area. If the problem with the motor nerve has continued over a length of time, muscle shrinkage (atrophy) or lack of muscle tone may be noticeable. Autonomic nerve damage is most noticeable when an individual stands upright and experiences problems such as light-headedness or changes in blood pressure. Other indicators of autonomic nerve damage are lack of sweat, tears, and saliva; **constipation**; urinary retention; and **impotence**. In some cases, heart beat irregularities and respiratory problems can develop.

Symptoms may appear over days, weeks, months, or years. Their duration and the ultimate outcome of the neuropathy are linked to the cause of the nerve damage. Potential causes include diseases, physical

injuries, poisoning, and malnutrition or alcohol **abuse**. In some cases, neuropathy is not the primary disorder, but a symptom of an underlying disease.

Disease

Diseases that cause peripheral neuropathies may either be acquired or inherited; in some cases, it is difficult to make that distinction. The diabetes-peripheral neuropathy link has been well established. A typical pattern of diabetes-associated neuropathic symptoms includes sensory effects that first begin in the feet. The associated pain or pins-and-needles, burning, crawling, or prickling sensations form a typical “stocking” distribution in the feet and lower legs. Other diabetic neuropathies affect the autonomic nerves and have potentially fatal cardiovascular complications.

Several other metabolic diseases have a strong association with peripheral neuropathy. Uremia, or **chronic kidney failure**, carries a 10–90% risk of eventually developing neuropathy, and there may be an association between liver failure and peripheral neuropathy. Accumulation of lipids inside blood vessels (**atherosclerosis**) can choke-off blood supply to certain peripheral nerves. Without oxygen and nutrients, the nerves slowly die. Mild polyneuropathy may develop in persons with low thyroid hormone levels. Individuals with abnormally enlarged skeletal extremities (acromegaly), caused by an overabundance of growth hormone, may also develop mild polyneuropathy.

Neuropathy can also result from severe vasculitides, a group of disorders in which blood vessels are inflamed. When the blood vessels are inflamed or damaged, blood supply to the nerve can be affected, injuring the nerve.

Both viral and bacterial infections have been implicated in peripheral neuropathy. Leprosy is caused by the bacteria *M. leprae*, which directly attack sensory nerves. Other bacterial illness may set the stage for an immune-mediated attack on the nerves. For example, one theory about Guillain-Barré syndrome involves complications following infection with *Campylobacter jejuni*, a bacterium commonly associated with **food poisoning**. This bacterium carries a protein that closely resembles components of myelin. The immune system launches an attack against the bacteria; but, according to the theory, the immune system confuses the myelin with the bacteria in some cases and attacks the myelin sheath as well. The underlying cause of neuropathy associated with Lyme disease is unknown; the bacteria may either promote

an immune-mediated attack on the nerve or inflict damage directly.

Infection with certain viruses is associated with extremely painful sensory neuropathies. A primary example of such a neuropathy is caused by **shingles**. After a case of **chickenpox**, the causative virus, varicella-zoster virus, becomes inactive in sensory nerves. Years later, the virus may be reactivated. Once reactivated, it attacks and destroys axons. Infection with HIV is also associated with peripheral neuropathy, but the type of neuropathy that develops can vary. Some HIV-linked neuropathies are noted for myelin destruction rather than axonal degradation. Also, HIV infection is frequently accompanied by other infections, both bacterial and viral, that are associated with neuropathy.

Several types of peripheral neuropathies are associated with inherited disorders. These inherited disorders may primarily involve the nervous system, or the effects on the nervous system may be secondary to an inherited metabolic disorder. Inherited neuropathies can fall into several of the principal syndromes, because symptoms may be sensory, motor, or autonomic. The inheritance patterns also vary, depending on the specific disorder. The development of inherited disorders is typically drawn out over several years and may herald a degenerative condition—that is, a condition that becomes progressively worse over time. Even among specific disorders, there may be a degree of variability in inheritance patterns and symptoms. For example, Charcot-Marie-Tooth disease is usually inherited as an autosomal dominant disorder, but it can be autosomal recessive or, in rare cases, linked to the X chromosome. Its estimated frequency is approximately one in 2,500 people. Age of onset and sensory nerve involvement can vary between cases. The main symptom is a degeneration of the motor nerves in legs and arms, and resultant muscle atrophy. Other inherited neuropathies have a distinctly metabolic component. For example, in familial amyloid polyneuropathies, protein components that make up the myelin are constructed and deposited incorrectly.

Physical injury

Accidental falls and mishaps during sports and recreational activities are common causes of physical injuries that can result in peripheral neuropathy. The common types of injuries in these situations occur from placing too much pressure on the nerve, exceeding the nerve's capacity to stretch, blocking adequate blood supply of oxygen and nutrients to the nerve, and tearing the nerve. Pain may not always be immediately noticeable, and obvious signs of damage may take a while to develop.

These injuries usually affect one nerve or a group of closely associated nerves. For example, a common injury encountered in contact sports such as football is the "burner," or "stinger," syndrome. Typically, a stinger is caused by overstretching the main nerves that span from the neck into the arm. Immediate symptoms are numbness, tingling, and pain that travels down the arm, lasting only a minute or two. A single incident of a stinger is not dangerous, but recurrences can eventually cause permanent motor and sensory loss.

Poisoning

The poisons, or toxins, that cause peripheral neuropathy include drugs, industrial chemicals, and environmental toxins. Neuropathy that is caused by drugs usually involves sensory nerves on both sides of the body, particularly in the hands and feet, and pain is a common symptom. Neuropathy is an unusual side effect of medications; therefore, most people can use these drugs safely. A few of the drugs that have been linked with peripheral neuropathy include metronidazole, an antibiotic; phenytoin, an anticonvulsant; and simvastatin, a cholesterol-lowering medication.

Certain industrial chemicals have been shown to be poisonous to nerves (neurotoxic) following work-related exposures. Chemicals such as acrylamide, allyl chloride, and carbon disulfide have all been strongly linked to development of peripheral neuropathy. Organic compounds, such as N-hexane and toluene, are also encountered in work-related settings, as well as in glue-sniffing and solvent abuse. Either route of exposure can produce severe sensorimotor neuropathy that develops rapidly.

Heavy metals are the third group of toxins that cause peripheral neuropathy. Lead, arsenic, thallium, and mercury usually are not toxic in their elemental form, but rather as components in organic or inorganic compounds. The types of metal-induced neuropathies vary widely. Arsenic poisoning may mimic Guillain-Barré syndrome; lead affects motor nerves more than sensory nerves; thallium produces painful sensorimotor neuropathy; and the effects of mercury are seen in both the CNS and PNS.

Malnutrition and alcohol abuse

Burning, stabbing pains and numbness in the feet, and sometimes in the hands, are distinguishing features of alcoholic neuropathy. The level of alcohol consumption associated with this variety of peripheral neuropathy has been estimated as approximately 3 L of beer or 300 mL of liquor daily for three years. However, it is unclear whether alcohol alone is responsible for the

neuropathic symptoms, because chronic alcoholism is strongly associated with malnutrition.

Malnutrition refers to an extreme lack of nutrients in the diet. It is unknown precisely which nutrient deficiencies cause peripheral neuropathies in alcoholics and famine and **starvation** patients, but it is suspected that the B **vitamins** have a significant role. For example, thiamine (vitamin B₁) deficiency is the cause of **beriberi**, a neuropathic disease characterized by **heart failure** and painful polyneuropathy of sensory nerves. **Vitamin E deficiency** seems to have a role in both CNS and PNS neuropathy.

Diagnosis

Clinical symptoms can indicate peripheral neuropathy, but an exact diagnosis requires a combination of medical history, medical tests, and possibly a process of exclusion. Certain symptoms can suggest a diagnosis, but more information is commonly needed. For example, painful, burning feet may be a symptom of alcohol abuse, diabetes, HIV infection, or an underlying malignant tumor, among other causes. Without further details, effective treatment would be difficult.

During a **physical examination**, an individual is asked to describe the symptoms very carefully. Detailed information about the location, nature, and duration of symptoms can help exclude some causes or even pinpoint the actual problem. The person's medical history may also provide clues as to the cause, because certain diseases and medications are linked to specific peripheral neuropathies. A medical history should also include information about diseases that run in the family, because some peripheral neuropathies are genetically linked. Information about hobbies, recreational activities, alcohol consumption, and work place activities can uncover possible injuries or exposures to poisonous substances.

The physical examination also includes blood tests, such as those that check levels of glucose and creatinine to detect diabetes and kidney problems, respectively. A blood count is also done to determine levels of different blood cell types. Iron, vitamin B₁₂, and other factors may be measured as well, to rule out malnutrition. More specific tests, such as an assay for heavy metals or poisonous substances, or tests to detect **vasculitis**, are not typically done unless there is reason to suspect a particular cause.

An individual with neuropathy may be sent to a doctor that specializes in nervous system disorders (neurologist). By considering the results of the physical examination and observations of the referring doctor, the neurologist may be able to narrow down

the possible diagnoses. Additional tests, such as nerve conduction studies and **electromyography**, which tests muscle reactions, can confirm that nerve damage has occurred and may also be able to indicate the nature of the damage. For example, some neuropathies are characterized by destruction of the myelin. This type of damage is shown by slowed nerve conduction. If the axon itself has suffered damage, the nerve conduction may be slowed, but it will also be diminished in strength. Electromyography adds further information by measuring nerve conduction and muscle response, which determines whether the symptoms are due to a neuropathy or to a muscle disorder.

In approximately 10% of peripheral neuropathy cases, a nerve biopsy may be helpful. In this test, a small part of the nerve is surgically removed and examined under a microscope. This procedure is usually the most helpful in confirming a suspected diagnosis, rather than as a diagnostic procedure by itself.

Treatment

Treat the cause

Attacking the underlying cause of the neuropathy can prevent further nerve damage and may allow for a better recovery. For example, in cases of bacterial infection such as leprosy or Lyme disease, **antibiotics** may be given to destroy the infectious bacteria. Viral infections are more difficult to treat, because antibiotics are not effective against them. Neuropathies associated with drugs, chemicals, and toxins are treated in part by stopping exposure to the damaging agent. Chemicals such as ethylenediaminetetraacetic acid (EDTA) are used to help the body concentrate and excrete some toxins. Diabetic neuropathies may be treated by gaining better control of blood sugar levels, but chronic kidney failure may require dialysis or even kidney transplant to prevent or reduce nerve damage. In some cases, such as compression injury or tumors, surgery may be considered to relieve pressure on a nerve.

In a crisis situation, as in the onset of Guillain-Barré syndrome, plasma exchange, intravenous immunoglobulin, and **steroids** may be given. Intubation, in which a tube is inserted into the trachea to maintain an open airway, and ventilation may be required to support the respiratory system. Treatment may focus more on symptom management than on combating the underlying cause, at least until a definitive diagnosis has been made.

Supportive care and long-term therapy

Some peripheral neuropathies cannot be resolved or require time for resolution. In these cases, long-term

KEY TERMS

Afferent—Refers to peripheral nerves that transmit signals to the spinal cord and the brain. These nerves carry out sensory function.

Autonomic—Refers to peripheral nerves that carry signals from the brain and that control involuntary actions in the body, such as the beating of the heart.

Autosomal dominant or autosomal recessive—

Refers to the inheritance pattern of a gene on a chromosome other than X or Y. Genes are inherited in pairs—one gene from each parent. However, the inheritance may not be equal, and one gene may overshadow the other in determining the final form of the encoded characteristic. The gene that overshadows the other is called the dominant gene; the overshadowed gene is the recessive one.

Axon—A long, threadlike projection that is part of a nerve cell.

Central nervous system (CNS)—The part of the nervous system that includes the brain and the spinal cord.

Efferent—Refers to peripheral nerves that carry signals away from the brain and spinal cord. These nerves carry out motor and autonomic functions.

Electromyography—A medical test that assesses nerve signals and muscle reactions. It can determine if there is a disorder with the nerve or if the muscle is not capable of responding.

Inheritance pattern—Refers to dominant or recessive inheritance.

Motor—Refers to peripheral nerves that control voluntary movements, such as moving the arms and legs.

Myelin—The protective coating on axons.

Nerve biopsy—A medical test in which a small portion of a damaged nerve is surgically removed and examined under a microscope.

Nerve conduction—The speed and strength of a signal being transmitted by nerve cells. Testing these factors can reveal the nature of nerve injury, such as damage to nerve cells or to the protective myelin sheath.

Neurotransmitter—Chemicals within the nervous system that transmit information from or between nerve cells.

Peripheral nervous system (PNS)—Nerves that are outside of the brain and spinal cord.

Sensory—Refers to peripheral nerves that transmit information from the senses to the brain.

monitoring and supportive care is necessary. Medical tests may be repeated to chart the progress of the neuropathy. If autonomic nerve involvement is a concern, regular monitoring of the cardiovascular system may be carried out.

Because pain is associated with many of the neuropathies, a **pain management** plan may need to be mapped out, especially if the pain becomes chronic. As in any chronic disease, **narcotics** are best avoided. Agents that may be helpful in neuropathic pain include amitriptyline, carbamazepine, and capsaicin cream. **Physical therapy** and physician-directed exercises can help maintain or improve function. In cases in which motor nerves are affected, braces and other supportive equipment can aid an individual's ability to move about.

Prognosis

The outcome for peripheral neuropathy depends heavily on the cause. Peripheral neuropathy ranges

from a reversible problem to a potentially fatal complication. In the best cases, a damaged nerve regenerates. Nerve cells cannot be replaced if they are killed, but they are capable of recovering from damage. The extent of recovery is tied to the extent of the damage and a person's age and general health status. Recovery can take weeks to years, because neurons grow very slowly. Full recovery may not be possible and it may also not be possible to determine the prognosis at the outset.

If the neuropathy is a degenerative condition, such as Charcot-Marie-Tooth disease, an individual's condition will become worse. There may be periods of time when the disease seems to reach a plateau, but cures have not yet been discovered for many of these degenerative diseases. Therefore, continued symptoms, potentially worsening to disabilities are to be expected.

A few peripheral neuropathies are eventually fatal. Fatalities have been associated with some cases

of **diphtheria**, botulism, and others. Some diseases associated with neuropathy may also be fatal, but the ultimate cause of **death** is not necessarily related to the neuropathy, such as with **cancer**.

Prevention

Peripheral neuropathies are preventable only to the extent that the underlying causes are preventable. Steps that a person can take to prevent potential problems include vaccines against diseases that cause neuropathy, such as **polio** and diphtheria. Treatment for physical injuries in a timely manner can help prevent permanent or worsening damage to nerves. Precautions when using certain chemicals and drugs are well advised in order to prevent exposure to neurotoxic agents. Control of chronic diseases such as diabetes may also reduce the chances of developing peripheral neuropathy.

Although not a preventive measure, genetic screening can serve as an early warning for potential problems. Genetic screening is available for some inherited conditions, but not all. In some cases, presence of a particular gene may not mean that a person will necessarily develop the disease, because there may be environmental and other components involved.

ORGANIZATIONS

American Diabetes Association, 1701 North Beauregard Street, Alexandria, VA, 22311, (800) 342-2383, AskADA @diabetes.org, <http://www.diabetes.org/>.
 The Myelin Project, P.O. Box 39, Pacific Palisades, 90272-0039, (310) 459-6218, (310) 230-4298, patti.chapman@myelin.org, <http://www.myelin.org/contact>.
 The Neuropathy Association, Inc, 60 East 42nd Street, Suite 942, New York, NY, 10165, (212) 692-0662, (212) 692-0668, info@neuropathy.org, <http://www.neuropathy.org>.

Julia Barrett

Peripheral vascular disease

Definition

Peripheral **vascular disease** is a narrowing of blood vessels that restricts blood flow. It mostly occurs in the legs, but is sometimes seen in the arms.

Description

Peripheral vascular disease includes a group of diseases in which blood vessels become restricted or blocked. Typically, the patient has peripheral vascular

disease from **atherosclerosis**. Atherosclerosis is a disease in which fatty plaques form in the inside walls of blood vessels. Other processes, such as **blood clots**, further restrict blood flow in the blood vessels. Both veins and arteries may be affected, but the disease is usually arterial. All the symptoms and consequences of peripheral vascular disease are related to restricted blood flow. Peripheral vascular disease is a progressive disease that can lead to **gangrene** of the affected area. Peripheral vascular disease may also occur suddenly if an **embolism** occurs or when a blot clot rapidly develops in a blood vessel already restricted by an atherosclerotic plaque, and the blood flow is quickly cut off.

Causes and symptoms

There are many causes of peripheral vascular disease. One major risk factor is **smoking** cigarettes. Other diseases predispose patients to develop peripheral vascular disease. These include diabetes, **Buerger's disease**, **hypertension**, and **Raynaud's disease**. The main symptom is **pain** in the affected area. Early symptoms include an achy, tired sensation in the affected muscles. Since this disease is seen mainly in the legs, these sensations usually occur when walking. The symptoms may disappear when resting. As the disease becomes worse, symptoms occur even during light exertion and, eventually, occur all the time, even at rest. In the severe stages of the disease the leg and foot may be cold to the touch and will feel numb. The skin may become dry and scaly. If the leg is even slightly injured, ulcers may form because, without a good blood supply, proper healing cannot take place. At the most severe stage of the disease, when the blood flow is greatly restricted, gangrene can develop in those areas lacking blood supply. In some cases, peripheral vascular disease occurs suddenly. This happens when an embolism rapidly blocks blood flow to a blood vessel. The patient will experience a sharp pain, followed by a loss of sensation in the affected area. The limb will become cold and numb, and lose color or turn bluish.

Diagnosis

Peripheral vascular disease can be diagnosed by comparing blood pressures taken above and below the point of pain. The area below the pain (downstream from the obstruction) will have a much lower or undetectable blood pressure reading. **Doppler ultrasonography** and **angiography** can also be used to diagnose and define this disease.

KEY TERMS

Embolism—The blockage of a blood vessel by air, blood clot, or other foreign body.

Plaque—A deposit, usually of fatty material, on the inside wall of a blood vessel.

Treatment

If the person is a smoker, they should stop smoking immediately. **Exercise** is essential to treating this disease. The patient should walk until pain appears, rest until the pain disappears, and then resume walking. The amount of walking a patient can do should increase gradually as the symptoms improve. Ideally, the patient should walk 30–60 minutes per day. Infections in the affected area should be treated promptly. Surgery may be required to attempt to treat clogged blood vessels. Limbs with gangrene must be amputated to prevent the **death** of the patient.

Prognosis

The prognosis depends on the underlying disease and the stage at which peripheral vascular disease is discovered. Removal of risk factors, such as smoking, should be done immediately. In many cases, peripheral vascular disease can be treated successfully but coexisting cardiovascular problems may ultimately prove to be fatal.

Resources

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- Zimring, Michael P., MD. *Healthy Travel: Don't Travel Without It!* Laguna Beach, CA: Basic Health Publications, Inc., 2009.

OTHER

- Avoid Deep Vein Thrombosis: Keep the Blood Flowing.* MedicineNet Website, 2010. www.medicinenet.com/script/main/art.asp?articlekey=40582.

ORGANIZATIONS

American Heart Association, 7272 Greenville Ave., Dallas, TX, 75231, (301) 223-2307, (800) 242-8721, <http://www.americanheart.org>.

Centers for Disease Control (CDC). Division for Heart Disease and Stroke Prevention, 4770 Buford Hwy NE, Atlanta, GA, 30341-3717, 770-488-2424, www.cdc.gov/cholesterol/faqs.htm.

National Heart, Lung, and Blood Institute, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (204) 629-3246, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov>.

Society of Interventional Radiology, 10201 Lee Highway, Suite 500, Fairfax, VA, 22030, 703-691-1805, <http://www.sirweb.org>.

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Peritoneal dialysis see **Dialysis, kidney**

Peritoneal endoscopy see **Laparoscopy**

Peritoneal fluid analysis see **Paracentesis**

Peritonitis

Definition

Peritonitis is an inflammation of the membrane which lines the inside of the abdomen and all of the internal organs. This membrane is called the peritoneum.

Description

Peritonitis may be primary (meaning that it occurs spontaneously, and not as the result of some other medical problem) or secondary (meaning that it results from some other condition). It is most often due to infection by bacteria, but may also be due to some kind of a chemical irritant (such as spillage of acid from the stomach, bile from the gall bladder and biliary tract, or enzymes from the pancreas during the illness called **pancreatitis**). Peritonitis has even been seen in patients who develop a reaction to the cornstarch used to powder gloves worn during surgery. Peritonitis with no evidence of bacteria, chemical irritant, or foreign body has occurred in such diseases as **systemic lupus erythematosus**, porphyria, and **familial Mediterranean fever**. When the peritoneum is contaminated by blood, the blood can both irritate the peritoneum and serve as a source of bacteria to cause an infection. Blood may leak into the abdomen due to a burst tubal **pregnancy**, an injury, or bleeding after surgery.

Causes and symptoms

Primary peritonitis usually occurs in people who have an accumulation of fluid in their abdomens (**ascites**). Ascites is a common complication of severe **cirrhosis** of the liver (a disease in which the liver grows increasingly scarred and dysfunctional). The fluid that accumulates creates a good environment for the growth of bacteria.

Secondary peritonitis most commonly occurs when some other medical condition causes bacteria to spill into the abdominal cavity. Bacteria are normal residents of a healthy intestine, but they should have no way to escape and enter the abdomen, where they could cause an infection. Bacteria can infect the peritoneum due to conditions in which a hole (perforation) develops in the stomach (due to an ulcer eating its way through the stomach wall) or intestine (due to a large number of causes, including a ruptured appendix or a ruptured diverticulum). Bacteria can infect the peritoneum due to a severe case of **pelvic inflammatory disease** (a massive infection of the female organs, including the uterus and fallopian tubes). Bacteria can also escape into the abdominal cavity due to an injury that causes the intestine to burst, or an injury to an internal organ which bleeds into the abdominal cavity.

Symptoms of peritonitis include **fever** and abdominal **pain**. An acutely ill patient usually tries to lie very still, because any amount of movement causes excruciating pain. Often, the patient lies with the knees bent, to decrease strain on the tender peritoneum. There is often **nausea and vomiting**. The usual sounds made by the active intestine and heard during examination with a stethoscope will be absent, because the intestine usually stops functioning. The abdomen may be rigid and boardlike. Accumulations of fluid will be notable in primary peritonitis due to ascites. Other signs and symptoms of the underlying cause of secondary peritonitis may be present.

Diagnosis

A diagnosis of peritonitis is usually based on symptoms. Discovering the underlying reason for the peritonitis, however, may require some work. A blood sample will be drawn in order to determine the **white blood cell count**. Because white blood cells are produced by the body in an effort to combat foreign invaders, the white blood cell count will be elevated in the case of an infection. A long, thin needle can be used to take a sample of fluid from the abdomen in an effort to diagnose primary peritonitis. The types of immune cells present are usually

KEY TERMS

Ascites—An accumulation of fluid within the abdominal cavity.

Cirrhosis—A progressive liver disease in which the liver grows increasingly more scarred. The presence of scar tissue then interferes with liver function.

Diverticulum—An outpouching of the intestine.

Laparotomy—An open operation on the abdomen.

Pancreatitis—An inflammation of the pancreas.

Perforation—A hole.

Peritoneum—The membrane that lines the inside of the abdominal cavity, and all of the internal organs.

characteristic in this form of peritonitis. X-ray films may be taken if there is some suspicion that a perforation exists. In the case of a perforation, air will have escaped into the abdomen and will be visible on the picture. When a cause for peritonitis cannot be found, an open exploratory operation on the abdomen (laparotomy) is considered to be a crucial diagnostic procedure, and at the same time provides the opportunity to begin treatment.

Treatment

Treatment depends on the source of the peritonitis, but an emergency laparotomy is usually performed. Any perforated or damaged organ is usually repaired at this time. If a clear diagnosis of pelvic inflammatory disease or pancreatitis can be made, however, surgery is not usually performed. Peritonitis from any cause is treated with **antibiotics** given through a needle in the vein, along with fluids to prevent **dehydration**.

Prognosis

Prognosis for untreated peritonitis is poor, usually resulting in **death**. With treatment, the prognosis is variable, dependent on the underlying cause.

Prevention

There is no way to prevent peritonitis, since the diseases it accompanies are usually not under the voluntary control of an individual. However, prompt treatment can prevent complications.

Resources

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Fauci, Anthony S., et al., eds. *Harrison's Principles of Internal Medicine*. 17th ed. New York: McGraw-Hill Professional, 2008.

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Permanent pacemakers see **Pacemakers**

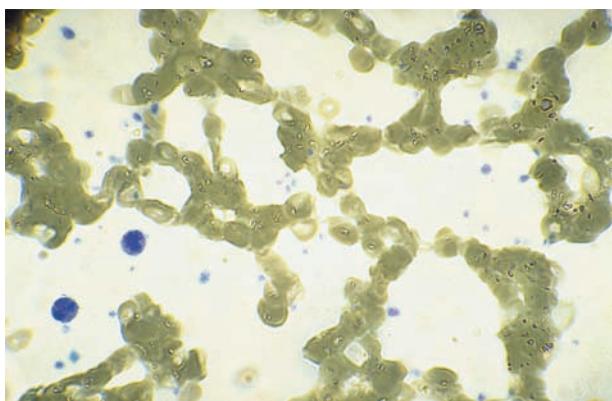
Pernicious anemia

Definition

Pernicious anemia is a disease in which the red blood cells are abnormally formed, due to an inability to absorb vitamin B₁₂. True pernicious anemia refers specifically to a disorder of atrophied parietal cells leading to absent intrinsic factor, resulting in an inability to absorb B₁₂.

Description

Vitamin B₁₂, or cobalamin, plays an important role in the development of red blood cells. It is found in significant quantities in liver, meats, milk and milk products, and legumes. During the course of the digestion of foods containing B₁₂, the B₁₂ becomes attached to a substance called intrinsic factor. Intrinsic factor is produced by parietal cells that line the stomach. The B₁₂-intrinsic factor complex then enters the intestine, where the vitamin is absorbed into the bloodstream. In fact, B₁₂ can only be absorbed when it is attached to intrinsic factor.



A smear of red blood cells indicating folic acid (vitamin B₁₂) deficiency. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

In pernicious anemia, the parietal cells stop producing intrinsic factor. The intestine is then completely unable to absorb B₁₂. So, the vitamin passes out of the body as waste. Although the body has significant amounts of stored B₁₂, this will eventually be used up. At this point, the symptoms of pernicious anemia will develop.

Pernicious anemia is most common among people from northern Europe and among African Americans. It is far less frequently seen among people from southern Europe and Asia. Pernicious anemia occurs in equal numbers in both men and women. Most patients with pernicious anemia are older, usually over 60. Occasionally, a child will have an inherited condition that results in defective intrinsic factor. Pernicious anemia seems to run in families, so that anyone with a relative diagnosed with the disease has a greater likelihood of developing it as well.

Causes and symptoms

Intrinsic factor is produced by specialized cells within the stomach called parietal cells. When these parietal cells shrink in size (atrophy), they produce less and less intrinsic factor. Eventually, the parietal cells stop functioning altogether. Other important products of parietal cells are also lessened, including stomach acid, and an enzyme involved in the digestion of proteins.

People with pernicious anemia seem to have a greater chance of having certain other conditions. These conditions include **autoimmune disorders**, particularly those affecting the thyroid, parathyroid, and adrenals. It is thought that the immune system, already out of control in these diseases, incorrectly becomes directed against the parietal cells. Ultimately, the parietal cells seem to be destroyed by the actions of the immune system.

As noted, true pernicious anemia refers specifically to a disorder of atrophied parietal cells leading to absent intrinsic factor, resulting in an inability to absorb B₁₂. However, there are other related conditions that result in decreased absorption of B₁₂. These conditions cause the same types of symptoms as true pernicious anemia. Other conditions that interfere with either the production of intrinsic factor, or the body's use of B₁₂, include conditions that require surgical removal of the stomach, or poisonings with corrosive substances which destroy the lining of the stomach. Certain structural defects of the intestinal system can result in an overgrowth of normal bacteria. These bacteria then absorb B₁₂ themselves, for use in their own growth. Intestinal worms (especially one

KEY TERMS

Anemia—A condition in which those elements of the blood responsible for oxygen delivery throughout the body (red blood cells, hemoglobin) are decreased in quantity or defective in some way.

Atrophy—Refers to the shrinking in size of an organ or cell.

Autoimmune disorder—A disorder in which the immune system, (responsible for fighting off such foreign invaders as bacteria and viruses), begins to attack and damage a part of the body as if it were foreign.

Hematopoietic system—The system in the body which is responsible for the production of blood cells.

Intrinsic factor—A substance produced by the parietal cells of the stomach. In order to be absorbed by the intestine, vitamin B₁₂ must form a complex with intrinsic factor.

Parietal cells—Specific cells which line the inside of the stomach. These cells are responsible for producing intrinsic factor and hydrochloric acid.

Reticulocyte—An early, immature form of a red blood cell. Over time, the reticulocyte develops to become a mature, oxygen-carrying red blood cell.

called fish tapeworm) may also use B₁₂, resulting in anemia. Various conditions that affect the first part of the intestine (the ileum), from which B₁₂ is absorbed, can also cause anemia due to B₁₂ deficiency. These ileum-related disorders include tropical sprue, Whipple's disease, **Crohn's disease**, **tuberculosis**, and the Zollinger-Ellison syndrome.

Symptoms of pernicious anemia and decreased B₁₂ affect three systems of the body: the system that is involved in the formation of blood cells (hematopoietic system); the gastrointestinal system; and the nervous system.

The hematopoietic system is harmed because B₁₂ is required for the proper formation of red blood cells. Without B₁₂, red blood cell production is greatly reduced. Those red blood cells that are produced are abnormally large and abnormal in shape. Because red blood cells are responsible for carrying oxygen around the body, decreased numbers (termed anemia) result in a number of symptoms, including **fatigue**, **dizziness**, ringing in the ears, pale or yellowish skin, fast heart rate, enlarged heart with an abnormal heart sound (murmur) evident on examination, and chest **pain**.

Symptoms that affect the gastrointestinal system include a sore and brightly red tongue, loss of appetite, weight loss, **diarrhea**, and abdominal cramping.

The nervous system is severely affected when pernicious anemia goes untreated. Symptoms include **numbness**, **tingling**, or burning in the arms, legs, hands, and feet; muscle weakness; difficulty and loss of balance while walking; changes in reflexes; irritability, confusion, and depression.

Diagnosis

Diagnosis of pernicious anemia is suggested when a blood test reveals abnormally large red blood cells. Many of these will also be abnormally shaped. The earliest, least mature forms of red blood cells (reticulocytes) will also be low in number. White blood cells and platelets may also be decreased in number. Measurements of the quantity of B₁₂ circulating in the bloodstream will be low.

Once these determinations are made, it will be important to diagnose the cause of the anemia. True pernicious anemia means that the parietal cells of the stomach are atrophied, resulting in decreased production of intrinsic factor. This diagnosis is made by the Schilling test. In this test, a patient is given radioactive B₁₂ under two different sets of conditions: once alone, and once attached to intrinsic factor. Normally, large amounts of B₁₂ are absorbed through the intestine, then circulate through the blood, and enter the kidneys, where a certain amount of B₁₂ is then passed out in the urine. When a patient has pernicious anemia, the dose of B₁₂ given by itself will not be absorbed by the intestine, so it will not pass into the urine. Therefore, levels of B₁₂ in the urine will be low. When the B₁₂ is given along with intrinsic factor, the intestine is able to absorb the vitamin. Urine levels of B₁₂ will therefore be higher.

Treatment

Treatment of pernicious anemia requires the administration of lifelong injections of B₁₂. Vitamin B₁₂ given by injection enters the bloodstream directly, and does not require intrinsic factor. At first, injections may need to be given several times a week, in

order to build up adequate stores of the vitamin. After this, the injections can be given on a monthly basis. Other substances required for blood cell production may also need to be given, iron and vitamin C.

Prognosis

Prognosis is generally good for patients with pernicious anemia. Many of the symptoms improve within just a few days of beginning treatment, although some of the nervous system symptoms may take up to 18 months to improve. Occasionally, when diagnosis and treatment have been delayed for a long time, some of the nervous system symptoms may be permanent.

Because an increased risk of **stomach cancer** has been noted in patients with pernicious anemia, careful monitoring is necessary, even when all the symptoms of the original disorder have improved.

Resources

BOOKS

Fauci, Anthony S., et al., eds. *Harrison's Principles of Internal Medicine*. 17th ed. New York: McGraw-Hill Professional, 2008.

Rosalyn Carson-DeWitt, MD

Peroneal muscular atrophy see
Charcot-Marie-Tooth disease

Peroxisomal disorders

Definition

Peroxisomal disorders are a group of congenital (existing from birth) diseases characterized by the absence of normal peroxisomes in the cells of the body. Peroxisomes are special parts (organelles) within a cell that contain enzymes responsible for critical cellular processes, including oxidation of fatty acids, biosynthesis of membrane phospholipids (plasmalogens), cholesterol, and bile acids, conversion of amino acids into glucose, reduction of hydrogen peroxide by catalase, and prevention of excess synthesis of oxalate (which can form crystals with **calcium**, resulting in **kidney stones**). Peroxisomal disorders are subdivided into two major categories. The first are disorders resulting from a failure to form intact, normal peroxisomes, resulting in multiple metabolic abnormalities, which are referred to as peroxisome biogenesis disorders (PBD) or as generalized

peroxisomal disorders. The second category includes those disorders resulting from the deficiency of a single peroxisomal enzyme. There are about 25 peroxisomal disorders known, although the number of diseases that are considered to be separate, distinct peroxisomal disorders varies among researchers and health care practitioners.

Description

A cell can contain several hundred peroxisomes, which are round or oval bodies with diameters of about 0.5 micron, that contain proteins that function as enzymes in metabolic processes. By definition, a peroxisome must contain catalase, which is an enzyme that breaks down hydrogen peroxide.

Approximately 50 different biochemical reactions occur entirely or partially within a peroxisome. Some of the processes are anabolic, or constructive, resulting in the synthesis of essential biochemical compounds, including bile acids, cholesterol, plasmalogens, and docosahexanoic acid (DHA), which is a long chain fatty acid that is a component of complex lipids, including the membranes of the central nervous system. Other reactions are catabolic, or destructive, and lead to the destruction of some fatty acids, including very long chain fatty acids (VLCFAs, fatty acids with more than 22 carbon atoms in their chains), phytanic acid, pipecolic acid, and the prostoglandins. The peroxisome is involved in breaking down VLCFAs to lengths that the body can use or get rid of.

When VLCFAs accumulate due to abnormal functioning of the peroxisomes, they are disruptive to the structure and stability of certain cells, especially those associated with the central nervous system and the myelin sheath, which is the fatty covering of nerve fibers. The peroxisomal disorders that include effects on the growth of the myelin sheath are considered to be part of a group of genetic disorders referred to as leukodystrophies. While metachromatic leukodystrophy (MLD) usually has its onset in infants or juveniles, there have been reports of its onset in young adults.

There are many other metabolic deficiencies that can occur in those who have peroxisomal disorders, which result in other types of detrimental effects, and together result in the abnormalities associated with the peroxisomal disorders. Unfortunately, it is not known how these abnormalities, and combinations of abnormalities, cause the disabilities seen in those afflicted with the disease.

Peroxisomal disorders form a heterogeneous disease group, with different degrees of severity. Included in the group referred to as PBD are:

- Zellweger syndrome (ZS), which is usually fatal within the first year of life,
- neonatal adrenoleukodystrophy (NALD), which is usually fatal within the first 10 years,
- infantile Refsum disease (IRD), which is not as devastating as ZS and NALD, as the children with this disorder with time and patience can develop some degree of motor, cognitive, and communication skills, although death generally occurs during the second decade of life.
- rhizomelic chondrodyplasia punctata (RCDP), which in its most severe form is fatal within the first year or two of life. However, survival into the teens has been known to occur. It is characterized by shortening of the proximal limbs (i.e., the legs from knee to foot, and the arms from elbow to hand).
- Zellweger-like syndrome, which is fatal in infancy, and is known to be a defect of three particular enzymes.

The differences among these disorders are continuous, with overlap between abnormalities. The range of disease abnormalities may be a result of a corresponding range of peroxisome failure; that is, in severe cases of ZS, the failure is nearly complete, while in IRD, there is some degree of peroxisome activity.

In peroxisomal single-enzyme disorders, the peroxisome is intact and functioning, but there is a defect in only one enzymatic process, with only one corresponding biochemical abnormality. However, these disorders can be as severe as those in which peroxisomal activity is nearly or completely absent.

X-linked **adrenoleukodystrophy** (X-ALD) is the most common of the peroxisomal disorders, affecting about one in 20,000 males. It is estimated that there are about 1,400 people in the United States with the disorder. In X-ALD there is a deficiency in the enzyme that breaks down VLCFAs, which then accumulate in the myelin and adrenal glands. Onset of X-ALD-related neurological symptoms occurs at about five–12 years of age, with **death** occurring within one to 10 years after onset of symptoms. In addition to physical abnormalities seen in other types of peroxisomal disorders, common symptoms of X-ALD also include behavioral changes such as abnormal withdrawal or aggression, poor memory, **dementia**, and poor academic performance. Other symptoms are muscle weakness and difficulties with hearing, speech, and vision. As the disease progresses, muscle tone deteriorates, swallowing becomes difficult and the patient becomes comatose. Unless treated with a diet that includes Lorenzo's oil, the disease will result in **paralysis**, **hearing loss**, blindness, **vegetative state**, and death. There are also milder forms of X-ALD: an adult onset ALD that typically begins between the

ages of 21 and 35, and a form that is occasionally seen in women who are carriers of the disorder. In addition to X-ALD, there are at least 10 other single-enzyme peroxisomal disorders, each with its own specific abnormalities.

Causes and symptoms

Most peroxisomal disorders are inherited autosomal recessive diseases, with X-ALD as an exception. They occur in all countries, among all races and ethnic groups. They are extremely rare, with frequencies reported at one in 30,000 to one in 150,000, although these numbers are only estimates.

In general, developmental delay, **mental retardation**, and vision and hearing impairment are common in those who have these disorders. Acquisition of speech appears to be especially difficult, and because of the reduced communication abilities, **autism** is common in those who live longer. Peroxisomal disorder patients have decreased muscle tone (hypotonia), which in the most severe cases is generalized, while in less severe cases, is usually restricted to the neck and trunk muscles. Sometimes this lack of control is only noticeable by a curved back in the sitting position. Head control and independent sitting is delayed, with most patients unable to walk independently.

Failure to thrive is a common characteristic of patients with peroxisomal disorder, along with an enlarged liver, abnormalities in liver enzyme function, and loss of fats in stools (steatorrhea).

Peroxisomal disorders are also associated with facial abnormalities, including high forehead, frontal bossing (swelling), small face, low set ears, and slanted eyes. These characteristics may not be prominent in some children, and are especially difficult to identify in an infant.

Diagnosis

Since hearing and vision deficiencies may be difficult to identify in infants, peroxisomal disorders are usually detected by observations of failure to thrive, hypotonia, mental retardation, widely open fontanel, abnormalities in liver enzymes, and an enlarged liver. If peroxisomal disorders are suspected, blood plasma assays for VLCFAs, phytanic acid, and pipecolic acid are conducted. Additional tests include plasmalogen biosynthesis potential.

Treatment

For many of the peroxisomal disorders, there is no standard course of treatment, with supportive treatment

KEY TERMS

Autosomal recessive inheritance—Two copies of an altered gene located on one of the autosomes must be present for an individual to be affected with the trait or condition determined by that gene. An affected individual (homozygote) has two parents who are unaffected but each parent carries the altered gene (heterozygote). The risk of two heterozygotes, or carriers, having an affected child is 25%, one in four, for each child that they have; similarly, there is a three in four chance that each child will not be affected. Males and females are at equal risk for being affected. Two affected individuals usually produce children, all of whom are affected as well.

Autosome—A chromosome not involved in sex determination.

Fontanel—One of the membranous intervals between the uncompleted angles of the parietal and neighboring bones of a fetal or young skull; so called because it exhibits a rhythmical pulsation.

Metabolic—Relating to the chemical changes in living cells.

Organelle—Specialized structure within a cell, which is separated from the rest of the cell by a membrane composed of lipids and proteins, where chemical and metabolic functions take place.

strategies focusing on alleviation of complications and symptoms. In general, most treatments that are attempted are dietary, whereby attempts are made to artificially correct biochemical abnormalities associated with the disorders. Therapies include supplementation of the diet with antioxidant **vitamins**, or limitation of intake of fatty acids, especially VLCFAs.

Another area of dietary therapy that is being investigated is the supplementation of the diet with pure DHA, given as early in life as possible, in conjunction with a normal well-balanced diet. Some results have indicated that if given soon enough during development, DHA therapy may prevent some of the devastating consequences of peroxisomal disorders, including brain damage and the loss of vision.

Other treatment strategies include addition of important missing chemicals. For example, in disorders where there is faulty adrenal function, replacement adrenal hormone therapy is used.

Any dietary changes should be monitored biochemically to determine if the supplements are having their desired effects and are not causing additional adverse effects.

Bone marrow transplants may be used to treat X-ALD, and can be effective if done early in the course of the childhood form of the disease.

Physical and psychological therapies are important for all types of peroxisomal disorders.

Alternative treatment

Patients with peroxisomal disorders, and particularly X-ALD, have been treated with a mixture of glycerol trioleate-glycerol trieucate (4:1 by volume),

prepared from olive and rapeseed oils, and referred to as Lorenzo's oil (developed by parents of a son, Lorenzo, who had X-ALD, whose story was documented in the 1992 movie, *Lorenzo's Oil*), to decrease the levels of VLCFA. Other **diets** that have been tried include dietary supplementation with plasmalogen precursors to increase plasmalogen levels and with cholic acid to normalize bile acids. However, there has been only limited success demonstrated with the use of these treatments. More research is needed to determine the long-term safety and effectiveness of these treatment strategies.

Prognosis

Peroxisomal disorders range from life-threatening to cases in which people may function with some degree of mental and motor retardation. There is not yet a cure. Enzyme replacement therapies, including enzyme infusion, transplantation, and **gene therapy**, may hold promise for future advances in the treatment of these disorders. Research is being conducted to increase scientific understanding of these disorders and to find ways to prevent, treat, and cure them.

Prevention

Unfortunately not enough is yet known about these diseases to develop comprehensive strategies for prevention. **Genetic counseling** is recommended for known or suspected carriers. As genes are identified that result in the disorders, **genetic testing** is being developed to identify carriers, who then can manage their reproduction to avoid the possibility of children being born with these deficiencies. As the genetic bases for the disorders are defined, prenatal diagnosis and

identification of carriers will be facilitated. For example, for X-ALD, diagnosis can be made from cultured skin fibroblasts or amniotic fluid cells. This allows prenatal diagnosis and carrier identification in 90% of those affected. More recently it has been shown that biochemical diagnosis can be performed through chorionic villi biopsy, a procedure performed very early in the first trimester of **pregnancy**.

Animal models of ZS and X-ADL have been developed and are providing researchers with methods to define pathogenic mechanisms and to evaluate new therapies.

Resources

PERIODICALS

Gallo, S., et al. "Late Onset MLD With Normal Nerve Conduction Associated With Two Novel Missense Mutations in the ASA Gene." *Journal of Neurology, Neurosurgery, and Psychiatry* April 2004: 655–658.

Moser, Hugo W. "Molecular Genetics of Peroxisomal Disorders." *Frontiers in Bioscience* 5 (March 1, 2001): 298–306.

OTHER

PeroxisomeDB Home. <http://www.peroxisomedb.org/>.

ORGANIZATIONS

National Institute of Neurological Disorders and Stroke (NINDS), NIH Neurological Institute, P. O. Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov/>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Judith Sims
Teresa G. Odle

Persantine-thallium heart scan see **Thallium heart scan**

deviant by the person's culture, and cause significant emotional **pain** and/or difficulties in relationships and occupational performance. In addition, the patient usually sees the disorder as being consistent with his or her self-image (ego-syntonic) and may blame others for his or her social, educational, or work-related problems.

Demographics

Personality disorders have their onset in late adolescence or early adulthood. Doctors rarely give a diagnosis of personality disorder to children on the grounds that children's personalities are still in the process of formation and may change considerably by the time they are in their late teens. In retrospect, however, many individuals with personality disorders could be judged to have shown evidence of the problems in childhood.

It is difficult to give close estimates of the percentage of the population that has personality disorders. Patients with certain personality disorders, including antisocial and borderline disorders, are more likely to get into trouble with the law or otherwise attract attention than are patients whose disorders chiefly affect their capacity for intimacy. On the other hand, some patients, such as those with narcissistic or obsessive-compulsive personality disorders, may be outwardly successful because their symptoms are useful within their particular occupations. It has, however, been estimated that about 15% of the general population of the United States has a personality disorder, with higher rates in poor or troubled neighborhoods. The rate of personality disorders among patients in psychiatric treatment is between 30% and 50%. It is possible for patients to have a so-called dual diagnosis; for example, they may have more than one personality disorder, or a personality disorder together with a substance-abuse problem.

Description

To meet the diagnosis of personality disorder, which is sometimes called character disorder, the patient's problematic behaviors must appear in two or more of the following areas:

- perception and interpretation of the self and other people
- intensity and duration of feelings and their appropriateness to situations
- relationships with others
- ability to control impulses

Personality disorders

Definition

Personality disorders are a group of mental disturbances defined by the fourth edition, text revision (2000) of the *Diagnostic and Statistical Manual of Mental Disorders (DSM-IV)* as "enduring pattern[s] of inner experience and behavior" that are sufficiently rigid and deep-seated to bring a person into repeated conflicts with his or her social and occupational environment. *DSM-IV* specifies that these dysfunctional patterns must be regarded as nonconforming or

The *DSM-IV* classifies personality disorders into three clusters based on symptom similarities:

- Cluster A (paranoid, schizoid, schizotypal): Patients appear odd or eccentric to others.
- Cluster B (antisocial, borderline, histrionic, narcissistic): Patients appear overly emotional, unstable, or self-dramatizing to others.
- Cluster C (avoidant, dependent, obsessive-compulsive): Patients appear tense and anxiety-ridden to others.

The *DSM-IV* clustering system does not mean that all patients can be fitted neatly into one of the three clusters. It is possible for patients to have symptoms of more than one personality disorder or to have symptoms from different clusters.

Some psychiatrists maintain that the *DSM-IV* classification is inadequate and should be expanded to include three additional categories: passive-aggressive personality disorder, characterized by a need to control or punish others through frustrating them or sabotaging plans; cyclothymic personality disorder, characterized by intense mood swings alternating between high spirits and moroseness or gloom; and depressive personality disorder, characterized by a negative and pessimistic approach to life.

Since the criteria for personality disorders include friction or conflict between the patient and his or her social environment, these syndromes are open to redefinition as societies change. Successive editions of *DSM* have tried to be sensitive to cultural differences, including changes over time, when defining personality disorders. One category that had been proposed for *DSM-III-R*, self-defeating personality disorder, was excluded from *DSM-IV* on the grounds that its definition reflected prejudice against women. *DSM-IV* recommends that doctors take a patient's background, especially recent immigration, into account before deciding that he or she has a personality disorder. One criticism that has been made of the general category of personality disorder is that it is based on Western notions of individual uniqueness. Its applicability to people from cultures with different definitions of human personhood is thus open to question. Furthermore, even within a culture, it can be difficult to define the limits of "normalcy."

The personality disorders defined by *DSM-IV* are as follows:

Paranoid

Patients with paranoid personality disorder are characterized by suspiciousness and a belief that

others are out to harm or cheat them. They have problems with intimacy and may join cults or groups with paranoid belief systems. Some are litigious, bringing lawsuits against those they believe have wronged them. Although not ordinarily delusional, these patients may develop psychotic symptoms under severe **stress**. It is estimated that 0.5–2.5% of the general population meet the criteria for paranoid personality disorder.

Schizoid

Schizoid patients are perceived by others as "loners" without close family relationships or social contacts. Indeed, they are aloof and really do prefer to be alone. They may appear cold to others because they rarely display strong emotions. They may, however, be successful in occupations that do not require personal interaction. About two percent of the general population has this disorder. It is slightly more common in men than in women.

Schizotypal

Patients diagnosed as schizotypal are often considered odd or eccentric because they pay little attention to their clothing and sometimes have peculiar speech mannerisms. They are socially isolated and uncomfortable in parties or other social gatherings. In addition, people with schizotypal personality disorder often have oddities of thought, including "magical" beliefs or peculiar ideas (for example, a belief in telepathy or UFOs) that are outside of their cultural norms. It is thought that three percent of the general population has schizotypal personality disorder. It is slightly more common in males. Schizotypal disorder should not be confused with **schizophrenia**, although there is some evidence that the disorders are genetically related.

Antisocial

Patients with antisocial personality disorder are sometimes referred to as sociopaths or psychopaths. They are characterized by lying, manipulativeness, and a selfish disregard for the rights of others; some may act impulsively. People with antisocial personality disorder are frequently chemically dependent and sexually promiscuous. It is estimated that three percent of males in the general population and one percent of females have antisocial personality disorder.

Borderline

Patients with **borderline personality disorder** (BPD) are highly unstable, with wide mood swings, a

history of intense but stormy relationships, impulsive behavior, and confusion about career goals, personal values, or sexual orientation. These often highly conflictual ideas may correspond to an even deeper confusion about their sense of self (identity). People with BPD frequently cut or burn themselves, or threaten or attempt **suicide**. Many of these patients have histories of severe childhood **abuse** or neglect. About two percent of the general population have BPD; 75% of these patients are female.

Histrionic

Patients diagnosed with this disorder impress others as overly emotional, overly dramatic, and hungry for attention. They may be flirtatious or seductive as a way of drawing attention to themselves, yet they are emotionally shallow. Histrionic patients often live in a romantic fantasy world and are easily bored with routine. About two to three percent of the population is thought to have this disorder. Although historically the disorder has been more associated with women in clinical settings, there may be bias toward diagnosing women with the histrionic personality disorder.

Narcissistic

Narcissistic patients are characterized by self-importance, a craving for admiration, and exploitative attitudes toward others. They have unrealistically inflated views of their talents and accomplishments, and may become extremely angry if they are criticized or outshone by others. Narcissists may be professionally successful but rarely have long-lasting intimate relationships. Fewer than one percent of the population has this disorder; about 75% of those diagnosed with it are male.

Avoidant

Patients with avoidant personality disorder are fearful of rejection and shy away from situations or occupations that might expose their supposed inadequacy. They may reject opportunities to develop close relationships because of their fears of criticism or humiliation. Patients with this personality disorder are often diagnosed with dependent personality disorder as well. Many also fit the criteria for social phobia. Between 0.5–1.0% of the population have avoidant personality disorder.

Dependent

Dependent patients are afraid of being on their own and typically develop submissive or compliant behaviors in order to avoid displeasing people. They

are afraid to question authority and often ask others for guidance or direction. Dependent personality disorder is diagnosed more often in women, but it has been suggested that this finding reflects social pressures on women to conform to gender stereotyping or bias on the part of clinicians.

Obsessive-compulsive

Patients diagnosed with this disorder are preoccupied with keeping order, attaining perfection, and maintaining mental and interpersonal control. They may spend a great deal of time adhering to plans, schedules, or rules from which they will not deviate, even at the expense of openness, flexibility, and efficiency. These patients are often unable to relax and may become “workaholics.” They may have problems in employment as well as in intimate relationships because they are very stiff and formal, and insist on doing everything their way. About one percent of the population has obsessive-compulsive personality disorder; the male/female ratio is about two to one.

Causes and symptoms

Personality disorders are thought to result from a bad interface, so to speak, between a child’s temperament and character on one hand and his or her family environment on the other. Temperament can be defined as a person’s innate or biologically shaped basic disposition. Human infants vary in their sensitivity to light or noise, their level of physical activity, their adaptability to schedules, and similar traits. Even such traits as **shyness** or novelty-seeking may be at least in part determined by the biology of the brain and the genes one inherits.

Character is defined as the set of attitudes and behavior patterns that the individual acquires or learns over time. It includes such personal qualities as work and study habits, moral convictions, neatness or cleanliness, and consideration of others. Since children must learn to adapt to their specific families, they may develop personality disorders in the course of struggling to survive psychologically in disturbed or stressful families. For example, nervous or high-strung parents might be unhappy with a baby who is very active and try to restrain him or her at every opportunity. The child might then develop an avoidant personality disorder as the outcome of coping with constant frustration and parental disapproval. As another example, **child abuse** is believed to play a role in shaping borderline personality disorder. One reason that some therapists use the term developmental damage instead of personality disorder is that it

takes the presumed source of the person's problems into account.

Some patients with personality disorders come from families that appear to be stable and healthy. It has been suggested that these patients are biologically hypersensitive to normal family stress levels. Levels of the brain chemical (neurotransmitter) dopamine may influence a person's level of novelty-seeking, and serotonin levels may influence aggression.

Other factors that have been cited as affecting children's personality development are the mass media and social or group **hysteria**, particularly after the events of September 11, 2001. Cases of so-called mass sociogenic illness have been identified, in which a group of children began to vomit or have other physical symptoms brought on in response to an imaginary threat. In two such cases, the children were reacting to the suggestion that toxic fumes were spreading through their school. Some authors believe that overly frequent or age-inappropriate discussions of terrorist attacks or bioterrorism may make children more susceptible to sociogenic illness as well as other distortions of personality.

Diagnosis

Diagnosis of personality disorders is complicated by the fact that affected persons rarely seek help until they are in serious trouble or until their families (or the law) pressure them to get treatment. The reason for this slowness is that the problematic traits are so deeply entrenched that they seem normal (ego-syntonic) to the patient. Diagnosis of a personality disorder depends in part on the patient's age. Although personality disorders originate during the childhood years, they are considered adult disorders. Some patients, in fact, are not diagnosed until late in life because their symptoms had been modified by the demands of their job or by marriage. After retirement or the spouse's **death**, however, these patients' personality disorders become fully apparent. In general, however, if the onset of the patient's problem is in mid-or late-life, the doctor will rule out **substance abuse** or personality change caused by medical or neurological problems before considering the diagnosis of a personality disorder. It is unusual for people to develop personality disorders "out of the blue" in mid-life.

There are no tests that can provide a definitive diagnosis of personality disorder. Most doctors will evaluate a patient on the basis of several sources of information collected over a period of time in order to determine how long the patient has been having difficulties, how many areas of life are affected, and how

severe the dysfunction is. These sources of information may include:

Interviews

The doctor may schedule two or three interviews with the patient, spaced over several weeks or months, in order to rule out an adjustment disorder caused by job loss, **bereavement**, or a similar problem. An office interview allows the doctor to form an impression of the patient's overall personality as well as obtain information about his or her occupation and family. During the interview, the doctor will note the patient's appearance, tone of voice, body language, eye contact, and other important non-verbal signals, as well as the content of the conversation. In some cases, the doctor may contact other people (family members, employers, close friends) who know the patient well in order to assess the accuracy of the patient's perception of his or her difficulties. It is quite common for people with personality disorders to have distorted views of their situations or to be unaware of the impact of their behavior on others.

Psychologic testing

Doctors use psychologic testing to help in the diagnosis of a personality disorder. Most of these tests require interpretation by a professional with specialized training. Doctors usually refer patients to a clinical psychologist for this type of test.

PERSONALITY INVENTORIES. Personality inventories are tests with true/false or yes/no answers that can be used to compare the patient's scores with those of people with known personality distortions. The single most commonly used test of this type is the **Minnesota Multiphasic Personality Inventory**, or MMPI. Another test that is often used is the Millon Clinical Multiaxial Inventory, or MCMI.

PROJECTIVE TESTS. Projective tests are unstructured. Unstructured means that instead of giving one-word answers to questions, the patient is asked to talk at some length about a picture that the psychologist has shown him or her, or to supply an ending for the beginning of a story. Projective tests allow the clinician to assess the patient's patterns of thinking, fantasies, worries or anxieties, moral concerns, values, and habits. Common projective tests include the Rorschach, in which the patient responds to a set of ten inkblots; and the **Thematic Apperception Test** (TAT), in which the patient is shown drawings of people in different situations and then tells a story about the picture.

Treatment

At one time psychiatrists thought that personality disorders did not respond very well to treatment. This opinion was derived from the notion that human personality is fixed for life once it has been molded in childhood, and from the belief among people with personality disorders that their own views and behaviors are correct, and that others are the ones at fault. More recently, however, doctors have recognized that humans can continue to grow and change throughout life. Most patients with personality disorders are now considered to be treatable, although the degree of improvement may vary. The type of treatment recommended depends on the personality characteristics associated with the specific disorder.

Hospitalization

Inpatient treatment is rarely required for patients with personality disorders, with two major exceptions: borderline patients who are threatening suicide or suffering from drug or alcohol withdrawal; and patients with paranoid personality disorder who are having psychotic symptoms.

Psychotherapy

Psychoanalytic **psychotherapy** is suggested for patients who can benefit from insight-oriented treatment. These patients typically include those with dependent, obsessive-compulsive, and avoidant personality disorders. Doctors usually recommend individual psychotherapy for narcissistic and borderline patients, but often refer these patients to therapists with specialized training in these disorders. Psychotherapeutic treatment for personality disorders may take as long as three to five years.

Insight-oriented approaches are not recommended for patients with paranoid or antisocial personality disorders. These patients are likely to resent the therapist and see him or her as trying to control or dominate them.

Supportive therapy is regarded as the most helpful form of psychotherapy for patients with schizoid personality disorder.

Cognitive-behavioral therapy

Cognitive-behavioral approaches are often recommended for patients with avoidant or dependent personality disorders. Patients in these groups typically have mistaken beliefs about their competence or likableness. These assumptions can be successfully challenged by cognitive-behavioral methods. More

recently, American psychiatrist Aaron Beck and his coworkers have successfully extended their approach to cognitive therapy to all ten personality disorders as defined by DSM-IV.

Group therapy

Group therapy is frequently useful for patients with schizoid or avoidant personality disorders because it helps them to break out of their social isolation. It has also been recommended for patients with histrionic and antisocial personality disorders. These patients tend to act out, and pressure from peers in group treatment can motivate them to change. Because patients with antisocial personality disorder can destabilize groups that include people with other disorders, it is usually best if these people meet exclusively with others who have APD in homogeneous groups.

Family therapy

Family therapy may be suggested for patients whose personality disorders cause serious problems for members of their families. It is also sometimes recommended for borderline patients from over-involved or possessive families.

Medications

Medications may be prescribed for patients with specific personality disorders. The type of medication depends on the disorder. In general, however, patients with personality disorders are helped only moderately by medications.

ANTIPSYCHOTIC DRUGS. **Antipsychotic drugs**, such as haloperidol (Haldol), may be given to patients with paranoid personality disorder if they are having brief psychotic episodes. Patients with borderline or schizotypal personality disorder are sometimes given antipsychotic drugs in low doses; however, the efficacy of these drugs in treating personality disorder is less clear than in schizophrenia.

MOOD STABILIZERS. Carbamazepine (Tegretol) is a drug that is commonly used to treat seizures, but is also helpful for borderline patients with rage outbursts and similar behavioral problems. Lithium and valproate may also be used as mood stabilizers, especially among people with borderline personality disorder.

ANTIDEPRESSANTS AND ANTI-ANXIETY MEDICATIONS.

Medications in these categories are sometimes prescribed for patients with schizoid personality disorder to help them manage **anxiety** symptoms while they are in psychotherapy. Antidepressants are also commonly used to treat people with borderline personality disorder.

KEY TERMS

Character—An individual's set of emotional, cognitive, and behavioral patterns learned and accumulated over time.

Character disorder—Another name for personality disorder.

Cognitive therapy—A form of psychotherapy that focuses on changing people's patterns of emotional reaction by correcting distorted patterns of thinking and perception.

Developmental damage—A term that some therapists prefer to personality disorder, on the grounds that it is more respectful of the patient's capacity for growth and change.

Ego-syntonic—Consistent with one's sense of self, as opposed to ego-alien or dystonic (foreign to one's sense of self). Ego-syntonic traits typify patients with personality disorders.

Neuroleptic—Another name for older antipsychotic medications, such as haloperidol. The term does not apply to such newer atypical agents as clozapine (Clozaril).

Personality—The organized pattern of behaviors and attitudes that makes a human being distinctive. Personality is formed by the ongoing interaction of temperament, character, and environment.

Projective tests—Psychological tests that probe into personality by obtaining open-ended responses to such materials as pictures or stories. Projective tests are often used to evaluate patients with personality disorders.

Rorschach test—A well-known projective test that requires the patient to describe what he or she sees in each of 10 inkblots. It is named for the Swiss psychiatrist who invented it.

Temperament—A person's natural or genetically determined disposition.

Treatment with medications is not recommended for patients with avoidant, histrionic, dependent, or narcissistic personality disorders. The use of potentially addictive medications should be avoided in people with borderline or antisocial personality disorders. However, some avoidant patients who also have social phobia may benefit from **monoamine oxidase inhibitors** (MAO inhibitors), a particular class of antidepressant.

Prognosis

The prognosis for recovery depends in part on the specific disorder. Although some patients improve as they grow older and have positive experiences in life, personality disorders are generally life-long disturbances with periods of worsening (exacerbations) and periods of improvement (remissions). Others, particularly schizoid patients, have better prognoses if they are given appropriate treatment. Beck and his coworkers estimate that effective cognitive therapy with patients with personality disorders takes two to three years on average. Patients with paranoid personality disorder are at some risk for developing delusional disorders or schizophrenia.

The personality disorders with the poorest prognoses are the antisocial and the borderline. Borderline patients are at high risk for developing substance abuse disorders or bulimia. About 80% of hospitalized borderline patients attempt suicide at some point

during treatment, and about five percent succeed in committing suicide. Borderline patients are also the most likely to sue their mental health professional for malpractice.

Prevention

The most effective preventive strategy for personality disorders is early identification and treatment of children at risk. High-risk groups include abused children, children from troubled families, children with close relatives diagnosed with personality disorders, children of substance abusers, and children who grow up in cults or extremist political groups.

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- American Psychiatric Association (APA), 1000 Wilson Blvd., Suite 1825, Arlington, VA, 22209, (703) 907–7300, apa@psych.org, <http://www.psych.org/>.
- American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002–4242, (202) 336–5700, <http://www.apa.org>.
- National Alliance on Mental Illness (NAMI), Colonial Place Three, 2107 Wilson Blvd., Suite 300, Arlington, VA, 22201, (703) 524–7600, (800) 950–NAMI (6264), (703) 524–9094, <http://www.nami.org/Hometemplate.cfm>.
- National Institute of Mental Health (NIMH), 6001 Executive Blvd., Room 8184, MSC 9663, Bethesda, MD, 20892, (301) 443–4513, (866) 615–6464, (301) 443–4279, nim.hinfo@nih.gov, <http://www.nimh.nih.gov/index.shtml>.
- National Mental Health Association (NMHA), 2000 N. Beauregard St., 6th Floor, Alexandria, VA, 22311, (703) 684–7722, (800) 969–NMHA, (703) 684–5968, <http://www1.nmha.org/>.

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Perthes disease see **Osteochondroses**

Pertussis see **Whooping cough**

Pervasive developmental disorders

Definition

Pervasive developmental disorders include five different conditions: Asperger's syndrome, autistic disorder, childhood disintegrative disorder (CDD),

pervasive developmental disorder not otherwise specified (PDDNOS), and Rett's syndrome. They are grouped together because of the similarities among them. The three most common shared problems involve communication skills, motor skills, and social skills. Since there are no clear diagnostic boundaries separating these conditions, it is sometimes difficult to distinguish one from the other for diagnostic purposes.

Demographics

Asperger's syndrome, autistic disorder, and childhood disintegrative disorder are four to five times more common in boys, and Rett's syndrome has been diagnosed primarily in girls. All of these disorders are rare.

Description

Asperger's syndrome

Children afflicted with Asperger's syndrome exhibit difficulties in social relationships and communication. They are reluctant to make eye contact, do not respond to social or emotional contacts, do not initiate play activities with peers, and do not give or receive attention or affection. To receive this diagnosis the individual must demonstrate normal development of language, thinking, and coping skills. Due to an impaired coordination of muscle movements, they appear to be clumsy. They usually become deeply involved in very few interests, which tend to occupy most of their time and attention.

Autistic disorder

Autistic disorder is frequently evident within the first year of life, and must be diagnosed before age three. It is associated with moderate **mental retardation** in three out of four cases. These children do not want to be held, rocked, cuddled, or played with. They are unresponsive to affection, show no interest in peers or adults, and have few interests. Other traits include avoidance of eye contact, an expressionless face, and the use of gestures to express needs. Their actions are repetitive, routine, and restricted. Rocking, hand and arm flapping, unusual hand and finger movements, and attachment to objects rather than pets and people are common. Speech, play, and other behaviors are repetitive and without imagination. They tend to be overactive, aggressive, and self-injurious. They are often highly sensitive to touch, noise, and smells and do not like changes in routine. **Autism** and several disorders classified with it have increased significantly in recent years so that they now are diagnosed more often in children than **spina bifida**, **cancer**, or **Down**

syndrome. This change may be due partly to improved recognition and diagnosis.

Childhood disintegrative disorder

Childhood disintegrative disorder is also called Heller's disease and most often develops between two and 10 years of age. Children with CDD develop normally until two to three years of age and then begin to disintegrate rapidly. Signs and symptoms include deterioration of the ability to use and understand language to the point where they are unable to carry on a conversation. This is accompanied by loss of control of the bladder and bowels. Any interest or ability to play and engage in social activities is lost. The behaviors are nearly identical with those that are characteristic of autistic disorder. However, childhood disintegrative disorder becomes evident later in life and results in developmental regression, or loss of previously attained skills, whereas autistic disorder can be detected as early as the first month of life and results in a failure to progress.

Pervasive developmental disorder not otherwise specified

The term pervasive developmental disorder not otherwise specified (PDDNOS) is also referred to as atypical personality development, atypical PDD, or atypical autism. Individuals with this disorder share some of the same signs and symptoms of autism or other conditions under the category of pervasive developmental disorders, but do not meet all of the criteria for diagnosis for any of the four syndromes included in this group of diseases. Because the children diagnosed with PDDNOS do not all exhibit the same combination of characteristics, it is difficult to do research on this disorder, but the limited evidence available suggests that patients are seen by medical professionals later in life than is the case for autistic children, and they are less likely to have intellectual deficits.

Rett's syndrome

Rett's syndrome was first described in 1966 and is found almost exclusively in girls. It is a disease in which cells in the brain experience difficulty in communicating with each other. At the same time the growth of the head falls behind the growth of the body so that these children are usually mentally retarded. These conditions are accompanied by deficits in movement (motor) skills and a loss of interest in social activities.

The course of the illness has been divided into four stages. In stage one the child develops normally for six to 18 months. In stage two, development slows down and stops. Stage three is characterized by a loss of the speech and motor skills already acquired. Typically this happens between nine months and three years of age. Stage four begins with a return to learning that will continue across the lifespan, but at a very slow rate. Problems with coordination and walking are likely to continue and even worsen. Other conditions that can occur with Rett's syndrome are convulsions, **constipation**, breathing problems, impaired circulation in the feet and legs, and difficulty chewing or swallowing.

Causes and symptoms

The causes of these disorders are unknown although brain structure abnormalities, genetic mutation, and alterations in brain function are believed to play a role. Still, no single brain abnormality or location has been connected to a cause. In 2004, scientists reported finding a gene mutation (on gene MECP2) that is present in 80% of people affected with Rett's syndrome. In 2004, a comprehensive review of research on twins revealed that interactions between multiple genes may play a role in the cause of autism. A number of neurological conditions, such as convulsions, are commonly found to accompany these disorders.

Diagnosis

The diagnosis of pervasive developmental disorder is made by medical specialists based on a thorough examination of the patient, including observing behavior and gathering information from parents and caregivers. Because many symptoms are common to more than one condition, distinctions between conditions must be carefully made. The following summary describes the distinction between three common disorders.

PDDNOS:

- impairment of two-way social interaction
- Repetitive and predictable behavior patterns and activities

Autism:

- all listed for PDDNOS
- severe impairment in communication
- abnormal social interaction and use of language for social communication or imaginative play before age of three
- not better accounted for by another psychiatric disorder

Asperger's disorder:

- all listed for PDDNOS
- clinically significant impairment in social, occupational, or other areas of functioning
- no general delay in language
- no delay in cognitive development, self-help skills, or adaptive behavior
- not better accounted for by another pervasive developmental disorder or schizophrenia

Rett's syndrome:

- a period of normal development between six and 18 months
- normal head circumference at birth, followed by a slowing of head growth
- mental retardation
- repetitive hand movements

CDD:

- normal development for at least two years
- loss of skills in at least two of the following areas: language, social skills, bowel or bladder control, play, movement skills
- abnormal functioning in at least two of the following areas: social interaction, communication, behavior patterns
- not better accounted for by another PDD or mental illness

Treatment

Treatment for children with pervasive developmental disorders is limited. Those who can be enrolled in educational programs will need a highly structured learning environment, a teacher-student ratio of not more than 1:2, and a high level of parental involvement that provides consistent care at home. **Psychotherapy** and social skills training can prove helpful to some. There is no specific medication available for treating the core symptoms of any of these disorders, though research is promising. Some psychiatric medications may be helpful in controlling particular behavior difficulties, such as agitation, mood instability, and self-injury. Music, massage, and **hydrotherapy** may exert a calming effect on behavior. Treatment may also include physical and **occupational therapy**.

Prognosis

In general, the prognosis in all of these conditions is tied to the severity of the illness.

The prognosis for Asperger's syndrome is more hopeful than that for other diseases in this cluster.

KEY TERMS

Hydrotherapy—This term literally means “water treatment” and involves the use of water in physical therapy as well as treatment of physical and emotional illness.

Mutation—A change in a gene. Since genes determine how a body is structured and functions, any change in a gene will produce some change in these areas.

Neurological conditions—A condition that has its origin in some part of the patient's nervous system.

These children are likely to grow up to be functional independent adults, but will always have problems with social relationships. They are also at greater risk for developing serious mental illness than the general population.

The prognosis for autistic disorder is not as good, although great strides have been made in recent years in its treatment. The higher the patient's IQ (intelligence quotient) and ability to communicate, the better the prognosis. However, many patients will always need some level of custodial care. In the past, most of these individuals were confined to institutions, but many are now able to live in group homes or supervised apartments. The prognosis for childhood disintegrative disorder is even less favorable. These children will require intensive and long-term care. Children diagnosed with PDDNOS have a better prognosis because their initial symptoms are usually milder, IQ scores are higher, and language development is stronger.

Prevention

The causes of pervasive developmental disorders are not understood, although research efforts are getting closer to understanding the problem. Until the causes are discovered, it will remain impossible to prevent these conditions.

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International Rett Syndrome Association (IRSA), 4600 Devitt Dr., Cincinnati, OH, 45246, (800) 818–7388, <http://www.rettsyndrome.org>.

Learning Disabilities Association of America (LDAA), 4156 Library Rd., Pittsburgh, PA, 15234, (412) 341–1515, <http://www.ldanatl.org>.

National Organization for Rare Disorders (NORD), 55 Kenosia Ave., PO Box 1968, Danbury, CT, 06813–1968, (800) 999–6673, <http://www.rarediseases.org>.

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PET scan see Positron emission tomography (PET)

Pet therapy

Definition

Animal-assisted therapy (AAT), also known as pet therapy, utilizes trained animals and handlers to achieve specific physical, social, cognitive, and emotional goals with patients.

KEY TERMS

Endorphins—A group of chemicals resembling opiates that are released in the body in response to trauma or stress. Endorphins react with opiate receptors in the brain to reduce pain sensations.

Purpose

Studies have shown that physical contact with a pet can lower high blood pressure, and improve survival rates for **heart attack** victims. There is also evidence that petting an animal can cause endorphins to be released. Endorphins are chemicals in the body that suppress the **pain** response. These are benefits that can be enjoyed from pet ownership, as well as from visiting therapeutic animals.

Many skills can be learned or improved with the assistance of a therapy animal. Patient **rehabilitation** can be encouraged by such activities as walking or running with a dog, or throwing objects for the animal to retrieve. Fine motor skills may be developed by petting, grooming, or feeding the animal. Patient communication is encouraged by the response of the animal to either verbal or physical commands. Activities such as writing or talking about the therapy animals or past pets also develop cognitive skills and communication. Creative inclusion of an animal in the life or therapy of a patient can make a major difference in the patient's comfort, progress, and recovery.

Description

Origins

The enjoyment of animals as companions dates back many centuries, perhaps even to prehistoric times. The first known therapeutic use of animals started in Gheel, Belgium in the ninth century. In this town, learning to care for farm animals has long been an important part of an assisted living program designed for people with disabilities.

Some of the earliest uses of animal-assisted healing in the United States were for psychiatric patients. The presence of the therapy animals produced a beneficial effect on both children and adults with mental health issues. It is only in the last few decades that AAT has been more formally applied in a variety of therapeutic settings, including schools and prisons, as well as hospitals, hospices, nursing homes, and outpatient care programs.



This autistic child is encouraged to interact with the guinea pig in an effort to improve his social interaction. (*Helen B. Senisi/Photo Researchers, Inc.*)

The way in which AAT is undertaken depends on the needs and abilities of the individual patient. Dogs are the most common visiting therapy animals, but cats, horses, birds, rabbits, and other domestic pets can be used as long as they are appropriately screened and trained.

For patients who are confined, small animals can be brought to the bed if the patient is willing and is not allergic to the animal. A therapeutic plan may include a simple interaction aimed at improving communication and small motor skills, or a demonstration with educational content to engage the patient cognitively.

If the patient is able to walk or move around, more options are available. Patients can walk small animals outside, or learn how to care for farm animals. Both of these activities develop confidence and motor abilities. Horseback riding has recently gained great therapeutic popularity. It offers an opportunity to work on balance, trunk control, and other skills. Many patients who walk with difficulty, or not at all, get great emotional benefit from interacting with and controlling a large animal.

One advantage of having volunteers provide this service is that cost and insurance are not at issue.

Precautions

AAT does not involve just any pet interacting with a patient. Standards for the training of the volunteers and their animals are crucial in order to promote a safe, positive experience for the patient. Trained volunteers will understand how to work with other medical professionals to set goals for the patient and keep records of progress. Animals that have been appropriately trained are well socialized to people, other animals, and medical equipment. They are not distracted by the food and odors that may be present in the therapy environment and will not chew inappropriate objects or mark territory.

Animals participating in AAT should be covered by some form of liability insurance.

Research and general acceptance

While the research evidence supporting the efficacy of AAT is slim, the anecdotal support is vast.

Although it may not be given much credence by medical personnel as a therapy with the potential to assist the progress of the patients, some institutions do at least allow it as something that will uplift the patients or distract them from their discomforts.

ORGANIZATIONS

Delta Society, 875 124th Ave NE #101, Bellevue, WA, 98005, (425) 679-5500, (425) 679-5539, info@DeltaSociety.org, <http://www.deltasociety.org>.

Judith Turner

Peyronie's disease

Definition

Peyronie's disease is an acquired inflammatory condition in which the erect penis is bent because of plaque—a hard lump of scar tissue—that prevents the area from stretching. Peyronie's is a variable and poorly understood urological condition. It is also called curvature of the penis or *induratio plastica penis*.

Demographics

Until recently Peyronie's disease was thought to be relatively uncommon, affecting less than 1% of men. However newer estimates range as high as 23%. One recent study found that 3.2% of German men between the ages of 30 and 80 were affected by Peyronie's. Although embarrassment prevents many men from seeking help, the number of diagnosed cases of Peyronie's disease has increased markedly in recent years. This is probably due to the availability of new drugs for treating **erectile dysfunction** (ED), which has encouraged many more men to seek treatment for sexual problems. Thus the number of diagnosed cases of Peyronie's disease is expected to continue to increase. The condition most often affects men over age 40.

Description

Penises vary in shape and size and erect penises are often slightly bent. However Peyronie's disease is characterized by a bent penis that causes **pain** and/or interferes with sex. In 1743 Francois Gigot de la Peyronie, personal physician to King Louis XV of France, wrote the first detailed description of the disorder that bears his name. Earlier writers had classified it as a form of **impotence** or ED.

The bulk of the penis consists of two tubular chambers called the corpora cavernosa that fill with blood during an erection, causing the penis to expand, stiffen, and straighten. The corpora cavernosa are enclosed within a sheath of elastic tissue called the tunica albuginea, which stretches during an erection. With Peyronie's disease flat fibrous scar tissue called plaque forms in the tunica albuginea and prevents stretching, so that the corpora cavernosa expand unevenly. This causes the penis to bend or curve in the direction of the plaque. Almost one third of men with Peyronie's disease have similar scar tissue on their hands, a disease called Dupuytren's contracture. However, although Dupuytren's contracture is fairly common in white men over age 50, only a small percentage of these men develop Peyronie's disease.

Peyronie's disease is frequently mild and does not progress past the inflammation stage, disappearing within months. However it can also develop into a severe condition in which sexual intercourse is painful or impossible. Peyronie's disease can affect sexual desire, as well as function, and may interfere with intimate relationships. Studies indicate that more than 75% of men with Peyronie's disease suffer from **anxiety** and emotional distress related to their condition.

Risk factors

Known risk factors for Peyronie's disease include:

- a father or brother with the disease
- connective tissue disorders such as Dupuytren's contracture
- age-related changes that cause tissues to be more easily injured and/or less readily healed
- diabetes
- tobacco use
- prostate surgery, catheterization, or pelvic injury

Causes and symptoms

The exact cause of Peyronie's disease is unclear. It most often seems to result from a minor trauma incurred during sexual activity, such as bending the penis during intercourse or pressure from the partner's pubic bone. It also can result from an accident or sports injury. Injury to the tunica albuginea can cause bleeding, inflammation, and tissue damage. If the wound does not heal properly, fibrosis—excess scar tissue formation—can occur, leading to inflexible plaques. Peyronie's disease that occurs suddenly and disappears without treatment is most often

due to such trauma that causes bleeding inside the penis. However many severe cases of Peyronie's disease develop slowly without apparent cause. Likewise, associated conditions such as Dupuytren's contracture do not appear to result from trauma. Other suggested causes of Peyronie's disease include vasculitis—an inflammation of blood or lymphatic vessels that leads to scar tissue formation—or other connective-tissue disorders involving the thickening or hardening of skin, cartilage, or bone. Some researchers have suggested that Peyronie's may be an autoimmune disorder. Because Peyronie's tends to run in families and because men with the disease have a particular immune cell marker, it has been suggested that the condition is inherited.

Symptoms of Peyronie's disease vary considerably. They can appear suddenly or develop gradually. Symptoms include:

- a bend in the erect penis
- narrowing of the penis with erection
- shortening of the penis
- lumps in the penis
- painful erections
- pain during intercourse
- soft erections
- ED—difficulty achieving or maintaining an erection
- difficult penetration due to curvature of the penis

Plaque formation may originate with a localized irritation and inflammation or swelling, which then hardens and reduces elasticity, causing the penis to bend in the direction of the plaque. Plaque also can cause indentation, shrinking, or shortening of the penis.

Plaque most often develops on the upper side of the penis, causing the erect penis to bend or curve upward, but it can also develop on the underside or lateral sides, causing a downward or sideways bend to the penis, respectively. Multiple plaques can cause complex curvatures. Sometimes there is a "hinge" effect, in which the erect penis bends sharply down at the base. Extensive plaque that encircles the penis usually results in an "hourglass," "waisting," or "bottleneck" deformity with a tight narrow band around the shaft rather than a curvature. In severe cases the plaque may accumulate **calcium** and become extremely hard.

The acute phase of Peyronie's disease lasts for six to 18 months. As the plaque forms, increasing penile curvature is accompanied by painful erections. However sometimes pain occurs whenever the penis is

KEY TERMS

Autoimmune disorder—Disorders such as rheumatoid arthritis that are caused by the immune system's antibodies or T cells attacking the body's own proteins, cells, or tissues.

Calcification—Hardening or stiffening due to calcium accumulation.

Catheterization—The placing of a flexible tube into the urethra or other body part.

Corpora cavernosa—Erectile tissues that form the bulk of the penis and become distended with blood during an erection.

Dupuytren's contracture—Shortening and thickening of connective tissue in the palm causing the fingers to pull in.

Erectile dysfunction (ED)—The consistent inability to achieve or maintain a penile erection.

Fibrosis—An abnormal thickening and scarring of connective tissue, most often following injury, infection, lack of oxygen, or surgery.

Plaque—A localized abnormal patch on a body part or surface.

Prostate—A gland that surrounds the outlet of the male bladder.

Tunica albuginea—The sheath of elastic tissue enclosing the corpora cavernosa.

touched, even if it is not erect, or only with orgasm. In the chronic phase of Peyronie's the deformity remains stable and erections are usually painless. However both acute and chronic Peyronie's can interfere with sexual activity and result in ED.

Diagnosis

Examination

Curvature of the penis can be diagnosed by a **physical examination**. Erection-producing drugs may be injected under **local anesthesia** or the physician may examine a picture of the erect penis. Erection-producing medication is also used to evaluate erectile function. Hard plaques or scar tissue can be felt under the skin of either a flaccid or erect penis as flat lumps or a band of hard tissue. The length of the penis may be measured to provide a baseline if the condition worsens and the penis shortens. A family doctor or general practitioner may refer the patient to a urologist.

Tests

The patient may be asked to complete a survey, such as the International Index of Erectile Function, to determine how the disorder is affecting sexual activity.

Procedures

Ultrasound or x-ray examination can be used to detect and characterize plaque and any calcification. Ultrasound following the injection of erection-producing medication can determine blood flow to the penis, detect any abnormalities, and measure the degree of penile curvature.

Treatment

Treatment for Peyronie's disease depends on the duration and severity of the condition. The goals of treatment are to relieve pain, restore normal penile anatomy so that intercourse is comfortable, and restore any lost erectile function. No treatment is required if:

- There is no pain.
- Plaques are small.
- The curvature is minor and no longer worsening
- Sexual function is satisfactory.

Traditional

Surgery is the traditional treatment for Peyronie's disease. Because the condition is so often lessens or disappears completely on its own and because surgery can lead to ED, it is usually only performed after at least two years, when the condition has stabilized—the curvature is no longer changing and erections have been painless for at least six months. Even then surgery is usually only performed if the penile deformity prevents satisfactory sexual activity.

There are several types of surgery for Peyronie's disease:

- Shortening the unaffected side: The Nesbit procedure or plication removes or pinches the tunica on the unaffected side to correct the bending. This results in an overall shortening of the erect penis, so it is generally used only when the penis is of adequate length and the curvature is less severe. These procedures are generally safe, relatively easy, and have a low risk of complications; however they cannot restore the length or girth of the penis.
- Lengthening of the affected side: The scar tissue may be cut or removed to allow the tunica albuginea to stretch and the penis to straighten. A graft of skin or

synthetic material replaces the removed tissue. This procedure is used when the penis is short or there is severe curvature or a complicated deformity. It is more difficult surgery and can worsen ED or cause penile numbness.

- Penile implants to replace the corpora cavernosa: Semi-rigid implants are bent up manually for intercourse and bent down to appear flaccid. Inflatable implants are inflated for an erection with a pump implanted in the groin or scrotum. Implants are generally when Peyronie's disease coexists with ED. The surgery usually includes incisions in the scar tissue to relieve tension in the tunica albuginea.

Non-surgical treatments for Peyronie's disease include:

- external penile traction therapy, which can improve girth, length, and curvature
- radiation therapy, which seems to reduce pain but has no effect on the plaque and may cause ED
- shock wave lithotripsy, which can break up the plaque

Drugs

Early-phase Peyronie's disease is treated with oral medications and/or plaque injections. Oral medications include colchicine (an anti-inflammatory), carnitine (an antioxidant), tamoxifen, and pentoxifylline. However none of these have been proven to be effective. Drugs may be injected directly into the plaque over a period of several months, under a local anesthetic, to attempt to soften the tissue, ease pain, and correct curvature. Sometimes these drugs are delivered into the plaque by ontophoresis, a painless electrical current:

- Verapamil is a calcium-channel blocker that is used to treat high blood pressure. It also disrupts collagen production. Collagen is a connective-tissue protein that is thought to be important in Peyronie's scar tissue formation.
- Interferon alpha-2b is a human protein that appears to disrupt production of collagen and promote its breakdown.
- Collagenase is an enzyme that breaks down collagen.
- Corticosteroids are sometimes injected but have undesirable side effects.

Alternative

There are a large number of alternative treatments for Peyronie's disease. Vitamin E, an antioxidant, has been a popular treatment for acute-stage Peyronie's since at least 1948. It inexpensive and has few side

effects, but there is little evidence for its effectiveness. Potassium aminobenzoate (Potaba) is a member of the vitamin B complex and has been shown to reduce plaque size but not curvature. However it is expensive, requires taking 24 pills daily for several months, and has gastrointestinal side effects. Other treatments include:

- topical verapamil
- high-intensity focused ultrasound
- hyperthermia

Home remedies

It is important to maintain an open and honest relationship with one's sexual partner while coping with Peyronie's disease and to explore alternative means for physical and emotional intimacy. A psychotherapist specializing in sexual function and personal relationships can be helpful in this regard.

Prognosis

Mild Peyronie's disease often resolves without treatment in six to 15 months. However in many cases, although pain decreases over time, the bend in the penis makes sexual intercourse difficult. Severe cases of Peyronie's disease can last for years and may continue to worsen, making intercourse impossible or leading to ED. Surgical treatments yield satisfactory results in 60–80% of cases. However surgery can result in complications and cannot correct problems such as penile shortening.

Prevention

There is no known prevention for Peyronie's disease.

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American Urological Association, 1000 Corporate Boulevard, Linthicum, MD, 21090, (410) 689-3700, (866) RING-AUA (746-4282), (410) 689-3800, aua@AUAnet.org, <http://www.auanet.org>.

Association of Peyronie's Disease Advocates, PO Box 62865, Colorado Springs, CO, 80962-2865, information @peyroniesassociation.org, <http://www.peyroniesassociation.org>.

National Kidney and Urologic Diseases Information Clearinghouse, 3 Information Way, Bethesda, MD, 20892-3580, (703) 738-4929, (800) 891-5390, (703) 738-4929, nkudic@info.niddk.nih.gov, <http://kidney.niddk.nih.gov>.

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Pharmacogenetics

Definition

Pharmacogenetics is the study of how the actions of and reactions to drugs vary with the patient's genes.

Description

Genes are the portions of chromosomes that determine many of the traits in every living thing. In humans, genes influence race, hair and eye color, gender, height, weight, aspects of behavior, and even the likelihood of developing certain diseases. Although some traits are a

combination of genetics and environment, researchers are still discovering new ways in which people are affected by their genes.

Pharmacogenetics is the study of how people respond to drug therapy. Although this science is still new, there have been many useful discoveries. It has long been known that genes influence the risk of developing certain diseases, or that genes could determine traits such as hair and eye color. Genes can also alter the risk of developing different diseases. It has long been known that people of African descent were more likely to have sickle cell anemia than people of other races. People of Armenian, Arab, and Turkish heritage are more prone to familiar Mediterranean **fever** than people of other nationalities. More recently, discoveries have shown that genes can determine other aspects of each individual, down to the level of the enzymes produced in the liver. Since these enzymes determine how quickly a drug is removed from the body, they can make major differences in the way people respond to drugs. Some of the most basic work concerns the way race and gender influence drug reactions—and race and gender are genetically determined.

Women often respond differently than men to drugs at the same dose levels. For example, women are more likely to have a good response to the **antidepressant drugs** that act as serotonin specific reuptake inhibitors (SSRIs, the group that includes Prozac and Paxil) than they are to the older group of **tricyclic antidepressants** (the group that includes Elavil and Tofranil). Women have a greater response to some narcotic **pain** relieving drugs than do men, but get less relief from some non-narcotic pain medications. Women may show a greater response to some steroid hormones than men do, but have a lower level of response to some anti-anxiety medications than men.

Race may also affect the way people respond to some medications. In this case, race implies specific genetic factors that are generally, but not always, found among members of specific ethnic groups. For example, the angiotensin II inhibitor enalopril (Vasotec), which is used to lower blood pressure, works better in Caucasians than in Blacks. Carvedilol (Coreg), a beta-adrenergic blocking agent that is also used to lower blood pressure, is more effective than other drugs in the same class when used to treat Black patients. Black patients with **heart failure** appear to respond better to a combination of hydralazine and isosorbide than do Caucasian patients using the same medication.

More specific research has identified individual genes that may influence drugs response, without

relying on group information such as gender and race. Specific genes have been identified that may determine how patients will respond to specific drugs. For example, some genes may determine whether people will get pain relief from codeine, or how well they will respond to drugs used to treat **cancer**.

Causes and symptoms

Genes alter responses to drugs because the genes influence many parts of the body itself. One of the simplest examples is the gene that influences body weight. Since many drugs are soluble in body fat, people with large amounts of fat will have these drug deposited into their fat stores. This means that there are lower levels of the drug that can reach the actual organs on which they work.

In the case of gender responses to antidepressants, women show greater response to serotonin specific antidepressants because women naturally have lower levels of serotonin than men do. This makes women more likely to develop a type of depression marked by low serotonin levels, but it also means that women will respond better to replacement of serotonin.

Because people of the same race carry similar genes, studies based on race were the earliest types of pharmacogenetic studies. One study evaluated the levels of alcohol dehydrogenase in people of different nationalities. This is an enzyme involved in the metabolism of alcohol. When people with high levels of this enzyme, or people in whom the enzyme acts more rapidly than in other people, drink alcohol, they are subject to facial flushing and slowing of the heartbeat. The activity of this enzyme is determined by genetics, and different levels can be seen in different races because these people belong to the same gene pools. Among Asiatic people, 85% have high levels of this enzyme, compared to 20% of Swiss people, and only 5–10% of British people.

Another trait that is influenced by genes is a liver enzyme, CYP2D6. This enzyme metabolizes some drugs, convert them to a form that can be removed from the body. Genes determine the level of this enzyme in the liver. People with low levels of CYP2D6 will metabolize drugs slowly. Slow metabolism means the drugs will act for a longer period of time. Slow metabolizers respond to smaller doses of medications that are eliminated by this enzyme, while fast metabolizers, people who have a lot of the enzyme, will need larger drug doses to get the same effects. At the same time, low levels of CYP2D6 means that people taking the drugs that are metabolized by

this enzyme will have higher drug levels, and are more likely to have unwanted side effects.

Another enzyme that can be important in drug dosing is called 2C9, and this enzyme is responsible for metabolizing the anticoagulant drug warfarin (Coumadin). Most people take warfarin in a dose of about 5 milligrams a day, but people who have low levels of 2C9 normally require a dose of only 1–5 milligrams a week.

Yet another mechanism of drug activity is the presence or absence of a specific drug receptor site. Drugs act by binding to specific chemicals, receptor sites, within body cells. Genes may help determine how many of these cells there are. The action of the widely used antipsychotic drug haloperidol (Haldol) depends on its ability to bind to the dopamine (D2) receptor site. The number of these sites are determined by genetics. In one study, 63% of patients whose genes caused a large number of these receptor sites had a response to treatment with haloperidol, while only about 29% of patients with a smaller number of dopamine (D2) receptor sites did well on the drug.

Other genetic studies indicate that genes may affect how people respond to foods as well as to drugs. An Australian study of **osteoporosis** (softening of the bones that often occurs in elderly people), reported that separate genes may affect response to vitamin D, **calcium**, and estrogens.

Implications

Although the study is still new, pharmacogenetics promises to offer great benefits in drug effectiveness and safety.

At the present time, most drug treatment is done by trial and error. Physicians prescribe medication, and the patient tries the drug. The drug may work, or it may not. It may cause adverse effects, or it may be safe. If the drug does not work, the dose is increased. If it causes harmful or unpleasant effects, a new drug is tried until, finally, the right drug is found. In some cases this procedure may take weeks or even months.

In other cases, drugs are carefully tested, and appear to be safe and effective. Only after they are approved for general use are reports of serious adverse effects that did not appear in the initial studies documented. This can occur if there is a rare gene that affects the way in which the drug acts, or the way in which the drug is metabolized.

With increasing understanding of how genes determine the way people respond to drugs, it will be possible to select drugs and doses based on a greater

KEY TERMS

Enzyme—Proteins produced by living cells that help produce specific biochemical reactions in the body.

Metabolism—The process by which foods and drugs are broken down for use and removal from the body.

Sickle cell anemia—A severe, inheritable disease, most common among people of African descent, marked by deformation and destruction of red blood cells, and by adherence of blood cells to the walls of blood vessels.

understanding of each individual patient. This promises more effective drug therapy, with greater safety and fewer treatment failures.

Physicians may be able to compare the person's genetic make-up with the properties of specific drugs, and make informed decisions about which drug in a group will work most effectively or most safely.

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ORGANIZATIONS

National Institute of General Medical Sciences, 45 Center Drive MSC 6200, Bethesda, MD, 20892-6200, (301) 496-7301, <http://www.nigms.nih.gov>.

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Pharyngeal pouch see **Esophageal pouches**

Pharyngitis see **Sore throat**

Phenelzine see **Monoamine oxidase inhibitors**

Phenobarbital see **Barbiturates**

Phenol see **Antiseptics**

Phenolphthalein see **Laxatives**

Phenylalaninemia see **Phenylketonuria**

Phenylketonuria

Definition

Phenylketonuria (PKU) can be defined as a rare metabolic disorder caused by a deficiency in the production of the hepatic (liver) enzyme phenylalanine hydroxylase (PAH). PKU is the most serious form of a class of diseases referred to as “hyperphenylalaninemia,” all of which involve above normal (elevated) levels of phenylalanine in the blood. The primary symptom of untreated PKU, **mental retardation**, is the result of consuming foods that contain the amino acid phenylalanine, which is toxic to brain tissue.

PKU is an inherited, autosomal recessive disorder. It is the most common genetic disease involving “amino acid metabolism.” PKU is incurable, but early, effective treatment can prevent the development of serious mental incapacity.

Description

PKU is a disease caused by the liver’s inability to produce a particular type of PAH enzyme. This enzyme converts (metabolizes) the amino acid called phenylalanine into another amino acid, tyrosine. This is the only role of PAH in the body. A lack of PAH results in the buildup of abnormally high phenylalanine concentrations (or levels) in the blood and brain. Above normal levels of phenylalanine are toxic to the cells that make up the nervous system and causes irreversible abnormalities in brain structure and function in PKU patients. Phenylalanine is a type of teratogen. Teratogens are any substance or organism that can cause **birth defects** in a developing fetus.

The liver is the body’s chief protein processing center. Proteins are one of the major food nutrients. They are generally very large molecules composed of strings of smaller building blocks or molecules called

Phenylketonuria (PKU) diet

A PKU diet is based on consuming foods low in protein. Foods that should be avoided include:

- | | |
|--|---|
| <ul style="list-style-type: none"> • Beans • Chocolate • Dairy products • Fish • Foods and beverages sweetened with aspartame | <ul style="list-style-type: none"> • Nuts or nut butters • Peas • Poultry • Red meat • Soy |
|--|---|

amino acids. About twenty amino acids exist in nature. The body breaks down proteins from food into individual amino acids and then reassembles them into “human” proteins. Proteins are needed for growth and repair of cells and tissues, and are the key components of enzymes, antibodies, and other essential substances.

PKU affects on the human nervous system

The extensive network of nerves in the brain and the rest of the nervous system are made up of nerve cells. Nerve cells have specialized extensions called dendrites and axons. Stimulating a nerve cell triggers nerve impulses, or signals, that speed down the axon. These nerve impulses then stimulate the end of an axon to release chemicals called “neurotransmitters” that spread out and communicate with the dendrites of neighboring nerve cells.

Many nerve cells have long, wire-like axons that are covered by an insulating layer called the myelin sheath. This covering helps speed nerve impulses along the axon. In untreated PKU patients, abnormally high phenylalanine levels in the blood and brain can produce nerve cells with “deformed” axons and dendrites, and cause imperfections in the myelin sheath referred to as hypomyelination and demyelination. This loss of myelin can “short circuit” nerve impulses (messages) and interrupt cell communication. A number of brain scan studies also indicate a degeneration of the “white matter” in the brains of older patients who have not maintained adequate dietary control.

PKU can also affect the production of one of the major neurotransmitters in the brain, called dopamine. The brain makes dopamine from the amino acid tyrosine. PKU patients who do not consume enough tyrosine in their diet cannot produce sufficient amounts of dopamine. Low dopamine levels in the brain disrupt normal communication between nerve cells, which results in impaired cognitive (mental) function.

Some preliminary research suggests that nerve cells of PKU patients also have difficulty absorbing tyrosine. This abnormality may explain why many PKU patients who receive sufficient dietary tyrosine still experience some form of learning disability.

Behavior and academic performance

IQ (intelligence quotient) tests provide a measure of cognitive function. The IQ of PKU patients is generally lower than the IQ of their healthy peers. Students with PKU often find academic tasks difficult and must struggle harder to succeed than their non-PKU peers.

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

They may require special tutoring and need to repeat some of their courses. Even patients undergoing treatment programs may experience problems with typical academic tasks as math, reading, and spelling. Visual perception, visual-motor skills, and critical thinking skills can also be affected. Ten years of age seems to be an important milestone for PKU patients. After age 10, variations in a patient's diet seems to have less influence on their IQ development.

People with PKU tend to avoid contact with others, appear anxious and show signs of depression. However, some patients may be much more expressive and tend to have hyperactive, talkative, and impulsive personalities. It is also interesting to note that people with PKU are less likely to display such "antisocial" habits as lying, teasing, and active disobedience. It should be emphasized that current research findings are still quite preliminary and more extensive research is needed to clearly show how abnormal phenylalanine levels in the blood and brain might affect behavior and academic performance.

One in fifty individuals in the United States have inherited a gene for PKU. About five million Americans are PKU carriers. About one in 15,000 babies test positive for PKU in the United States. Studies indicate that the incidence of this disease in Caucasian and Native American populations is higher than in African-American, Hispanic, and Asian populations.

Causes and symptoms

PKU symptoms are caused by alterations or "mutations" in the genetic code for the PAH enzyme. Mutations in the PAH gene prevent the liver from producing adequate levels of the PAH enzyme needed to break down phenylalanine. The PAH gene and its PKU mutations are found on chromosome 12 in the human genome. In more detail, PKU mutations can involve many different types of changes, such as deletions and insertions, in the DNA of the gene that codes for the PAH enzyme.

PKU is described as an inherited, autosomal recessive disorder. The term autosomal means that the gene for PKU is not located on either the X or Y sex chromosome. The normal PAH gene is dominant to recessive PKU mutations. A recessive genetic trait, such as PKU, is one that is expressed—or shows up—only when two copies are inherited (one from each parent).

A person with one normal and one PKU gene is called a carrier. A carrier does not display any symptoms of the disease because their liver produces normal quantities of the PAH enzyme. However, PKU

carriers can pass the PKU genetic mutation onto their children. Two carrier parents have a 25% chance of producing a baby with PKU symptoms, and a 50% chance having a baby that is a carrier for the disease. Although PKU conforms to these basic genetic patterns of inheritance, the actual expression, or phenotype, of the disease is not strictly an "either/or" situation. This is because there are at least 400 different types of PKU mutations. Although some PKU mutations cause rather mild forms of the disease, others can initiate much more severe symptoms in untreated individuals. The more severe the PKU mutation, the greater the effect on cognitive development and performance (mental ability).

Untreated PKU patients develop a broad range of symptoms related to severely impaired cognitive function, sometimes referred to as mental retardation. Other symptoms can include extreme patterns of behavior, delayed speech development, seizures, a characteristic body odor, and light body pigmentation. The light pigmentation is due to a lack of melanin, which normally colors the hair, skin and eyes. Melanin is made from the amino acid tyrosine, which is lacking in untreated cases of PKU. Physiologically, PKU patients show high levels of phenylalanine and low levels of tyrosine in the blood. Babies do not show any visible symptoms of the disease for the first few months of life. However, typical PKU symptoms usually do show up by a baby's first birthday.

Diagnosis

The primary diagnostic test for PKU is the measurement of phenylalanine levels in a drop of blood taken from the heel of a newborn baby's foot. This screening procedure is referred to as the Guthrie test (Guthrie bacterial inhibition assay). In this test, PKU is confirmed by the appearance of bacteria growing around high concentrations of phenylalanine in the blood spot. PKU testing was introduced in the early 1960s and is the largest genetic screening program in the United States. It is required by law in all 50 states. Early diagnosis is critical. It ensures early the treatment PKU babies need to develop normally and avoid the ravages of PKU.

The American Academy of Pediatrics recommends that this test should be performed on infants between 24 hours and seven days after birth. The preferred time for testing is after the baby's first feeding. If the initial PKU test produces a positive result, then follow-up tests are performed to confirm the diagnosis and to determine if the elevated phenylalanine levels may be caused by some medical condition other than PKU. Treatment for PKU is recommended

KEY TERMS

Amino acid—Organic compounds that form the building blocks of protein. There are 20 types of amino acids (eight are “essential amino acids” which the body cannot make and must therefore be obtained from food).

Axon—Skinny, wire-like extension of nerve cells.

Enzyme—A protein that catalyzes a biochemical reaction or change without changing its own structure or function.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Genetic disease—A disease that is (partly or completely) the result of the abnormal function or expression of a gene; a disease caused by the inheritance and expression of a genetic mutation.

IQ—Abbreviation for Intelligence Quotient. Compares an individual’s mental age to his/her true or chronological age and multiplies that ratio by 100.

Metabolism—The total combination of all of the chemical processes that occur within cells and tissues of a living body.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease, and can be transmitted to offspring.

Myelin—A fatty sheath surrounding nerves in the peripheral nervous system, which help them conduct impulses more quickly.

Nervous system—The complete network of nerves, sense organs, and brain in the body.

Phenylalanine—An essential amino acid that must be obtained from food since the human body cannot manufacture it.

Protein—Important building blocks of the body, composed of amino acids, involved in the formation of body structures and controlling the basic functions of the human body.

Recessive—Genetic trait expressed only when present on both members of a pair of chromosomes, one inherited from each parent.

for babies that show a blood phenylalanine level of 7–10 mg/dL or higher for more than a few consecutive days. Another, more accurate test procedure for PKU measures the ratio (comparison) of the amount of phenylalanine to the amount of tyrosine in the blood.

Newer diagnostic procedures (called mutation analysis and genotype determination) can actually identify the specific types of PAH gene mutations inherited by PKU infants. Large-scale studies have helped to clarify how various mutations affect the ability of patients to process phenylalanine. This information can help doctors develop more effective customized treatment plans for each of their PKU patients.

Treatment

The severity of the PKU symptoms experienced by people with this disease is determined by both lifestyle as well as genetic factors. In the early 1950s, researchers first demonstrated that phenylalanine-restricted diets could eliminate most of the typical PKU symptoms—except for mental retardation. Today, dietary therapy (also called **nutrition** therapy)

is the most common form of treatment for PKU patients. PKU patients who receive early and consistent dietary therapy can develop fairly normal mental capacity to within about five IQ points of their healthy peers. By comparison, untreated PKU patients generally have IQ scores below 50.

Infants with PKU should be put on a specialized diet as soon as they are diagnosed to avoid progressive brain damage and other problems caused by an accumulation of phenylalanine in the body. A PKU diet helps patients maintain very low blood levels of phenylalanine by restricting the intake of natural foods that contain this amino acid. Even breast milk is a problem for PKU babies. Special PKU dietary mixtures or formulas are usually obtained from medical clinics or pharmacies.

Phenylalanine is actually an essential amino acid. This means that it has to be obtained from food because the body cannot produce this substance on its own. Typical diets prescribed for PKU patients provide very small amounts of phenylalanine and higher quantities of other amino acids, including tyrosine. The amount of allowable phenylalanine can be increased slightly as a child becomes older.

In addition, PKU diets include all the nutrients normally required for good health and normal growth, such as carbohydrates, fats, **vitamins**, and **minerals**. High protein foods like meat, fish, chicken, eggs, nuts, beans, milk, and other dairy products are banned from PKU diets. Small amounts of moderate protein foods (such as grains and potatoes) and low protein foods (some fruits and vegetables, low protein breads and pastas) are allowed. Sugar-free foods, such as diet soda, which contain the artificial sweetener aspartame, are also prohibited foods for PKU patients. That is because aspartame contains the amino acid phenylalanine.

Ideally, school-age children with PKU should be taught to assume responsibility for managing their diet, recording food intake, and for performing simple blood tests to monitor their phenylalanine levels. Blood tests should be done in the early morning when phenylalanine levels are highest. Infants and young children require more frequent blood tests than older children and adults. The amount of natural foods allowed in a diet could be adjusted to ensure that the level of phenylalanine in the blood is kept within a safe range—two to 6 mg/dL before 12 years of age and 2–15 mg/dL for PKU patients over 12 years old.

A specialized PKU diet can cause abnormal fluctuations in tyrosine levels throughout the day. Thus, some health professionals recommend adding time released tyrosine that can provide a more constant supply of this amino acid to the body. It should be noted that some PKU patients show signs of learning disabilities even with a special diet containing extra tyrosine. Research studies suggests that these PKU patients may not be able to process tyrosine normally.

For PKU caregivers, providing a diet that is appealing as well as healthy and nutritious is a constant challenge. Many PKU patients, especially teenagers, find it difficult to stick to the relatively bland PKU diet for extended periods of time. Some older patients decide to go off their diet plan simply because they feel healthy. However, many patients who abandon careful nutritional management develop cognitive problems, such as difficulties remembering, maintaining focus, and paying attention. Many PKU health professionals contend that all PKU patients should adhere to a strictly controlled diet for life.

One promising line of PKU research involves the synthesis (manufacturing) of a new type of enzyme that can break down phenylalanine in food consumed by the patient. This medication would be taken orally and could prevent the absorption of digested phenylalanine into the patient's bloodstream.

In general, medical researchers express concern about the great variation in treatment programs currently available to PKU patients around the world. They have highlighted the urgent need for new, consistent international standards for proper management of PKU patients, which should emphasize comprehensive psychological as well as physiological monitoring and assessment.

PKU and Pregnancy

Women with PKU must be especially careful with their diets if they want to have children. They should ensure that phenylalanine blood levels are under control before conception and throughout her **pregnancy**. Mothers with elevated (higher than normal) phenylalanine levels are high risk for having babies with significant birth defects, such as microencephaly (smaller than normal head size), and **congenital heart disease** (abnormal heart structure and function), stunted growth, mental retardation, and psychomotor (coordination) difficulties. This condition is referred to as maternal PKU and can even affect babies who do not have the PKU disease.

Prognosis

Early newborn screening, careful monitoring, and a life-long strict dietary management can help PKU patients to live normal, healthy, and long lives.

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American Academy of Allergy, Asthma & Immunology, 555 East Wells Street, Suite 1100, Milwaukee, WI, 53202-3823, (414) 272-6071, <http://www.aaaai.org>.

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, New York, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Children's PKU Network (CPN), 3790 Via De La Valle, Ste 120, Del Mar, CA, 92014, (858) 509-0767, (858) 509-0768, (800) 377-6677, pkunetwork@aol.com, <http://www.pkunetwork.org/>.

March of Dimes Birth Defects Foundation, 1275 Mamaroneck Ave., White Plains, NY, 10605, (914) 997-4488, <http://www.modimes.org>.

National Society for Phenylketonuria, PO Box 26642, London, England, N14 4ZF, 440208364 3010, info@nspku.org, <http://www.nspku.org/>.

University of Washington PKU Clinic, CHDD, Box 357920, University of Washington, Seattle, WA, 206 685-3015, <http://depts.washington.edu/pku/contact.html>.

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Phenylpropanolamine see **Decongestants**

Phenytoin see **Anticonvulsant drugs**

Pheochromocytoma

Definition

Pheochromocytoma is a tumor of special cells (called chromaffin cells), most often found in the middle of the adrenal gland.

Description

Because pheochromocytomas arise from chromaffin cells, they are occasionally called chromaffin tumors. Most (90%) are benign tumors so they do not spread to other parts of the body. However, these tumors can cause many problems and if they are not treated and can result in **death**.

Pheochromocytomas can be found anywhere chromaffin cells are found. They may be found in the heart and in the area around the bladder, but most (90%) are found in the adrenal glands. Every individual has two adrenal glands that are located above the kidneys in the back of the abdomen. Each adrenal

gland is made up of two parts: the outer part (called the adrenal cortex) and the inner part (called the adrenal medulla). Pheochromocytomas are found in the adrenal medulla. The adrenal medulla normally secretes two substances, or hormones, called norepinephrine and epinephrine. These two substances, when considered together, are known as adrenaline. Adrenaline is released from the adrenal gland, enters the bloodstream and helps to regulate many things in the body including blood pressure and heart rate. Pheochromocytomas cause the adrenal medulla to secrete too much adrenaline, which in turn causes high blood pressure. The high blood pressure usually causes the other symptoms of the disease.

Pheochromocytomas are rare tumors. They have been reported in babies as young as five days old as well as adults 92 years old. Although they can be found at any time during life, they usually occur in adults between 30 and 40 years of age. Pheochromocytomas are somewhat more common in women than in men.

Causes and symptoms

The cause of most pheochromocytomas is not known. A small minority (about 10-20%) of pheochromocytomas arise because a person has an inherited susceptibility to them. Inherited pheochromocytomas are associated with four separate syndromes: Multiple Endocrine Neoplasia, type 2A (MEN2A), Multiple Endocrine Neoplasia, type 2B (MEN2B), von Hippel-Lindau disease (VHL) and **Neurofibromatosis** type 1 (NF1).

Individuals with pheochromocytomas as part of any of these four syndromes usually have other medical conditions as well. People with MEN2A often have **cancer** (usually **thyroid cancer**) and other hormonal problems. Individuals with MEN2B can also have cancer and hormonal problems, but also have other abnormal physical features. Both MEN2A and MEN2B are due to genetic alterations or mutations in a gene called RET, found at chromosome 10q11.2. Individuals with VHL often have other benign tumors of the central nervous system and pancreas, and can sometimes have renal cell cancer. This syndrome is caused by a mutation in the VHL gene, found at chromosome 3p25-26. Individuals with NF1 often have neurofibromas (benign tumors of the peripheral nervous system). NF1 is caused by mutations in the NF1 gene, found at chromosome 17q11.

All of these disorders are inherited in an autosomal dominant inheritance pattern. With autosomal dominant inheritance, men and women are equally likely to inherit the syndrome. In addition, children

of individuals with the disease are at 50% risk of inheriting it. **Genetic testing** is available for these four syndromes (MEN2A, MEN2B, VHL and NF1) but, due to the complexity, **genetic counseling** should be considered before testing.

Most people (90%) with pheochromocytoma have **hypertension**, or high blood pressure. The other symptoms of the disease are extremely variable. These symptoms usually occur in episodes (or attacks) called paroxysms and include:

- headaches
- excess sweating
- racing heart
- rapid breathing
- anxiety/nervousness
- nervous shaking
- pain in the lower chest or upper abdomen
- nausea
- heat intolerance

The episodes can occur as often as 25 times a day or, as infrequently as once every few months. They can last a few minutes, several hours, or days. Usually, the attacks occur several times a week and last for about 15 minutes. After the episode is over, the person feels exhausted and fatigued.

Between the attacks, people with pheochromocytoma can experience the following:

- increased sweating
- cold hands and feet
- weight loss
- constipation

Diagnosis

If a pheochromocytoma is suspected, urine and/or a blood test are usually recommended. A test called “24-hour urinary catecholamines and metanephrines” will be done. This test is designed to look for adrenaline and the break-down products of adrenaline. Since the body gets rid of these hormones in the urine, those testing will need to collect their urine for 24 hours. The laboratory will determine whether or not the levels of hormones are too high. This test is very good at making the diagnosis of pheochromocytoma. Another test called “serum catecholamines” measures the level of adrenaline compounds in the blood. It is not as sensitive as the 24-hour urine test, but can still provide some key information if it shows that the level of adrenaline compounds is too high.

One of the difficulties with these tests is that a person needs to have an attack of symptoms either during the 24-hour urine collection time period or shortly before the blood is drawn for a serum test to ensure the test’s accuracy. If a person did not have an episode during that time, the test can be a “false negative.” If a doctor suspects the patient has had a “false negative” test, additional tests called “pharmacologic tests” can be ordered. During these tests, a specific drug is given to the patient (usually through an IV) and the levels of hormones are monitored from the patient’s blood. These types of tests are only done rarely.

Once a person has been diagnosed with a pheochromocytoma, he or she will undergo tests to identify exactly where in the body the tumor is located. The imaging techniques used are usually computed tomography scan (CT scan) and magnetic resonance imaging (MRI). A CT scan creates pictures of the interior of the body from computer-analyzed differences in x rays passing through the body. CT scans are performed at a hospital or clinic and take only a few minutes. An MRI is a computerized scanning method that creates pictures of the interior of the body using radio waves and a magnet. An MRI is usually performed at a hospital and takes about 30 minutes.

Treatment

Once a pheochromocytoma is found, more tests will be done to see if the tumor is benign (not cancer) or malignant (cancer). If the tumor is malignant, tests will be done to see how far the cancer has spread. There is no accepted staging system for pheochromocytoma; but an observation of the tumor could provide one of these four indications:

- Localized benign pheochromocytoma means that the tumor is found only in one area, is not cancer, and cannot spread to other tissues of the body.
- Regional pheochromocytoma means that the tumor is malignant and has spread to the lymph nodes around the original cancer. Lymph nodes are small structures found all over the body that make and store infection-fighting cells.
- Metastatic pheochromocytoma means that the tumor is malignant and has spread to other, more distant parts of the body.
- Recurrent pheochromocytoma means that a malignant tumor that was removed has come back.

Treatment in all cases begins with surgical removal of the tumor. Before surgery, medications such as alpha-adrenergic blockers are given to block the effect of the hormones and normalize blood

KEY TERMS

Adrenal medulla—The central core of the adrenal gland.

Laparoscope—An instrument used to examine body cavities during certain types of surgery; for example, surgeries to remove fibroid tumors, or gall bladders, are often removed through the navel rather than cutting into the body.

Paroxysm—A sudden attack of symptoms.

pressure. These medications are usually started seven to 10 days prior to surgery. The surgery of choice is laparoscopic laparotomy, which is a minimally invasive outpatient procedure performed under general or **local anesthesia**. A small incision is made in the abdomen, the laparoscope is inserted, and the tumor is removed. The patient can usually return to normal activities within two weeks. If a laparoscopic laparotomy cannot be done, a traditional laparotomy will be performed. This is a more invasive surgery done under spinal or **general anesthesia** and requires five to seven days in the hospital. Usually patients are able to return to normal activities after four weeks. After surgery, blood and urine tests will be done to make sure hormone levels return to normal. If the hormone levels are still above normal, it may mean that some tumor tissue was not removed. If not all tumor can be removed (as in malignant pheochromocytoma, for example) drugs will be given to control high blood pressure.

If a pheochromocytoma is malignant, **radiation therapy** and/or **chemotherapy** may be used. Radiation therapy uses high-energy x rays to kill cancer cells and shrink tumors. Because there is no evidence that radiation therapy is effective in the treatment of malignant pheochromocytoma, it is not often used for treatment. However, it is useful in the treatment of painful bone metastases if the tumor has spread to the bones. Chemotherapy uses drugs to kill cancer cells. Like radiation therapy, it has not been shown to be effective in the treatment of malignant pheochromocytoma. Chemotherapy, therefore, is only used in rare instances.

Untreated pheochromocytoma can be fatal due to complications of the high blood pressure. In the vast majority of cases, when the tumor is surgically removed, pheochromocytoma is cured. In the minority of cases (10%) where pheochromocytoma is malignant, prognosis depends on how far the cancer has spread, and the patient's age and general health. The

overall median five-year survival from the initial time of surgery and diagnosis is approximately 43%.

Prevention

Unfortunately, little is known about environmental and other causes of pheochromocytoma. Some of the tumors are due to inherited predisposition. Because of these factors, pheochromocytoma can not be prevented.

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Phimosis

Definition

A tightening of the foreskin of the penis that may close the opening of the penis.

Description

The foreskin of a newborn boy is always closely contracted around the penis head (glans). Only a small passage allows the urine to pass through. In the first months the foreskin is stuck to the glans and cannot be pulled back and one should not attempt to do so. During the first couple of years, the foreskin will become gradually looser and in many boys it can in time be pulled back without trouble. Half of all three-year-olds can pull back their foreskin. It is not advisable to try pulling the foreskin back using force, since this may cause small cuts in the foreskin with **scars** which could finally cause a regular foreskin contraction.

Foreskin contraction, called phimosis, can last throughout life and not cause any trouble at all. It is a voluntary decision whether to have a **circumcision** operation or not. If any problems do arise, they happen after **puberty**. The contraction may occur for the

first time as an adult and usually requires circumcision, the surgical removal of the foreskin.

Causes & symptoms

Phimosis is caused by the inability of the foreskin to retract from around the opening of the penis. In adults, phimosis can lead to chronic inflammation and cancer.

Diagnosis

A physician usually diagnoses phimosis when there are persistent problems urinating, when there are recurrent infections under the foreskin, or when the opening to the penis is completely blocked by the foreskin. Phimosis is a tight ring of foreskin often made of scar tissue preventing retraction of the foreskin. It may be primary, or secondary to recurrent infection. It may produce urinary obstruction with ballooning of the foreskin. Phimosis is different than having a non-retractable foreskin, which is normal in many boys.

Treatment

If the foreskin cannot be pulled back into place treatment should be sought. If the blood flow to the penis is restricted then emergency treatment is required and if the foreskin cannot be pulled back a surgical cut to the trapped foreskin may be required. Failure to seek treatment can result in permanent damage to the penis. Once phimosis is diagnosed, the available treatments include topical **corticosteroids**, manual stretching, foreskin surgical repair or **plastic surgery**, and circumcision. Conservative treatments should be tried in the first instance and surgery used as the treatment of last resort.

A number of studies show that phimosis can be safely and effectively treated by the application of topical **steroids** in 80–90% of cases. Betamethasone cream 0.05% should be applied to the exterior and interior of the tip of the foreskin two or three times a day. The treatment should be discontinued as ineffective after three months if the foreskin has not become retractile during this time.

A number of corrections are available for the adult or adolescent non-retractable foreskin. These include surgery to repair the foreskin, in which an incision is made through the constrictive band of the foreskin. The underlying tissue is spread with forceps to expose the Buck's fascia (the deep, connective tissue of the penis) and the incision is closed with absorbable sutures. This procedure has less risk of disease and

KEY TERMS

Balanitis xerotica obliterans (Bxo)—A chronic, progressive, hardening skin inflammation of the penis.

Buck's fascia—The deep connective tissue of the penis.

Circumcision—The removal of all or part of the foreskin from the penis.

Corticosteroids—A synthetic drug similar or identical to a natural corticosteroid, used to reduce inflammation.

Glans—The head of the penis.

Paraphimosis—The entrapment of a retracted foreskin behind the coronal sulcus, a groove that separates the shaft and head of the penis.

infection than circumcision, and allows the foreskin to be retained.

Circumcision is very traumatic to a child. It is essentially irreversible and should be the treatment of last resort. Phimosis due to *balanitis xerotica obliterans* (Bxo), a chronic, progressive, hardening skin inflammation of the penis, has been considered the one common absolute indication for circumcision.

Alternative treatment

There are no alternative medicine treatments for phimosis.

Prognosis

In most men, phimosis is not a serious problem and will not require treatment. However, it is not expected to improve on its own. With treatment, phimosis in most males can be managed or corrected.

Prevention

Proper hygiene is the most important preventative measure. The American Academy of Pediatrics recommends that the immature foreskin of boys not be forced back for cleaning. The only person who should clean and retract the foreskin is the boy himself. Bubble bath products and other chemical irritants can cause the foreskin to tighten and it is recommended they should be avoided by males with foreskins.

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Phlebitis see **Thrombophlebitis**

Phlebotomy

Definition

Phlebotomy is the act of drawing or removing blood from the circulatory system through a cut (incision) or puncture in order to obtain a sample for analysis and diagnosis. Phlebotomy is also done as part of the patient's treatment for certain blood disorders.

Purpose

Treatment

Phlebotomy that is part of treatment (therapeutic phlebotomy) is performed to treat **polycythemia vera**, a condition that causes an elevated red blood cell volume (**hematocrit**). Phlebotomy is also prescribed for patients with disorders that increase the amount of iron in their blood to dangerous levels, such as **hemochromatosis**, **hepatitis B**, and **hepatitis C**. Patients with **pulmonary edema** may undergo phlebotomy procedures to decrease their total blood volume.

Diagnosis

Phlebotomy is also used to remove blood from the body during **blood donation** and for analysis of the substances contained within it.

Precautions

Patients who are anemic or have a history of cardiovascular disease may not be good candidates for phlebotomy.

Description

Phlebotomy, which is also known as venesection, is performed by a nurse or a technician known as a phlebotomist. Blood is usually taken from a vein on the back of the hand or inside of the elbow. Some blood tests, however, may require blood from an artery. The skin over the area is wiped with an anti-septic, and an elastic band is tied around the arm. The band acts as a tourniquet, slowing the blood flow in the arm and making the veins more visible. The patient is asked to make a fist, and the technician feels the veins in order to select an appropriate one. When a vein is selected, the technician inserts a needle into the vein and releases the elastic band. The appropriate amount of blood is drawn and the needle is withdrawn from the vein. The patient's pulse and blood pressure may be monitored during the procedure.

For some tests requiring very small amounts of blood for analysis, the technician uses a finger stick. A lance, or small needle, makes a small cut in the surface of the fingertip, and a small amount of blood is collected in a narrow glass tube. The fingertip may be squeezed to get additional blood to surface.

The amount of blood drawn depends on the purpose of the phlebotomy. Blood donors usually contribute a unit of blood (500 mL) in a session. The volume of blood needed for laboratory analysis varies widely with the type of test being conducted. Therapeutic phlebotomy removes a larger amount of blood than donation and blood analysis require. Phlebotomy for treatment of hemochromatosis typically involves removing a unit of blood—or 250 mg of iron—once a week. Phlebotomy sessions are required until iron levels return to a consistently normal level, which may take several months to several years. Phlebotomy for polycythemia vera removes enough blood to keep the patient's hematocrit below 45%. The frequency and duration of sessions depends on the patient's individual needs.

Preparation

Patients having their blood drawn for analysis may be asked to discontinue medications or to avoid food (to fast) for a period of time before the blood test. Patients donating blood will be asked for a brief medical history, have their blood pressure taken, and have

KEY TERMS

Finger stick—A technique for collecting a very small amount of blood from the fingertip area.

Hemochromatosis—A genetic disorder known as iron overload disease. Untreated hemochromatosis may cause osteoporosis, arthritis, cirrhosis, heart disease, or diabetes.

Thrombocytosis—A vascular condition characterized by high blood platelet counts.

Tourniquet—Any device that is used to compress a blood vessel to stop bleeding or as part of collecting a blood sample. Phlebotomists usually use an elastic band as a tourniquet.

Venesection—Another name for phlebotomy.

their hematocrit checked with a finger stick test prior to donation.

Aftercare

After blood is drawn and the needle is removed, pressure is placed on the puncture site with a cotton ball to stop bleeding, and a bandage is applied. It is not uncommon for a patient to feel dizzy or nauseated during or after phlebotomy. The patient may be encouraged to rest for a short period once the procedure is completed. Patients are also instructed to drink plenty of fluids and eat regularly over the next 24 hours to replace lost blood volume. Patients who experience swelling of the puncture site or continued bleeding after phlebotomy should get medical help at once.

Risks

Most patients will have a small bruise or mild soreness at the puncture site for several days. Therapeutic phlebotomy may cause **thrombocytosis** and chronic iron deficiency (anemia) in some patients. As with any invasive procedure, infection is also a risk. This risk can be minimized by the use of prepackaged sterilized equipment and careful attention to proper technique.

Normal results

Normal results include obtaining the needed amount of blood with the minimum of discomfort to the patient.

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Phobias

Definition

A phobia is an intense but unrealistic fear that can interfere with the ability to socialize, work, or go about everyday life, brought on by an object, event, or situation.

Demographics

Phobias occur in all races, with social phobia being the most common type of phobia and **agoraphobia** being the least common. Women are more likely to suffer from a phobia than men at a ratio of about two to one. Simple and social phobias appear earlier in life (at a median age of 15 and 16) than agoraphobia (which appears at a median age of 29).

Description

Just about everyone is afraid of something such as an upcoming job interview or being alone outside after dark, but about 18% of all Americans are tormented by irrational fears that interfere with their daily lives. They are not "crazy"—they know their fear is unreasonable, but they cannot control the fear. These people have phobias.

Phobias belong to a large group of mental problems known as **anxiety disorders** that include **obsessive-compulsive disorder (OCD)**, **panic disorder**, and **post-traumatic stress disorder (PTSD)**. Phobias themselves can be divided into three types:

- specific phobias (formerly called "simple phobias")
- social phobia
- agoraphobia

Specific phobias

As its name suggests, a specific phobia is the fear of a particular situation or object, including anything from airplane travel to dental visits. Found in one out of every 10 Americans, specific phobias seem to run in families and are roughly twice as likely to appear in females. If the person rarely encounters the feared

object, the phobia does not cause much harm. However, if the feared object or situation is common, it can seriously disrupt everyday life. Common examples of specific phobias, which can begin at any age, include fear of snakes, flying, dogs, escalators, elevators, high places, or open spaces.

Social phobia

People with social phobia have deep fears of being watched or judged by others and being embarrassed in public. This may extend to a general fear of social situations or be more specific or circumscribed, such as a fear of giving speeches or of performing (stage fright). More rarely, people with social phobia may have trouble using a public restroom, eating in a restaurant, or signing their name in front of others.

Social phobia is not the same as **shyness**. Shy people may feel uncomfortable with others, but they do not experience severe **anxiety**, do not worry excessively about social situations beforehand, and do not avoid events that make them feel self-conscious. On the other hand, people with social phobia may not be shy; they may feel perfectly comfortable with people except in specific situations. Social phobias may be only mildly irritating, or they may significantly interfere with daily life. It is not unusual for people with social phobia to turn down job offers or avoid relationships because of their fears.

Agoraphobia

Agoraphobia is the intense fear of feeling trapped and having a panic attack in a public place. This type of phobia usually begins between ages 15 and 35, and affects three times as many women as men, or about 3% of the population.

An episode of spontaneous panic is usually the initial trigger for the development of agoraphobia. After an initial panic attack, the person becomes afraid of experiencing a second one. Individuals “fear the fear,” and worry incessantly about when and where the next attack may occur. As they begin to avoid the places or situations in which the panic attack occurred, their fear generalizes. Eventually the person completely avoids public places. In severe cases, people with agoraphobia can no longer leave their homes for fear of experiencing a panic attack.

Causes and symptoms

Experts do not really know why phobias develop, although research suggests the tendency to develop phobias may be a complex interaction between heredity and environment. Some hypersensitive people have

unique chemical reactions in the brain that cause them to respond much more strongly to **stress**. These people also may be especially sensitive to **caffeine**, which triggers certain brain chemical responses.

While experts believe the tendency to develop phobias runs in families and may be hereditary, a specific stressful event usually triggers the development of a specific phobia or agoraphobia. For example, someone predisposed to develop phobias who experiences severe turbulence during a flight might go on to develop a phobia about flying. What scientists do not understand is why some people who experience a frightening or stressful event develop a phobia and others do not.

Social phobia typically appears in childhood or adolescence, sometimes following an upsetting or humiliating experience. Certain vulnerable children who have had unpleasant social experiences (e.g., being rejected) or who have poor social skills may develop social phobias. The condition also may be related to low self-esteem, unassertive personality, and feelings of inferiority.

A person with agoraphobia may have a panic attack at any time, for no apparent reason. While the attack may last only a minute or so, the person remembers the feelings of panic so strongly that the possibility of another attack becomes terrifying. For this reason, people with agoraphobia avoid places where they might not be able to escape if a panic attack occurs. As the fear of an attack escalates, the person’s world narrows.

While the specific trigger may differ, the symptoms of different phobias are remarkably similar. These include feelings of terror and impending doom, rapid heartbeat (tachycardia) and rapid breathing, sweaty palms, and other features of a panic attack. Individuals may experience severe anxiety symptoms in anticipating a phobic trigger. For example, someone who is afraid to fly may begin having episodes of pounding heart and sweating palms at the mere thought of getting on a plane in two weeks.

Diagnosis

A mental health professional can diagnose phobias after a detailed interview and discussion of both mental and physical symptoms. Social phobia is often associated with other anxiety disorders, depression, or **substance abuse**.

Treatment

People who have a specific phobia that is easy to avoid (e.g. fear of snakes) and that does not interfere much with their lives may not need to get help. When phobias do interfere with a person’s daily life, a

KEY TERMS

Agoraphobia—An intense fear of being trapped in a crowded, open, or public space where it may be hard to escape, combined with the dread of having a panic attack.

Benzodiazepine—A class of drugs that have a hypnotic and sedative action, used mainly as tranquilizers to control symptoms of anxiety.

Beta blockers—A group of drugs that are usually prescribed to treat heart conditions, but that also are used to reduce the physical symptoms of anxiety and phobias, such as sweating and palpitations.

MAO-B inhibitors—Inhibitors of the enzyme monoamine oxidase B. MAO-B helps break down dopamine; inhibiting it prolongs the action of dopamine

in the brain. MAOs can be used to treat social phobia.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, raising the levels of serotonin. SSRIs include Prozac, Zoloft, and Paxil.

Serotonin—5-Hydroxytryptamine; a substance that occurs throughout the body with numerous effects including neurotransmission in the brain. Inadequate amounts of serotonin are implicated in some forms of depression, obsessive-compulsive disorder, and anxiety disorders.

Social phobia—Fear of being judged or ridiculed by others; fear of being embarrassed in public.

combination of **psychotherapy** and medication can be quite effective in reducing the phobia to manageable levels.

Psychotherapy

Cognitive-behavioral therapy adds a cognitive approach to more traditional behavioral therapy. It teaches individuals how to change their thoughts, behavior, and attitudes, while providing techniques to lessen anxiety, such as deep breathing, muscle relaxation, and refocusing. One cognitive-behavioral approach is desensitization (also known as exposure therapy), in which people gradually are exposed to the frightening object or event until they become used to it and their physical symptoms decrease. For example, someone who is afraid of snakes might first be shown a photo of a snake. Once the person can look at a photo without anxiety, he or she might then be shown a video of a snake. Each step is repeated until the physical symptoms of fear, such as pounding heart and sweating palms, disappear. Eventually, the person might reach the point where he or she can touch a live snake. Three-fourths of patients are significantly improved with this type of treatment.

Another more dramatic cognitive-behavioral approach is called flooding. It exposes the person immediately to the feared object or situation. The person remains in the situation until the anxiety lessens.

Drugs

Medication can block the feelings of panic and when combined with cognitive-behavioral therapy,

can be effective in reducing phobias. Drug therapy is individualized based on the age of the patient, severity of the phobia, co-existing physical and/or mental disorders, and history of drug or alcohol addition.

Several drugs are used to treat specific phobias and social phobia by controlling symptoms and helping to prevent panic attacks. Treating agoraphobia is more difficult than treating other phobias because there are often so many fears involved, such as open spaces, traffic, elevators, and escalators.

Drugs often used to treat phobias include anti-anxiety drugs such as buspirone (BuSpar) and **benzodiazepines** such as alprazolam (Xanax), lorazepam (Ativan), clonazepam (Klonopin), and diazepam (Valium). Antihypertensive beta-blockers (drugs that lower blood pressure), such as propranolol (Inderal), atenolol (Tenormin), and nadolol (Corgard), appear to work well in the treatment of circumscribed social phobia when anxiety gets in the way of performance, such as public speaking. These drugs reduce overstimulation, thereby controlling the physical symptoms of anxiety.

In addition, some antidepressants may be effective when used together with cognitive-behavioral therapy. These include venlafaxine (Effexor), imipramine (Tofranil), desipramine (Norpramin), nortriptyline (Pamelor), duloxetine (Cymbalta), clomipramine (Anafranil), citalopram (Celexa), escitalopram (Lexapro), fluoxetine (Prozac), paroxetine (Paxil), sertraline (Zoloft) and fluvoxamine (Luvox). The monoamine oxidase inhibitor (MAO inhibitor) phenelzine (Nardil) may be used when treatment with antidepressants fails or is not tolerated.

Home remedies

In all types of phobias, symptoms may be eased by lifestyle changes, such as:

- eliminating caffeine
- reducing or eliminating alcohol use
- eating a healthy diet
- getting plenty of exercise
- reducing stress

Prognosis

Phobias are among the most treatable mental health problems. Depending on the severity of the condition and the type of phobia, most properly treated individuals can go on to lead normal lives. Research suggests that once a person overcomes the phobia, the problem may not return for many years, if at all.

Although phobias are highly treatable, only about 20% of specific phobias will go away without treatment, and agoraphobia will get worse with time if untreated. Social phobias tend to be chronic and are not likely go away without treatment. Moreover, untreated phobias can lead to other problems, including depression, **alcoholism**, and feelings of shame and low self-esteem. Unfortunately, only about 25% of people with phobias seek help to deal with their condition.

Prevention

There is no known way to prevent the development of phobias. Medication and cognitive-behavioral therapy may help prevent the recurrence of symptoms once they have been diagnosed.

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Anxiety Disorders Association of America, 8730 Georgia Ave., Suite 600, Silver Spring, MD, 20910, (240) 485-1001, (240) 485-1035, information@adaa.org, <http://www.adaa.org>.

National Anxiety Foundation, 3135 Custer Dr., Lexington, KY, 40517, 606-272-7166, <http://www.lexington-online.com/naf.html>.

National Institute of Mental Health, 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD, 20892-9663, (301) 443-4513. TTY (301) 443-8431, (866) 615-6464 TTY (866) 415-8051 , (301) 443-4279, nimhinfo@nimh.nih.gov, <http://www.nih.nih.gov>.

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Phospholipidosis see **Pulmonary alveolar proteinosis**

Phosphorus imbalance

Definition

Phosphorus imbalance refers to conditions in which the element phosphorus is present in the body at too high a level (hyperphosphatemia) or too low a level (hypophosphatemia).

Description

Almost all of the phosphorus in the body occurs as phosphate (phosphorus combined with four oxygen atoms), and most of the body's phosphate (85%) is located in the skeletal system, where it combines with **calcium** to give bones their hardness. The remaining amount (15%) exists in the cells of the body, where it plays an important role in the formation of key nucleic acids, such as DNA, and in the process by which the body turns food into energy (metabolism). The body regulates phosphate levels in the blood through the controlled release of parathyroid hormone (PTH) from the parathyroid gland and calcitonin from the thyroid gland. PTH keeps phosphate levels from becoming too high by stimulating the excretion of phosphate in urine and causing the release of calcium from bones (phosphate blood levels are inversely proportional to calcium blood levels). Calcitonin keeps phosphate blood levels in check by moving phosphates

out of the blood and into the bone matrix to form a mineral salt with calcium.

Most phosphorus imbalances develop gradually and are the result of other conditions or disorders, such as **malnutrition**, poor kidney function, or a malfunctioning gland.

Causes and symptoms

Hypophosphatemia

Hypophosphatemia (low blood phosphate) has various causes. **Hyperparathyroidism**, a condition in which the parathyroid gland produces too much PTH, is one primary cause. Poor kidney function, in which the renal tubules do not adequately reabsorb phosphorus, can result in hypophosphatemia, as can overuse of **diuretics**, such as theophylline, and **antacids** containing aluminum hydroxide. Problems involving the intestinal absorption of phosphate, such as chronic **diarrhea** or a deficiency of vitamin D (needed by the intestines to properly absorb phosphates) can cause the condition. Malnutrition due to chronic **alcoholism** can result in an inadequate intake of phosphorus. Recovery from conditions such as **diabetic ketoacidosis** or severe **burns** can provoke hypophosphatemia, since the body must use larger-than-normal amounts of phosphate. Respiratory alkylosis, brought on by hyperventilation, can also result in temporary hypophosphatemia.

Symptoms generally occur only when phosphate levels have decreased profoundly. They include muscle weakness, **tingling** sensations, **tremors**, and bone weakness. Hypophosphatemia may also result in confusion and **memory loss**, seizures, and **coma**.

Hyperphosphatemia

Hyperphosphatemia (high blood phosphate) also has various causes. It is most often caused by a decline in the normal excretion of phosphate in urine as a result of kidney failure or impaired function. **Hypoparathyroidism**, a condition in which the parathyroid gland does not produce enough PTH, or pseudoparathyroidism, a condition in which the kidneys lose their ability to respond to PTH, can also contribute to decreased phosphate excretion. Hyperphosphatemia can also result from the overuse of **laxatives** or **enemas** that contain phosphate. **Hypocalcemia** (abnormally low blood calcium) can cause phosphate blood levels to increase abnormally. A side-effect of hyperphosphatemia is the formation of calcium-phosphate crystals in the blood and soft tissue.

Hyperphosphatemia is generally asymptomatic; however, it can occur in conjunction with hypocalcemia, the symptoms of which are **numbness and tingling** in the extremities, **muscle cramps** and spasms, depression, memory loss, and convulsions. When calcium-phosphate crystals build up in the blood vessels, they can cause arteriosclerosis, which can lead to heart attacks or strokes. When the crystals build up in the skin, they can cause severe **itching**.

Diagnosis

Disorders of phosphate metabolism are assessed by measuring serum or plasma levels of phosphate and calcium. Hypophosphatemia is diagnosed if the blood phosphate level is less than 2.5 milligrams per deciliter of blood. Hyperphosphatemia is diagnosed if the blood phosphate level is above 4.5 milligrams per deciliter of blood. Appropriate tests are also used to determine if the underlying cause of the imbalance, including assessments of kidney function, dietary intake, and appropriate hormone levels.

Treatment

Treatment of phosphorus imbalances focuses on correcting the underlying cause of the imbalance and restoring equilibrium. Treating the underlying condition may involve surgical removal of the parathyroid gland in the case of hypophosphatemia caused by hyperparathyroidism; initiating hormone therapy in cases of hyperphosphatemia caused by hypoparathyroidism; ceasing intake of drugs or medications that contribute to phosphorus imbalance; or instigating measures to restore proper kidney function.

Restoring phosphorus equilibrium in cases of mild hypophosphatemia may include drinking a prescribed solution that is rich in phosphorus; however, since this solution can cause diarrhea, many doctors recommend that patients drink 1 qt (0.9 L) of skim milk per day instead, since milk and other dairy products are significant sources of phosphate. Other phosphate-rich foods include green, leafy vegetables; peas and beans; nuts; chocolate; beef liver; turkey; and some cola drinks. Severe hypophosphatemia may be treated with the administration of an intravenous solution containing phosphate.

Restoring phosphorus equilibrium in cases of mild hyperphosphatemia involves restricting intake of phosphorus-rich foods and taking a calcium-based antacid that binds to the phosphate and blocks its absorption in the intestines. In cases of severe hyperphosphatemia, an intravenous infusion of calcium gluconate may be administered. Dialysis may also be

required in severe cases to help remove excess phosphate from the blood.

Prognosis

The prognosis for treating hyperphosphatemia and hypophosphatemia are excellent, though in cases where these problems are due to genetic disease, life-long hormone treatment may be necessary.

Prevention

Phosphorus imbalances caused by hormonal disorders or other genetically determined conditions cannot be prevented. Hypophosphatemia resulting from poor dietary intake can be prevented by eating foods rich in phosphates, and hypophosphatemia caused by overuse of diuretics or antacids can be prevented by strictly following instructions concerning proper dosages, as can hyperphosphatemia due to excessive use of enemas or laxative. Finally, patients on dialysis or who are being fed intravenously should be monitored closely to prevent phosphorus imbalances.

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Tom Brody, PhD

Photoallergy see **Photosensitivity**

called porfimer sodium (Photofrin) was first approved as a treatment for **esophageal cancer** in 1995. The Food and Drug Administration (FDA) then extended its approval of this drug to cover **non-small cell lung cancer** in 1998. The FDA has also approved porfimer sodium for the treatment of tumors located in the bronchi of the lungs and for palliative treatment of advanced cancers of the esophagus. Some cancer centers in the United States administer PDT with porfimer sodium for the treatment of certain types of skin cancer (squamous cell carcinoma, **basal cell carcinoma**, and Bowen's disease), recurrences of **breast cancer** following **mastectomy**, colorectal cancer, and cancers of the vulva and cervix, but these applications of PDT are still considered experimental as of early 2010.

In December 1999, the FDA approved a compound called aminolevulinic acid (ALA or Levulan Kerastick) for the treatment of actinic keratosis, a precancerous skin disorder caused by sun exposure. Verteporfin (Visudyne) was approved in 2000 as a photosensitizing agent for the treatment of eye disorders.

Porfimer sodium, ALA, and verteporfin are the only photosensitizing agents approved by the FDA for use in the United States as of 2010. Several newer drugs for PDT are being tested in cancer centers in the United States and Europe. The most important of these will be described below.

In addition to cancer therapy, PDT is used to treat such conditions as wet **macular degeneration**, an eye disorder that can lead to blindness, as well as such benign skin conditions as **psoriasis**, **acne**, and skin disorders caused by the **human papilloma virus**. In addition, PDT is under investigation as a possible treatment for certain forms of **coronary artery disease**.

Precautions

Precautions for porfimer sodium (Photofrin):

- Porfimer sodium cannot be used in patients who are allergic to hematoporphyrin, a blood pigment used to make the drug.
- It cannot be used in pregnant or nursing women because its safety during pregnancy or lactation has not been established.
- It cannot be used to treat children.
- Lung tumors treated with Photofrin must be located in an airway where the doctor can reach them with a bronchoscope.
- Photofrin cannot be used to treat tumors in the esophagus or bronchi that are beginning to break into the patient's windpipe or a major blood vessel.

Photodynamic therapy

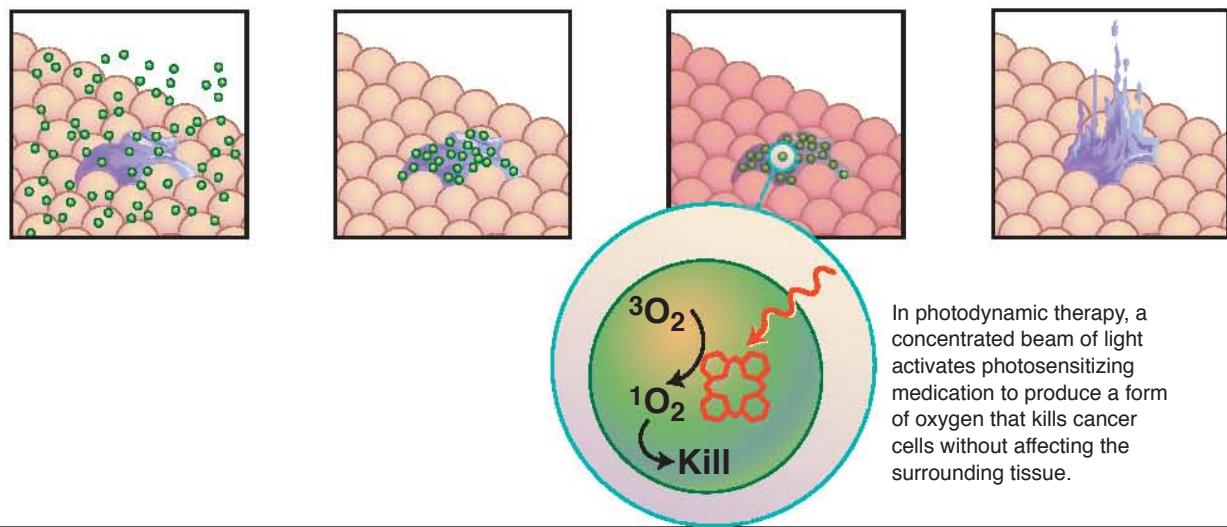
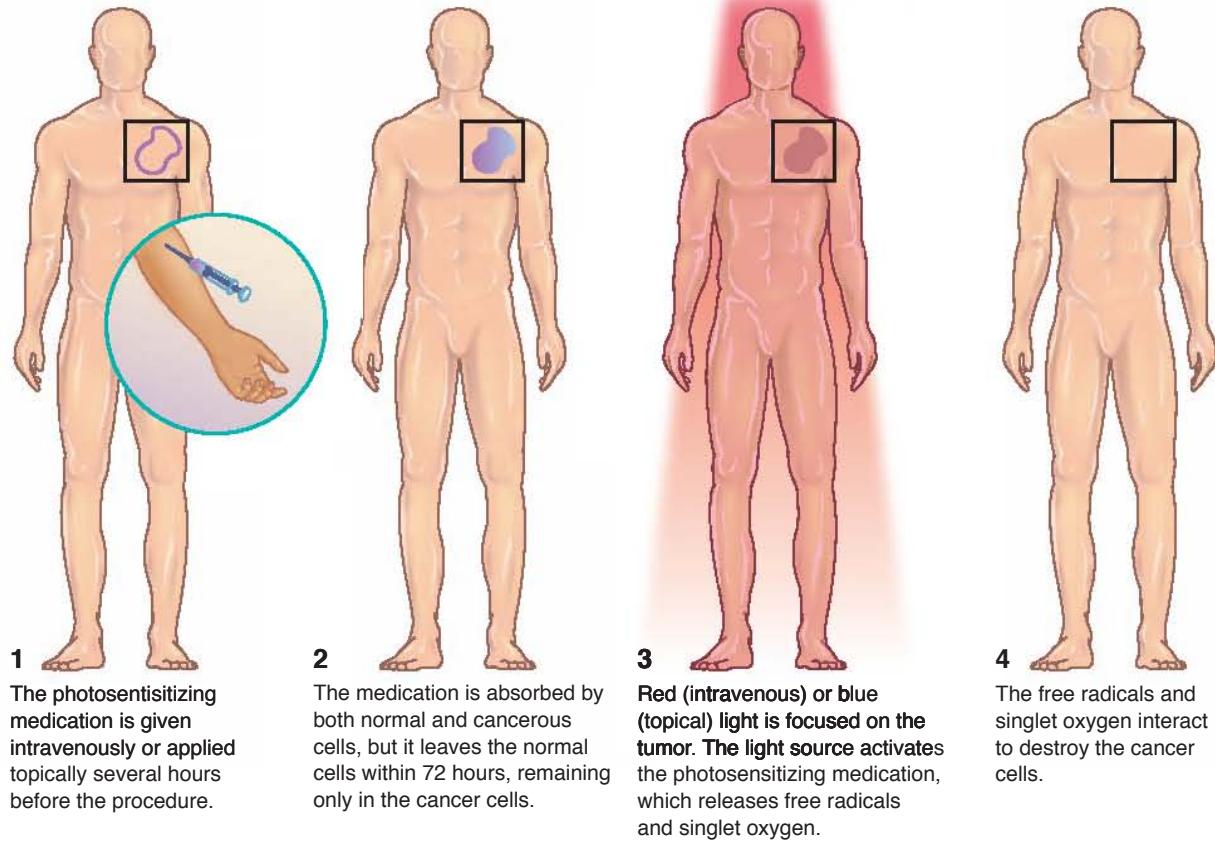
Definition

Photodynamic therapy (PDT) is a form of non-surgical **cancer** treatment available since the early 1990s that combines a photosensitizing medication with exposure to a laser or other specific light wavelength to kill cancer cells. It can be used before or after surgery and other forms of cancer treatment. In some cases, PDT can even be administered during surgery to kill any cancer cells that were not removed by excision.

Purpose

Photodynamic therapy is still evolving, both in terms of the types of cancer it is approved to treat and the specific drugs that are used. PDT with a drug

How photodynamic therapy works



(Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

KEY TERMS

Actinic keratosis (plural, keratoses)—A type of pre-cancerous skin growth with a scaly or bumpy surface caused by overexposure to the sun.

Barrett's esophagus—A precancerous condition of the esophagus that may develop as a complication of gastroesophageal reflux disease (GERD).

Bronchi (singular, bronchus)—The larger air passages inside the lungs.

Fiberoptics—Bundles of specially treated glass or plastic fibers that intensify light from a light source by internal reflection. Fiberoptics can be attached to lasers for use in PDT.

Free radicals—Molecules that contain at least one unpaired electron. They are highly reactive and can destroy cells by disrupting their normal biological processes. Free radicals are released during PDT and help to kill tumor cells.

Hematoporphyrin—A dark reddish-purple pigment found in blood. A purified form of hematoporphyrin is used to make porfimer sodium.

Nanometer—A measurement of length equal to 10^{-9} meters, or one billionth of a meter. It is used as a unit of measurement for light waves.

Orphan drug—A drug that treats a rare disease—“rare disease” being defined by the Food and Drug Association as one affecting fewer than 200,000 Americans. The category of orphan drug includes experimental as well as approved medications. Some photosensitizing drugs used in Europe are considered orphan drugs in the United States.

Palliative—Referring to treatment used to relieve the symptoms of a disease or disorder rather than to cure it.

Photosensitizer—A chemical compound that can be excited (activated) by light of a specific wavelength.

Singlet oxygen—A highly reactive form of the oxygen molecule (O_2) formed during PDT that helps to destroy cancer cells by attacking their cell membranes.

The drug should also be used cautiously in treating bronchial tumors that could block the airway if they develop inflammation following PDT.

- Patients who are receiving radiotherapy should not have PDT with porfimer sodium until four weeks after their last radiation treatment. They should also not be treated with radiotherapy until two to four weeks after a PDT treatment.

Precautions for aminolevulinic acid (ALA):

- Patients being treated with ALA must protect their skin from exposure to sunlight or bright indoor light in the short time period between application of the drug to the skin and the PDT treatment.
- ALA should be used cautiously in pregnant women or nursing mothers.
- If a second treatment is necessary, it should not be done before eight weeks after the first treatment.

Description

How PDT works

Photodynamic therapy is based on a series of chemical reactions involving a specific wavelength of visible light, a photosensitizing drug, and oxygen. There is no standard wavelength of light, light source, exposure period, or method of administering the

medication that covers all forms of PDT. Most photosensitizing drugs are given intravenously, but some are applied to the skin or taken by mouth. Photosensitizers given by injection are activated by light in the red portion of the visible light spectrum, around 630–700 nanometers (nm; a nanometer is a measure of length, one billionth of a meter), while those applied to the skin are usually activated by blue light.

In general, cancerous tumors inside the body need more concentrated doses of light than abnormal growths on the body surface. Lasers are usually used to deliver highly concentrated light at one specific wavelength, while light sources that provide a larger area of illumination, such as light-emitting diodes (LEDs), are more efficient for treating skin tumors.

In contrast to their uses in surgery, lasers are not used in PDT to remove tissue or seal blood vessels with heat; rather they are used to start a chemical reaction. As a result, they do not become hot enough to burn tissue. The burning or stinging sensation that some patients experience during PDT is caused by the release of oxygen stimulating nearby nerve endings rather than heat from the laser itself.

Lasers can be attached to fiberoptics for treating tumors inside the body. Fiberoptics are thin strands of plastic or glass with special optical properties that can

be threaded through a bronchoscope or endoscope, which are special tubes that allow the doctor to see into the patient's lungs or esophagus. Light from the laser is then transmitted along the special fibers to the tumor, thus allowing the doctor to activate the photosensitizing medication in a very small area of tissue without damaging normal tissue nearby.

PDT is a two-step form of therapy. First, the photosensitizing medication is injected into a vein or applied to the skin several days or hours before the scheduled treatment. The drug is absorbed by all body tissues but remains in cancer cells longer than in normal cells because the cancer cells are multiplying faster. After the medication has had time to collect in the malignant cells, the doctor directs a light source of the proper wavelength on the targeted area. When the light source strikes tissue containing the photosensitizing medication in the presence of oxygen, the medication is activated and produces free radicals and a highly reactive form of oxygen called singlet oxygen. The free radicals and singlet oxygen interact with the cell membranes of the cancer cells to destroy the energy-producing structures inside the cancer cells. In addition to killing the cancer cells directly, PDT works by closing blood vessels inside the tumor, thereby shutting off its supply of nutrients, and by stimulating the immune system to produce interleukins (nonantibody proteins) and other substances that attack the cancer.

Photosensitizing drugs

PORFIMER SODIUM. Porfimer sodium, or Photofrin, was the first medication used for PDT. It is a purified derivative of hematoporphyrin, a dark reddish-purple pigment found in blood. Photofrin is activated by red light at a wavelength of 630 nm; one disadvantage of this short wavelength is that it cannot penetrate tissue deeper than about a third of an inch, thus making Photofrin unsuitable for treating large solid tumors or tumors that lie deep beneath the surface. The light used to activate Photofrin is usually generated by a laser.

Porfimer sodium has several other disadvantages for PDT: It is a complex chemical mixture that tends to break down over time; it has limited ability to penetrate tissue; and it takes four to six weeks to be cleared from the skin, thus leaving patients susceptible to a **photosensitivity** reaction for a long period of time after their PDT treatment. A photosensitivity reaction occurs when sensitized skin is exposed to sunlight or other bright light and is characterized by redness, swelling, and blistering of the exposed skin. As a result of Photofrin's disadvantages, researchers have been

studying other photosensitizers with the following characteristics:

- They are single compounds rather than mixtures of chemicals.
- They are more effective in absorbing the red region of the visible light spectrum.
- They are more selective in targeting malignant tissue.
- They are more efficient in generating singlet oxygen.

AMINOLEVULINIC ACID. Aminolevulinic acid, or ALA, is a short-lived photosensitizer that is applied to the skin as a 5–20% oil-in-water mixture. It is activated by either a special blue light illuminator or by light at 630–635 nm.

SECOND-GENERATION PHOTOSENSITIZERS. Newer photosensitizing agents include:

- HPPH (2-[1-hexyloethyl]-2-devinyl-pyropheophorbide-a). HPPH, also called Photochlor, is a photosensitizer that is activated by light more efficiently than Photofrin. In addition, patients treated with HPPH do not have the long-term photosensitivity reactions associated with Photofrin. HPPH has been used experimentally since 2003 at the Roswell Park Cancer Institute in Buffalo, New York, to treat esophageal cancer, Barrett's esophagus, basal cell carcinoma, and recurrent breast cancer following mastectomy. It is also undergoing clinical trials in schools of veterinary medicine as a possible treatment for cancers in cats and dogs. Like Photofrin, HPPH is given intravenously. There are 12 clinical trials of HPPH under way as of 2010.
- Verteporfin (also known as BPD-MA [benzoporphyrin derivative monoacid ring A]; brand name Visudyne). Verteporfin is a second-generation photosensitizer used primarily to treat eye disorders, including age-related macular degeneration, other abnormal formations of blood vessels within the eye, and histoplasmosis (an eye infection caused by a fungus). It was approved for these uses by the FDA in 2000. Verteporfin is also being investigated as a possible treatment for skin cancer and psoriasis.
- Temoporfin (Meta-tetra hydroxyphenyl chlorin; brand name Foscan). Temoporfin is a chlorin-type photosensitizer developed in the United Kingdom. It was approved by the European Union in 2001 for the treatment of head and neck cancers and certain types of lung cancer, but is categorized as an orphan drug in the United States. The FDA lists temoporfin as an orphan drug for the palliative treatment of inoperable head and neck cancers. There are two clinical trials of temoporfin under way as of 2010.

- Motexafin lutetium (brand name Lu-Tex). Lu-Tex is an injectable dye that has been used in clinical trials to treat malignant melanoma. It has a high degree of selectivity for cancer cells. It also shows promise as a treatment for recurrent breast cancer and atherosclerosis. There were three clinical trials of Lu-Tex in the treatment of cervical and prostate cancer being conducted as of 2010.

Clinical trials

As of 2010, the National Cancer Institute (NCI) was conducting 213 different clinical studies of PDT as a treatment for macular degeneration and for cancers of the brain, skin, prostate, cervix, liver, gallbladder, urinary bladder, and the abdominal cavity. Other researchers are investigating photosensitizers that are stronger than Photofrin or better able to penetrate large solid tumors. Still another area of research is improving the effectiveness of PDT in treating metastatic cancers. At present, photodynamic therapy is usually used only to treat primary cancers.

Preparation

PDT for skin conditions

A patient receiving PDT for skin cancer or a precancerous skin disorder will have ALA applied to the affected area three to six hours before the scheduled treatment. The skin may or may not be covered with a dressing. The patient does not need to fast or make any other special preparations. If the affected area of skin is on the face, the patient may be given goggles to wear to protect the eyes from the blue light used to activate the drug.

PDT for internal cancers

The photosensitizing agents used for PDT or palliative treatment of esophageal or lung cancers are given by injection, usually two to three days before treatment. The patient may return home after the injection, but must avoid sunlight and bright light indoors before the light treatment. The patient does not need to fast or discontinue other medications, but should cover the windows and skylights in his or her home before receiving the light treatment to prevent exposure to bright light after returning home.

Patients undergoing PDT for esophageal or lung cancers are given a local or general anesthetic before the doctor inserts the bronchoscope or endoscope. They may also be given a mild tranquilizer to relieve anxiety.

Aftercare

Aftercare following PDT with porfimer sodium involves four to six weeks of protection from sunlight and other sources of bright light, including **tanning** lamps or the examination lamps found in doctors' and dentists' offices. During this period, the patient should wear dark glasses; long-sleeved shirts of light-color, and tightly woven fabric; long pants or slacks; and a wide-brimmed hat to protect the skin and eyes outdoors for at least 30 days after treatment. Sunscreen creams and lotions do not provide enough protection. It is best to run necessary errands after sundown or ask someone else in the household to drive the car. Women should not use helmet-type hair dryers or hand-held dryers on a high setting, as the drug remains in the scalp for several weeks and may cause **burns** if exposed to high heat. Exposure to low levels of indoor light is necessary, however, in order to break down the Photofrin remaining in the skin. After 30 days, the doctor will give the patient instructions on testing the skin for any remaining sensitivity to light.

Patients who have received PDT for cancers in the lining of the bronchi must return two days after the treatment for a follow-up **bronchoscopy**, in which the doctor will remove dead tumor cells and other pieces of tissue from the treated area. This follow-up procedure is necessary to prevent inflammation and possible blockage of the patient's airway. Treated tumor sites require between four and eight weeks for complete healing.

Patients who receive PDT with ALA do not need to take special precautions regarding sun exposure after treatment because the drug is short-lived. The treated skin will usually form a crust or scale for several days before healing completely.

Risks

Porfimer sodium

Risks of PDT with porfimer sodium include photosensitivity reactions if the patient fails to observe the guidelines for aftercare; chest **pain** or a burning sensation in the chest or throat; difficulty swallowing; **itching**; the formation of ulcers or scar tissue; and discomfort in the eyes when exposed to sunlight, bright lights, or car headlights. Breast cancer and lung cancer patients who have severe chest pain after PDT can be given medications to control the pain.

Aminolevulinic acid

Some patients experience a stinging or burning sensation in the skin during the blue light treatment,

but this usually goes away as soon as the light is turned off. Some patients also report temporary swelling or redness of the skin in the treated areas, or minor changes in the pigmentation of their skin.

Normal results

Normal results of PDT of the esophagus or the lining of the bronchi are shrinkage of the tumor and destruction of cancer cells. Normal results of palliative treatment for cancer of the esophagus are sufficient shrinkage of the tumor to allow the patient to swallow again.

Normal results for PDT of the skin include shrinkage and destruction of the tumor, although large skin tumors may require a second treatment for complete removal.

Abnormal results

Abnormal results include allergic reactions to the photosensitizing medication or failure of the tumor to respond to PDT.

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Rebecca Frey, Ph.D.
 Brenda W. Lerner

Photokeratitis see **Keratitis**

Photorefractive keratectomy and laser-assisted in-situ keratomileusis

Definition

Photorefractive keratectomy (PRK) and laser-assisted in-situ keratomileusis (LASIK) are two similar surgical techniques that use an excimer laser to correct nearsightedness (**myopia**) by reshaping the cornea. The cornea is the clear outer structure of the eye that lies in front of the colored part of the eye (iris). PRK and LASIK are two forms of vision-correcting (refractive) surgery. The two techniques differ in how the surface layer of the cornea is treated. As of mid 1998, two excimer lasers (Summit and Visx) had been approved for laser vision correction (refractive surgery using a laser) in the PRK procedure. Since then, Visx, Summit, and other lasers have received approval by the Food and Drug Administration (FDA) for use in LASIK procedures.

Purpose

The purpose of both LASIK and PRK is to correct nearsightedness in persons who don't want to, or can't, wear **eye glasses** or **contact lenses**. Most patients are able to see well enough to pass a driver's license exam without glasses or contact lenses after the operation. After approximately age 40, the lens in the

eye stiffens making it harder to focus up close. Because laser vision correction only affects the cornea, the procedures do not eliminate the need for reading glasses. Patients should be wary of any ads that “guarantee” 20/20 vision. Patients should also make sure that the laser being used is approved by the FDA.

Precautions

Patients should be over 18 years of age, have healthy corneas, and have vision that has been stable for the past year. People who may not be good candidates for these procedures are pregnant women or women who are **breastfeeding** (vision may not be stable); people with scarred corneas or macular disease; people with autoimmune diseases (i.e., **systemic lupus erythematosus** or **rheumatoid arthritis**); or people with diabetes. Patients with glaucoma should not have LASIK because the intraocular pressure (IOP) of the eye is raised during the procedure. A patient with persistent lid infections (i.e., blepharitis) may not be a good candidate because of an increased risk of infection. An ophthalmologist who specializes in laser vision correction can determine who would be likely to benefit from the operation and suggest which of the two operations might be more appropriate for any given patient.

If a patient is thinking of having **cataract surgery**, they should discuss it with the doctor. During cataract surgery an intracocular lens (IOL) will be inserted and that alone may correct distance vision.

Description

PRK and LASIK are both performed with an excimer laser, which uses a cold beam of ultraviolet light to sculpt or reshape the cornea so that light will focus properly on the retina. The cornea is the major focusing structure of the eye. The retina sends the image focused on it to the brain. In myopia, the cornea is either too steep or the eye is too long for a clear image to be focused on the retina. PRK and LASIK flatten out the cornea so that the image will focus more precisely on the retina.

In PRK, the surface of the cornea is removed by the laser. In LASIK, the outer layer of the cornea is sliced, lifted, moved aside while the cornea is reshaped with the laser, then replaced to speed healing. Both procedures cause the cornea to become flatter, which corrects the nearsighted vision.

At least one laser has been approved to treat mild **astigmatism**. Correcting farsightedness (**hyperopia**) may be possible in the future.

These laser vision-correcting procedures are rapidly replacing **radial keratotomy** (RK), an earlier form of refractive surgery that involved cutting the cornea with a scalpel in a pattern of radiating spokes. RK has declined in popularity since the approval of the excimer laser in 1995, falling from a high of 250,000 procedures performed per year in 1994 to 50,000 in 1997.

For both LASIK and PRK, the patient’s eye is numbed with anesthetic drops. No injections are necessary. The patient is awake and relaxed during the procedure.

LASIK is sometimes referred to as a “flap and zap” procedure because a thin flap of tissue is temporarily removed from the surface of the cornea and the underlying cornea is then “zapped” with a laser. Prior to the surgery, the surface of the cornea is marked with a dye marker so that the flap of cornea can be precisely aligned when it is replaced. The doctor places a suction ring on the eye to hold it steady. During this part of the operation, which lasts only a few seconds, the patient is not able to see. A surgical instrument called a microkeratome is passed over the cornea to create a very thin flap of tissue. The IOP is increased at this time which is why it is contraindicated in patients with glaucoma. This thin tissue layer is folded back. The cornea is reshaped with the laser beam and the cell layer is replaced. Because the cell layer is not permanently removed, patients have a faster recovery time and experience far less discomfort than with PRK. An antibiotic drop is put in and the eye is patched until the following day’s checkup.

In PRK, a small area of the surface layer of the cornea is vaporized. It takes about three days for the surface cells to grow back and vision will be blurred. Some patients describe it as “looking through Vaseline.” PRK is generally recommended for patient’s with mild to moderate myopia (usually under -5.00 diopters).

With both PRK and LASIK, there is a loud tapping sound from the laser and a burning smell as the cornea is reshaped. The surgery itself is painless and takes only a minute or two. Patients are usually able to return home immediately after surgery. Most patients wait (up to six months) before they have the second one done. This allows the first eye to heal and to see if there were complications from the surgery.

The cost of these procedures can vary with geographic area and the doctor. In general, the procedure costs \$1,350–\$2,500 per eye for PRK and about \$500 more per eye for LASIK. PRK and LASIK are generally not covered by insurance. However, insurance

KEY TERMS

Blepharitis—An inflammation of the eyelid.

Cataract—A condition in which the lens of the eye turns cloudy and interferes with vision.

Cornea—The clear, curved tissue layer in front of the eye. It lies in front of the colored part of the eye (iris) and the black hole in the center of the iris (pupil).

Diopter (D)—A unit of measure of the power or strength of a lens.

Excimer laser—An instrument that is used to vaporize tissue with a cold, coherent beam of light with a single wavelength in the ultraviolet range.

Intraocular lens (IOL) implant—A small, plastic device (IOL) that is usually implanted in the lens capsule of the eye to correct vision after the lens of the eye is removed. This is the implant used in cataract surgery.

Macular degeneration—A condition usually associated with age in which the area of the retina called the

macula is impaired due to hardening of the arteries (arteriosclerosis). This condition interferes with vision.

Microkeratome—A precision surgical instrument that can slice an extremely thin layer of tissue from the surface of the cornea.

Myopia—A vision problem in which distant objects appear blurry. Myopia results when the cornea is too steep or the eye is too long and the light doesn't focus properly on the retina. People who are myopic or nearsighted can usually see near objects clearly, but not far objects.

Refractive surgery—A surgical procedure that corrects visual defects.

Retina—The sensory tissue in the back of the eye that is responsible for collecting visual images and sending them to the brain.

may cover these procedures for people in certain occupations, such as police officers and firefighters.

Preparation

If a patient wears contact lenses, they should not be worn for a few weeks prior to surgery. It also is important to discontinue contact lens wear prior to the visual exams to make sure vision is stable. The doctor should be advised of contact lens wear.

Upon arrival at the doctor's office on the day of surgery, patients are given some eye drops and a sedative, such as Valium, to relax them. Their vision is tested. They rest while waiting for the sedative to take effect. Immediately before the surgery, patients are given local anesthetic eye drops.

Aftercare

After surgery, antibiotic drops are placed in the eye and the eye may be patched. The patient returns for a follow-up visit the next day. The patient is usually given a prescription for eyedrops (usually antibiotic and anti-inflammatory). Patients who have had PRK usually feel mild discomfort for one to three days after the procedure. They may need a bandage contact lens. Patients who have had LASIK generally have less, or even no discomfort after the surgery. After LASIK, antibiotic and anti-inflammatory drops are generally necessary for one week. After PRK, steroid eye

drops may be necessary for months. Because **steroids** may increase the possibility of glaucoma or **cataracts**, it is a big drawback to the procedure. The patient should speak with the doctor to see how long follow-up medications will be necessary.

Most patients return to work within one to three days after the procedure, although visual recovery from PRK may take as long as four weeks. An eye shield may be used for about one week at night and patients may be sensitive to bright light for a few days. Patients may be asked by their doctor to keep water out of their eye for a week and to avoid mascara or eyeliner during this period.

Risks

There is a risk of under- or over-correction with either of these procedures. If vision is under-corrected, a second procedure can be performed to achieve results that may be closer to 20/20 vision. About 5–10% of PRK patients return for an adjustment, as do 10–25% of LASIK patients. People with higher degrees of myopia have vision that is harder to correct and usually have LASIK surgery rather than PRK. This may account for the higher incidence of adjustments for LASIK patients. Patients with very high myopia (over -15.00 diopters) may experience improvement after LASIK, but they are not likely to achieve 20/40 vision without glasses. However, their glasses will not need to be as thick or heavy after the surgery. However, most

patients, especially those with less extreme myopia, do not need glasses after the surgery.

Haze is another possible side effect. Although hazy vision is unlikely, it is more likely to occur after PRK than after LASIK. This haze usually clears up. Corneal scarring, halos, or glare at night, or an irritating bump on the cornea are other possible side effects. As with any eye surgery, infection is possible, but rare. Loss of vision is possible with these procedures, but this complication is extremely rare.

Most complications from LASIK are related to the creation and realignment of the flap. The microkeratome must be in good-working order and sharp. LASIK requires a great deal of skill on the part of the surgeon and the complication rate is related to the experience level of the surgeon. In one study, the rate of LASIK complications declined from 3% for surgeons during their first three months using this technique, to 1% after a year's experience in the technique, to 0% after 18 months experience.

Normal results

Most patients experience improvement in their vision immediately after the operation and about half of LASIK patients are able to see 20/30 within one day of the surgery. Vision tends to become sharper over the next few days and then stabilizes; however, it is possible to have shifts in myopia for the next few months. Vision clears and stabilizes faster after LASIK than after PRK. Final vision is achieved within three to six months with LASIK and six to eight months with PRK. The vast majority of patients (95% for people with low to moderate myopia and 75% for people with high levels of myopia) are able to see 20/40 after either of these procedures and are able to pass a driver's license test without glasses or contact lenses.

LASIK is more complicated than PRK because of the addition of the microkeratome procedure. However, LASIK generally has faster recovery time, less pain, and less chance of halos and scarring than PRK. LASIK can treat higher degrees of myopia (-5.00–25.00 diopters). LASIK also requires less use of steroids. Patients need to speak with qualified, experienced eye surgeons to help in choosing the procedure that is right for them.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, (415) 561-8500, <http://www.aoa.org>.

American Society of Cataract and Refractive Surgery, 4000 Legato Road, Suite 700, Fairfax, VA, 22033, (703) 591-2220, (703) 591-0614, <http://www.ascrs.org>.

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Photosensitivity

Definition

Photosensitivity refers to any increase in the reactivity of the skin to sunlight.

Description

The skin is a carefully designed interface between our bodies and the outside world. It is infection-proof when intact, nearly waterproof, and filled with protective mechanisms. Sunlight threatens the health of the skin. Normal skin is highly variable in its ability to resist sun damage. Natural skin pigmentation is its main protection. The term photosensitivity refers to any increase beyond what is considered normal variation.

Causes and symptoms

There are over three dozen diseases, two dozen drugs, a variety of herbal preparations, and several perfume and cosmetic components that can cause photosensitivity. There are also several different types of reaction to sunlight—phototoxicity, photoallergy, and polymorphous light eruption. In addition, prolonged exposure to sunlight, even in normal skin,



A skin rash on the front of a woman's neck caused by a photosensitive reaction to sunlight. (Dr. P. Marazzi/Photo Researchers, Inc.)



This person had a phototoxic reaction after taking an antibiotic drug. (Photo Researchers, Inc.)

leads to skin aging and **cancer**. These effects are accelerated in patients who have photosensitivity.

- Phototoxicity is a severely exaggerated reaction to sunlight caused by a new chemical in the skin. The primary symptom is sunburn, which is rapid and can be severe enough to blister (a second degree burn). The chemicals associated with phototoxicity are usually drugs. The list includes several common antibiotics—quinolones, sulfonamides, and tetracyclines; diuretics (water pills); major tranquilizers; oral diabetes medication; and cancer medicines. There are also some dermatologic drugs, both topical and oral, that can sensitize skin.
- Photoallergy produces an intense itching rash on exposure to sunlight. Patients develop chronic skin changes (lichen simplex) as a result of scratching. Some of the agents that cause phototoxicity can also cause photoallergy. Some cosmetic and perfume ingredients, including a compound that was formerly used in sunscreens—para-amino benzoic acid (PABA)—can do this. Most sunscreen preparations in the early 2000s, however, no longer include PABA.
- Polymorphous light eruption (PLE) resembles photoallergy in its production of intensely itching rashes in sunlight. However, this condition lessens with continued light exposure, and so is seen mostly in the spring. Also, there does not seem to be an identifiable chemical involved. PLE is most likely to develop in fair-skinned individuals. It is estimated to affect about 10% of the United States population compared to 21% of the Swedish population. The female: male ratio is 2.5: 1, but it is thought that the imbalance may be due to the fact that women are more likely than men to seek treatment for PLE.
- There is a form of inherited PLE that affects Native Americans. The inheritance pattern is autosomal dominant.

KEY TERMS

Albino—A person or animal lacking normal coloring in the eyes, hair, and skin due to a hereditary inability to produce the skin pigment melanin. The condition itself is called albinism.

Biopsy—Surgical removal of tissue for examination.

Rosacea—A chronic skin disease characterized by persistent redness of the skin and periodic outbreaks of pustules, usually affecting the middle third of the face.

Diseases of several kinds increase skin sensitivity:

- A hereditary disease called xeroderma pigmentosum includes a defect in repair mechanisms that greatly accelerates skin damage from sunlight.
- A family of metabolic diseases called porphyrias produce chemicals (porphyrins) that absorb sunlight in the skin and thereby cause damage.
- Albinos lack skin pigment through a genetic defect and are thus very sensitive to light.
- Malnutrition, specifically a deficiency of niacin known as pellagra, sensitizes the skin.
- Several diseases like acne, systemic lupus erythematosus, rosacea, and herpes simplex (fever blisters) decrease the resistance of the skin to sun damage. Rosacea is sometimes described as a photoaggravated skin disorder because its symptoms increase in severity when patients are exposed to sunlight.
- Photosensitivity is increasingly recognized as a common development in HIV-positive patients. Risk factors for photosensitivity in this group include African American ethnicity and treatment with highly active antiretroviral therapy (HAART).

Diagnosis

The pattern of appearance on the skin, a history of drug or chemical exposure, and the timing of the symptoms often suggests a diagnosis. A **skin biopsy** may be needed for further clarification.

Treatment

Removal of the offending drug or chemical is primary. Direct sunlight exposure should be limited. Some people must avoid sunlight altogether, while others can tolerate some direct sunlight with the aid of **sunscreens**.

Prevention

A sunscreen with an SPF of 15 or greater protects most skin from damage. Such protective garments as hats and long-sleeved shirts are highly recommended in addition.

Resources

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United States Food and Drug Administration (FDA), 10903 New Hampshire Ave, Silver Spring, MD, 02993-0002, (888) 463-6332, <http://www.fda.gov>.

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Phototherapy

Definition

Phototherapy, or **light therapy**, is the administration of doses of bright light in order to normalize the body's internal clock and/or relieve depression.

Purpose

Phototherapy is prescribed primarily to treat **seasonal affective disorder** (SAD), a mood disorder characterized by depression in the winter months, and is occasionally employed to treat **insomnia** and **jet lag**.

The exact mechanisms by which the treatment works are not known, but the bright light employed in phototherapy may act to readjust the body's circadian (daily) rhythms, or internal clock. Other popular theories are that light triggers the production of serotonin, a neurotransmitter believed to be related to **depressive disorders**, or that it influences the body's production of melatonin, a hormone derived from serotonin that may be related to circadian rhythms.

Precautions

Patients with eye problems should see an ophthalmologist regularly, both before and during phototherapy. Because some ultraviolet rays are emitted by the light boxes used in phototherapy, patients taking photosensitizing medications (medications making the skin more sensitive to light) and those who have sun-sensitive skin should consult with their physician before beginning treatment. Patients with medical conditions that make them sensitive to ultraviolet rays should also be seen by a physician before starting phototherapy. Patients who have a history of mood swings or **mania** should be monitored closely, since phototherapy may cause excessive mood elevation in some individuals.

Description

Phototherapy is generally administered at home. The most commonly used phototherapy equipment is a portable lighting device known as a light box. The box may be mounted upright to a wall, or slanted downwards towards a table. The patient sits in front of the box for a prescribed period of time (anywhere from 15 minutes to several hours). Some patients with SAD undergo phototherapy sessions two or three times a day, others only once. The time of day and number of times treatment is administered depend on the physical needs and lifestyle of the individual patient. If phototherapy has been prescribed for the treatment of SAD, it typically begins in the fall months as the days begin to shorten, and continues throughout the winter and possibly the early spring.

The light from a slanted light box is designed to focus on the table it sits upon, so patients may look down to read or do other sedentary activities during therapy. Patients using an upright light box must face the light source (although they need not look directly into the light). The light sources in these light boxes typically range from 2,500–10,000 lux. (In contrast, average indoor lighting is 300–500 lux; a sunny summer day is about 100,000 lux).

KEY TERMS

Circadian rhythm—The rhythmic repetition of certain phenomena in living organisms at about the same time each day.

Lux—A standard unit of measure for illumination.

Neurotransmitter—A chemical in the brain that transmits messages between neurons, or nerve cells.

Photosensitivity—An abnormally heightened reaction to light.

Seasonal affective disorder (SAD)—A mood disorder characterized by depression during the winter months. An estimated 11 million Americans experience SAD.

Phototherapy prescribed for the treatment of SAD may be covered by insurance. Individuals requiring phototherapy should check with their insurance company to see if the cost of renting or purchasing a light box is covered.

Aftercare

Patients beginning light therapy for SAD may need to adjust the length, frequency, and timing of their phototherapy sessions to achieve the maximum benefit. These patients should keep their doctor informed of their progress and the status of their depressive symptoms. Occasionally, antidepressants and/or **psychotherapy** may be recommended as an adjunct to phototherapy.

Risks

An abnormally elevated or expansive mood (hypomania) may occur, but it is usually temporary. Some patients undergoing phototherapy treatment report side effects of eyestrain, headaches, insomnia, **fatigue**, **sunburn**, and dry eyes or nose. Most of these effects can be managed by adjusting the timing and duration of the phototherapy sessions. A strong sun block and eye and nose drops can alleviate the other problems. Long-term studies have shown no negative effects to the eye function of individuals undergoing phototherapy treatments.

Normal results

Patients with SAD typically report an alleviation of depressive symptoms within two to 14 days after beginning phototherapy.

ORGANIZATIONS

National Institute of Mental Health (NIMH).
Society for Light Treatment and Biological Rhythms, P.O. Box 591687, 174 Cook St, San Francisco, CA, 4159-1687, <http://www.sltbr.org>.

Paula Anne Ford-Martin

Phototoxic reaction see **Photosensitivity**

Phycomycosis see **Mucormycosis**

Physical allergy

Definition

Physical **allergies** are allergic reactions to cold, sunlight, heat, or minor injury.

Description

The immune system is designed to protect the body from harmful invaders such as germs. Occasionally, it goes awry and attacks harmless or mildly noxious agents, doing more harm than good. This event is termed allergy if the target is from the outside—like pollen or bee venom—and autoimmunity if it is caused by one of the body's own components.

The immune system usually responds only to certain kinds of chemicals, namely proteins. However, non-proteins can trigger the same sort of response, probably by altering a protein to make it look like a target. Physical allergy refers to reactions in which a protein is not the initial inciting agent.

Sometimes it takes a combination of elements to produce an allergic reaction. A classic example is drugs that are capable of sensitizing the skin to sunlight. The result is phototoxicity, which appears as an increased sensitivity to sunlight or as localized skin **rashes** on sun-exposed areas.

Causes and symptoms

- Minor injury, such as scratching, causes itchy welts to develop in about 5% of people. The presence of itchy welts (urticaria) is a condition called dermographism.
- Cold can change certain proteins in the blood so that they induce an immune reaction. This may indicate that there are abnormal proteins in the blood from a disease of the bone marrow. The reaction may also involve the lungs and circulation, producing wheezing and fainting.

KEY TERMS

Antihistamine—Drugs that block histamine, a major cause of itching.

Hemolysis—Destruction of red blood cells.

Inflammation—Heat, redness, swelling, and pain caused by an immune response.

- Heat allergies can be caused by exercise or even strong emotions in sensitive people.
- Sunlight, even without drugs, causes immediate urticaria in some people. This may be a symptom of porphyria—a genetic metabolic defect.
- Elements like nickel and chromium, although not proteins, commonly cause skin rashes, and iodine allergy causes skin rashes and sores in the mouth in allergic individuals.
- Pressure or vibration can also cause urticaria.
- Water contact can cause aquagenic urticaria, presumably due to chlorine or some other trace chemical in the water, although distilled water has been known to cause this reaction.

When the inflammatory reaction involves deeper layers of the skin, urticaria becomes angioedema. The skin, especially the lips and eyelids, swells. The tongue, throat, and parts of the digestive tract may also be involved. Angioedema may be due to physical agents. Often the cause remains unknown.

Diagnosis

Visual examination of the symptoms usually diagnoses the reaction. Further skin tests and review of the patient's **photosensitivity** may reveal a cause.

Treatment

Removing the offending agent is the first step to treatment. If sun is involved, shade and **sunscreens** are necessary. The reaction can usually be controlled with epinephrine, **antihistamines**, or cortisone-like drugs. Urticaria may be treated with antihistamines such as diphenhydramine (Benadryl) or desloratadine (Claritin). Claritin is non-sedating, meaning it will not make patients drowsy. **Itching** can be controlled with cold packs or commercial topical agents that contain menthol, camphor, eucalyptus oil, aloe, antihistamines, or cortisone preparations.

Prognosis

If the causative agent has been diagnosed, avoidance of or protection against the allergen cures the allergy. Usually, allergies can be managed through treatment.

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Teresa G. Odle

Physical examination

Definition

A physical examination is an evaluation of the body and its functions using inspection, palpation (feeling with the hands), percussion (tapping with the fingers), and auscultation (listening). A complete health assessment also includes gathering information about a person's medical history and lifestyle, doing laboratory tests, and screening for disease.

Purpose

The annual physical examination has been replaced by the periodic health examination. How often this is done depends on the patient's age, sex, and risk factors for disease. The United States Preventative Services Task Force (USPSTF) has developed guidelines for preventative health examinations that health care professionals widely follow. Organizations that promote detection and prevention of specific diseases, like the American **Cancer** Society, generally recommend more intensive or frequent examinations.

A comprehensive physical examination provides an opportunity for the healthcare professional to obtain baseline information about the patient for future use, and to establish a relationship before problems happen. It provides an opportunity to answer questions and teach good health practices. Detecting a problem in its early stages can have good long-term results.

Precautions

The patient should be comfortable and treated with respect throughout the examination. As the

examination proceeds, the examiner should explain what he or she is doing and share any relevant findings.

Description

A complete physical examination usually starts at the head and proceeds all the way to the toes. However, the exact procedure will vary according to the needs of the patient and the preferences of the examiner. An average examination takes about 30 minutes. The cost of the examination will depend on the charge for the professional's time and any tests that are done. Most health plans cover routine physical examinations including some tests.

The examination

First, the examiner will observe the patient's appearance, general health, and behavior, along with measuring height and weight. The vital signs—including pulse, breathing rate, body temperature, and blood pressure—are recorded.

With the patient sitting up, the following systems are reviewed:

- Skin. The exposed areas of the skin are observed; the size and shape of any lesions are noted.
- Head. The hair, scalp, skull, and face are examined.
- Eyes. The external structures are observed. The internal structures can be observed using an ophthalmoscope (a lighted instrument) in a darkened room.
- Ears. The external structures are inspected. A lighted instrument called an otoscope may be used to inspect internal structures.
- Nose and sinuses. The external nose is examined. The nasal mucosa and internal structures can be observed with the use of a penlight and a nasal speculum.
- Mouth and pharynx. The lips, gums, teeth, roof of the mouth, tongue, and pharynx are inspected.
- Neck. The lymph nodes on both sides of the neck and the thyroid gland are palpated (examined by feeling with the fingers).
- Back. The spine and muscles of the back are palpated and checked for tenderness. The upper back, where the lungs are located, is palpated on the right and left sides and a stethoscope is used to listen for breath sounds.
- Breasts and armpits. A woman's breasts are inspected with the arms relaxed and then raised. In both men and women, the lymph nodes in the armpits are felt with the examiner's hands. While the patient is still sitting, movement of the joints in

the hands, arms, shoulders, neck, and jaw can be checked.

Then while the patient is lying down on the examining table, the examination includes:

- Breasts. The breasts are palpated and inspected for lumps.
- Front of chest and lungs. The area is inspected with the fingers, using palpation and percussion. A stethoscope is used to listen to the internal breath sounds.

The head should be slightly raised for:

- Heart. A stethoscope is used to listen to the heart's rate and rhythm. The blood vessels in the neck are observed and palpated.

The patient should lie flat for:

- Abdomen. Light and deep palpation is used on the abdomen to feel the outlines of internal organs including the liver, spleen, kidneys, and aorta, a large blood vessel.
- Rectum and anus. With the patient lying on the left side, the outside areas are observed. An internal digital examination (using a finger), is usually done if the patient is over 40 years old. In men, the prostate gland is also palpated.
- Reproductive organs. The external sex organs are inspected and the area is examined for hernias. In men, the scrotum is palpated. In women, a pelvic examination is done using a speculum and a Papanicolaou test (Pap test) may be taken.
- Legs. With the patient lying flat, the legs are inspected for swelling, and pulses in the knee, thigh, and foot area are found. The groin area is palpated for the presence of lymph nodes. The joints and muscles are observed.
- Musculoskeletal system. With the patient standing, the straightness of the spine and the alignment of the legs and feet is noted.
- Blood vessels. The presence of any abnormally enlarged veins (varicose), usually in the legs, is noted.

In addition to evaluating the patient's alertness and mental ability during the initial conversation, additional inspection of the nervous system may be indicated:

- Neurologic screen. The patient's ability to take a few steps, hop, and do deep knee bends is observed. The strength of the hand grip is felt. With the patient sitting down, the reflexes in the knees and feet can be tested with a small hammer. The sense of touch in the hands and feet can be evaluated by testing reaction to pain and vibration.

KEY TERMS

Auscultation—The process of listening to sounds that are produced in the body. Direct auscultation uses the ear alone, such as when listening to the grating of a moving joint. Indirect auscultation involves the use of a stethoscope to amplify the sounds from within the body, like a heartbeat.

Hernia—The bulging of an organ, or part of an organ, through the tissues normally containing it; also called a rupture.

Inspection—The visual examination of the body using the eyes and a lighted instrument if needed. The sense of smell may also be used.

Ophthalmoscope—Lighted device for studying the interior of the eyeball.

Otoscope—An instrument with a light for examining the internal ear.

Palpation—The examination of the body using the sense of touch. There are two types: light and deep.

Percussion—An assessment method in which the surface of the body is struck with the fingertips to obtain sounds that can be heard or vibrations that can be felt. It can determine the position, size, and consistency of an internal organ. It is done over the chest to determine the presence of normal air content in the lungs, and over the abdomen to evaluate air in the loops of the intestine.

Reflex—An automatic response to a stimulus.

Speculum—An instrument for enlarging the opening of any canal or cavity in order to facilitate inspection of its interior.

Stethoscope—A Y-shaped instrument that amplifies body sounds such as heartbeat, breathing, and air in the intestine. Used in auscultation.

Varicose veins—The permanent enlargement and twisting of veins, usually in the legs. They are most often seen in people with occupations requiring long periods of standing, and in pregnant women.

- Sometimes additional time is spent examining the 12 nerves in the head (cranial) that are connected directly to the brain. They control the sense of smell, strength of muscles in the head, reflexes in the eye, facial movements, gag reflex, and muscles in the jaw. General muscle tone and coordination, and the reaction of the abdominal area to stimulants like pain, temperature, and touch would also be evaluated.

Preparation

Before visiting the health care professional, the patient should write down important facts and dates about his or her own medical history, as well as those of family members. He or she should have a list of all medications with their doses or bring the actual bottles of medicine along. If there are specific concerns about anything, writing them down is a good idea.

Before the physical examination begins, the bladder should be emptied and a urine specimen can be collected in a small container. For some blood tests, the patient may be told ahead of time not to eat or drink after midnight.

The patient usually removes all clothing and puts on a loose-fitting hospital gown. An additional sheet is provided to keep the patient covered and comfortable during the examination.

Aftercare

Once the physical examination has been completed, the patient and the examiner should review what laboratory tests have been ordered and how the results will be shared with the patient. The medical professional should discuss any recommendations for treatment and follow-up visits. Special instructions should be put in writing. This is also an opportunity for the patient to ask any remaining questions about his or her own health concerns.

Risks

Other than discovering an unknown condition or health problem, which is the reason for performing a physical examination, there are no risks associated with the procedure.

Normal results

Normal results of a physical examination correspond to the healthy appearance and normal functioning of the body. For example, appropriate reflexes will be present, no suspicious lumps or lesions will be found, and vital signs will be normal.

Abnormal results

Abnormal results of a physical examination include any findings that indicated the presence of a

disorder, disease, or underlying condition. For example, the presence of lumps or lesions, **fever**, muscle weakness or lack of tone, poor reflex response, heart arrhythmia, or swelling of lymph nodes will point to a possible health problem.

Resources

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ORGANIZATIONS

- American Academy of Family Physicians, 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2672, (913) 906-6000, fp@aafp.org, <http://www.aafp.org>.
- American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007-1098, (847) 434-4000, (847) 434-8000, kidsdoc@aap.org, <http://www.aap.org>.
- American College of Physicians, 190 N. Independence Mall West, Philadelphia, PA, 19106-1572, (215) 351-2600, (800) 523-1546, <http://www.acponline.org>.
- American Medical Association, 515 N. State Street, Chicago, IL, 60610, (312) 464-5000, <http://www.ama-assn.org>.

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Physical therapy

Definition

Physical therapy, also called physiotherapy, is the prevention and treatment of medical conditions by physical and mechanical means, including **exercise**, body manipulation, water, light, heat, and electricity.

Purpose

The purpose of physical therapy is to restore function, improve mobility, relieve **pain**, and prevent or limit permanent physical disabilities, with the goal of improving a patient's functioning at school or work and in daily life. Physical therapists treat patients with a variety of conditions and diseases, including:

- all types of injuries, including sprains, strains, fractures, and head injuries
- back and neck pain
- knee pain
- shoulder pain
- repetitive stress and overuse injuries
- poor posture
- arthritis
- heart disease
- stroke
- diabetes
- osteoporosis
- lymphedema

Common applications of physical therapy include:

- retraining muscles and adjusting to the use of artificial joints

- strengthening leg muscles following a hip fracture
- assessing and fitting walking aids such as canes and walkers
- strengthening arm muscles for using walking aids
- treating pain from tendinitis/bursitis and arthritis to avoid the use of prescription pain medications in patients at risk for heart disease
- rehabilitating stroke victims for walking safely, with or without a walking aid

Pediatric physical therapy is commonly used for children with:

- birth defects such as spina bifida
- genetic disorders
- prenatal drug or alcohol exposure
- cerebral palsy
- orthopedic disabilities and limb deficiencies
- developmental delays
- heart and lung conditions
- muscle diseases
- head injuries
- acute trauma

Demographics

The use of physical therapy to treat patients of all ages—from newborns to the elderly—is becoming increasingly widespread:

- As of 2008, there were some 33.1 million non-institutionalized adult Americans with some degree of difficulty in physical functioning—15% of the non-institutionalized adult population.
- Lower back pain affects as many as 80% of all Americans at some point in their lives.
- Improper use or fitting of walking aids by senior citizens accounts for 47,000 emergency rooms visits annually in the United States.
- An estimated 17.9 million American children and adults have been diagnosed with diabetes.
- Secondary lymphedema resulting in fluid retention and arm swelling affects 71% of women who undergo surgery for breast cancer.
- As of 2008, there were about 185,000 physical therapy jobs in the United States.
- By 2018 employment of physical therapists is expected to have grown by 30%, much more than the average for all occupations.

There are several reasons for the growth of physical therapy:

- The increasingly elderly population in the United States is especially vulnerable to chronic and debilitating conditions that require physical therapy.
- The huge generation of baby boomers is reaching the age of susceptibility to heart attacks and strokes, which require physical rehabilitation.
- Medical and technological developments have led to greatly increased survival of newborns with birth defects and casualties of war and other traumas, many of whom require physical therapy, often for the remainder of their lives.
- Advances in physical therapy have led to treatments for many disabling conditions that were previously untreatable.
- The federally mandated Individuals with Disabilities Education Act guarantees student access to physical therapy.
- A growing number of employers are using physical therapists to evaluate worksites, develop exercise programs, and teach safe work habits to reduce injuries.

Description

Physical therapy is performed by physical therapists, physical therapy assistants, and physical therapy aides. Physical therapists take medical histories, perform physical exams, and assess the ability of patients to function independently. They use a variety of tests and measurements for evaluation. Range of motion is determined using a goniometer—an instrument that measures the largest angle through which a joint can move. The therapist determines whether restricted motion is due to tight muscles or tight ligaments and tendons. The therapist also evaluates:

- strength
- coordination and balance
- posture
- motor function
- muscle performance
- respiration

Based on these evaluations, physical therapists develop treatment plans, including purpose, strategy, and anticipated outcomes. During the course of physical therapy a patient's progress is tracked with periodic examinations and tests.

Physical therapists may treat a wide range of conditions or specialize in certain areas. They often consult or collaborate with physicians, nurses, dentists, occupational therapists, speech-language pathologists, audiologists, educators, and/or social workers. Working under the direct supervision of a physical

therapist, physical therapy assistants often implement treatment plans and record patient responses.

About 60% of physical therapists work in hospitals or in the offices of other healthcare practitioners, especially physicians. Physical therapy also can take place in:

- nursing homes
- outpatient clinics
- dedicated physical therapy and rehabilitation facilities
- adult daycare facilities
- private physical therapy practices
- child care centers, preschools, and schools
- sports and fitness facilities
- recreation centers
- workplaces
- the patient's home

Physical therapy exercises are aimed at improving flexibility, range of motion, muscle strength, balance, coordination, ambulation (walking), and/or endurance. Patients are often taught exercises to perform at home. Activities can include water walking and swimming. Physical therapy also teaches patients to use assistive and adaptive devices such as crutches, wheelchairs, and prostheses.

For range-of-motion stretches the muscles are often first warmed with heat to improve effectiveness and reduce pain. Tight ligaments or tendons require gentle stretching, whereas the joint can be stretched more vigorously if tight muscles are causing poor range of motion. An affected joint must be moved beyond the point of pain, but should not cause residual pain after the movement is stopped. Sustained moderate stretching may be applied with weights and pulleys. There are three types of range-of-motion exercises:

- active exercise for patients who can move their limbs and exercise a muscle or joint without assistance
- active-assistive exercise for patients who need some help moving their limbs and exercising muscles or for whom moving joints is painful
- passive exercise in which the therapist moves the limbs

There are a variety of other physical therapy exercises:

- There are many muscle-strengthening exercises, all of which progressively increase resistance. Muscle-strengthening exercises also increase muscle mass and endurance. Movement against gravity is used for very weak muscles and the resistance is gradually increased using stretchy bands or weights.

- Rehabilitation from a stroke or brain damage often requires coordination exercises that involve specific tasks that work multiple joints and muscles, such as picking up an object.
- Rehabilitation may also require balance exercises, beginning with shifting one's weight from side to side and front to back using parallel bars.
- Once a patient can balance while standing, ambulation exercises begin with walking using parallel bars and progressing to a walker, crutches or a cane, possibly wearing a brace or assistive belt to prevent falls.
- Once a patient can walk on a level surface, ambulation exercises involve stepping over curbs or climbing stairs. This may include teaching family members and caregivers how to correctly support the patient.
- General conditioning exercises combine range-of-motion, muscle-strengthening, and ambulation exercises to counter the effects of prolonged bed rest or immobilization, improve cardiovascular fitness, and maintain flexibility and muscle strength.

Transfer training—moving safely and independently from bed to chair, chair to toilet, or chair to standing—is a critical component of physical therapy. It is often required for patients who have had a hip fracture, **amputation**, or **stroke**. Transfer training techniques depend on whether the patient:

- can bear weight on one or both legs
- can balance well
- is paralyzed on one side
- can use assistive devices

Tilt tables are used for patients who have had strict bed rest for several weeks or have had a **spinal cord injury**, since they can become dizzy when standing up. Tilt tables retrain blood vessels to narrow and widen appropriately with changes in posture. The patient lies face-up on a padded table with a footboard and is held in place with a safety belt as the table is slowly tilted.

To decrease lower back pain and restore mobility, physical therapy utilizes:

- manual therapies, including spinal manipulation, to improve the mobility of joints and soft tissues
- specific strengthening and/or flexibility exercises
- training for sitting, sleeping, bending, lifting, and performing chores
- education about back care

Physical therapy for diabetes includes:

- testing sensation in the feet
- teaching patients how to protect feet that have lost sensation

- recommending footwear or assistive devices
- adapting shoes or orthotic devices for walking
- decreasing cramping during walking
- caring for skin ulcers and sores
- supervising exercise programs

Physical therapists design exercise programs to prevent knee or other injuries, beginning with an evaluation of body traits that predispose a patient to injury. Physical therapy can also help prevent and treat **osteoporosis**. A 2009 study found that physical therapy started soon after **breast cancer** surgery, including massage and shoulder exercises, can reduce or prevent the common complication of **lymphedema**.

Physical therapy uses a variety of techniques to reduce swelling and relieve pain, including:

- hot and cold packs
- paraffin baths
- electrical stimulation
- ultrasound
- massage, including deep-tissue massage
- traction

Pediatric physical therapists evaluate children in the context of their daily routines and activities. Evaluation may include:

- mobility
- analyzing gait (walking and running)
- sensory and neuromotor development
- muscle and joint function
- strength and endurance
- posture and balance
- cardiopulmonary status
- oral motor skills
- use of assistive technologies

Physical therapy for children can include:

- movement and mobility
- posture, positioning, and lifting
- strengthening
- motor learning
- coordination and balance
- cardiopulmonary endurance
- developmental activities
- adapting daily routines and activities
- fitting and use of assistive technology
- orthotics and prosthetics
- burn and wound care
- safety, health, and prevention programs

Pediatric physical therapy utilizes many of the same evaluation and therapeutic techniques as adult physical therapy, but often includes toys and pediatric therapy gyms with balls, benches, swings, and slides. Pediatric physical therapy may also include:

- identifying existing and potential problems
- developmental activities such as crawling and walking
- adaptive play
- aquatic therapy
- recommending safe sports and other activities
- consulting with medical, psychiatric, and school personnel on individual education plans

Benefits

Physical therapy can help patients gain and maintain mobility, independence, and quality of life. It can help prevent and manage medical conditions and motivate patients to improve on their own. A study published in 2008 found that exercises developed by physical therapists could reduce the risk of athletic injuries by 41%. Physical therapy can prevent loss of mobility by designing exercise programs based on individual characteristics. Physical therapy does not usually require a referral from a physician.

Physical therapy often can eliminate the need for prescription drugs or surgery. A 2008 study found that physical therapy and medical management were as effective as knee surgery for relieving stiffness and pain from moderate to severe **osteoarthritis**.

Precautions

Physical therapy can be painful and patients often must do much of the hard work on their own. For some conditions, such as tight ligaments or tendons, range of motion often cannot be increased by gentle stretching until after surgical intervention.

Preparation

Patient attitude and cooperation are key to successful physical therapy. Patients must be active participants in their treatment, aware of the short-term and long-term goals of their therapy, and able to communicate with their therapists.

Aftercare

Physical therapy often requires patients to follow a specially designed exercise program. Practicing on one's own can be an essential component of successful physical therapy.

KEY TERMS

Ambulation—Moving from place to place.

Goniometer—An instrument for measuring angles of a joint.

Lymphedema—Swelling (edema) due to damaged lymphatic drainage.

Orthotics—Support or bracing of weak or ineffectual muscles or joints.

Prosthesis—An artificial device that replaces or augments a body part.

Repetitive stress injury; repetitive strain injury (RSI)—Any of various musculoskeletal disorders—such as tendonitis or carpal tunnel syndrome—that are caused by cumulative damage to muscles, tendons, ligaments, nerves, or joints from highly repetitive movements, such as of the hand, wrist, arm, or shoulder.

Stroke—A sudden diminishing or loss of consciousness, sensation, or voluntary movement from a rupture or obstruction of a blood vessel in the brain.

Tilt table; tiltboard—An apparatus for rotating a person from horizontal to an oblique or vertical position.

Traction—Pulling force exerted on a skeletal structure by a special device or piece of equipment.

Risks

Risks of physical therapy can include pain, falls, bruising, or injury.

Training and certification

Physical therapists have master's degrees or clinical doctorates in physical therapy. As of 2009 there were 212 accredited physical therapist education programs in the United States—12 awarding master's degrees and 200 awarding doctoral degrees. In 2008 more than 75% of all physical therapy graduates were doctors of physical therapy (DPTs). The programs include basic medical and clinical coursework and supervised clinical experience. Physical therapists are required to pass a national licensure exam and must be licensed in each state in which they practice. Physical therapists participate in continuing education courses and workshops. Some physical therapists are board-certified in cardiovascular and pulmonary, clinical electrophysiologic, geriatric, neurologic, orthopedic, pediatric, sports, or **women's health** specialties.

Physical therapy assistants usually have an associate degree from an accredited physical therapist assistant program. They are also required to have clinical and first-aid experience and certification in **cardiopulmonary resuscitation** (CPR). Physical therapy aides are usually required to have a high school diploma and are trained on the job. Because they are not licensed, aides are able to perform only a limited range of tasks.

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ORGANIZATIONS

AGS Foundation for Health in Aging, The Empire State Building, 350 Fifth Avenue, Suite 801, New York, NY, 10118, (212) 755-6810, (800) 563-4916, (212) 832-8646, www.healthinaging.org.

American Physical Therapy Association, 1111 North Fairfax Street, Fairfax, VA, 22314-1488, (703) 684-APTA (2782), (800) 999-APTA (2782), (703) 684-7343, <http://www.apta.org>.

National Rehabilitation Information Center, 8201 Corporate Drive, Suite 600, Landover, MD, 20785, (301) 459-5900, (800) 346-2742, (301) 459-4263, naricinfo@heitechservices.com, <http://www.naric.com/>.

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Pica

Definition

Pica is the persistent craving and compulsive eating of nonfood substances. The Diagnostic and Statistical Manual of Mental Disorders, fourth edition, classifies it as a feeding and eating disorder of childhood.

Description

The puzzling phenomenon of pica has been recognized and described since ancient times. Pica has been observed in ethnic groups worldwide, in both primitive and modernized cultures, in both sexes, and in all age groups. The word pica comes from the Latin name for magpie, a bird known for its unusual and indiscriminate eating habits. In addition to humans, pica has been observed in other animals, including the chimpanzee.

Pica in humans has many different subgroups, defined by the substance that is ingested. Some of the most commonly described types of pica are eating earth, soil or clay (geophagia), ice (pagophagia) and starch (amylophagia). However, pica involving dozens of other substances, including cigarette butts and ashes, hair, paint chips, and paper have also been reported. In one unusual case, the patient ingested transdermal patches of fentanyl, an opioid medication given for severe pain. Eating the skin patch increased the patient's dose of the drug by a factor of 10.

Although pica can occur in individuals of any background, a higher incidence of pica is associated with:

- pregnancy
- developmental delay and mental retardation
- psychiatric disease and autism
- early childhood
- poor nutrition or low blood levels of iron and other minerals
- certain cultural or religious traditions

Causes and symptoms

Evidence suggests that there may be several causes of pica. One widely held theory points to iron deficiency as a major cause of pica. Several reports have described pica in individuals with documented iron deficiency, although there has been uncertainty as to whether the iron deficiency was a cause of pica or a result of it. Because some substances, such as clay, are believed to block the absorption of iron into the bloodstream, it was thought that low blood levels of

iron could be the direct result of pica. However, some studies have shown that pica cravings in individuals with iron deficiency stop once iron supplements are given to correct the deficiency. Another study looked specifically at the rate of iron absorption during pica conditions and normal dietary behavior, and showed that the iron absorption was not decreased by pica. In addition, low blood levels of iron commonly occur in pregnant women and those with poor nutrition, two populations at higher risk for pica. Such findings offer strong support of iron deficiency as a cause, rather than result, of pica.

Other reports suggest that pica may have a psychological basis and may even fall into the spectrum of obsessive-compulsive disorder. Pica has a higher incidence in populations with an underlying diagnosis involving mental functioning. These diagnoses include psychiatric conditions like schizophrenia, developmental disorders including autism, and conditions with mental retardation. These conditions are not characterized by iron deficiency, which supports a psychological component in the cause of pica.

Cultural and religious traditions may also play a role in pica behavior. In some cultures, nonfood substances are believed to have positive health or spiritual effects. Among some African Americans in the south, ingesting a particular kind of white clay is believed to promote health and reduce morning sickness during pregnancy. Other cultures practice pica out of belief that eating a particular substance may promote fertility or bring good luck.

The hallmark feature of pica, consistently consuming nonfood substances, often does not present publicly. People may be embarrassed to admit to these unusual eating habits, and may hide it from their family and physician. In other cases, an individual may not report the pica to a physician simply because of a lack of knowledge of pica's potential medical significance.

Because the eating behaviors of pica are not usually detected or reported, it is the complications of the behavior that bring it to attention. Complications vary, depending on the type of pica. Geophagia has potential side effects that most commonly affect the intestine and bowel. Complications can include constipation, cramping, pain, obstruction caused by formation of an indigestible mass, perforation from sharp objects like rocks or gravel, and contamination and infection from soil-dwelling parasites.

Amylophagia usually involves the consumption of cornstarch and, less frequently, laundry starch. The high caloric content of starch can cause excessive

weight gain, while at the same time leading to **malnutrition**, as starch contributes “empty” calories lacking **vitamins** and **minerals**. Amylophagia during pregnancy can mimic **gestational diabetes** in its presentation and even in its potential harmful effects on the fetus.

Pica involving the ingestion of substances such as lead-based paint or paper containing mercury can cause symptoms of toxic **poisoning**. Compulsive consumption of even a seemingly harmless substance like ice (pagophagia) can have negative side effects, including decreased absorption of nutrients by the gut.

Diagnosis

In order for the diagnosis of pica to be made, there must be a history of persistent consumption of a non-food substance continuing for a minimum period of one month. Infants and toddlers are typically excluded from this diagnosis since mouthing objects is a normal developmental behavior at that age. Individuals with mental retardation who function at or below an approximate cognitive level of 18 months may also be exempt from this diagnosis.

Pica is most often diagnosed when a report of such behaviors can be provided by the patient or documented by another individual. In other cases, pica is diagnosed after studies have been performed to assess the presenting symptoms. For example, imaging studies ordered to assess severe gastrointestinal complaints may reveal intestinal blockage with an opaque substance; such a finding is suggestive of pica. Biopsy of intestinal contents can also reveal findings, such as parasitic infection, consistent with pica. Pica may also be suspected if abnormal levels of certain minerals or chemicals are detected in the blood.

Pica in pregnant women is sometimes diagnosed after **childbirth** because of a health problem in the newborn caused by the substance(s) ingested by the mother. In one instance reported in Chicago, a newborn girl was treated for **lead poisoning** caused by her mother’s eating fragments of lead-glazed pottery during pregnancy.

Treatment

Treatment of pica will often depend on the cause and type of pica. Conventional medical treatment may be appropriate in certain situations. For example, supplementation with iron-containing vitamins has been shown to cause the unusual cravings to subside in some iron-deficient patients.

Medical complications and health threats, including high lead levels, bowel perforation or intestinal

KEY TERMS

Amylophagia—The compulsive eating of purified starch, typically cornstarch or laundry starch.

Geophagia—The compulsive eating of earthy substances, including sand, soil, and clay.

Pagophagia—The compulsive eating of ice.

obstruction, will require additional medical management, beyond addressing the underlying issue of pica.

Alternative treatment

Because most cases of pica do not have an obvious medical cause, treatment with counseling, education, and nutritional management is often more successful and more appropriate than treatment with medication. Some therapists specializing in **eating disorders** may have expertise in treating pica.

Prognosis

The prognosis for individuals with pica varies greatly, according to the type and amount of substance ingested, the extent of presenting side effects, and the success of treatment. Many of the side effects and complications of pica can be reversed once the behavior is stopped, while other complications, including infection and bowel perforation, pose significant health threats and if not successfully treated may result in **death**.

When seen in children, pica behavior tends to lessen with age. However, individuals with a history of pica are more likely to experience it again. Counseling and nutritional education can reduce the risk of recurrence.

Prevention

There are no known methods of preventing pica. However, once pica is known or suspected, measures can be taken to reduce further ingestion of nonfood substances. Removing the particular substance from readily accessible areas can be helpful. Close observation of the individual with pica may limit inappropriate eating behaviors.

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ORGANIZATIONS

- American Academy of Child and Adolescent Psychiatry (AACAP), 3615 Wisconsin Ave. NW, Washington, DC, 20013-3007, (202) 966-7300, (202) 966-2891, communications@aacap.org, <http://www.aacap.org/>.

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Pickwickian syndrome

Definition

A group of symptoms that generally accompany massive **obesity**.

Description

Pickwickian syndrome is a complex of symptoms that primarily affect patients with extreme obesity. The syndrome is named after a character in a Charles Dickens novel, *The Pickwick Papers*, who seemed to show some of the traits of this disease.

The major health problem that occurs in patients with this disease is **sleep apnea**. This is caused in part by the excess amounts of fatty tissue surrounding the chest muscles. This excess fat places a strain on the heart, lungs, and diaphragm of the patient, making it difficult to breathe.

KEY TERMS

Latency—The period of inactivity between the time a stimulus is provided and the time a response occurs.

Obesity—Exceeding one's normal weight by 20%. A person suffering from extreme obesity would exceed their normal weight by a much higher percentage.

Pulmonary system—Lungs and respiratory system of the body.

Causes and symptoms

The major cause of Pickwickian syndrome is extreme obesity. This obesity places an excessive load on the pulmonary system. The role of genetics is also being studied. Symptoms of Pickwickian syndrome include excessive daytime sleepiness, **shortness of breath** due to elevated blood carbon dioxide pressure, disturbed sleep at night, and flushed face. The skin can also have a bluish tint, and the patient may have high blood pressure, an enlarged liver, and an abnormally high red blood cell count.

Diagnosis

Some tests that can be used to diagnose this condition include **echocardiography** to determine heart enlargement or **pulmonary hypertension**. Giving the patient multiple sleep latency tests can help give an objective measurement of daytime sleepiness. **Magnetic resonance imaging** (MRI), computed tomography (CT) scans, or fiberoptic evaluation of the upper airway may also be used.

Treatment

The primary treatment for Pickwickian syndrome is focused on weight loss and increased physical activity. Also, medroxyprogesterone may help improve the condition.

Prognosis

Pickwickian syndrome is entirely reversible if it is diagnosed and treated properly. If the problem goes undiagnosed, the outcome can be fatal.

Prevention

Prevention of Pickwickian syndrome can be achieved by maintaining a healthy body weight and

getting the proper amount of **exercise**. For prevention of the sleep apnea that generally accompanies Pickwickian syndrome, there are several possible treatments. If the sleep apnea is only present when the patient is flat on their back, a tennis ball can be sewn into the sleep clothes to remind the patient not to sleep on their back. For more severe cases of sleep apnea, a **tonsillectomy** or the use of dental appliances may be recommended.

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PID see **Pelvic inflammatory disease**



Piercing and tattoos

Definition

Piercing and tattoos are popular forms of body art that have been practiced throughout history. Piercing involves punching a hole in the earlobe or another body part for the insertion of jewelry. Tattoos are temporary or permanent writing or designs anywhere on the body. Permanent tattoos involve the insertion of pigments through pricks in the skin.

Demographics

Although piercing of the female earlobe and male tattooing have long been common in Western societies, in recent decades the piercing of various body parts, as well as tattooing, have become increasingly popular among both males and females, although body piercing is more common in females. The reported prevalence of body piercing ranges from 6.8–14% in the general population and from 4.3–51% among teenagers and young adults. Increases in piercing and tattooing have resulted in an increase in medical complications. A Mayo Clinic study reported that 17% of college students with piercings suffered complications such as infection or tearing. African Americans often develop thick **scars** called **keloids** after body piercing.

Tattoo artist Michael Wilson displaying his own tattoos and piercings. (Susan McCartney/Photo Researchers, Inc.)

Description

Body piercing and tattooing have been practiced throughout human history as emblems of beauty and as symbols. The 4,000-year-old body of a tattooed man was discovered preserved in an Alpine glacier in 1992. Egyptians tattoos in the period from 4000–2000 BC symbolized fertility and nobility. Body piercing often connoted royalty and courage. In some hunting and gathering societies body piercing and tattoos have long been used in initiation rites and as socialization/enculturation symbols. In today's industrialized societies piercing and tattoos are a popular art form practiced by people of all ages. They can signify beauty, independence, defiance, a psychology of **self-mutilation**, or membership in social groups, such as prison or gang cultures. Oral and genital piercing are sometimes thought to enhance sexuality.

Piercing and tattooing are performed by amateurs on themselves or others or by professionals in tattoo

or beauty parlors or shops selling jewelry. In the United States commercial piercing and tattooing are usually regulated at the state or local level.

Although earlobe piercing remains the most common form, other popular piercing sites include the cartilage of the ears, nasal septa, eyebrows, tongue, lips, cheeks, the uvula (the fleshy lobe at the back of the palate), nipples, the naval, labia, and the penis. After cleaning the skin the needle and jewelry are quickly inserted through the tissue in one motion, without anesthesia. Earlobes are normally pierced using a sterile, single-use, spring-loaded piercing gun. Other body parts are pieced using hollow, six-gauge to 18-gauge needles.

The word "tattoo" comes from the Tahitian word "tattau," meaning "to mark." Permanent tattoos are colored inks injected into small deep holes in the skin. This process can take several hours and cause a small amount bleeding and some degree of **pain**. None of the more than 50 pigments and shades in tattoo inks has been approved by the U.S. Food and Drug Administration (FDA) for injection into the skin. Many of them are not even approved for contact with skin and some are industrial-grade colors for printers' ink or automobile paint.

There are various types of permanent tattoos:

- Professional tattoos are produced by a tattoo artist with a special electric needle gun that repeatedly punctures the skin and inserts tiny drops of ink.
- Amateur tattoos are most often India ink imbedded beneath the skin with a pin. Pen ink, charcoal, or ashes are also used as pigment. Amateur tattoos are often crude.
- Cultural tattoos are produced according to specific ethnic or cultural traditions.
- Cosmetic tattoos or permanent makeup replace the use of a cosmetic such as an eyebrow pencil, eyeliner, lip liner, or blush. Cosmetic tattoos can also replace a nipple after breast surgery or cover up a non-pigmented patch of skin or another tattoo.
- Medical tattoos are placed by a physician, usually as a guide for radiation therapy.
- Traumatic tattoos are pigmentation remaining after dirt or other debris becomes imbedded in the skin as the result of an accident or puncture wound.

Temporary tattoos are produced with either stickers or a natural plant dye called henna or mehndi. Sticker-type tattoos are designs on coated paper that are applied to the skin with water or by rubbing. They last only a few days. In the United States they contain only colors that are approved for use in cosmetics.

Henna tattoos are drawn on the skin and last for two or three weeks. The FDA has not approved henna for use on the skin.

Although they can be attractive body adornments, piercing and tattoos can also be problematical:

- The most common medical problems are infections, keloids, and allergic reactions.
- Fashions, personal taste, or the body itself can change, making a piercing or tattoo undesirable; however a piercing may leave a permanent hole and tattoos can be difficult or impossible to remove.
- If tattoo pigments are injected too deeply, they can migrate and blur the design.

Risk factors

Several recent studies have suggested that piercing and tattoos may be associated with negative and anti-social behaviors teenagers and young adults. Body piercing has been associated with a variety of potentially harmful behaviors, including alcohol and drug use, **smoking**, problem gambling, high-risk sex, and Russian roulette. Body piercing has been found to be more common in young females who are compulsive, thrill-seeking, and/or emotionally negative. A survey of 1753 American college students found that those with at least four tattoos, at least seven piercings, or piercings in the nipples or genitals were significantly more likely to use **marijuana** regularly, to occasionally use other illegal drugs, and to have been arrested for a crime. They were also more likely to cheat in school, binge drink, and report having multiple sex partners.

Causes and symptoms

Piercing and tattooing under non-sterile conditions can transmit infections. The most common bacterial skin infections are caused by *Streptococcus* and *Staphylococcus aureus* and antibiotic-resistant "staph" has been reported among tattoo recipients. Transmission of **tetanus** and **tuberculosis** from ear piercing has been reported. Piercing and tattooing can theoretically transmit serious viral infections, such as hepatitis and HIV, although there has never been a known HIV transmission by a professional experienced tattoo artist.

Piercing and tattoos can cause scarring and keloid or granuloma formation. Granulomas are nodules that can form around foreign material, such as particles of tattoo pigment.

Other potential complications of piercing include:

- abscesses from serious infection
- infections that spread throughout the body

- endocarditis, a serious inflammation of the heart lining and valves
- weakening and tearing of fleshy tissue leading to disfigurement
- damage to the delicate cartilage of the upper ear, sometimes requiring surgical intervention
- contact dermatitis
- allergic reactions to jewelry containing brass plating or a nickel alloy
- skin that grows over the jewelry, often from studs that are too tight
- disfigurement from the forcible removal of jewelry
- urethral rupture from genital piercing

Oral piercing carries additional risks including:

- gum injury, receding tissues, or damage to teeth or fillings from contact with jewelry
- drooling or interference with speech, chewing, or swallowing from excessive saliva production
- nerve damage from tongue piercing, causing numbness or changes in the sense of taste
- tongue swelling that can block the airway
- severe blood loss from tongue piercing
- slow healing due to constant tongue movement
- unfastened jewelry that is a choking hazard or can be swallowed, injuring the digestive tract
- jewelry that interferes with dental x rays

Allergic reactions can occur to compounds used in tattoo pigments, including organic dyes and oxides of iron, mercury, chromium, cadmium, and cobalt. Sometimes an allergic reaction can develop to a tattoo that has been in place for years. Allergic reactions can cause serious problems because the pigments are hard to remove. Tattoos also occasionally interfere with **magnetic resonance imaging (MRI)** or other medical tests.

Although henna tattoos do not pierce the skin, black henna, which contains para-phenylenediamine, frequently causes **contact dermatitis**. It can be absorbed through the skin of some people and has been known to cause renal (kidney) failure and even **death**. It is particularly dangerous for young children.

Symptoms of a localized bacterial infection include redness, swelling, pain, and pus. Allergic skin reactions include swelling, redness, and severe **itching**.

Diagnosis

Examination

Bacterial infection or allergic reactions in a pierced or tattooed area are usually apparent upon

KEY TERMS

Contact dermatitis—Skin inflammation resulting from contact with an allergen or other substance.

Endocarditis—Inflammation of the heart lining and valves.

Granuloma—A nodule or mass of chronically inflamed tissue.

Henna—Mehndi; a reddish-brown dye from the leaves of the henna plant; used for hair dye and temporary tattoos.

Hepatitis—Inflammation of the liver, often caused by a virus.

Keloid—A thick scar.

Socialization—Process by which new members are integrated into a social group.

physical examination. However signs of blood-borne infections may not be obvious.

Treatment

Traditional

- Keloids may require surgery, including cryosurgery.
- Pus-producing granulomas must also be surgically removed.
- Plastic surgery may be required to correct holes or disfigurements from piercing.
- Damage to teeth from oral jewelry may require restorative dentistry.

Methods for removing tattoos include:

- laser surgery
- excision (surgical cutting)
- dermabrasion—sanding the skin with a wire brush
- salabrasion—soaking with a salt solution
- scarification—using an acid solution to replace the tattoo with scar tissue
- cosmetic over-tattooing

Drugs

- over-the-counter antibiotic ointments for minor infections
- oral antibiotics for serious infections
- topical steroids or other medications for allergic reactions
- steroid or interferon injections for keloids

Home remedies

New piercings should be cleaned with a medicated cleanser while gently moving the jewelry around. The area should also be cleaned twice daily with soap and warm water. An antibacterial mouth rinse should be used after meals with oral piercing.

The bandage should be removed from a new tattoo after 24 hours. The skin should be kept clean with plain soap and water and patted dry. Antibiotic ointment should be applied to the tattoo during healing. A mild moisturizer should be applied to newly tattooed skin several times a day.

Minor localized infections can be treated with warm compresses or by soaking with mild sea salt. Jewelry should remain in an infected piercing to ensure proper drainage and prevent **abscess** formation.

Prognosis

Healing time for piercing ranges from a few months to two years. Tattoos take up to two weeks to heal. Minor infections respond well to antibiotic therapy and can usually be treated without losing the piercing. However blood-borne infections can have life-altering and life-threatening consequences. Disfigurements from piercing may be correctable with **plastic surgery**. Allergic reactions are only rarely life-threatening but can lead to permanent scarring or altered pigmentation. Tattoo removal is expensive, usually involves several treatments, is not always successful, and rarely leaves the skin as pristine as before the tattoo.

Prevention

- Piercing and tattooing should be performed steriley by an experienced professional who complies with local regulations and inspections and always wears a new pair of sterile gloves.
- New sterile needles and tubes should be unwrapped in front of the customer.
- All non-disposable equipment should be heat sterilized in an autoclave.
- Drawer handles, tables, and sinks should be washed with a commercial disinfectant or bleach solution after each use.
- Piercing guns should be single-use or take sterile disposable cassettes.
- Piercing guns should be used only on the ear.
- Piercing should be completed with smoothly polished jewelry made of 14-carat or 18-carat gold, titanium, surgical steel, or niobium.
- Jewelry should be handled as little as possible.

- Jewelry should never be pulled.
- A new tattoo should not be exposed to sunlight for at least a few weeks.
- Clothing should not be allowed to stick to a new tattoo.
- Scabs should not be picked.

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- American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168, (847) 240-1280, (866) 503-SKIN (7546), (847) 240-1859, <http://www.aad.org>.
- American Academy of Family Physicians, 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2680, (913) 906-6000, (800) 274-6000, (913) 906-6075, <http://www.aafp.org/online/en/home.html>.
- American Dental Association, 211 East Chicago Ave., Chicago, IL, 60611-2678, (312) 440-2500, <http://www.ada.org>.

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Pilates

Definition

Pilates or Physical Mind method, is a series of non-impact exercises designed by Joseph Pilates to develop strength, flexibility, balance, and inner awareness.

Purpose

Pilates is a form of strength and flexibility training that can be done by someone at any level of fitness. The exercises can also be adapted for people who have limited movement or who use wheel chairs. It is an engaging **exercise** program that people want to do. Pilates promotes a feeling of physical and mental well-being and also develops inner physical awareness. Since this method strengthens and lengthens the muscles without creating bulk, it is particularly beneficial for dancers and actors. Pilates is also helpful in preventing and rehabilitating from injuries, improving posture, and increasing flexibility, circulation, and balance. Pregnant women who do these exercises can develop body alignment, improve concentration, and develop body shape and tone after **pregnancy**. According to Joseph Pilates, “You will feel better in 10 sessions, look better in 20 sessions and have a completely new body in 30 sessions.”

Although Pilates is often associated with dancers, athletes, and younger people in general who are interested in improving their physical strength and flexibility, a simplified version of some Pilates exercises is also being used to lower the risk of hospital-related deconditioning in older adults. A Canadian study of hospitalized patients over the age of 70 found that those who were given a set of Pilates exercises that could be performed in bed recovered more rapidly than a control group given a set of passive range-of-motion exercises.



Woman performing Pilates exercises with the aid of a Pilates reformer. (© Jim Cummins/Corbis.)

Description

Origins

Joseph Pilates (pronounced pie-LAH-tes), the founder of the Pilates method (also simply referred to as “the method”) was born in Germany in 1880. As a frail child with **rickets**, **asthma**, and **rheumatic fever**, he was determined to become stronger. He dedicated himself to building both his body and his mind through practices which included **yoga**, zen, and ancient Roman and Greek exercises. His conditioning regime worked and he became an accomplished gymnast, skier, boxer, and diver.

While interned in England during World War I for being a German citizen, Pilates became a nurse. During this time, he designed a unique system of hooking springs and straps to a hospital bed in order to help his disabled and immobilized patients regain strength and movement. It was through these experiments that he recognized the importance of training the core abdominal and back muscles to stabilize the torso and allow the entire body to move freely. This experimentation provided the foundation for his style of conditioning and the specialized exercise equipment associated with the Pilates method.

Pilates emigrated to the United States in 1926 after the German government invited him to use his conditioning methods to train the army. That same year he opened the first Pilates studio in New York City. Over the years, dancers, actors, and athletes flocked to his studio to heal, condition, and align their bodies.

Joseph Pilates died at age 87 in a fire at his studio. Although his strength enabled him to escape the flames by hanging from the rafters for over an hour,

he died from **smoke inhalation**. He believed that ideal fitness is “the attainment and maintenance of a uniformly developed body with a sound mind fully capable of naturally, easily, and satisfactorily performing our many and varied daily tasks with spontaneous zest and pleasure.”

During the initial meeting, an instructor will analyze the client’s posture and movement and design a specific training program. Once the program has been created, the sessions usually follow a basic pattern. A session generally begins with mat work and passive and active stretching. In passive stretching, the instructor moves and presses the client’s body to stretch and elongate the muscles. During the active stretching period, the client performs the stretches while the instructor watches their form and breathing. These exercises warm up the muscles in preparation for the machine work. The machines help the client to maintain the correct positioning required for each exercise.

There are over 500 exercises that were developed by Joseph Pilates. “Classical” exercises, according to the Pilates Studio in New York involve several principles. These include concentration, centering, flowing movement, and breath. Some instructors teach only the classical exercises originally taught by Joseph Pilates. Others design new exercises that are variations upon these classical forms in order to make the exercises more accessible for a specific person.

There are two primary exercise machines used for Pilates, the Universal Reformer and the Cadillac, and several smaller pieces of equipment. The Reformer resembles a single bed frame and is equipped with a carriage that slides back and forth and adjustable springs that are used to regulate tension and resistance. Cables, bars, straps, and pulleys allow the exercises to be done from a variety of positions. Instructors usually work with their clients on the machines for 20–45 minutes. During this time, they are observing and giving feedback about alignment, breathing, and precision of movement. The exercises are done slowly and carefully so that the movements are smooth and flowing. This requires focused concentration and muscle control. The session ends with light stretching and a cool-down period.

Once the basics are learned from an instructor, from either one-on-one lessons or in a class, it is possible to train at home using videos. Exercise equipment for use at home is also available and many exercises can be performed on a mat.

KEY TERMS

Yoga—A system of physical, mental, and breathing exercises developed in India.

Zen—A form of meditation that emphasizes direct experience.

A private session costs between \$45–75 dollars, depending on the part of the country one is in. This method is not specifically covered by insurance although it may be covered when the instructor is a licensed physical therapist.

Precautions

The Pilates method is not a substitute for good **physical therapy**, although it has been increasingly used and recommended by physical therapists since the mid-1980s. People with chronic injuries are advised to see a physician.

Research and general acceptance

As of early 2004, several physical therapists and gerontologists have done research studies on the Pilates method, although much more work needs to be done in this area. One recent finding is that the method should not be used by patients with lower back **pain**, as it appears to be ineffective in treating this condition.

The appeal of the Pilates method to a wide population, coupled with a new interest in it on the part of **rehabilitation** therapists, suggests that further studies may soon be underway. Dancers and actors originally embraced the Pilates method as a form of strength training that did not create muscle bulk. Professional and amateur athletes also use these exercises to prevent reinjury. Sedentary people find Pilates to be a gentle, non-impact approach to conditioning. Pilates equipment and classes can be found in hospitals, health clubs, spas, and gyms.

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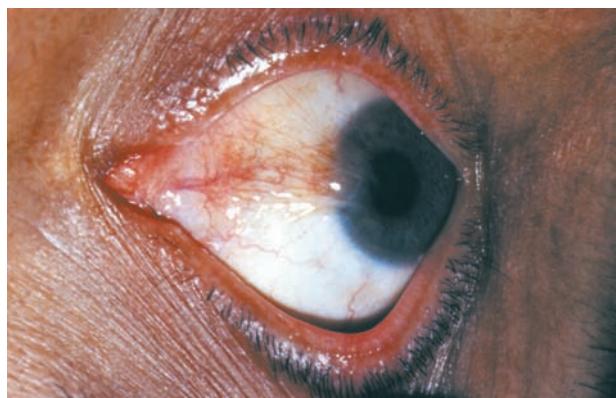
ORGANIZATIONS

Pilates Method Alliance, P.O. Box 370906, Miami, FL, 33137-0906, (305) 573-4461, (866) 573-4945, info@pilatesmethodalliance.org, http://www.pilatesmethodalliance.org.

United States Pilates Association, 1500 East Broward Blvd. Suite 250, Ft. Lauderdale, FL, 33301, (888) 484-8772, info@unitedstatespilatesassociation.com, http://www.unitedstatespilatesassociation.com.

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Piles see **Hemorrhoids**



Pterygium, an overgrowth of the cornea, is usually on the inner side of the eye by thickened and degenerative conjunctiva. (Photo Researchers, Inc.)

may also grow over the cornea and may therefore affect vision.

Causes and symptoms

Causes

The cause or causes of these disorders are unknown, but they are more frequent in people who live in sunny and windy climates and people whose jobs expose them to ultraviolet (UV) light (for example, farmers and arc welders). Pingueculae and pterygia also occur in older people. It is thought these growths are the result of UV or infrared light and irritation. It is also believed that prolonged exposure to these risk factors (that is, UV light) increases the chances of occurrence.

Symptoms

Although some people with pinguecula constantly feel like they have a foreign body in their eye, most are asymptomatic. Because the lids can no longer spread the tears over a smooth area, dry areas may result. Some people with a pterygium are also asymptomatic; some feel like they have a foreign body in their eye. Because a pterygium can stretch and distort the cornea, some people acquire **astigmatism** from a pterygium.

Diagnosis

An eye doctor (ophthalmologist or optometrist) can usually diagnose pingueculae and pterygia by external observation, generally using an instrument called a slit lamp. A slit lamp is a microscope with a

Pinguecula and pterygium

Definition

Pinguecula and pterygium are both non-malignant, slow-growing proliferations of conjunctival connective tissue in the eye. Pterygia, but not pingueculae, extend over the cornea.

Description

The outer layer of the eyeball consists of the tough white sclera and the transparent cornea. The cornea lies in front of the colored part of the eye (iris). Overlying the sclera is a transparent mucous membrane called the conjunctiva. The conjunctiva lines the inside of the lids (palpebral conjunctiva) and covers the sclera (bulbar conjunctiva).

Pingueculae and pterygia are common in adults, and their incidence increases with age. Pterygia are less common than pingueculae.

Pingueculae are seen as small, raised, thickenings of the conjunctiva. They may be yellow, gray, white, or colorless. They are almost always to one side of the iris—not above or below—and usually on the side closest to the nose. A pinguecula may develop into a pterygium.

Pterygia are conjunctival thickenings that may have blood vessels associated with them. They often have a triangular-shaped appearance. The pterygia

KEY TERMS

Astigmatism—Asymetric vision problems due to irregularities in the cornea.

Beta radiation—Streams of electrons emitted by beta emitters like carbon-14 and radium.

Conjunctiva—The mucous membrane that covers the white part of the eyes and lines the eyelids.

Cornea—The clear outer covering of the front of the eye. It is in front of the colored part of the eye (iris) and the iris's central black hole (pupil).

light source and magnifies the structures of the eye for the examiner. However, because pingueculae and pterygia can sometimes look similar to more serious eye growths, it is important for people to have them checked by an eye care professional.

Treatment

Usually, no treatment is needed. Artificial tears can be used to relieve the sensation of a foreign body in the eye and to protect against dryness. Surgery to remove the pinguecula or pterygium is advisable when the effect on the cornea causes visual defects or when the thickening is causing excessive and recurrent discomfort or inflammation. Sometimes surgical removal is also performed for cosmetic reasons. However, healing from this type of surgery, although usually painless, takes many weeks, and there is a high rate of recurrence (as high as 50–60% in some regions). Accordingly, surgery is avoided unless problems due to the pinguecula or pterygium are significant.

Several methods have been used to attempt to reduce the recurrence of the pinguecula or pterygium after surgery. One method that should be abandoned is beta radiation. Although it is effective at slowing the regrowth of pingueculae and pterygia, it can cause **cataracts**. A preferable method is the topical application of the anticancer drug, mitomycin-C.

Prognosis

Most pingueculae and pterygia grow slowly and almost never cause significant damage, so the prognosis is excellent. Again, a diagnosis must be made to rule out other more serious disorders.

Prevention

There is nothing that has been clearly shown to prevent these disorders, or to prevent a pinguecula from progressing to a pterygium. However, the presence of pingueculae and pterygia have been linked to exposure to UV radiation. For that reason, UV exposure should be reduced. The American Optometric Association (AOA) suggests that sunglasses should block 99–100% of UV-A and UV-B rays. Patients should speak to their eye care professionals about protective coatings on sunglasses or regular spectacles. Protecting the eyes from sunlight, dust, and other environmental irritants is a good idea.

ORGANIZATIONS

New York University Department of Ophthalmology, 462 First Avenue, NBV 5N 18, New York, NY, 10016, (212) 263-6434, (212) 263-8749, <http://www.med.nyu.edu>.

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Pinkeye see **Conjunctivitis**

Pinta

Definition

A bacterial infection of the skin which causes red to bluish-black colored spots.

Description

Pinta is a skin infection caused by the bacterium *Treponema carateum*, a relative of the bacterium which causes **syphilis**. The word “pinta” comes from the Spanish and means “painted.” Pinta is also known as “azula” (blue), and “mal de pinto” (pinto sickness). It is one of several infections caused by different *Treponema* bacteria, which are called “endemic” or “non-venereal” treponematoses.

Pinta is primarily found in rural, poverty-stricken areas of northern South America, Mexico, and the Caribbean. The disease is usually acquired during childhood and is spread from one person to another by direct skin-to-skin contact. The bacteria enter the skin through a small cut, scratch, or other skin damage. Once inside the skin, the warmth and moisture allow the bacteria to multiply. The bacterial infection causes red, scaly lesions on the skin.

KEY TERMS

Lesion—An abnormal change in skin due to disease.

Causes and symptoms

Pinta is caused by an infection with the bacterium *Treponema carateum*. Persons at risk for pinta are those who live in rural, poverty-stricken, overcrowded regions of South America, Mexico, and the Caribbean. Symptoms of pinta occur within two to four weeks after exposure to the bacteria. The first sign of infection is a red, scaly, slowly enlarging bump on the skin. This is called the “primary lesion.” The primary lesion usually appears at the site where the bacteria entered the skin. This is often on the arms, legs, or face. The smaller lesions which form around the primary lesion are called “satellite lesions.” Lymph nodes located near the infected area will become enlarged, but are painless.

The second stage of pinta occurs between one and 12 months after the primary lesion stage. Many flat, red, scaly, itchy lesions called “pintids” occur either near the primary lesion, or scattered around the body. Pintid lesions progress through a range of color changes, from red to bluish-black. The skin of older lesions will become depigmented (loss of normal color).

Diagnosis

Pinta can be diagnosed by dermatologists (doctors who specialize in skin diseases) and **infectious disease** specialists. The appearance of the lesions helps in the diagnosis. A blood sample will be taken from the patient’s arm to test for antibodies to *Treponema carateum*. A scraping of a lesion will be examined under the microscope to look for *Treponema* bacteria. The results of these tests should be available within one to two days.

Treatment

Pinta is treated with benzathine penicillin G (Bicillin), given as a single injection.

Prognosis

Treatment will result in a complete cure but will not undo any skin damage caused by the late stages of

disease. Spread of pinta to the eyes can cause eyelid deformities.

Prevention

Good personal hygiene and general health may help prevent infections. In general, avoid physical contact with persons who have **skin lesions**.

Belinda Rowland, PhD

Pinworm infection see **Enterobiasis**

Pituitary adenoma see **Pituitary tumors**

Pituitary dwarfism

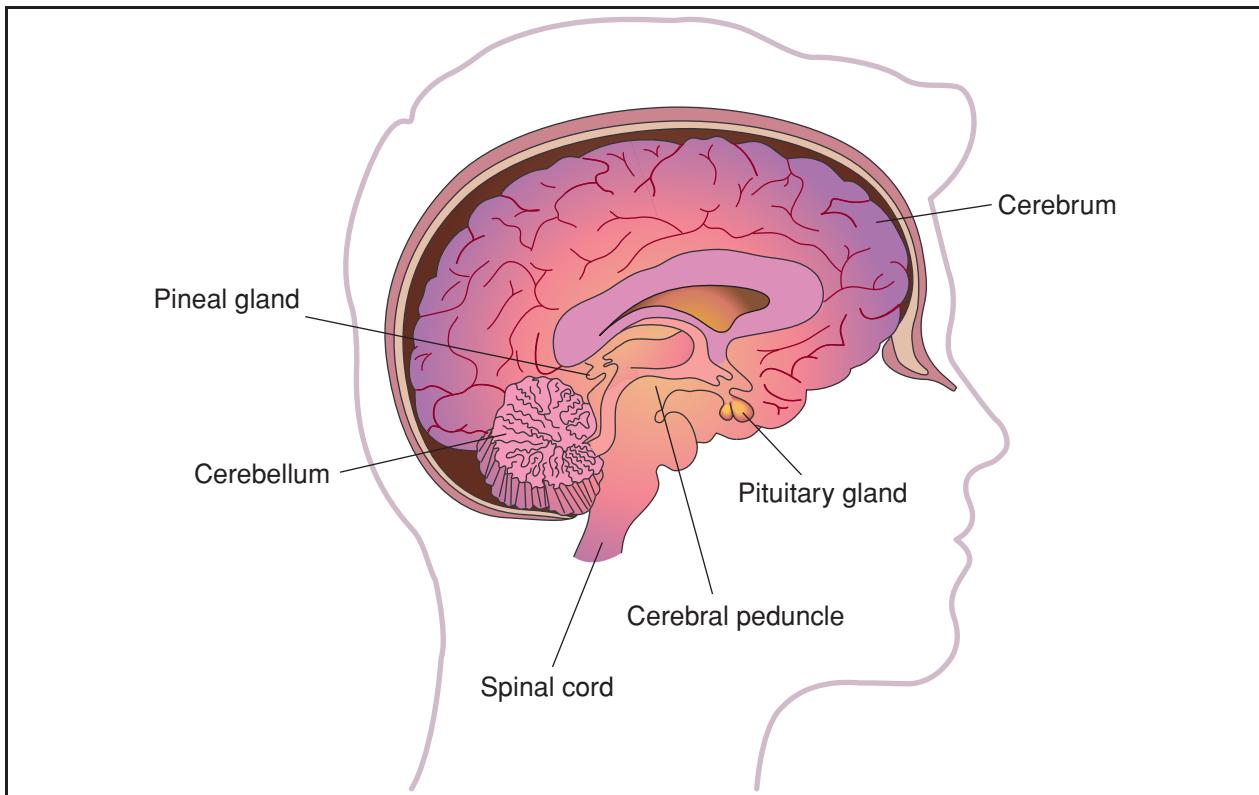
Definition

Dwarfism is a condition in which the growth of the individual is very slow or delayed. There are many forms of dwarfism. The word pituitary is in reference to the pituitary gland in the body. This gland regulates certain chemicals (hormones) in the body. Therefore, pituitary dwarfism is decreased bodily growth due to hormonal problems. The end result is a proportionate little person, because the height as well as the growth of all other structures of the individual are decreased.

Description

Pituitary dwarfism is caused by problems arising in the pituitary gland. The pituitary gland is also called the hypophysis. The pituitary gland is divided into two halves: the anterior (front) and posterior (back) halves. The anterior half produces six hormones: growth hormone, adrenocorticotropin (corticotropin), thyroid stimulating hormone (thyrotropin), prolactin, follicle stimulating hormone, and lutenizing hormone. The posterior pituitary gland only produces two hormones. It produces antidiuretic hormone (vasopressin) and oxytocin.

Most forms of dwarfism are a result of decreased production of hormones from the anterior half of the pituitary gland. The most common form is due to decreases of growth hormone which will be discussed here. These decreases during childhood cause the individual’s arms, legs, and other structures to develop normal proportions for their bodies, but at a decreased rate.



Pituitary dwarfism is a condition of growth retardation characterized by patients who are very short but have normal body proportions. It is caused by a dysfunction of the pituitary gland, the pea-sized mass of tissue located at the base of the brain. ((Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

When all of the hormones of the anterior pituitary gland are not produced, this is called panhypopituitarism. Another type of dwarfism occurs when only the growth hormone is decreased. Dwarfism can also result from a lack of somatomedin C (also called insulin like growth factor, IGF-1) production. Somatomedin C is a hormone produced in the liver that increases bone growth when growth hormone is present. The African pygmy and the Levi-Lorain dwarfs lack the ability to produce somatomedin C in response to growth hormone. All causes of dwarfism lead to a proportionate little person.

Growth is the body's response to different hormones. The forebrain contains a small organ called the hypothalamus, which is responsible for releasing hormones in response to the body's needs for purposes of regulation. Growth hormone is produced in the anterior pituitary gland when growth hormone-releasing hormone (GHRH), is released by the hypothalamus. Growth hormone is then released and stimulates the liver to produce IGF-1. In return, IGF-1 stimulates the long bones to grow in length. Thus, growth can be slowed down or stopped if there is a problem making

any of these hormones or if there is a problem with the cells receiving these hormones.

Some estimates show that there are between 10,000 and 15,000 children in the United States who have growth problems due to a deficiency of growth hormone.

Causes and symptoms

Pituitary dwarfism has been shown to run in families. New investigations are underway to determine the specific cause and location of the gene responsible for dwarfism. The human cell contains 46 chromosomes arranged in 23 pairs. Most of the genes in the two chromosomes of each pair are identical or almost identical with each other. However, with dwarfism, there appears to be disruption on different areas of chromosome 3 and 7. Some studies have isolated defects for the production of pituitary hormones to the short arm (the "p" end) of chromosome 3 at a specific location of 3p11. Other studies have found changes on the short arm of chromosome 7.

KEY TERMS

Adrenocorticotropin (corticotrophin)—A hormone that acts on cells of the adrenal cortex, causing them to produce male sex hormones and hormones that control water and mineral balance in the body.

Antidiuretic hormone (vasopressin)—A hormone that acts on the kidneys to regulate water balance.

Craniopharyngioma—A tumor near the pituitary gland in the craniopharyngeal canal that often results in intracranial pressure.

Deprivational dwarfism—A condition where emotional disturbances are associated with growth failure and abnormalities of pituitary function.

Follicle-stimulating hormone (FSH)—A hormone that in females stimulates estrogen and in males stimulates sperm production.

Growth hormone—A hormone that eventually stimulates growth. Also called somatotropin.

Hormone—A chemical messenger produced by the body that is involved in regulating specific bodily

functions such as growth, development, and reproduction.

Luteinizing hormone—A hormone secreted by the pituitary gland that regulates the menstrual cycle and triggers ovulation in females. In males it stimulates the testes to produce testosterone.

Oxytocin—A hormone that stimulates the uterus to contract during child birth and the breasts to release milk.

Panhypopituitarism—Generalized decrease of all of the anterior pituitary hormones.

Prolactin—A hormone that helps the breast prepare for milk production during pregnancy.

Puberty—Point in development when the gonads begin to function and secondary sexual characteristics begin to appear.

Thyroid stimulating hormone (thyrotropin)—A hormone that stimulates the thyroid gland to produce hormones that regulate metabolism.

A child with a growth hormone deficiency is often small with an immature face and chubby body build. The child's growth will slow down and not follow the normal growth curve patterns. In cases of tumor, most commonly **craniopharyngioma** (a tumor near the pituitary gland), children and adolescents may present with neurological symptoms such as headaches, **vomiting**, and problems with vision. The patient may also have symptoms of double vision. Symptoms such as truly bizarre and excessive drinking behaviors (polydipsia) and sleep disturbances may be common.

Diagnosis

The primary symptom of pituitary dwarfism is lack of height. Therefore, a change in the individual's growth habits will help lead to a diagnosis. Another diagnostic technique uses an x ray of the child's hand to determine the child's bone age by comparing this to the child's actual chronological age. The bone age in affected children is usually two years or more behind the chronological age. This means that if a child is ten years old, his or her bones will look like they are those of an eight-year-old child. The levels of growth hormone and somatomedin C must also be measured with blood tests.

Hypopituitarism may be gained or acquired following birth for several reasons. It could be due to trauma to the pituitary gland such as a fall or following surgery to the brain for removal of a tumor. It may also be due to the child's environment (deprivational dwarfism).

On examination by the doctor there may be optic nerve atrophy, if the dwarfism is due to a type of tumor. X rays of the area where the pituitary gland is located (sella turcica) or more advanced imaging such as **magnetic resonance imaging (MRI)** or computed tomography CT may show changes of the pituitary gland itself. Computed tomography, is an advanced form of x ray that will help determine the integrity of the bone and how much calcification the tumor is producing. Magnetic resonance imaging will also help in the diagnosis. MRI is a type of imaging device that can visualize soft tissues such as muscle and fat.

If the dwarfism is due to environmental and emotional problems, the individual may be hospitalized to monitor hormone levels. Following a few days of hospitalization, hormone levels may become normal due to avoidance of the original environment.

Treatment

The main course of therapy is growth **hormone replacement therapy** when there is lack of growth hormone in the body. A pediatric endocrinologist, a doctor specializing in the hormones of children, usually administers this type of therapy before a child's growth plates have fused or joined together. Once the growth plates have fused, GH replacement therapy is rarely effective.

Growth hormone used to be collected from recently deceased humans. However, frequent disease complications resulting from human growth hormone collected from deceased bodies, lead to the banning of this method. In the mid-1980s, techniques were discovered that could produce growth hormones in the lab. Now, the only growth hormone used for treatment is that made in a laboratory.

A careful balancing of all of the hormones produced by the pituitary gland is necessary for patients with panhypopituitarism. This form of dwarfism is very difficult to manage.

Prognosis

The prognosis for each type of dwarfism varies. A panhypopituitarism dwarf does not pass through the initial onset of adult sexual development (**puberty**) and never produces enough gonadotropic hormones to develop adult sexual function. These individuals also have several other medical conditions. Dwarfism due to only growth hormone deficiency has a different prognosis. These individuals do pass through puberty and mature sexually, however, they remain proportionately small in stature.

If the individual is lacking only growth hormone then growth hormone replacement therapy can be administered. The success of treatment with growth hormone varies however. An increase in height of 4–6 in (10–15 cm) can occur in the first year of treatment. Following this first year, the response to the hormone is not as successful. Therefore the amount of growth hormone administered must be tripled to maintain this rate. Long-term use is considered successful if the individual grows at least 0.75 in (2 cm) per year more than they would without the hormone. However, if the growth hormone treatment is not administered before the long bones—such as the legs and arms—fuse, then the individual will never grow. This fusion is completed by adult age.

Improvement for individuals with dwarfism due to other causes such as a tumor, varies greatly. If the dwarfism is due to deprevalent causes, then

removing a child from that environment should help to alleviate the problem.

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ORGANIZATIONS

The Human Growth Foundation, 997 Glen Cove Ave., Suite 5, Glen Head, NY, 11545, (516) 671-4055, (800) 451-6434, <http://www.hgfound.org/>.

Little People of America, Inc., 250 El Camino Real, Suite 201, Tustin, CA, 92780, (714) 368-3689, (714) 368-3367, 888 LPA-2001 (572-2001), <http://www.lpaonline.org>.

Jason S. Schliesser, DC

Pituitary gland removal see **Hypophysectomy**

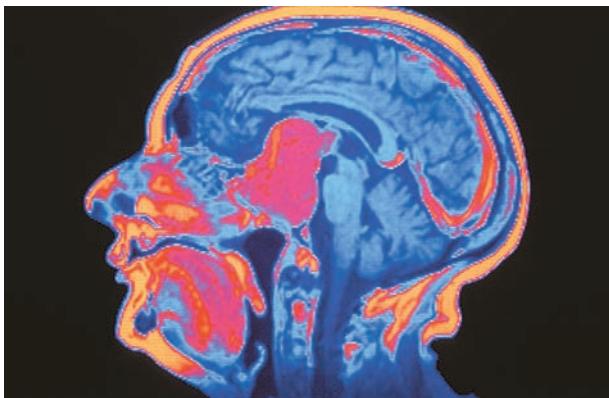
Pituitary tumors

Definition

Pituitary tumors are abnormal growths on the pituitary gland. Some tumors secrete hormones normally made by the pituitary gland.

Description

Located in the center of the brain, the pituitary gland manufactures and secretes hormones that regulate growth, sexual development and functioning, and the fluid balance of the body. About 10% of all cancers in the skull are pituitary tumors. Pituitary adenomas (adenomas are tumors that grow from gland tissues) and pituitary tumors in children and adolescents (craniopharyngiomas) are the most common types of



Colorized MRI showing large pituitary tumor at center in pink.
(Mehau Kulyk/Photo Researchers, Inc.)

pituitary tumors. They are usually benign and grow slowly. Even malignant pituitary tumors rarely spread to other parts of the body.

Pituitary adenomas do not secrete hormones but are likely to be larger and more invasive than tumors that do. Craniopharyngiomas are benign tumors that are extremely difficult to remove. Radiation does not stop them from spreading throughout the pituitary gland. Craniopharyngiomas account for less than 5% of all brain tumors. Pituitary tumors usually develop between the ages of 30 and 40, but half of all craniopharyngiomas occur in children, with symptoms most often appearing between the ages of five and ten.

Causes and symptoms

The cause of pituitary tumors is not known, but may be genetic. Symptoms related to tumor location, size, and pressure on neighboring structures include:

- persistent headache on one or both sides, or in the center of the forehead
- blurred or double vision; loss of peripheral vision
- drooping eyelid caused by pressure on nerves leading to the eye
- seizures

Symptoms related to hormonal imbalance include:

- excessive sweating
- loss of appetite
- loss of interest in sex
- inability to tolerate cold temperatures
- nausea
- high levels of sodium in the blood
- menstrual problems

- excessive thirst
- frequent urination
- dry skin
- constipation
- premature or delayed puberty
- delayed growth in children
- galactorrhea (milk secretion in the absence of pregnancy or breast feeding)
- low blood pressure
- low blood sugar

Diagnosis

As many as 40% of all pituitary tumors do not release excessive quantities of hormones into the blood. Known as clinically nonfunctioning, these tumors are difficult to distinguish from tumors that produce similar symptoms. They may grow to be quite large before they are diagnosed.

Endocrinologists and neuroendocrinologists base the diagnosis of pituitary tumors on:

- the patient's own observations and medical history
- physical examination
- laboratory studies of the patient's blood and cerebrospinal fluid
- x rays of the skull and other studies that provide images of the inside of the brain (CT, MRI)
- vision tests
- urinalysis

Treatment

Some pituitary tumors stabilize without treatment, but a neurosurgeon will operate at once to remove the tumor (adenectomy) or pituitary gland (**hypophysectomy**) of a patient whose vision is deteriorating rapidly. Patients who have pituitary apoplexy may experience very severe headaches, have symptoms of stiff neck, and sensitivity to light. This condition is considered an emergency. **Magnetic resonance imaging** (MRI) is the best imaging technique for patients with these symptoms. If the tumor is small, surgery may be done through the nose. If the tumor is large, it may require opening the skull for **tumor removal**. Selected patients do well with proton beam radiosurgery (the use of high energy particles in the form of a high energy beam to destroy an overactive gland).

Treatment is determined by the type of tumor and by whether it has invaded tissues adjacent to the pituitary gland. Hormone-secreting tumors can be successfully treated with surgery, radiation, bromocriptine (Parlodel), Sandostatin (Octreotide), or other

KEY TERMS

Agonist—A drug that increases the effectiveness of another drug.

Analogue—A drug that is similar to the drug from which it is derived.

somatostatin analogues (drugs similar to somatostatin). Surgery is usually used to remove all or part of a tumor within the gland or the area surrounding it, and may be combined with **radiation therapy** to treat tumors that extend beyond the pituitary gland. Removal of the pituitary gland requires life-long **hormone replacement therapy**.

Radiation therapy can provide long-term control of the disease if it recurs after surgery, and radioactive pellets can be implanted in the brain to treat craniopharyngiomas. CV205-502, a new dopamine agonist (a drug that increases the effect of another, in this instance dopamine) can control symptoms of patients who do not respond to bromocriptine.

Prognosis

Pituitary tumors are usually curable. Following surgery, adults may gradually resume their normal activities, and children may return to school when the effects of the operation have diminished, and appetite and sense of well-being have returned. Patients should wear medical identification tags identifying their condition and the hormonal replacement medicines they take.

ORGANIZATIONS

American Brain Tumor Association, 2720 River Road, Des Plaines, IL, 60018, (847) 827-9910, (847) 827-9918, (800) 886-2282, info@abta.org, <http://www.abta.org/>.
Pituitary Network Association, P.O. Box 1958, Thousand Oaks, CA, 91358, (805) 499-9973, (805) 480-0633, info@pituitary.org, <http://www.pituitary.org>.

Maureen Haggerty



The torso of a man covered with pityriasis rosea. The cause of this disorder is thought to be due to a viral infection. It often appears on the torso and upper parts of the limbs of young people and may be contagious. (Dr. P. Marazzi/SPL/Photo Researchers, Inc.)

followed later by a rash of colored spots on the body and upper arms.

Description

Pityriasis rosea is most common in young adults, and appears up to 50% more often in women. Its cause is unknown; however, some scientists believe that the rash is an immune response to some type of infection in the body.

Causes and symptoms

Doctors do not think that pityriasis rosea is contagious, but the cause is unknown. Some experts suspect the rash, which is most common in spring and fall, may be triggered by a virus, but no infectious agent has ever been found.

KEY TERMS

Antihistamines—A group of drugs that block the effects of histamine, a chemical released during an allergic reaction.

Steroids—A group of drugs that includes the corticosteroids, similar to hormones produced by the adrenal glands, and used to relieve inflammation and itching.

It is not sexually transmitted, and does not appear to be contagious from one person to the next.

Sometimes, before the symptoms appear, people experience preliminary sensations including **fever**, malaise, **sore throat**, or **headache**. Symptoms begin with a single, large round spot called a “herald patch” on the body, followed days or weeks later by slightly raised, scaly-edged round or oval pink-copper colored spots on the trunk and upper arms. The spots, which have a wrinkled center and a sharp border, sometimes resemble a Christmas tree. They may be mild to severely itchy, and they can spread to other parts of the body.

Diagnosis

A physician can diagnose the condition with blood tests, skin scrapings, or a biopsy of the lesion.

Treatment

The rash usually clears up on its own, although a physician should rule out other conditions that may cause a similar rash (such as **syphilis**).

Treatment includes external and internal medications for **itching** and inflammation. Mild inflammation and itching can be relieved with antihistamine drugs or calamine lotion, zinc oxide, or other mild lubricants or anti-itching creams. Gentle, soothing strokes should be used to apply the ointments, since vigorous rubbing may cause the lesions to spread. More severe itching and inflammation is treated with topical **steroids**. Moderate exposure to sun or ultraviolet light may help heal the lesions, but patients should avoid being sunburned.

Soap makes the rash more uncomfortable; patients should bathe or shower with plain lukewarm water, and apply a thin coating of bath oil to freshly-dried skin afterwards.

Prognosis

These spots, which may be itchy, last for 3–12 weeks. Symptoms rarely recur.

ORGANIZATIONS

American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168-4014, (847) 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

Carol A. Turkington

PKU see **Phenylketonuria**

Placenta previa

Definition

Placenta previa is a condition that occurs during **pregnancy** when the placenta is abnormally placed, and partially or totally covers the cervix.

Description

The uterus is the muscular organ that contains the developing baby during pregnancy. The lowest segment of the uterus is a narrowed portion called the cervix. The cervix has an opening (the os) that leads into the vagina, or birth canal. The placenta is the organ that attaches to the wall of the uterus during pregnancy. The placenta allows nutrients and oxygen from the mother’s blood circulation to pass into the developing baby (the fetus) via the umbilical cord.

During labor, the muscles of the uterus contract repeatedly. This allows the cervix to begin to grow thinner (called effacement) and more open (dilatation). Eventually, the cervix will become completely effaced and dilated, and the baby can leave the uterus and enter the birth canal. Under normal circumstances, the baby will emerge through the mother’s vagina during birth.

In placenta previa, the placenta develops in an abnormal location. Normally, the placenta should develop relatively high up in the uterus, on the front or back wall. In about one in 200 births, the placenta will be located low in the uterus, partially or totally covering the os. This causes particular problems in late pregnancy, when the lower part of the uterus begins to take on a new formation in preparation for delivery. As the cervix begins to efface and dilate, the attachments of the placenta to the uterus are damaged, resulting in bleeding.

Causes and symptoms

While the actual cause of placenta previa is unknown, certain factors increase the risk of a woman developing the condition. These factors include:

- having abnormalities of the uterus
- being older in age
- having had other babies
- having a prior delivery by cesarean section
- smoking cigarettes

When a pregnancy involves more than one baby (twins, triplets, etc.), the placenta will be considerably larger than for a single pregnancy. This also increases the chance of placenta previa.

Placenta previa may cause a number of problems. It is thought to be responsible for about 5% of all miscarriages. It frequently causes very light bleeding (spotting) early in pregnancy. Sometime after 28 weeks of pregnancy (most pregnancies last about 40 weeks), placenta previa can cause episodes of significant bleeding. Usually, the bleeding occurs suddenly and is bright red. The woman rarely experiences any accompanying **pain**, although about 10% of the time the placenta may begin separating from the uterine wall (called abruptio placentae), resulting in pain. The bleeding usually stops on its own. About 25% of such patients will go into labor within the next several days. Sometimes, placenta previa does not cause bleeding until labor has already begun.

Placenta previa puts both the mother and the fetus at high risk. The mother is at risk of severe and uncontrollable bleeding (hemorrhage), with dangerous blood loss. If the mother's bleeding is quite severe, this puts the fetus at risk of becoming oxygen deprived. The fetus' only source of oxygen is the mother's blood. The mother's blood loss, coupled with certain changes that take place in response to that blood loss, decreases the amount of blood going to the placenta, and ultimately to the fetus. Furthermore, placenta previa increases the risk of preterm labor, and the possibility that the baby will be delivered prematurely.

Diagnosis

Diagnosis of placenta previa is suspected whenever bright red, painless vaginal bleeding occurs during the course of a pregnancy. The diagnosis can be confirmed by performing an ultrasound examination. This will allow the location of the placenta to be evaluated.

While many conditions during pregnancy require a pelvic examination, in which the health care

KEY TERMS

Cesarean section—Delivery of a baby through an incision in the mother's abdomen instead of through the vagina.

Labor—The process during which the uterus contracts, and the cervix opens to allow the passage of a baby into the vagina.

Placenta—The organ that provides oxygen and nutrition from the mother to the baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the baby via the umbilical cord.

Umbilical cord—The blood vessels that allow the developing baby to receive nutrition and oxygen from its mother; the blood vessels also eliminate the baby's waste products. One end of the umbilical cord is attached to the placenta and the other end is attached to the baby's belly button (umbilicus).

Vagina—The birth canal; the passage from the cervix of the uterus to the opening leading outside of a woman's body.

provider's fingers are inserted into the patient's vagina, such an examination should never be performed if there is any suspicion of placenta previa. Such an examination can disturb the already susceptible placenta, resulting in hemorrhage.

Sometimes placenta previa is found early in a pregnancy, during an ultrasound examination performed for another reason. In these cases, it is wise to have a repeat ultrasound performed later in pregnancy (during the last third of the pregnancy, called the third trimester). A large percentage of these women will have a low-lying placenta, but not a true placenta previa where some or all of the os is covered.

Treatment

Treatment depends on how far along in the pregnancy the bleeding occurs. When the pregnancy is less than 36 weeks along, the fetus is not sufficiently developed to allow delivery without a high risk of complications. Therefore, a woman with placenta previa is treated with bed rest, blood transfusions as necessary, and medications to prevent labor. After 36 weeks, the baby can be delivered via **cesarean section**. This is almost always the preferred method of delivery in order to avoid further bleeding from the low-lying placenta.

Prognosis

In cases of placenta previa, the prognosis for the mother is very good. However, there is a 15–20% chance the infant will not survive. This is 10 times the **death** rate associated with normal pregnancies. About 60% of these deaths occur because the baby delivered was too premature to survive.

Prevention

There are no known ways to insure the appropriate placement of the placenta in the uterus. However, careful treatment of the problem can result in the best chance for a good outcome for both mother and baby.

ORGANIZATIONS

American College of Obstetricians and Gynecologists (ACOG), PO Box 96920, Washington, DC, 20090-6920, (202) 638-5577, <http://www.acog.org>.

Rosalyne Carson-DeWitt, MD

Placental abruption

Definition

Placental abruption occurs when the placenta separates from the wall of the uterus prior to the birth of the baby. This can result in severe, uncontrollable bleeding (hemorrhage).

Description

The uterus is the muscular organ that contains the developing baby during **pregnancy**. The lowest segment of the uterus is a narrowed portion called the cervix. The cervix has an opening (the os) that leads into the vagina, or birth canal. The placenta is the organ that attaches to the wall of the uterus during pregnancy. The placenta allows nutrients and oxygen from the mother's blood circulation to pass into the developing baby (the fetus) via the umbilical cord.

During labor, the muscles of the uterus contract repeatedly. This allows the cervix to begin to grow thinner (called effacement) and more open (dilatation). Eventually, the cervix will become completely effaced and dilated, and the baby can leave the uterus and enter the birth canal. Under normal circumstances, the baby will go through the mother's vagina during birth.

During a normal labor and delivery, the baby is born first. Several minutes to 30 minutes later, the placenta separates from the wall of the uterus and is delivered. This sequence is necessary because the baby relies on the placenta to provide oxygen until he or she begins to breathe independently.

Placental abruption occurs when the placenta separates from the uterus before the birth of the baby. Placental abruption occurs in about one out of every 200 deliveries. African-American and Latin-American women have a greater risk of this complication than do Caucasian women. It was once believed that the risk of placental abruption increased in women who gave birth to many children, but this association is still being researched.

Causes and symptoms

The cause of placental abruption is unknown. However, a number of risk factors have been identified. These factors include:

- older age of the mother
- history of placental abruption during a previous pregnancy
- high blood pressure
- certain disease states (diabetes, collagen vascular diseases)
- the presence of a type of uterine tumor called a leiomyoma
- twins, triplets, or other multiple pregnancies
- cigarette smoking
- heavy alcohol use
- cocaine use
- malformations of the uterus
- malformations of the placenta
- injury to the abdomen (as might occur in a car accident)

Symptoms of placental abruption include bleeding from the vagina, severe **pain** in the abdomen or back, and tenderness of the uterus. Depending on the severity of the bleeding, the mother may experience a drop in blood pressure, followed by symptoms of organ failure as her organs are deprived of oxygen. Sometimes, there is no visible vaginal bleeding. Instead, the bleeding is said to be "concealed." In this case, the bleeding is trapped behind the placenta, or there may be bleeding into the muscle of the uterus. Many patients will have abnormal contractions of the uterus, particularly extremely hard, prolonged contractions. Placental abruption can be

total (in which case the fetus will almost always die in the uterus), or partial.

Placental abruption can also cause a very serious complication called consumptive coagulopathy. A series of reactions begin that involve the elements of the blood responsible for clotting. These clotting elements are bound together and used up by these reactions. This increases the risk of uncontrollable bleeding and may contribute to severe bleeding from the uterus, as well as causing bleeding from other locations (nose, urinary tract, etc.).

Placental abruption is risky for both the mother and the fetus. It is dangerous for the mother because of blood loss, loss of clotting ability, and oxygen deprivation to her organs (especially the kidneys and heart). This condition is dangerous for the fetus because of oxygen deprivation, too, since the mother's blood is the fetus' only source of oxygen. Because the abrupting placenta is attached to the umbilical cord, and the umbilical cord is an extension of the fetus' circulatory system, the fetus is also at risk of hemorrhaging. The fetus may die from these stresses, or may be born with damage due to oxygen deprivation. If the abruption occurs well before the baby was due to be delivered, early delivery may cause the baby to suffer complications of premature birth.

Diagnosis

Diagnosis of placental abruption relies heavily on the patient's report of her symptoms and a **physical examination** performed by a health care provider. Ultrasound can sometimes be used to diagnose an abruption, but there is a high rate of missed or incorrect diagnoses associated with this tool when used for this purpose. Blood will be taken from the mother and tested to evaluate the possibility of life-threatening problems with the mother's clotting system.

Treatment

The first line of treatment for placental abruption involves replacing the mother's lost blood with blood transfusions and fluids given through a needle in a vein. Oxygen will be administered, usually by a mask or through tubes leading to the nose. When the placental separation is severe, treatment may require prompt delivery of the baby. However, delivery may be delayed when the placental separation is not as severe, and when the fetus is too immature to insure a healthy baby if delivered. The baby is delivered vaginally when possible. However, a

KEY TERMS

Cesarean section—Delivery of a baby through an incision in the mother's abdomen, instead of through the vagina.

Labor—The process during which the uterus contracts, and the cervix opens to allow the passage of a baby into the vagina.

Placenta—The organ that provides oxygen and nutrition from the mother to the baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the baby via the umbilical cord.

Umbilical cord—The blood vessels that allow the developing baby to receive nutrition and oxygen from its mother; the blood vessels also eliminate the baby's waste products. One end of the umbilical cord is attached to the placenta and the other end is attached to the baby's belly button (umbilicus).

Uterus—The muscular organ that contains the developing baby during pregnancy.

Vagina—The birth canal; the passage from the cervix of the uterus to the opening leading outside of a woman's body.

Cesarean section may be performed to deliver the baby more quickly if the abruption is quite severe or if the baby is in distress.

Prognosis

The prognosis for cases of placental abruption varies, depending on the severity of the abruption. The risk of **death** for the mother ranges up to 5%, usually due to severe blood loss, **heart failure**, and kidney failure. In cases of severe abruption, 50–80% of all fetuses die. Among those who survive, nearly half will have lifelong problems due to oxygen deprivation in the uterus and premature birth.

Prevention

Some of the causes of placental abruption are preventable. These include cigarette **smoking**, alcohol **abuse**, and **cocaine** use. Other causes of abruption may not be avoidable, like diabetes or high blood pressure. These diseases should be carefully treated. Patients with conditions known to increase the risk of placental abruption should be carefully monitored for signs and symptoms of this complication.

ORGANIZATIONS

American College of Obstetricians and Gynecologists (ACOG), PO Box 96920, Washington, DC, 20090-6920, (202) 638-5577, <http://www.acog.org>.

Rosalyn Carson-DeWitt, MD

Plague

Definition

Plague is a serious, potentially life-threatening **infectious disease** that is usually transmitted to humans by the **bites** of rodent fleas. It was one of the scourges of early human history. There are three major forms of the disease: bubonic, septicemic, and pneumonic.

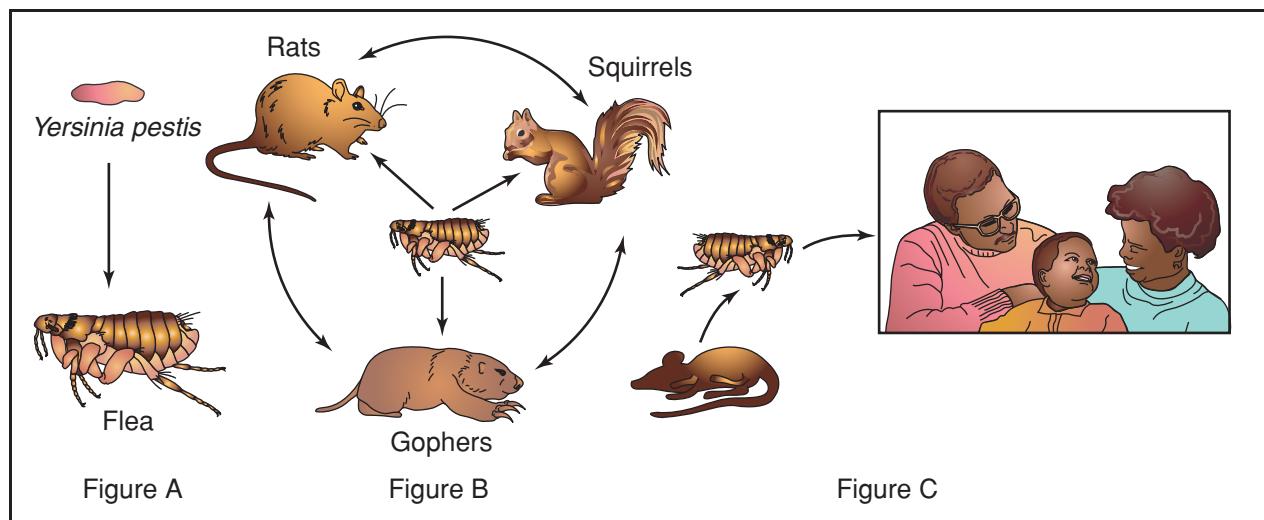
Description

Plague has been responsible for three great world pandemics, which caused millions of deaths and significantly altered the course of history. A pandemic is a disease occurring in epidemic form throughout the entire population of a country, a people, or the world. Although the cause of the plague was not identified until the third pandemic in 1894, scientists are virtually certain that the first two pandemics were plague because a number of the survivors wrote about their experiences and described the symptoms.

The first great pandemic appeared in AD 542 and lasted for 60 years. It killed millions of citizens, particularly people living along the Mediterranean Sea. This sea was the busiest coastal trade route at that time and connected what is now southern Europe, northern Africa, and parts of coastal Asia. This pandemic is sometimes referred to as the Plague of Justinian, named for the great emperor of Byzantium who was ruling at the beginning of the outbreak. According to the historian Procopius, this outbreak of plague killed 10,000 people per day at its height just within the city of Constantinople.

The second pandemic occurred during the fourteenth century, and was called the Black **Death** because its main symptom was the appearance of black patches (caused by bleeding) on the skin. It was also a subject found in many European paintings, drawings, plays, and writings of that time. The connections between large active trading ports, rats coming off the ships, and the severe outbreaks of the plague were understood by people at the time. This was the most severe of the three, beginning in the mid-1300s with an origin in central Asia and lasting for 400 years. Between a fourth and a third of the entire European population died within a few years after plague was first introduced. Some smaller villages and towns were completely wiped out.

The final pandemic began in northern China, reaching Canton and Hong Kong by 1894. From there, it spread to all continents, killing millions.



Plague is a serious infectious disease transmitted by the bites of rat fleas. There are three major forms of plague: bubonic, pneumonic, and septicemic. As illustrated above, fleas carry the bacterium *Yersinia pestis*. When a flea bites an infected rodent, it becomes a vector and then passes the plague bacteria when it bites a human. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

The great pandemics of the past occurred when wild rodents spread the disease to rats in cities, and then to humans when the rats died. Another route for infection came from rats coming off ships that had traveled from heavily infected areas. Generally, these were busy coastal or inland trade routes. Plague was introduced into the United States during this pandemic and it spread from the West towards the Midwest and became endemic in the Southwest of the United States.

About 10–15 Americans living in the southwestern United States contract plague each year during the spring and summer. The last rat-borne epidemic in the United States occurred in Los Angeles in 1924–25. Since then, all plague cases in this country have been sporadic, acquired from wild rodents or their fleas. Plague can also be acquired from ground squirrels and prairie dogs in parts of Arizona, New Mexico, California, Colorado, and Nevada. Around the world, there are between 1,000 and 2,000 cases of plague each year. Recent outbreaks in humans occurred in Africa, South America, and Southeast Asia.

Some people and/or animals with bubonic plague go on to develop **pneumonia** (pneumonic plague). This can spread to others via infected droplets during coughing or sneezing.

Plague is one of three diseases still subject to international health regulations. These rules require that all confirmed cases be reported to the World Health Organization (WHO) within 24 hours of diagnosis. According to the regulations, passengers on an international voyage who have been to an area where there is an epidemic of pneumonic plague must be placed in **isolation** for six days before being allowed to leave.

While plague is found in several countries, there is little risk to United States travelers within endemic areas (limited locales where a disease is known to be present) if they restrict their travel to urban areas with modern hotel accommodations.

Over the past few years, this infection primarily of antiquity has become a modern issue. This change has occurred because of the concerns about the use of plague as a weapon of biological warfare or terrorism (bioterrorism). Along with **anthrax** and **smallpox**, plague is considered to be a significant risk. In this scenario, the primary manifestation is likely to be pneumonic plague transmitted by clandestine aerosols. It has been reported that during World War II

the Japanese dropped “bombs” containing plague-infected fleas in China as a form of biowarfare.

Causes and symptoms

Fleas carry the bacterium *Yersinia pestis*, formerly known as *Pasteurella pestis*. The plague bacillus can be stained with Giemsa stain and typically looks like a safety pin under the microscope. When a flea bites an infected rodent, it swallows the plague bacteria. The bacteria are passed on when the fleas, in turn, bite a human. Interestingly, the plague bacterium grows in the gullet of the flea, obstructing it and not allowing the flea to eat. Transmission occurs during abortive feeding with regurgitation of bacteria into the feeding site. Humans also may become infected if they have a break or cut in the skin and come in direct contact with body fluids or tissues of infected animals.

More than 100 species of fleas have been reported to be naturally infected with plague; in the western United States, the most common source of plague is the golden-mantled ground squirrel flea. Chipmunks and prairie dogs have also been identified as hosts of infected fleas.

Since 1924, there have been no documented cases in the United States of human-to-human spread of plague from droplets. All but one of the few pneumonic cases have been associated with handling infected cats. While dogs and cats can become infected, dogs rarely show signs of illness and are not believed to spread disease to humans. However, plague has been spread from infected coyotes (wild dogs) to humans. In parts of central Asia, gerbils have been identified as the source of cases of bubonic plague in humans.

Bubonic plague

Two to five days after infection, patients experience a sudden **fever**, chills, seizures, and severe headaches, followed by the appearance of swellings or “buboies” in armpits, groin, and neck. The most commonly affected sites are the lymph glands near the site of the first infection. As the bacteria multiply in the glands, the lymph node becomes swollen. As the nodes collect fluid, they become extremely tender. Occasionally, the bacteria will cause an ulcer at the point of the first infection.

Septicemic plague

Bacteria that invade the bloodstream directly (without involving the lymph nodes) cause septicemic plague. (Bubonic plague also can progress to septicemic plague if not treated appropriately.) Septicemic

plague that does not involve the lymph glands is particularly dangerous because it can be hard to diagnose the disease. The bacteria usually spread to other sites, including the liver, kidneys, spleen, lungs, and sometimes the eyes, or the lining of the brain. Symptoms include fever, chills, prostration, abdominal **pain**, **shock**, and bleeding into the skin and organs.

Pneumonic plague

Pneumonic plague may occur as a direct infection (primary) or as a result of untreated bubonic or septicemic plague (secondary). Primary pneumonic plague is caused by inhaling infective drops from another person or animal with pneumonic plague. Symptoms, which appear within one to three days after infection, include a severe, overwhelming pneumonia, with **shortness of breath**, high fever, and blood in the phlegm. If untreated, half the patients will die; if blood poisoning occurs as an early complication, patients may die even before the buboes appear.

Life-threatening complications of plague include shock, high fever, problems with blood clotting, and convulsions.

Diagnosis

Plague should be suspected if there are painful buboes, fever, exhaustion, and a history of possible exposure to rodents, rabbits, or fleas in the West or Southwest. The patient should be isolated. Chest x rays are taken, as well as blood cultures, antigen testing, and examination of lymph node specimens. Blood cultures should be taken 30 minutes apart, before treatment.

A group of German researchers reported in 2004 on a standardized enzyme-linked immunosorbent assay (ELISA) kit for the rapid diagnosis of plague. The test kit was developed by the German military and has a high degree of accuracy as well as speed in identifying the plague bacillus. The kit could be useful in the event of a bioterrorist attack as well as in countries without advanced microbiology laboratories.

Treatment

As soon as plague is suspected, the patient should be isolated, and local and state departments notified. Drug treatment reduces the risk of death to less than 5%. The preferred treatment is streptomycin administered as soon as possible. Alternatives include gentamicin, chloramphenicol, tetracycline, or trimethoprim/sulfamethoxazole.

KEY TERMS

Bioterrorism—The use of disease agents to terrorize or intimidate a civilian population.

Buboes—Smooth, oval, reddened, and very painful swellings in the armpits, groin, or neck that occur as a result of infection with the plague.

Endemic—A disease that occurs naturally in a geographic area or population group.

Epidemic—A disease that occurs throughout part of the population of a country.

Pandemic—A disease that occurs throughout a regional group, the population of a country, or the world.

Septicemia—The medical term for blood poisoning, in which bacteria have invaded the bloodstream and circulates throughout the body.

Prognosis

Plague can be treated successfully if it is caught early; the mortality rate for treated disease is 1–15% but 40–60% in untreated cases. Untreated pneumonic plague is almost always fatal, however, and the chances of survival are very low unless specific antibiotic treatment is started within 15–18 hours after symptoms appear. The presence of plague bacteria in a blood smear is a grave sign and indicates septicemic plague. Septicemic plague has a mortality rate of 40% in treated cases and 100% in untreated cases.

Prevention

Anyone who has come in contact with a plague pneumonia victim should be given **antibiotics**, since untreated pneumonic plague patients can pass on their illness to close contacts throughout the course of the illness. All plague patients should be isolated for 48 hours after antibiotic treatment begins. Pneumonic plague patients should be completely isolated until sputum cultures show no sign of infection.

Residents of areas where plague is found should keep rodents out of their homes. Anyone working in a rodent-infested area should wear insect repellent on skin and clothing. Pets can be treated with insecticidal dust and kept indoors. Handling sick or dead animals (especially rodents and cats) should be avoided.

Plague vaccines have been used with varying effectiveness since the late nineteenth century. Experts

believe that **vaccination** lowers the chance of infection and the severity of the disease. However, the effectiveness of the vaccine against pneumonic plague is not clearly known.

Vaccinations against plague are not required to enter any country. Because immunization requires multiple doses over a 6–10 month period, plague vaccine is not recommended for quick protection during outbreaks. Moreover, its unpleasant side effects make it a poor choice unless there is a substantial long-term risk of infection. The safety of the vaccine for those under age 18 has not been established. Pregnant women should not be vaccinated unless the need for protection is greater than the risk to the unborn child. Even those who receive the vaccine may not be completely protected. The inadequacy of the vaccines available as of the early 2000s explains why it is important to protect against rodents, fleas, and people with plague. A team of researchers in the United Kingdom reported in the summer of 2004 that an injected subunit vaccine is likely to offer the best protection against both bubonic and pneumonic forms of plague.

Resources

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Jones, Abby, Catharine Bosio, Angela Duffy, Andrew Goodyear, Martin Schriefer, Steven Dow. *Vaccine* (August 16, 2010): 5924–5929.

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Infectious Diseases Weblink. <http://webpages.charter.net/deziel/>.

International Society of Travel Medicine. <http://www.istm.org>.
World Health Organization. <http://www.who.ch/>.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

National Institute of Allergies and Infectious Diseases, 6610 Rockledge Drive, MSC 6612, Bethesda, MD, 20892-6612, (301) 496-5717, (301) 402-3573, (866)

284-4107, ocpostoffice@niaid.nih.gov, <http://www.niaid.nih.gov>.

World Health Organization (WHO), Avenue Appia 201211, Geneva, Switzerland, 27, 4122791-2111, info@who.int, <http://www.who.int>.

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Rebecca J. Frey, PhD

Plaque see **Skin lesions**

Plasma cell myeloma see **Multiple myeloma**

Plasma renin activity

Definition

Renin is an enzyme released by the kidney to help control the body's sodium-potassium balance, fluid volume, and blood pressure.

Purpose

Plasma renin activity (PRA), also called plasma renin assay, may be used to screen for high blood pressure (**hypertension**) of kidney origin, and may help plan treatment of essential hypertension, a genetic disease often aggravated by excess **sodium** intake. PRA is also used to further evaluate a diagnosis of excess aldosterone, a hormone secreted by the adrenal cortex, in a condition called Conn's syndrome.

Precautions

Patients taking **diuretics**, antihypertensives, **vasodilators**, **oral contraceptives**, and licorice should discontinue use of these substances for two to four weeks before the test. It should be noted that renin is increased in **pregnancy** and in **diets** with reduced salt intake. Also, since renin is affected by body position, as well as by diurnal (daily) variation, blood samples should be drawn in the morning, and the position of the patient (sitting or lying down) should be noted.

Description

When the kidneys release the enzyme renin in response to certain conditions (high blood potassium, low blood sodium, decreased blood volume), it is the first step in what is called the renin-angiotensin-aldosterone cycle. This cycle includes the conversion of angiotensinogen to angiotensin I, which in turn is

KEY TERMS

Aldosteronism—A disorder caused by excessive production of the hormone aldosterone, which is produced by a part of the adrenal glands called the adrenal cortex. Causes include a tumor of the adrenal gland (Conn's syndrome), or a disorder reducing the blood flow through the kidney. This leads to overproduction of renin and angiotensin, and in turn causes excessive aldosterone production.

Symptoms include hypertension, impaired kidney function, thirst and muscle weakness.

Conn's syndrome—A disorder caused by excessive aldosterone secretion by a benign tumor of one of the adrenal glands. This results in malfunction of the body's salt and water balance and subsequently causes hypertension. Symptoms include thirst, muscle weakness, and excessive urination.

converted to angiotensin II, in the lung. Angiotensin II is a powerful blood vessel constrictor, and its action stimulates the release of aldosterone from an area of the adrenal glands called the adrenal cortex. Together, angiotensin and aldosterone increase the blood volume, the blood pressure, and the blood sodium to re-establish the body's sodium-potassium and fluid volume balance. Primary aldosteronism, the symptoms of which include hypertension and low blood potassium (**hypokalemia**), is considered "low-renin aldosteronism."

Renin itself is not actually measured in the PRA test, because renin can be measured only with great difficulty even in research laboratories. In the most commonly used renin assay, the test actually determines, by a procedure called radioimmunoassay, the rate of angiotensin I generation per unit time, while the PRC (plasma renin concentration) measures the maximum renin effect.

Both the PRA and the PRC are extremely difficult to perform. Not only is renin itself unstable, but the patient's body position and the time of day affect the results. Also, the sample must be collected properly: drawn into a chilled syringe and collection tube, placed on ice, and sent to the performing laboratory immediately. Even if all these procedures are followed, results can vary significantly.

A determination of the PRA and a measurement of the plasma aldosterone level are used in the differential diagnosis of primary and secondary **hyperaldosteronism**. Patients with primary hyperaldosteronism (caused by an adrenal tumor that overproduces aldosterone) will have an increased aldosterone level with decreased renin activity. Conversely, patients with secondary hyperaldosteronism (caused by certain types of **kidney disease**) will have increased levels of renin.

Renin stimulation test

The renin stimulation test is performed to help diagnose and distinguish the two forms of hyperaldosteronism. With the patient having been on a low-salt diet and lying down for the test, a blood sample for PRA is obtained. The PRA is repeated with the patient still on the low salt diet but now standing upright. In cases of primary hyperaldosteronism, the blood volume is greatly expanded, and a change in position or reduced salt intake does not result in decreased kidney blood flow or decreased blood sodium. As a result, renin levels do not increase. However, in secondary hyperaldosteronism, blood sodium levels decrease with a lowered salt intake, and when the patient is standing upright, the kidney blood flow decreases as well. Consequently, renin levels do increase.

Captopril test

The captopril test is a screening test for hypertension of kidney origin (**renovascular hypertension**). For this test, a baseline PRA test is done first, then the patient receives an oral dose of captopril, which is an angiotensin-converting enzyme (ACE) inhibitor. Blood pressure measurements are taken at this time and again at 60 minutes when another PRA test is done. Patients with kidney-based hypertension demonstrate greater falls in blood pressure and increases in PRA after captopril administration than do those with essential hypertension. Consequently, the captopril test is an excellent screening procedure to determine the need for a more invasive radiographic evaluation such as renal arteriography.

Preparation

This test requires a blood sample. For the PRA, the patient should maintain a normal diet with a restricted amount of sodium (approximately 3 g per day) for three days before the test. It is recommended

that the patient be **fasting** (nothing to eat or drink) from midnight the day of the test.

Risks

Risks for this test are minimal, but may include slight bleeding from the puncture site, **fainting** or feeling lightheaded after venipuncture, or hematoma (blood accumulating under the puncture site).

Normal results

Reference values for the PRA test are laboratory-specific and depend upon the kind of diet (sodium restricted or normal), the age of the patient, and the patient's posture at the time of the test. Values are also affected if renin has been stimulated or if the patient has received an ACE inhibitor, like captopril.

Abnormal results

Increased PRA levels are seen in essential hypertension (uncommon), malignant hypertension, and kidney-based (renovascular) hypertension. Renin-producing renal tumors, while rare, can also cause elevated levels, as can **cirrhosis**, low blood volume due to hemorrhage, and diminished adrenal function (**Addison's disease**). Decreased renin levels may indicate increased blood volume due to a high-sodium diet, salt-retaining **steroids**, primary aldosteronism, licorice ingestion syndrome, or essential hypertension with low renin levels.

Resources

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Janis O. Flores

Plasmapheresis

Definition

Plasmapheresis is a blood purification procedure used to treat several autoimmune diseases. It is also known as therapeutic plasma exchange.

Purpose

In an autoimmune disease, the immune system attacks the body's own tissues. In many autoimmune diseases, the chief weapons of attack are antibodies,

proteins that circulate in the bloodstream until they meet and bind with the target tissue. Once bound, they impair the functions of the target, and signal other immune components to respond as well.

Plasmapheresis is used to remove antibodies from the bloodstream, thereby preventing them from attacking their targets. It does not directly affect the immune system's ability to make more antibodies, and therefore may only offer temporary benefit. This procedure is most useful in acute, self-limited disorders such as **Guillain-Barré syndrome**, or when chronic disorders, such as **myasthenia gravis**, become more severe in symptoms. In these instances, a rapid improvement could save the patient's life. Neurologic diseases comprise 90% of the diseases that could profit from plasmapheresis.

Precautions

Patients with clotting disorders may not be suitable candidates for plasmapheresis.

Description

The basic procedure consists of removal of blood, separation of blood cells from plasma, and return of these blood cells to the body's circulation, diluted with fresh plasma or a substitute. Because of concerns over viral infection and allergic reaction, fresh plasma is not routinely used. Instead, the most common substitute is saline solution with sterilized human albumin protein. During the course of a single session, two to three liters of plasma is removed and replaced.

Plasmapheresis requires insertion of a venous catheter, either in a limb or central vein. Central veins allow higher flow rates and are more convenient for repeat procedures, but are more often the site of complications, especially bacterial infection.

When blood is outside the body, it must be treated to prevent it from clotting. While most of the anticoagulant agent is removed from the blood during treatment, some is returned to the patient.

Three procedures are available:

- “Discontinuous flow centrifugation.” Only one venous catheter line is required. Approximately 300 mL of blood is removed at a time and centrifuged to separate plasma from blood cells.
- “Continuous flow centrifugation.” Two venous lines are used. This method requires slightly less blood volume to be out of the body at any one time.
- “Plasma filtration.” Two venous lines are used. The plasma is filtered using standard hemodialysis equipment.

KEY TERMS

Anaphylaxis—Also called anaphylactic shock, it is a severe allergic reaction to a foreign substance that the patient has had contact with. Penicillin is an example of a substance that causes severe allergic reactions for some people.

Antibody—Chemicals produced by the body to defend it against bacteria, viruses, or other cells foreign to the body (antigens). Each specific antibody reacts against a specific foreign body. Antibodies are also termed immunoglobulins.

Autoimmune—Autoimmune refers to the body's development of intolerance of the antigens on its own cells.

Hemodialysis—A method to take out unwanted parts of the blood. The patient's blood is run through a catheter and tubing into a machine called a dialyzer, which filters out the unwanted blood component.

Plasma—Plasma makes up 50% of human blood. It is a watery fluid that carries red cells, white cells, and platelets throughout the body.

It requires less than 100 mL of blood to be outside the body at one time.

A single plasmapheresis session may be effective, although it is more common to have several sessions per week over the course of two weeks or more.

Preparation

Good **nutrition** and plenty of rest make the procedure less stressful. The treating physician determines which of the patient's medications should be discontinued before the plasmapheresis session.

Aftercare

The patient may experience **dizziness**, **nausea**, **numbness**, **tingling**, or lightheadedness during or after the procedure. These effects usually pass quickly, allowing the patient to return to normal activities the same day.

Risks

Reinfusion (replacement) with human plasma may cause **anaphylaxis**, a life threatening allergic reaction. All procedures may cause a mild allergic reaction, leading to **fever**, chills, and rash. Bacterial infection is a risk, especially when a central venous catheter is used. Reaction to the citrate anticoagulant used may cause cramps and numbness, though these usually resolve on their own. Patients with impaired kidney function may require drug treatment for the effects of citrate metabolism.

Plasma contains clotting agents, chemicals that allow the blood to coagulate into a solid clot. Plasma exchange removes these. Bleeding complications are rare following plasmapheresis, but may require replacement of clotting factors.

Normal results

Plasmapheresis is an effective temporary treatment for:

- Guillain-Barré syndrome (an acute neurological disorder following a viral infection that produces progressive muscle weakness and paralysis)
- Myasthenia gravis (an autoimmune disease that causes muscle weakness)
- chronic inflammatory demyelinating polyneuropathy (a chronic neurological disorder caused by destruction of the myelin sheath of peripheral nerves, which produces symptoms similar to Guillain-Barré syndrome)
- thrombotic thrombocytopenic purpura (a rare blood disorder)
- Paraproteinemic peripheral neuropathies (a neurological disorder affecting the peripheral nerves)
- blood that is too thick (hyperviscosity)

Other conditions may respond to plasmapheresis as well. Beneficial effects are usually seen within several days. Effects commonly last up to several months, although longer-lasting changes are possible, presumably by inducing shifts in immune response.

Resources

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Richard Robinson

Plasmodium infection see **Malaria**

Plastic, reconstructive, and cosmetic surgery

Definition

Plastic, reconstructive, and cosmetic surgery procedures are a variety of operations performed in order to repair or restore body parts to look normal, or to change a body part to look better. These types of surgery are highly specialized. They are characterized by careful preparation of a person's skin and tissues, by precise cutting and suturing techniques, and by care taken to minimize scarring. Recent advances in the development of miniaturized instruments, new materials for artificial limbs and body parts, and improved surgical techniques have expanded the range of plastic surgery procedures that can be performed.

Purpose

Although these three types of surgery share some common techniques and approaches, they have somewhat different emphases. Plastic surgery is usually performed to treat **birth defects** and to remove skin blemishes such as **warts**, **acne scars**, or **birthmarks**.

Cosmetic surgery procedures are performed to make persons look younger or enhance their appearance in other ways. Reconstructive surgery is used to reattach body parts severed in combat or accidents, to perform skin grafts after severe **burns**, or to reconstruct parts of person's body that were missing at birth or removed by surgery. Reconstructive surgery is the oldest form of plastic surgery, having developed out of the need to treat wounded soldiers in wartime.

Demographics

The top 10 most commonly performed elective cosmetic surgeries in the United States include the following:

- liposuction
- breast augmentation
- eyelid surgery
- face lift
- tummy tuck
- collagen injections
- chemical peel
- laser skin resurfacing



A patient undergoing abdominoplasty. (Photo Researchers, Inc.)

Top elective cosmetic surgeries and procedures in the United States

Surgical procedures	Nonsurgical procedures
Breast augmentation (311,957)	Botulinum toxin type A injection (2,557,068)
Liposuction (283,785)	Hyaluronic acid injection (1,313,038)
Cosmetic eyelid surgery (149,943)	Laser hair removal (1,280,031)
Nose reshaping (138,258)	Microdermabrasion (621,943)
Tummy tuck (127,923)	Chemical peel (529,285)
Breast reduction (113,511)	Laser skin resurfacing (512,318)
Breast lift (98,279)	Sclerotherapy (452,924)
Facelift (94,247)	IPL Laser treatment (452,210)
Forehead lift (30,789)	Noninvasive tightening (275,119)
Cosmetic ear surgery (21,817)	Laser treatment of leg veins (119,939)

SOURCE: The American Society for Aesthetic Plastic Surgery (ASAPS), 2009 ASAPS Statistics. Available online at: <http://www.surgery.org/media/statistics> (accessed June 9, 2010).

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

- rhinoplasty
- forehead lift

There were approximately 31 million surgical procedures performed in the United States in 2006. Because many plastic and reconstructive surgical procedures are performed in private professional offices or as outpatient procedures, accurate statistics concerning the number of procedures performed are not available.

Description

Plastic surgery

Plastic surgery includes a number of different procedures that usually involve skin. Operations to remove excess fat from the abdomen ("tummy tucks"), dermabrasion to remove acne scars or **tattoos**, and reshaping the cartilage in children's ears (otoplasty) are common applications of plastic surgery.

Cosmetic surgery

Most cosmetic surgery is done on the face. It is intended either to correct disfigurement or to enhance a person's features. The most common cosmetic procedure for children is correction of a **cleft lip** or palate. In adults, the most common procedures are remodeling of the nose (**rhinoplasty**), removal of baggy skin around the eyelids (**blepharoplasty**), face lifts (rhytidectomy), or changing the size or shape of the breasts (mammoplasty). Although many people still think of cosmetic surgery as only for women, growing numbers

of men are choosing to have face lifts and eyelid surgery, as well as hair transplants and "tummy tucks."

Reconstructive surgery

Reconstructive surgery is often performed on burn and accident victims. It may involve the rebuilding of severely fractured bones, as well as **skin grafting**. Reconstructive surgery includes such procedures as the reattachment of an amputated finger or toe, or implanting a prosthesis. Prostheses are artificial structures and materials that are used to replace missing limbs or teeth, or arthritic hip and knee joints.

Diagnosis/Preparation

General preparation

Preparation for nonemergency plastic or reconstructive surgery includes individual education, as well as medical considerations. Some operations, such as nose reshaping or the removal of warts, small birthmarks, and tattoos can be done as outpatient procedures under **local anesthesia**. Most plastic and reconstructive surgery, however, involves a stay in the hospital and **general anesthesia**.

Medical preparation

Preparation for plastic surgery includes the surgeon's detailed assessment of the parts of an individual's body that will be involved. Skin grafts require evaluating suitable areas of skin for the right color and texture to match the skin at the graft site. Face lifts and cosmetic surgery in the eye area require very close attention to the texture of the skin and the placement of surgical cuts (incisions).

Persons scheduled for plastic surgery under general anesthesia will be given a **physical examination**, blood and urine tests, and other tests to make sure that they do not have any previously undetected health problems or blood clotting disorders. The surgeon will check the list of prescription medications that the prospective patient may be taking to make sure that none of them will interfere with normal blood clotting or interact with the anesthetic.

Individuals are asked to avoid using **aspirin** or medications containing aspirin for a week to two weeks before surgery, because these drugs lengthen the time of blood clotting. Smokers are asked to stop **smoking** two weeks before surgery because smoking interferes with the healing process. For some types of plastic surgery, individuals may be asked to donate several units of their own blood before the procedure, in case a **transfusion** is needed during the operation.

KEY TERMS

Blepharoplasty—Surgical reshaping of the eyelid.

Dermabrasion—A technique for removing the upper layers of skin with planing wheels powered by compressed air.

Face lift—Plastic surgery performed to remove sagging skin and wrinkles from an individual's face.

Liposuction—A surgical technique for removing fat from under the skin by vacuum suctioning.

Mammoplasty—Surgery performed to change the size or shape of breasts.

Rhinoplasty—Surgery performed to change the shape of the nose.

The prospective patient will be asked to sign a consent form before the operation.

Personal education

The surgeon will meet with the prospective patient before the operation is scheduled, in order to explain the procedure and to be sure that the individual is realistic about the expected results. This consideration is particularly important for people undergoing cosmetic surgery.

Medical considerations

Some people should not have plastic surgery because of certain medical risks. These groups include:

- persons recovering from a heart attack, severe infection (for example, pneumonia), or other serious illnesses
- people with infectious hepatitis or HIV infections
- individuals with cancer whose cancer might spread (metastasize)
- people who are extremely overweight (Individuals who are more than 30% overweight should not have liposuction.)
- persons with blood clotting disorders

Psychological

Plastic, cosmetic, and reconstructive surgeries have an important psychological dimension because of the high value placed on outward appearance in Western society. Many people who are born with visible deformities or disfigured by accidents later in life develop emotional problems related to social rejection. Other people work in fields such as acting,

modeling, media journalism, and even politics, where their employment depends on how they look. Some people have unrealistic expectations of cosmetic surgery and think that it will solve all their life problems. It is important for anyone considering non-emergency plastic or cosmetic surgery to be realistic about its results. One type of psychiatric disorder, called **body dysmorphic disorder**, is characterized by an excessive preoccupation with imaginary or minor flaws in appearance. Persons with this disorder frequently seek unnecessary plastic surgery.

Aftercare

Medical

Medical aftercare following plastic surgery under general anesthesia includes bringing patients to a recovery room, monitoring their vital signs, and giving medications to relieve **pain** as necessary. Persons who have had fat removed from the abdomen may be kept in bed for as long as two weeks. Individuals who have had mammoplasties, **breast reconstruction**, and some types of facial surgery typically remain in the hospital for a week after the operation. Those who have had **liposuction** or eyelid surgery are usually sent home in a day or two.

People who have had outpatient procedures are usually given **antibiotics** to prevent infection and are sent home as soon as their vital signs are normal.

Psychological

Some individuals may need follow-up **psychotherapy** or counseling after plastic or reconstructive surgery. These people typically include children whose schooling and social relationships have been affected by birth defects, as well as persons of any age whose deformities or disfigurements were caused by trauma from accidents, war injuries, or violent crimes.

Risks

The risks associated with plastic, cosmetic, and reconstructive surgery include the postoperative complications that can occur with any surgical operation under anesthesia. These complications include wound infection, internal bleeding, **pneumonia**, and reactions to the anesthesia.

In addition to these general risks, some plastic, cosmetic, and reconstructive surgical procedures carry specific risks:

- formation of undesirable scar tissue
- development of persistent pain, redness, or swelling in the area of the surgery

- infection inside the body related to inserting a prosthesis (These infections can result from contamination at the time of surgery or from bacteria migrating into the area around the prosthesis at a later time.)
- anemia or fat embolisms from liposuction
- rejection of skin grafts or tissue transplants
- loss of normal feeling or function in the area of the operation (For example, it is not unusual for women who have had mammoplasties to lose sensation in their nipples.)
- complications resulting from unforeseen technological problems (The best-known example of this problem was the discovery in the mid-1990s that breast implants made with silicone gel could leak into the recipient's body.)

Normal results

Normal results include an individual's recovery from the surgery with satisfactory results and without complications.

Morbidity and mortality rates

Morbidity and mortality rates vary with the complexity and severity of different procedures. Mortality is similar to that associated with all surgical procedures. Morbidity is influenced by personal expectations. From a surgical perspective, most morbidity is due to errors associated with anesthesia, procedure, pain medications, and after care. From an individual's perspective, morbidity involves the degree to which actual results compared to expected outcomes. The latter distinction is very subjective.

Alternatives

Alternatives to plastic, reconstructive, and cosmetic surgical procedures include using various products that may be affixed to articles of clothing or the surface of the body.

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ORGANIZATIONS

- American Academy of Facial Plastic and Reconstructive Surgery, 310 S. Henry Street, Alexandria, VA, 22314, (703) 299-9291, <http://www.aafprs.org>.
- American Board of Plastic Surgery, Seven Penn Center, Suite 400, 1635 Market Street, Philadelphia, PA, 19103-2204, (215) 587-9322, <http://www.abplsurg.org>.
- American Society for Aesthetic Plastic Surgery, 11081 Winners Circle, Los Alamitos, CA, 90720, (888) 272-7711, <http://www.surgery.org>.
- American Society of Plastic Surgeons, 444 E. Algonquin Road, Arlington Heights, IL, 60005, (847) 228-9900, <http://www.plasticsurgery.org>.

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Platelet aggregation test

Definition

Platelets are disk-shaped blood cells that are also called thrombocytes. They play a major role in the blood-clotting process. The platelet aggregation test is a measure of platelet function.

Purpose

The platelet aggregation test aids in the evaluation of bleeding disorders by measuring the rate and degree to which platelets form a clump (aggregate) after the addition of a chemical that stimulates clumping (aggregation).

Precautions

There are many medications that can affect the results of the platelet aggregation test. The patient should discontinue as many as possible beforehand. Some of the drugs that can decrease platelet aggregation include **aspirin**, some **antibiotics**, **beta blockers**, dextran (Macrodex), alcohol, heparin (Lipo-Hepin), **nonsteroidal anti-inflammatory drugs** (NSAIDs), **tricyclic antidepressants**, and warfarin (Coumadin).

Description

There are many factors involved in blood clotting (coagulation). One of the first steps in the process involves small cells in the bloodstream called platelets, which are produced in the bone marrow. Platelets gather at the site of an injury and clump together to form a plug, or aggregate, that helps to limit the loss of blood and promote healing.

Inherited bleeding disorders (e.g., **hemophilia** or von Willebrand's disease) and acquired bleeding problems that occur because of another disorder or a medication can affect the number of platelets and their level of function. When these problems are present, the result is a drop in platelet aggregation and a lengthened **bleeding time**.

The platelet aggregation test uses a machine called an aggregometer to measure the cloudiness (turbidity) of blood plasma. Several different substances called agonists are used in the test. These agonists include adenosine diphosphate, epinephrine, thrombin, collagen, and ristocetin. The addition of an agonist to a plasma sample causes the platelets to clump together, making the fluid more transparent. The aggregometer then measures the increased light transmission through the specimen.

KEY TERMS

Aggregation—The blood cell clumping process that is measured in the platelet aggregation test.

Agonist—A chemical that is added to the blood sample in the platelet aggregation test to stimulate the clumping process.

Hemophilia—An inherited bleeding disorder caused by a deficiency of factor VIII, one of a series of blood proteins essential for blood clotting.

Platelets—Small, round, disk-shaped blood cells that are involved in clot formation. The platelet aggregation test measures the clumping ability of platelets.

Turbidity—The cloudiness or lack of transparency of a solution.

von Willebrand's disease—An inherited lifelong bleeding disorder caused by an abnormal gene, similar to hemophilia. The gene defect results in a decreased blood concentration of a substance called von Willebrand's factor (vWF).

Preparation

The test requires a blood sample. The patient should either avoid food and drink altogether for eight hours before the test, or eat only nonfat foods. High levels of fatty substances in the blood can affect test results.

Because the use of aspirin and/or aspirin compounds can directly affect test results, the patient should avoid these medications for two weeks before the test. If the patient must take aspirin and the test cannot be postponed, the laboratory should be notified and asked to verify the presence of aspirin in the blood plasma. If the results are abnormal, aspirin use must be discontinued and the test repeated in two weeks.

Aftercare

Because the platelet aggregation test is ordered when some type of bleeding problem is suspected, the patient should be cautioned to watch the puncture site for signs of additional bleeding.

Risks

Risks for this test are minimal in normal individuals. Patients with bleeding disorders, however, may have prolonged bleeding from the puncture wound or the formation of a bruise (hematoma) under the skin where the blood was withdrawn.

Normal results

The normal time for platelet aggregation varies somewhat depending on the laboratory, the temperature, the shape of the vial in which the test is performed, and the patient's response to different agonists. For example, the difference between the response to ristocetin and other products should be noted because ristocetin triggers aggregation through a different mechanism than other agonists.

Abnormal results

Prolonged platelet aggregation time can be found in such congenital disorders as hemophilia and von Willebrand's disease, as well as in some connective tissue disorders. Prolonged aggregation times can also occur in leukemia or myeloma; after recent heart/lung bypass or **kidney dialysis**; and after taking certain drugs.

Resources

BOOKS

Pagana, Kathleen Deska, and Timothy J. Pagana. *Mosby's Manual of Diagnostic and Laboratory Tests*. 4th ed. St. Louis: Mosby, 2009.

Janis O. Flores

Platelet count

Definition

A platelet count is a diagnostic test that determines the number of platelets in the patient's blood. Platelets, which are also called thrombocytes, are small disk-shaped blood cells produced in the bone marrow and involved in the process of blood clotting. There are normally between 150,000–450,000 platelets in each microliter of blood. Low platelet counts or abnormally shaped platelets are associated with bleeding disorders. High platelet counts sometimes indicate disorders of the bone marrow.

Purpose

The primary functions of a platelet count are to assist in the diagnosis of bleeding disorders and to monitor patients who are being treated for any disease involving bone marrow failure. Patients who have leukemia, **polycythemia vera**, or **aplastic anemia** are given periodic platelet count tests to monitor their

health or to ascertain the bone marrow's response to treatment.

Description

Blood collection and storage

Platelet counts use a freshly-collected blood specimen to which a chemical called EDTA has been added to prevent clotting before the test begins. About 5 mL of blood are drawn from a vein in the patient's inner elbow region. Blood drawn from a vein helps to produce a more accurate count than blood drawn from a fingertip. Collection of the sample takes only a few minutes.

After collection, the mean platelet volume of EDTA-blood will increase over time. This increase is caused by a change in the shape of the platelets after removal from the body. The changing volume is relatively stable for a period of one to three hours after collection. This period is the best time to count the sample when using electronic instruments, because the platelets will be within a standard size range.

Counting methods

Platelets can be observed in a direct blood smear for approximate quantity and shape. A direct smear is made by placing a drop of blood onto a microscope slide and spreading it into a thin layer. After staining to make the various blood cells easier to see and distinguish, a laboratory technician views the smear through a light microscope. Accurate assessment of the number of platelets requires other methods of counting. There are three methods used to count platelets; hemacytometer, voltage-pulse counting, and electro-optical counting.

HEMACYTOMETER COUNTING. The microscopic method uses a phase contrast microscope to view blood on a hemacytometer slide. A sample of the diluted blood mixture is placed in a hemacytometer, which is an instrument with a grid etched into its surface to guide the counting. For a proper count, the platelets should be evenly distributed in the hemacytometer. Counts made from samples with platelet clumping are considered unreliable. Clumping can be caused by several factors, such as clotting before addition of the anticoagulant and allowing the blood to remain in contact with a capillary blood vessel during collection. Errors in platelet counting are more common when blood is collected from capillaries than from veins.

ELECTRONIC COUNTING. Electronic counting of platelets is the most common method. There are two

types of electronic counting, voltage-pulse and electro-optical counting systems. In both systems, the collected blood is diluted and counted by passing the blood through an electronic counter. The instruments are set to count only particles within the proper size range for platelets. The upper and lower levels of the size range are called size exclusion limits. Any cells or material larger or smaller than the size exclusion limits will not be counted. Any object in the proper size range is counted, however, even if it is not a platelet. For these instruments to work properly, the sample must not contain other material that might mistakenly be counted as platelets. Electronic counting instruments sometimes produce artificially low platelet counts. If a platelet and another blood cell pass through the counter at the same time, the instrument will not count the larger cell because of the size exclusion limits, which will cause the instrument to accidentally miss the platelet. Clumps of platelets will not be counted because clumps exceed the upper size exclusion limit for platelets. In addition, if the patient has a high **white blood cell count**, electronic counting may yield an unusually low platelet count because white blood cells may filter out some of the platelets before the sample is counted. On the other hand, if the red blood cells in the sample have burst, their fragments will be falsely counted as platelets.

Preparation

There is no specific preparation required for this test.

Aftercare

Because platelet counts are sometimes ordered to diagnose or monitor bleeding disorders, patients with these disorders should be cautioned to watch the puncture site for signs of additional bleeding.

Risks

Risks for a platelet count test are minimal in normal individuals. Patients with bleeding disorders, however, may have prolonged bleeding from the puncture wound or the formation of a bruise (hematoma) under the skin where the blood was withdrawn.

Results

Normal results

The normal range for a platelet count is 150,000–450,000 platelets per microliter of blood.

KEY TERMS

Capillaries—The smallest of the blood vessels that bring oxygenated blood to tissues.

EDTA—A colorless compound used to keep blood samples from clotting before tests are run. Its chemical name is ethylenediaminetetraacetic acid.

Hemocytometer—An instrument used to count platelets or other blood cells.

Phase contrast microscope—A light microscope in which light is focused on the sample at an angle to produce a clearer image.

Thrombocyte—Another name for platelet.

Thrombocytopenia—An abnormally low platelet count.

Thrombocytosis—An abnormally high platelet count. It occurs in polycythemia vera and other disorders in which the bone marrow produces too many platelets.

Abnormal results

An abnormally low platelet level (**thrombocytopenia**) is a condition that may result from increased destruction of platelets, decreased production, or increased usage of platelets. In **idiopathic thrombocytopenic purpura** (ITP), platelets are destroyed at abnormally high rates. **Hypersplenism** is characterized by the collection (sequestration) of platelets in the spleen. Disseminated intravascular coagulation (DIC) is a condition in which **blood clots** occur within blood vessels in a number of tissues. All of these diseases produce reduced platelet counts.

Abnormally high platelet levels (**thrombocytosis**) may indicate either a benign reaction to an infection, surgery, or certain medications; or a disease like polycythemia vera, in which the bone marrow produces too many platelets too quickly.

Resources

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Platelet function disorders

Definition

Platelets are cells within the bloodstream that recognize and cling to damaged areas inside blood vessels. When they do this, the platelets trigger a series of chemical changes that result in the formation of a blood clot. There are certain hereditary disorders that affect platelet function and impair their ability to start the process of blood clot formation. One result is the possibility of excessive bleeding from minor injuries or from menstrual flow.

Description

Platelets are formed in the bone marrow—a spongy tissue located inside the long bones of the body—as fragments of a large precursor cell (a megakaryocyte). These fragments circulate in the bloodstream and form the first line of defense against blood escaping from injured blood vessels.

Damaged blood vessels release a chemical signal that increases the stickiness of platelets in the area of the injury. The sticky platelets adhere to the damaged area and gradually form a platelet plug. At the same time, the platelets release a series of chemical signals that prompt other factors in the blood to reinforce the platelet plug. Between the platelet and its reinforcements, a sturdy clot is created that acts as a patch while the damaged area heals.

There are several hereditary disorders characterized by some impairment of the platelet's action. Examples include von Willebrand disease, Glanzmann's thrombasthenia, and Bernard–Soulier syndrome. Vulnerable aspects of platelet function include errors in the production of the platelets themselves or errors in the formation, storage, or release of their chemical signals. These defects can prevent platelets from responding to injuries or from prompting the action of other factors involved in clot formation.

Causes and symptoms

Platelet function disorders can be inherited, but they may also occur as a symptom of acquired diseases or as a side effect of certain drugs, including **aspirin** and **nonsteroidal anti-inflammatory drugs** (NSAIDS). The most common inherited bleeding disorder is von Willebrand disease, a relatively minor condition, which is thought to affect as many as one in every 1,000 people. There are several variants of this disorder.

KEY TERMS

Anemia—A condition in which inadequate quantities of hemoglobin and red blood cells are produced.

Bone marrow—A spongy tissue located within the body's flat bones, including the hip and breast bones and the skull. Marrow contains stem cells, the precursors to platelets and red and white blood cells.

Hemoglobin—The substance inside red blood cells that enables them to carry oxygen.

Megakaryocyte—A large bone marrow cell with a lobed nucleus that is the precursor cell of blood platelets.

Platelets—Fragments of a large precursor cell (a megakaryocyte) found in the bone marrow. These fragments adhere to areas of blood vessel damage and release chemical signals that direct the formation of a blood clot.

Symptoms of platelet function disorders vary in severity depending on the etiology of the condition and can include bleeding from the nose, gums, vagina, or anus; pinpoint **bruises** and purplish patches on the skin; and abnormally heavy menstrual bleeding.

Diagnosis

In diagnosing platelet function disorders, specific tests are needed to determine whether the problem is caused by low numbers of platelets or impaired platelet function. A blood smear, a **platelet count**, and **bleeding time** are common screening tests. If these tests confirm that the symptoms are due to impaired platelet function, further tests are done—such as platelet aggregation or an analysis of the platelet proteins—that pinpoint the exact nature of the defect.

Treatment

Treatment is intended to prevent bleeding and stop it quickly when it occurs. For example, patients are advised to be careful when they brush their teeth to reduce damage to the gums. They are also warned against taking medications that interfere with platelet function. Some patients may require iron and folate supplements to counteract potential anemia. Some patients diagnosed with immune or idiopathic platelet disorders can be treated with **corticosteroids**. Platelet

transfusions may be necessary to prevent life-threatening hemorrhaging in some cases. Hormone therapy is useful in treating heavy menstrual bleeding. Von Willebrand's disease can be treated with desmopressin (DDAVP, Stimate).

Prognosis

Outcome depends on the specific disorder and the severity of its symptoms. Platelet function disorders range from life-threatening conditions to easily treated or little-noticed problems.

Prevention

Inherited platelet function disorders cannot be prevented except by **genetic counseling**; however, some acquired function disorders may be guarded against by avoiding substances that trigger the disorder.

Resources

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Pleural biopsy

Definition

The pleura is the membrane that lines the lungs and chest cavity. A pleural biopsy is the removal of pleural tissue for examination.

Purpose

Pleural biopsy is done to differentiate between benign and malignant disease, to diagnose viral, fungal, or parasitic diseases, and to identify a condition called collagen vascular disease of the pleura. It is also ordered when a **chest x ray** indicates a pleural-based tumor, reaction, or thickening of the lining.

Precautions

Because pleural biopsy is an invasive procedure, it is not recommended for patients with severe bleeding disorders.

Description

Pleural biopsy is usually ordered when pleural fluid obtained by another procedure called **thoracentesis** (aspiration of pleural fluid) suggests infection, signs of **cancer**, or **tuberculosis**. Pleural biopsies are 85–90% accurate in diagnosing these diseases.

The procedure most often performed for pleural biopsy is called a percutaneous (passage through the skin by needle puncture) needle biopsy. The procedure takes 30–45 minutes, although the biopsy needle itself remains in the pleura for less than one minute. This type of biopsy is usually performed by a physician at bedside, if the patient is hospitalized, or in the doctor's office under local anesthetic.

The actual procedure begins with the patient in a sitting position, shoulders and arms elevated and supported. The skin overlying the biopsy site is anesthetized and a small incision is made to allow insertion of the biopsy needle. This needle is inserted with a cannula (a plastic or metal tube) until fluid is removed. Then the inner needle is removed and a trocar (an instrument for withdrawing fluid from a cavity) is inserted to obtain the actual biopsy specimen. As many as three separate specimens are taken from different sites during the procedure. These specimens are then placed into a fixative solution and sent to the laboratory for tissue (histologic) examination.

Preparation

Preparations for this procedure vary, depending on the type of procedure requested. Pleural biopsy can be performed in several ways: percutaneous needle biopsy (described above), by **thoracoscopy** (insertion of a visual device called a laparoscope into the pleural space for inspection), or by open pleural biopsy, which requires **general anesthesia**.

Aftercare

Potential complications of this procedure include bleeding or injury to the lung, or a condition called **pneumothorax**, in which air enters the pleural cavity (the space between the two layers of pleura lining the lungs and the chest wall). Because of these possibilities, the patient is to report any **shortness of breath**, and to note any signs of bleeding, decreased blood pressure, or increased pulse rate.

Risks

Risks for this procedure include respiratory distress on the side of the biopsy, as well as bleeding, possible shoulder **pain**, pneumothorax (immediate) or **pneumonia** (delayed).

Normal results

Normal findings indicate no evidence of any pathologic or disease conditions.

Abnormal results

Abnormal findings include tumors called neoplasms (any new or abnormal growth) that can be either benign or malignant. Pleural tumors are divided into two classifications: primary (**mesothelioma**), or metastatic (arising from cancer sites elsewhere in the body). These tumors are often associated with an accumulation of fluid between the pleural layers called a **pleural effusion**, which itself may be caused by pneumonia, **heart failure**, cancer, or blood clot in the lungs (**pulmonary embolism**).

Other causes of abnormal findings include viral, fungal, or parasitic infections, and tuberculosis.

Resources

BOOKS

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Janis O. Flores

smoothly against one another during breathing movements. Any extra fluid is taken up by blood and lymph vessels, maintaining a balance. When either too much fluid forms or something prevents its removal, the result is an excess of pleural fluid—an effusion. The most common causes are disease of the heart or lungs, and inflammation or infection of the pleura.

Pleural effusion itself is not a disease as much as a result of many different diseases. For this reason, there is no "typical" patient in terms of age, sex, or other characteristics. Instead, anyone who develops one of the many conditions that can produce an effusion may be affected.

There are two types of pleural effusion: the transudate and the exudate. This is a very important point because the two types of fluid are very different, and which type is present points to what sort of disease is likely to have produced the effusion. It also can suggest the best approach to treatment.

Transudates

A transudate is a clear fluid, similar to blood serum, that forms not because the pleural surfaces themselves are diseased, but because the forces that normally produce and remove pleural fluid at the same rate are out of balance. When the heart fails, pressure in the small blood vessels that remove pleural fluid is increased and fluid "backs up" in the pleural space, forming an effusion. Or, if too little protein is present in the blood, the vessels are less able to hold the fluid part of blood within them and it leaks out into the pleural space. This can result from disease of the liver or kidneys, or from **malnutrition**.

Exudates

An exudate—which often is a cloudy fluid, containing cells and much protein—results from disease of the pleura itself. The causes are many and varied. Among the most common are infections such as bacterial **pneumonia** and **tuberculosis**; **blood clots** in the lungs; and connective tissue diseases, such as **rheumatoid arthritis**. **Cancer** and disease in organs such as the pancreas also may give rise to an exudative pleural effusion.

Special types of pleural effusion

Some of the pleural disorders that produce an exudate also cause bleeding into the pleural space. If the effusion contains half or more of the number of red blood cells present in the blood itself, it is called hemothorax. When a pleural effusion has a milky

Pleural effusion

Definition

Pleural effusion occurs when too much fluid collects in the pleural space (the space between the two layers of the pleura). It is commonly known as "water on the lungs." It is characterized by **shortness of breath**, chest **pain**, gastric discomfort (**dyspepsia**), and **cough**.

Description

There are two thin membranes in the chest, one (the visceral pleura) lining the lungs, and the other (the parietal pleura) covering the inside of the chest wall. Normally, small blood vessels in the pleural linings produce a small amount of fluid that lubricates the opposed pleural membranes so that they can glide

appearance and contains a large amount of fat, it is called chylothorax. Lymph fluid that drains from tissues throughout the body into small lymph vessels finally collects in a large duct (the thoracic duct) running through the chest to empty into a major vein. When this fluid, or chyle, leaks out of the duct into the pleural space, chylothorax is the result. Cancer in the chest is a common cause.

Causes and symptoms

Causes of transudative pleural effusion

Among the most important specific causes of a transudative pleural effusion are:

- Congestive heart failure. This causes pleural effusions in about 40% of patients and is often present on both sides of the chest. Heart failure is the most common cause of bilateral (two-sided) effusion. When only one side is affected it usually is the right (because patients usually lie on their right side).
- Pericarditis. This is an inflammation of the pericardium, the membrane covering the heart.
- Too much fluid in the body tissues, which spills over into the pleural space. This is seen in some forms of kidney disease; when patients have bowel disease and absorb too little of what they eat; and when an excessive amount of fluid is given intravenously.
- Liver disease. About 5% of patients with a chronic scarring disease of the liver called cirrhosis develop pleural effusion.

Causes of exudative pleural effusions

A wide range of conditions may be the cause of an exudative pleural effusion:

- Pleural tumors account for up to 40% of one-sided pleural effusions. They may arise in the pleura itself (mesothelioma), or from other sites, notably the lung.
- Tuberculosis in the lungs may produce a long-lasting exudative pleural effusion.
- Pneumonia affects about three million persons each year, and four of every ten patients will develop pleural effusion. If effective treatment is not provided, an extensive effusion can form that is very difficult to treat.
- Patients with any of a wide range of infections by a virus, fungus, or parasite that involve the lungs may have pleural effusion.
- Up to half of all patients who develop blood clots in their lungs (pulmonary embolism) will have pleural effusion, and this sometimes is the only sign of embolism.

- Connective tissue diseases, including rheumatoid arthritis, lupus, and Sjögren's syndrome may be complicated by pleural effusion.
- Patients with disease of the liver or pancreas may have an exudative effusion, and the same is true for any patient who undergoes extensive abdominal surgery. About 30% of patients who undergo heart surgery will develop an effusion.
- Injury to the chest may produce pleural effusion in the form of either hemothorax or chylothorax.

Symptoms

The key symptom of a pleural effusion is shortness of breath. Fluid filling the pleural space makes it hard for the lungs to fully expand, causing the patient to take many breaths so as to get enough oxygen. When the parietal pleura is irritated, the patient may have mild pain that quickly passes or, sometimes, a sharp, stabbing pleuritic type of pain. Some patients will have a dry cough. Occasionally a patient will have no symptoms at all. This is more likely when the effusion results from recent abdominal surgery, cancer, or tuberculosis. Tapping on the chest will show that the usual crisp sounds have become dull, and on listening with a stethoscope the normal breath sounds are muted. If the pleura is inflamed, there may be a scratchy sound called a "pleural friction rub."

Diagnosis

When pleural effusion is suspected, the best way to confirm it is to take chest x rays, both straight-on and from the side. The fluid itself can be seen at the bottom of the lung or lungs, hiding the normal lung structure. If **heart failure** is present, the x-ray shadow of the heart will be enlarged. An ultrasound scan may disclose a small effusion that caused no abnormal findings during chest examination. A computed tomography scan is very helpful if the lungs themselves are diseased.

In order to learn what has caused the effusion, a needle or catheter is often used to obtain a fluid sample, which is examined for cells and its chemical make-up. This procedure, called a **thoracentesis**, is the way to determine whether an effusion is a transudate or exudate, giving a clue as to the underlying cause. In some cases—for instance when cancer or bacterial infection is present—the specific cause can be determined and the correct treatment planned. Culturing a fluid sample can identify the bacteria that cause tuberculosis or other forms of pleural infection. The next diagnostic step is to take a tissue sample, or **pleural biopsy**, and examine it under a microscope. If the effusion is caused by lung disease, placing a

KEY TERMS

Culture—A test that exposes a sample of body fluid or tissue to special material to see whether bacteria or another type of microorganism is present.

Dyspepsia—A vague feeling of being too full and having heartburn, bloating, and nausea. Usually felt after eating.

Exudate—The type of pleural effusion that results from inflammation or other disease of the pleura itself. It features cloudy fluid containing cells and proteins.

Pleura or pleurae—A delicate membrane that encloses the lungs. The pleura is divided into two areas separated by fluid—the visceral pleura, which covers the lungs, and the parietal pleura, which lines the chest wall and covers the diaphragm.

Pleural cavity—The area of the thorax that contains the lungs.

Pleural space—The potential area between the visceral and parietal layers of the pleurae.

Pneumonia—An acute inflammation of the lungs, usually caused by bacterial infection.

Sclerosis—The process by which an irritating material is placed in the pleural space in order to inflame the pleural membranes and cause them to stick together, eliminating the pleural space and recurrent effusions.

Thoracentesis—Placing a needle, tube, or catheter in the pleural space to remove the fluid of pleural effusion. Used for both diagnosis and treatment.

Transudate—The type of pleural effusion seen with heart failure or other disorders of the circulation. It features clear fluid containing few cells and little protein.

viewing tube (bronchoscope) through the large air passages will allow the examiner to see the abnormal appearance of the lungs.

Treatment

The best way to clear up a pleural effusion is to direct treatment at what is causing it, rather than treating the effusion itself. If heart failure is reversed or a lung infection is cured by antibiotics, the effusion will usually resolve. However, if the cause is not known, even after extensive tests, or no effective treatment is at hand, the fluid can be drained away by placing a large-bore needle or catheter into the pleural space, just as in diagnostic thoracentesis. If necessary, this can be repeated as often as is needed to control the amount of fluid in the pleural space. If large effusions continue to recur, a drug or material that irritates the pleural membranes can be injected to deliberately inflame them and cause them to adhere close together—a process called sclerosis. This will prevent further effusion by eliminating the pleural space. In the most severe cases, open surgery with removal of a rib may be necessary to drain all the fluid and close the pleural space.

Prognosis

When the cause of pleural effusion can be determined and effectively treated, the effusion itself will reliably clear up and should not recur. In many other cases, sclerosis will prevent sizable effusions from recurring. Whenever a large effusion causes a patient to be short of breath,

thoracentesis will make breathing easier, and it may be repeated if necessary. To a great extent, the outlook for patients with pleural effusion depends on the primary cause of effusion and whether it can be eliminated. Some forms of pleural effusion, such as that seen after abdominal surgery, are only temporary and will clear without specific treatment. If heart failure can be controlled, the patient will remain free of pleural effusion. If, on the other hand, effusion is caused by cancer that cannot be controlled, other effects of the disease probably will become more important.

Prevention

Because pleural effusion is a secondary effect of many different conditions, the key to preventing it is to promptly diagnose the primary disease and provide effective treatment. Timely treatment of infections such as tuberculosis and pneumonia will prevent many effusions. When effusion occurs as a drug side-effect, withdrawing the drug or using a different one may solve the problem. On rare occasions, an effusion occurs because fluid meant for a vein is mistakenly injected into the pleural space. This can be prevented by making sure that proper technique is used.

ORGANIZATIONS

American Lung Association, 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20001, (202) 758-3355, (202) 452-1805, (800) 548-8252, info@lungusa.org, <http://www.lungusa.org>.

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

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Pleural fluid analysis see **Thoracentesis**

Pleurisy

Definition

Pleurisy is an inflammation of the membrane that surrounds and protects the lungs (the pleura). Inflammation occurs when an infection or damaging agent irritates the pleural surface. As a consequence, sharp chest pains are the primary symptom of pleurisy.

Demographics

Pleurisy may affect any individual but a higher incidence is observed in those with an underlying lung condition such as a lung tumor or **abscess**, **tuberculosis**, or **pneumonia**.

Description

Pleurisy, also called pleuritis, is a condition that generally stems from an existing respiratory infection, disease, or injury. In people who have otherwise good health, respiratory infections or pneumonia are the main causes of pleurisy. This condition used to be more common, but with the advent of **antibiotics** and modern disease therapies, pleurisy has become less prevalent.

The pleura is a double-layered structure made up of an inner membrane, which surrounds the lungs, and an outer membrane, which lines the chest cavity. The pleural membranes are very thin, close together, and have a fluid coating in the narrow space between them. This liquid acts as a lubricant, so that when the lungs inflate and deflate during breathing, the pleural surfaces can easily glide over one another.

Pleurisy occurs when the pleural surfaces rub against one another, due to irritation and inflammation. Infection within the pleural space is the most common irritant, although the abnormal presence of air, blood, or cells can also initiate pleurisy. These disturbances all act to displace the normal pleural fluid, which forces the membranes to rub, rather than glide, against one another. This rubbing irritates

nerve endings in the outer membrane and causes **pain**. Pleurisy also causes a chest noise that ranges from a faint squeak to a loud creak. This characteristic sound is called a “friction rub.”

Pleurisy cases are classified either as having **pleural effusion** or as being “dry.” Pleural effusion is more common and refers to an accumulation of fluid within the pleural space; dry pleurisy is inflammation without fluid build-up. Less pain occurs with pleural effusion because the fluid forces the membrane surfaces apart. However, pleural effusion causes additional complications because it places pressure on the lungs. This leads to respiratory distress and possible lung collapse.

Causes and symptoms

A variety of conditions can give rise to pleurisy. The following list represents the most common sources of pleural inflammation.

- infections, including pneumonia, tuberculosis, and other bacterial or viral respiratory infections
- immune disorders, including systemic lupus erythematosus, rheumatoid arthritis, and sarcoidosis
- diseases, including cancer, pancreatitis, liver cirrhosis, and heart or kidney failure
- injury, from a rib fracture, collapsed lung, esophagus rupture, blood clot, or material such as asbestos
- drug reactions, from certain drugs used to treat tuberculosis (isoniazid), cancer (methotrexate, procarbazine), or the immune disorders mentioned above (hydralazine, procainamide, phenytoin, quinidine)

Symptomatic pain

The hallmark symptom of pleurisy is sudden, intense chest pain that is usually located over the area of inflammation. Although the pain can be constant, it is usually most severe when the lungs move during breathing, coughing, sneezing, or even talking. The pain is usually described as shooting or stabbing, but in minor cases it resembles a mild cramp. When pleurisy occurs in certain locations, such as near the diaphragm, the pain may be felt in other areas such as the neck, shoulder, or abdomen (referred pain). Another indication of pleurisy is that holding one's breath or exerting pressure against the chest causes pain relief.

Breathing difficulties

Pleurisy is also characterized by certain respiratory symptoms. In response to the pain, pleurisy

patients commonly have a rapid, shallow breathing pattern. Pleural effusion can also cause **shortness of breath**, as excess fluid makes expanding the lungs difficult. If severe breathing difficulties persist, patients may experience a blue colored complexion (**cyanosis**).

Additional symptoms of pleurisy are specific to the illness that triggers the condition. Thus, if infection is the cause, then chills, **fever**, and **fatigue** will be likely pleurisy symptoms.

Diagnosis

The distinctive pain of pleurisy is normally the first clue physicians use for diagnosis. Doctors usually feel the chest to find the most painful area, which is the likely site of inflammation. A stethoscope is also used to listen for abnormal chest sounds as the patient breathes. If the doctor hears the characteristic friction rub, the diagnosis of pleurisy can be confirmed. Sometimes, a friction rub is masked by the presence of pleural effusion and further examination is needed for an accurate diagnosis.

Identifying the actual illness that causes pleurisy is more difficult. To make this diagnosis, doctors must evaluate the patient's history, additional symptoms, and laboratory test results. A **chest x ray** may also be taken to look for signs of accumulated fluid and other abnormalities. Possible causes, such as pneumonia, fractured ribs, esophagus rupture, and lung tumors may be detected on an x ray. Computed tomography scan (CT scan) and ultrasound scans are more powerful diagnostic tools used to visualize the chest cavity. Images from these techniques more clearly pinpoint the location of excess fluid or other suspected problems.

The most helpful information in diagnosing the cause of pleurisy is a fluid analysis. Once the doctor knows the precise location of fluid accumulation, a sample is removed using a procedure called **thoracentesis**. In this technique, a fine needle is inserted into the chest to reach the pleural space and extract fluid. The fluid's appearance and composition is thoroughly examined to help doctors understand how the fluid was produced. Several laboratory tests are performed to analyze the chemical components of the fluid. These tests also determine whether infection-causing bacteria or viruses are present. In addition, cells within the fluid are identified and counted. Cancerous cells can also be detected to learn whether the pleurisy is caused by a malignancy.

In certain instances, such as dry pleurisy, or when a fluid analysis is not informative, a biopsy of the pleura may be needed for microscopic analysis. A sample of pleural tissue can be obtained several ways: with a biopsy needle, by making a small incision in the chest wall, or by using a thoracoscope (a video-assisted instrument for viewing the pleural space and collecting samples).

Treatment

Pain management

The pain of pleurisy is usually treated with analgesic and anti-inflammatory drugs, such as **acetaminophen**, ibuprofen, and indomethacin. People with pleurisy may also receive relief from lying on the painful side. Sometimes, a painful **cough** will be controlled with codeine-based cough syrups. However, as the pain eases, a person with pleurisy should try to breathe deeply and cough to clear any congestion; otherwise pneumonia may occur. Rest is also important to aid in the recovery process.

Treating the source

The treatment used to cure pleurisy is ultimately defined by the underlying cause. Thus, pleurisy from a bacterial infection can be successfully treated with antibiotics, while no treatment is given for viral infections that must run their course. Specific therapies designed for more chronic illnesses can often cause pleurisy to subside. For example, tuberculosis pleurisy is treated with standard anti-tuberculosis drugs. With some illnesses, excess fluid continues to accumulate and causes severe respiratory distress. In these individuals, the fluid may be removed by thoracentesis, or the doctor may insert a chest tube to drain large amounts. If left untreated, a more serious infection may develop within the fluid, called **empyema**.

Alternative treatment

Alternative treatments can be used in conjunction with conventional treatment to help heal pleurisy. **Acupuncture** and botanical medicines are alternative approaches for alleviating pleural pain and breathing problems. An herbal remedy commonly recommended is pleurisy root (*Asclepias tuberosa*), so named because of its use by early American settlers who learned of this medicinal plant from Native Americans. Pleurisy root helps to ease pain, inflammation, and breathing difficulties brought on by pleurisy. This herb is often used in conjunction with mullein (*Verbascum thapsus*) or elecampane

KEY TERMS

Effusion—The accumulation of fluid within a cavity, such as the pleural space.

Empyema—An infection that causes pus to accumulate in the pleural space. The pus may cause a tear in the pleural membrane, which allows the infection to spread to other areas in the body. Intravenous antibiotics are often given to control the infection.

Inflammation—An accumulation of fluid and cells within tissue that is often caused by infection and the immune response that occurs as a result.

Pneumonia—A condition caused by bacterial or viral infection that is characterized by inflammation of the lungs and fluid within the air passages. Pneumonia is often an underlying cause of pleurisy.

Referred pain—The presence of pain in an area other than where it originates. In some pleurisy cases, referred pain occurs in the neck, shoulder, or abdomen.

(*Inula helenium*), which serve as **expectorants** to clear excess mucus from the lungs. In addition, there are many other respiratory herbs that are used as expectorants or for other actions on the respiratory system. Herbs thought to combat infection, such as **echinacea** (*Echinacea spp.*) are also included in herbal pleurisy remedies. Antiviral herbs, such as *Lomatium dissectum* and *Ligusticum porteri*, can be used if the pleurisy is of viral origin. **Traditional Chinese medicine** uses the herb ephedra (*Ephedra sinica*), which acts to open air passages and alleviate respiratory difficulties in pleurisy patients. Dietary recommendations include eating fresh fruits and vegetables, adequate protein, and good quality fats (**omega-3 fatty acids** are anti-inflammatory and are found in fish and flax oil). Taking certain **nutritional supplements**, especially large doses of vitamin C, may also provide health benefits to people with pleurisy. Contrast **hydrotherapy** applied to the chest and back, along with compresses (cloths soaked in an herbal solution) or poultices (crushed herbs applied directly to the skin) of respiratory herbs, can assist in the healing process. Homeopathic treatment, guided by a trained practitioner, can be effective in resolving pleurisy. Alternative treatments should be used with care, as the benefits of many such treatments have not been confirmed by scientific research.

Prognosis

Prompt diagnosis, followed by appropriate treatment, ensures a good recovery for most pleurisy patients. Generally speaking, the prognosis for pleurisy is linked to the seriousness of its cause. Therefore, the outcome of pleurisy caused by a disease such as **cancer** will vary depending on the type and location of the tumor.

Prevention

Preventing pleurisy is often a matter of providing early medical attention to conditions that can cause pleural inflammation. Along this line, appropriate antibiotic treatment of bacterial respiratory infections may successfully prevent some cases of pleurisy. Maintaining a healthy lifestyle and avoiding exposure to harmful substances (for example, asbestos) are more general preventative measures.

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ORGANIZATIONS

American Lung Association (ALA), 1301 Pennsylvania Ave. NW, Washington, DC, 20004, (202) 785 3355, (202) 452 1805, <http://www.lungusa.org>.

American Thoracic Society (ATS), 61 Broadway, 6th Floor, New York, NY, 10000, (212) 315-8600, <http://www.thoracic.org>.

Centers for Disease Control and Prevention (CDC), 1600 Clifton Rd., Atlanta, GA, 30333, (404) 639-3311, (800) 311-3435, <http://www.cdc.gov>.

National Heart Lung and Blood Institute (NHLBI), PO Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573. TTY: (240) 629-3255, <http://www.nhlbi.nih.gov>.

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Pleuritis see **Pleurisy**

Plumbism see **Lead poisoning**

PMDD see **Premenstrual dysphoric disorder**

PMS see **Premenstrual syndrome**

Pneumococcal pneumonia

Definition

Pneumococcal **pneumonia** is a common but serious infection and inflammation of the lungs. It is caused by the bacterium *Streptococcus pneumoniae*.

Description

The gram-positive, spherical bacteria, *Streptococcus pneumoniae*, is the cause of many human diseases, including pneumonia. Although the bacteria can normally be found in the nose and throat of healthy individuals, it can grow and cause infection when the immune system is weakened. Infection usually begins with the upper respiratory tract and then travels into the lungs. Pneumonia occurs when the bacteria find their way deep into the lungs, to the area called the alveoli, or air sacs. This is the functional part of the lungs where oxygen is absorbed into the blood. Once in the alveoli, *Streptococcus pneumoniae* begin to grow and multiply. White blood cells and immune proteins from the blood also accumulate at the site of infection in the alveoli. As the alveoli fill with these substances and fluid, they can no longer function in the exchange of oxygen. This fluid filling of the lungs is how pneumonia is defined.

Those people most at risk of developing pneumococcal pneumonia have a weakened immune system. This includes the elderly, infants, **cancer** patients, **AIDS** patients, post-operative patients, alcoholics, and those with diabetes. Pneumococcal pneumonia is a disease that has a high rate of hospital transmission, putting hospital patients at greater risk. Prior lung infections also makes someone more likely to develop pneumococcal pneumonia. The disease can be most severe in patients who have had their spleen removed. It is the spleen that is responsible for removing the bacteria from the blood. Cases of pneumonia, which is spread by close contact, seem to occur most often between November through April. If not treated, the disease can spread, causing continually decreasing lung function, heart problems, and arthritis.

Causes and symptoms

Symptoms of bacterial pneumonia include a **cough**, sputum (mucus) production that may be pus-like or bloody, shaking and chills, **fever**, and chest **pain**. Symptoms often have an abrupt beginning and occur after an upper respiratory infection such as a cold. Symptoms may differ somewhat in the elderly, with minimal cough, no sputum and no fever, but

rather tiredness and confusion leading to **hypothermia** and **shock**.

Diagnosis

The presence of symptoms and a physical exam that reveals abnormal lung sounds usually suggest the presence of pneumonia. Diagnosis is typically made from an x ray of the lungs, which indicates the accumulation of fluid. Additional tests that may be done include a **complete blood count**, a sputum sample for microscopic examination and culture for *Streptococcus pneumoniae*, and possibly blood cultures.

Treatment

Depending on the severity of the disease, **antibiotics** are given either at home or in the hospital. Historically, the treatment for pneumococcal pneumonia has been penicillin. An increasing number of cases of pneumococcal pneumonia have become partially or completely resistant to penicillin, making it less effective in treating this disease. Other effective antibiotics include amoxicillin and erythromycin. If these antibiotics are not effective, vancomycin or cephalosporin may alternatively be used.

Symptoms associated with pneumococcal pneumonia can also be treated. For instance, fever can be treated with **aspirin** or **acetaminophen**. Supplemental oxygen and intravenous fluids may help. Patients are advised to get plenty of rest and take increased amounts of fluids. Coughing should be promoted because it helps to clear the lungs of fluid.

Alternative treatment

Being a serious, sometimes fatal disease, pneumococcal pneumonia is best treated as soon as possible with antibiotics. However, there are alternative treatments that both support this conventional treatment and prevent recurrences. Maintaining a healthy immune system is important. One way to do this is by taking the herb, **echinacea** (*Echinacea spp.*). Getting plenty of rest and reducing **stress** can help the body heal. Some practitioners feel that mucus-producing foods (including dairy products, eggs, gluten-rich grains such as wheat, oats, rye, as well as sugar) can contribute to the lung congestion that accompanies pneumonia. Decreasing these foods and increasing the amount of fresh fruits and vegetables may help to decrease lung congestion. Adequate protein in the diet is also essential for the body to produce antibodies. Contrast and constitutional **hydrotherapy** can be very helpful in treating cases of pneumonia. Other alternative therapies, including **acupuncture**, Chinese herbal

KEY TERMS

Acetaminophen—A drug used for pain relief as well as to decrease fever. A common trade name for the drug is Tylenol.

Aspirin—A commonly used drug for pain relief and to decrease fever.

Bronchi—Two main branches of the trachea that go into the lungs. This then further divides into the bronchioles and alveoli.

Sputum—A substance that comes up from the throat when coughing or clearing the throat. It is important since it contains materials from the lungs.

medicine, and homeopathy, can be very useful during the recovery phase, helping the body to rebuild after the illness and contributing to the prevention of recurrences.

Prognosis

Simple, uncomplicated cases of pneumococcal pneumonia will begin to respond to antibiotics in 48 to 72 hours. Full recovery from pneumonia, however, is greatly dependent on the age and overall health of the individual. Normally, healthy and younger patients can recover in only a few days, while the elderly or otherwise weakened individuals may not recover for several weeks. Complications may develop which give a poorer prognosis. Even when promptly and properly diagnosed, such weakened patients may die of their pneumonia.

Prevention

Vaccination

Recently, a **vaccination** has become available for the prevention of pneumococcal pneumonia. This vaccination is generally recommended for people with a high likelihood of developing pneumococcal infection or for those in whom a serious complication of infection is likely to develop. This would include persons over the age of 65, as well as those with:

- chronic pulmonary disease
- advanced cardiovascular disease
- diabetes mellitus
- alcoholism
- cirrhosis

- chronic kidney disease
- spleen dysfunction, or removal of spleen
- immunosuppression (cancer, organ transplant or AIDS)
- sickle cell anemia

Unfortunately, those people for whom the vaccination is most recommended are also those who are least likely to respond favorably to a vaccination. Therefore, the overall effectiveness of this vaccine remains questionable.

Antibiotics

The use of oral penicillin to prevent infection may be recommended for some patients at high risk, such as children with **sickle cell disease** and those with a spleen removed. This treatment, however, must be weighed with the increased likelihood of developing penicillin-resistant infections.

ORGANIZATIONS

American Lung Association, 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20001, (202) 758-3355, (202) 452-1805, (800) 548-8252, info@lungusa.org, <http://www.lungusa.org>.

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Cindy L. A. Jones, PhD

Pneumocystis pneumonia

Definition

Pneumocystis **pneumonia** is a lung infection that occurs primarily in people with weakened immune systems—especially people who are HIV-positive. The disease agent is an organism whose biological classification is still uncertain. *Pneumocystis carinii* was originally thought to be a one-celled organism (a protozoan), but more recent research suggests that it is a fungus. Although its life cycle is known to have three stages, its method of reproduction is not yet completely understood. The complete name of the disease is *Pneumocystis carinii* pneumonia, often shortened to PCP. It is also sometimes called pneumocystosis.

Description

Pneumonia as a general term refers to a severe lung inflammation. In pneumocystis pneumonia, this

inflammation is caused by the growth of *Pneumocystis carinii*, a fungus-like organism that is widespread in the environment. PCP is ordinarily a rare disease, affecting only people with weakened immune systems. Many of these people are patients receiving drugs for organ transplants or **cancer** treatment. With the rising incidence of **AIDS**, however, PCP has become primarily associated with AIDS patients. In fact, as many as 75% of AIDS patients have developed PCP. It has also been the leading cause of **death** in AIDS patients.

Transmission

The organism that causes PCP is widely distributed in nature and is transmitted through the air. When the organism is inhaled, it enters the upper respiratory tract and infects the tiny air sacs at the ends of the smaller air tubes (bronchioles) in the lungs. These tiny air sacs are called alveoli. Under a microscope, alveoli look like groups of hollow spheres resembling grape clusters. The exchange of oxygen with the blood takes place in the alveoli. It appears that *P. carinii* lives in the fluid in the lining of the alveoli.

Person-to-person infection does not appear to be very common; however, clusters of PCP outbreaks in hospitals and groups of immunocompromised people indicate that patients with active PCP should not be exposed to others with weakened immune systems. It is thought that many people actually acquire mild *Pneumocystis carinii* infections from time to time, but are protected by their immune systems from developing a full-blown case of the disease.

Causes and symptoms

Causes

P. carinii is an opportunistic organism. This means that it causes disease only under certain conditions, as when a person is immunocompromised. Under these circumstances, *P. carinii* can multiply and cause pneumonia. The mechanisms of the organism's growth within the alveoli are not fully understood. As the pneumocystis organism continues to replicate, it gradually fills the alveoli. As the pneumonia becomes more severe, fluid accumulates and tissue scarring occurs. These changes result in decreased respiratory function and lower levels of oxygen in the blood.

High-risk groups

Some patients are at greater risk of developing PCP. These high-risk groups include:

- premature infants
- patients with immunodeficiency diseases, including severe combined immunodeficiency disease (SCID) and acquired immunodeficiency syndrome (AIDS),
- patients receiving immunosuppressive drugs, especially cortisone-like drugs (corticosteroids)
- Patients with protein malnutrition.

AIDS is currently the most common risk factor for PCP in the United States. PCP is, however, also found in countries with widespread hunger and poor hygiene.

Symptoms

The incubation period of PCP is not definitely known, but is thought to be between four and eight weeks. The major symptoms include **shortness of breath**, **fever**, and a nonproductive **cough**. Less common symptoms include production of sputum, blood in the sputum, difficulty breathing, and chest **pain**. Most patients will have symptoms for one to two weeks before seeing a physician. Occasionally, the disease will spread outside of the lung to other organs, including the lymph nodes, spleen, liver, or bone marrow.

Diagnosis

The diagnosis of PCP begins with a thorough **physical examination** and blood tests. Although imaging studies are helpful in identifying abnormal areas in the lungs, the diagnosis of PCP must be confirmed by microscopic identification of the organism in the lung. Samples may be taken from the patient's sputum, or may be obtained via **bronchoscopy** or **lung biopsy**. Because of the severity of the disease, many physicians will proceed to treat patients with symptoms of pneumocystis pneumonia if they belong to a high-risk group, without the formality of an actual diagnosis. The severity of PCP can be measured by x-ray studies and by determining the amount of oxygen and carbon dioxide present in the patient's blood.

Treatment

Treatment for PCP involves the use of **antibiotics**. These include trimethoprim-sulfamethoxazole (TMP-SMX, Bactrim, Septra) and pentamidine isoethionate (Nebupent, Pentam 300). Both of these anti-microbial drugs are equally effective. AIDS patients are typically treated for 21 days, whereas non-AIDS patients are treated for 14 days. TMP-SMX may be highly toxic in AIDS patients, causing severe side effects that include fever, rash, decreased numbers of white blood cells and platelets, and hepatitis. Pentamidine also causes

KEY TERMS

Alveoli—Small, hollow air sacs found in the lungs at the end of the smaller airways (bronchioles). Air exchange occurs in the alveoli.

Azotemia—The presence of excess nitrogenous wastes in the blood.

Biopsy—A procedure in which a piece of tissue is obtained for microscopic study.

Bronchoscopy—A procedure that uses a fiber-optic scope to view the airways in the lung.

Fungus—A member of a group of simple organisms related to yeasts and molds.

Pentamidine isoethionate—An antibiotic used to treat and prevent PCP.

Pneumocystosis—Another name for active PCP infection.

Protozoan—A microorganism belonging to the Protista, which includes the simplest one-celled organisms.

Sputum—A substance obtained from the lungs and bronchial tubes by clearing the throat or coughing. Sputum can be tested for evidence of PCP infection.

Trimethoprim-sulfamethoxazole (TMP-SMX)—An antibiotic used to treat and prevent PCP.

side effects in immunocompromised patients. These side effects include decreased blood pressure, irregular heart beats, the accumulation of nitrogenous waste products in the blood (azotemia), and electrolyte imbalances. Pentamidine can be given in aerosol form to minimize side effects. Alternative drugs can be used for patients experiencing these side effects.

P. carinii appears to be developing resistance to TMP-SMX. In addition, some patients are allergic to the standard antibiotics given for PCP. As a result, other antibiotics for the treatment of PCP are continually under investigation. Some drugs proven to be effective against *P. carinii* include dapsone (DDS) with trimethoprim (Trimpex), clindamycin (Cleocin) with primaquine, as well as atovaquone (Mepron). Paradoxically, **corticosteroids** have been found to improve the ability of TMP-SMX or pentamidine to treat PCP. As a treatment of last resort, trimetrexate with leucovorin (Wellcovorin) can also be used.

Prognosis

If left untreated, PCP will cause breathing difficulties that will eventually cause death. The prognosis for this disease depends on the amount of damage to the patient's lungs prior to treatment. Prognosis is usually better at a facility that specializes in caring for AIDS patients. Antibiotic treatment of PCP is about 80% effective.

Prevention

Medications

For patients at serious risk for PCP infection, low doses of TMP-SMX, given daily or three times a week, are effective in preventing PCP. The drug is, however,

highly toxic. Researchers are currently evaluating the effectiveness and toxicity of aerosol pentamidine and dapsone in preventing PCP.

Lifestyle modifications

Patients who have previously had PCP often experience a recurrence. Healthy lifestyle choices, including exercising, eating well, and giving up **smoking** may keep the disease at bay.

ORGANIZATIONS

American Lung Association, 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20001, (202) 758-3355, (202) 452-1805, (800) 548-8252, info@lungusa.org, <http://www.lungusa.org>.

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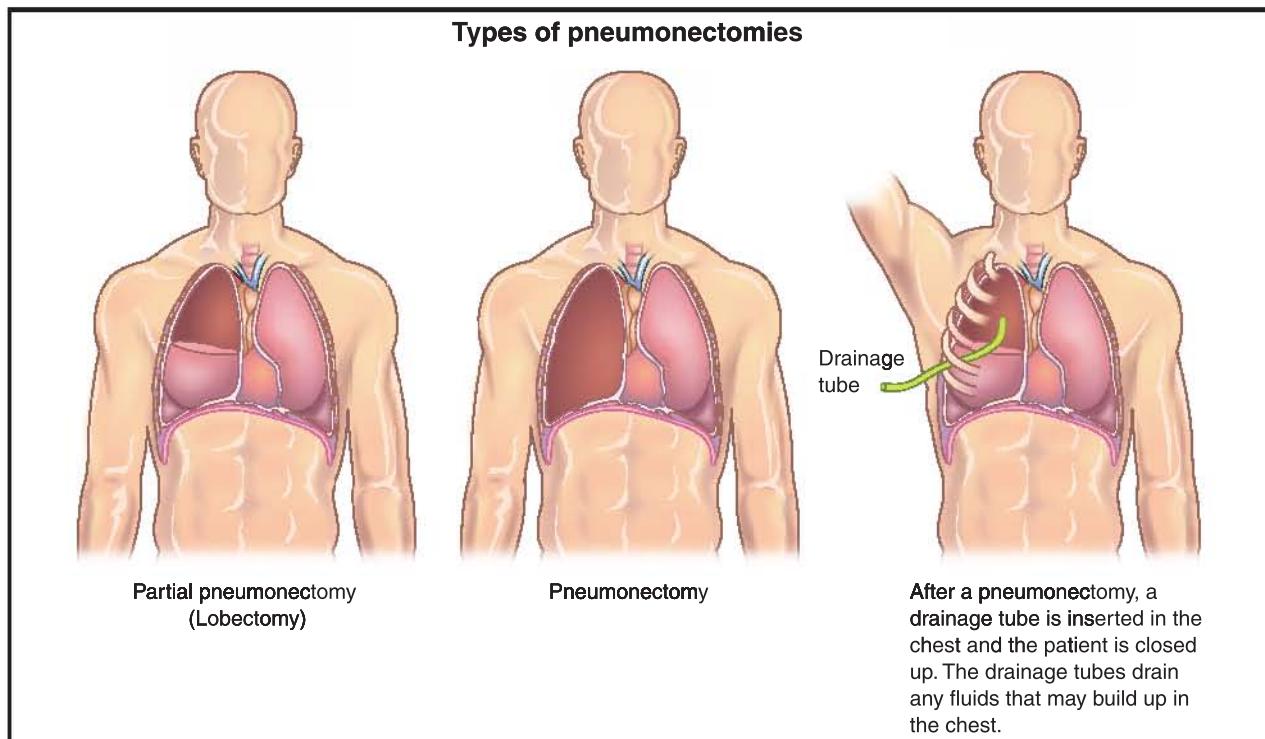
Pneumonectomy

Definition

Pneumonectomy is the medical term for the surgical removal of a lung.

Purpose

A pneumonectomy is most often used to treat lung **cancer** when less radical surgery cannot achieve satisfactory results. It may also be the most appropriate treatment for a tumor located near the center of the lung that affects the pulmonary artery or veins, which transport



(Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

blood between the heart and lungs. In addition, pneumonectomy may be the treatment of choice when the patient has a traumatic chest injury that has damaged the main air passage (bronchus) or the lung's major blood vessels so severely that they cannot be repaired.

Demographics

Pneumonectomies are usually performed on patients with lung cancer, as well as patients with such noncancerous diseases as **chronic obstructive pulmonary disease** (COPD), which includes **emphysema** and chronic **bronchitis**. These diseases cause airway obstruction.

Approximately 342,000 Americans die of lung disease every year. Lung disease is responsible for one in seven deaths in the United States, according to the American Lung Association. This makes lung disease America's number three killer. More than 35 million Americans are now living with chronic lung disease.

Lung cancer

Lung cancer is the leading cause of cancer-related deaths in the United States, accounting for a third of all cancer-related deaths. It is projected to claim more

than 1.3 million lives worldwide in 2010, 170,000 of these deaths in the United States. Lung cancer kills more people than cancers of the breast, prostate, colon, and pancreas combined. Cigarette **smoking** accounts for nearly 90% of cases of lung cancer in the United States.

Lung cancer is the second most common cancer among both men and women and is the leading cause of **death** from cancer in both sexes. In addition to the use of tobacco as a major cause of lung cancer among smokers, second-hand smoke contributes to the development of lung cancer among nonsmokers. Exposure to asbestos and other hazardous substances is also known to cause lung cancer. Air pollution is also a probable cause, but makes a relatively small contribution to incidence and mortality rates. Indoor exposure to radon may also make a small contribution to the total incidence of lung cancer in certain geographic areas of the United States.

In each of the major racial/ethnic groups in the United States, the rates of lung cancer among men are about two to three times greater than the rates among women. Among men, age-adjusted lung cancer incidence rates (per 100,000) range from a low of about 14 among Native

KEY TERMS

Bronchodilator—A drug that relaxes bronchial muscles resulting in expansion of the bronchial air passages.

Bronchopleural fistula—An abnormal connection between an air passage and the membrane that covers the lungs.

Corticosteroids—Any of various adrenal-cortex steroids used as anti-inflammatory agents.

Emphysema—A chronic disease characterized by loss of elasticity and abnormal accumulation of air in lung tissue.

Empyema—An accumulation of pus in the lung cavity, usually as a result of infection.

Malignant mesothelioma—A cancer of the pleura (the membrane lining the chest cavity and covering the lungs) that typically is related to asbestos exposure.

Pleural space—The small space between the two layers of the membrane that covers the lungs and lines the inner surface of the chest.

Pulmonary embolism—Blockage of a pulmonary artery by a blood clot or foreign matter.

Pulmonary rehabilitation—A program to treat COPD, which generally includes education and counseling, exercise, nutritional guidance, techniques to improve breathing, and emotional support.

Americans to a high of 117 among African Americans, an eight-fold difference. For women, the rates range from approximately 15 per 100,000 among Japanese Americans to nearly 51 among Native Alaskans, only a three-fold difference.

Chronic obstructive pulmonary disease

The following are risk factors for COPD:

- current smoking or a long-term history of heavy smoking
- employment that requires working around dust and irritating fumes
- long-term exposure to second-hand smoke at home or in the workplace
- a productive cough (with phlegm or sputum) most of the time
- shortness of breath during vigorous activity

- shortness of breath that grows worse even at lower levels of activity
- a family history of early COPD (before age 45)

Diagnosis/Preparation

Diagnosis

In some cases, the diagnosis of a lung disorder is made when the patient consults a physician about chest pains or other symptoms. The symptoms of lung cancer vary somewhat according to the location of the tumor; they may include persistent coughing, coughing up blood, **wheezing**, **fever**, and weight loss. In cases involving direct trauma to the lung, the decision to perform a pneumonectomy may be made in the emergency room. Before scheduling a pneumonectomy, however, the surgeon reviews the patient's medical and surgical history and orders a number of tests to determine how successful the surgery is likely to be.

In the case of lung cancer, blood tests, a **bone scan**, and **computed tomography scans** of the head and abdomen indicate whether the cancer has spread beyond the lungs. **Positron emission tomography (PET)** scanning is also used to help stage the disease. Cardiac screening indicates how well the patient's heart will tolerate the procedure, and extensive pulmonary testing (e.g., breathing tests and quantitative ventilation/perfusion scans) predicts whether the remaining lung will be able to make up for the patient's diminished ability to breathe.

Preparation

A patient who smokes must stop as soon as a lung disease is diagnosed. Patients should not take **aspirin** or ibuprofen for seven to 10 days before surgery. Patients should also consult their physician about discontinuing any blood-thinning medications such as Coumadin or warfarin. The night before surgery, patients should not eat or drink anything after midnight.

Description

In a conventional pneumonectomy, the surgeon removes only the diseased lung itself. In a partial pneumonectomy, one or more lobes of a lung are removed. In an extrapleural pneumonectomy, the surgeon removes the lung, part of the membrane covering the heart (pericardium), part of the diaphragm, and the membrane lining the chest cavity (parietal pleura). Either operation is extensive, and require that the patient be given **general anesthesia**. An intravenous line inserted into one arm supplies fluids and

medication throughout the operation, which usually lasts one to three hours.

The surgeon begins the operation by cutting a large opening on the same side of the chest as the diseased lung. This posterolateral thoracotomy incision extends from a point below the shoulder blade around the side of the patient's body along the curvature of the ribs at the front of the chest. Sometimes the surgeon removes part of the fifth rib in order to have a clearer view of the lung and greater ease in removing the diseased organ.

A surgeon performing a traditional pneumonectomy then:

- deflates (collapses) the diseased lung
- ties off the lung's major blood vessels to prevent bleeding into the chest cavity
- clamps the main bronchus to prevent fluid from entering the air passage
- cuts through the bronchus
- removes the lung
- staples or sutures the end of the bronchus that has been cut
- makes sure that air is not escaping from the bronchus
- inserts a temporary drainage tube between the layers of the pleura (pleural space) to draw air, fluid, and blood out of the surgical cavity
- closes the chest incision

Aftercare

Chest tubes drain fluid from the incision and a respirator helps the patient breathe for at least 24 hours after the operation. The patient may be fed and medicated intravenously. If no complications arise, the patient is transferred from the surgical intensive care unit to a regular hospital room within one to two days.

A patient who has had a conventional pneumonectomy will usually leave the hospital within 10 days. Aftercare during hospitalization is focused on:

- relieving pain
- monitoring the patient's blood oxygen levels
- encouraging the patient to walk in order to prevent formation of blood clots
- encouraging the patient to cough productively in order to clear accumulated lung secretions

If the patient cannot **cough** productively, the doctor uses a flexible tube (bronchoscope) to remove the lung secretions and fluids.

Recovery is usually a slow process, with the remaining lung gradually taking on the work of the lung that has been removed. The patient may gradually resume normal non-strenuous activities. A pneumonectomy patient who does not experience postoperative problems may be well enough within eight weeks to return to a job that is not physically demanding; however, 60% of all pneumonectomy patients continue to struggle with **shortness of breath** six months after having surgery.

Risks

The risks for any surgical procedure requiring anesthesia include reactions to the medications and breathing problems. The risks for any surgical procedure include bleeding and infection.

Between 40% and 60% of pneumonectomy patients experience such short-term postoperative difficulties as:

- prolonged need for a mechanical respirator
- abnormal heart rhythm (cardiac arrhythmia); heart attack (myocardial infarction); or other heart problem
- pneumonia
- infection at the site of the incision
- a blood clot in the remaining lung (pulmonary embolism)
- an abnormal connection between the stump of the cut bronchus and the pleural space due to a leak in the stump (bronchopleural fistula)
- accumulation of pus in the pleural space (empyema)
- kidney or other organ failure

Over time, the remaining organs in the patient's chest may move into the space left by the surgery. This condition is called postpneumonectomy syndrome; the surgeon can correct it by inserting a fluid-filled prosthesis into the space formerly occupied by the diseased lung.

Normal results

The doctor will probably advise the patient to refrain from strenuous activities for a few weeks after the operation. The patient's rib cage will remain sore for some time.

A patient whose lungs have been weakened by noncancerous diseases like emphysema or chronic bronchitis may experience long-term shortness of breath as a result of this surgery. On the other hand, a patient who develops a fever, chest pain, persistent cough, or shortness of breath, or whose incision bleeds

or becomes inflamed, should notify his or her doctor immediately.

Morbidity and mortality rates

In the United States, the immediate survival rate from surgery for patients who have had the left lung removed is between 96% and 98%. Due to the greater risk of complications involving the stump of the cut bronchus in the right lung, between 88% and 90% of patients survive removal of this organ. Following lung volume reduction surgery, most investigators now report mortality rates of 5–9%.

Alternatives

Lung cancer

The treatment options for lung cancer are surgery, **radiation therapy**, and **chemotherapy**, either alone or in combination, depending on the stage of the cancer.

After the cancer is found and staged, the cancer care team discusses the treatment options with the patient. In choosing a treatment plan, the most significant factors to consider are the type of lung cancer (small cell or non-small cell) and the stage of the cancer. It is very important that the doctor order all the tests needed to determine the stage of the cancer. Other factors to consider include the patient's overall physical health; the likely side effects of the treatment; and the probability of curing the disease, extending the patient's life, or relieving his or her symptoms.

Chronic obstructive pulmonary disease

Although surgery is rarely used to treat COPD, it may be considered for people who have severe symptoms that have not improved with medication therapy. A significant number of patients with advanced COPD face a miserable existence and are at high risk of death, despite advances in medical technology. This group includes patients who remain symptomatic despite the following:

- smoking cessation
- use of inhaled bronchodilators
- treatment with antibiotics for acute bacterial infections, and inhaled or oral corticosteroids
- use of supplemental oxygen with rest or exertion
- pulmonary rehabilitation

After the severity of the patient's airflow obstruction has been evaluated, and the foregoing interventions implemented, a pulmonary disease specialist should examine him or her, with consideration given to surgical treatment.

Surgical options for treating COPD include laser therapy or the following procedures:

- **Bullectomy.** This procedure removes the part of the lung that has been damaged by the formation of large air-filled sacs called bullae.
- **Lung volume reduction surgery.** In this procedure, the surgeon removes a portion of one or both lungs, making room for the remaining lung tissue to work more efficiently. Its use is considered experimental, although it has been used in selected patients with severe emphysema.
- **Lung transplant.** In this procedure a healthy lung from a donor who has recently died is given to a person with COPD.

Resources

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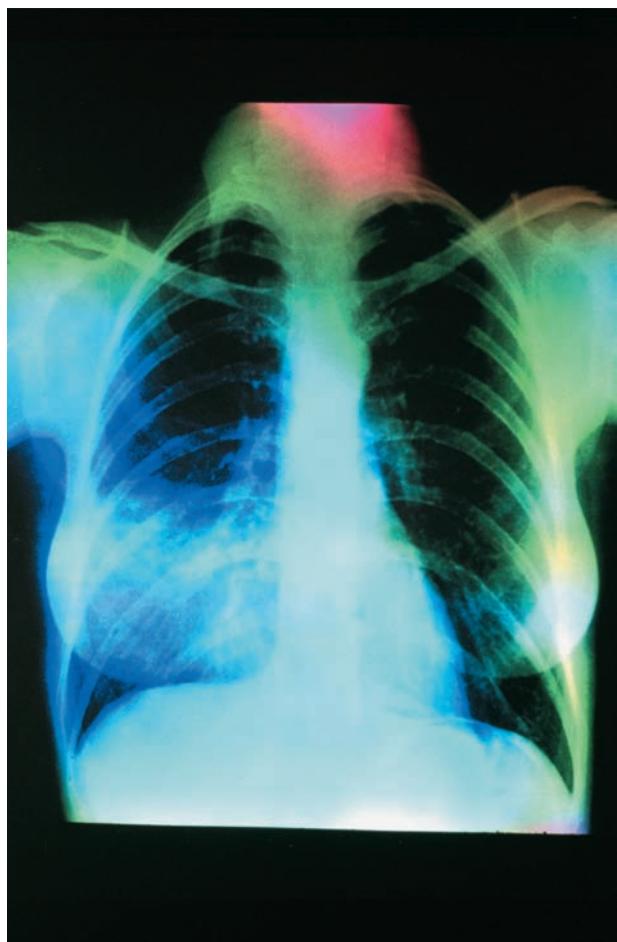
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ORGANIZATIONS

- American Cancer Society., 1599 Clifton Road, N.E, Atlanta, GA, 30329-4251, (800) 227-2345, www.cancer.org.
- American Lung Association, 1301 Pennsylvania Ave. NW, Washington, DC, 20004, (202) 785-3355, www.lungusa.org.
- National Cancer Institute (NCI), Building 31, Room 10A03, 31 Center Drive, Bethesda, MD, 20892-2580, (301) 435-3848, (800) 4-CANCER, www.nci.nih.gov.
- National Comprehensive Cancer Network, 275 Commerce Drive, Suite 300, Fort Washington, PA, 19034, (215) 690-0300, www.nccn.org.
- National Heart, Lung and Blood Institute (NHLBI), 6701 Rockledge Drive, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, www.nhlbi.nih.gov.

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Pneumonia

Definition

Pneumonia is an infection of the lung that can be caused by nearly any class of organism known to cause human infections. These include bacteria, amoebae, viruses, fungi, and parasites. Pneumonia may also result from non-infectious causes, such as inhalation of food, liquids, gases, or dust. Pneumonia often develops as a complication of a pre-existing condition or infection or when a patient's immune system is weakened by a condition such as a simple viral respiratory tract infection or by **influenza**. Pneumonia and influenza together are ranked as the eighth leading cause of **death** in the United States, with pneumonia accounting for most of those deaths. In the elderly, pneumonia is the fourth leading cause of death and the leading infectious cause of death. In 2006, 55,477 people in the United States died of pneumonia.

When a person has pneumonia, the air sacs in the lungs become filled with pus and other liquids, and oxygen transfer from the lungs to the blood stream is inhibited. Without sufficient oxygen, body cells cannot function properly. Lobar pneumonia affects a

A chest x ray showing lobar pneumonia in the lower lobe of a patient's right lung. The alveoli (air sacs) of the lung become blocked with pus, which forces air out and causes the lung to become solidified. (Photo Researchers, Inc.)

section (lobe) of a lung while bronchial pneumonia affects patches throughout both lungs.

Description

Anatomy of the lung

To better understand pneumonia, it is important to understand the basic anatomic features of the respiratory system. The human respiratory system begins at the nose and mouth, where air is breathed in (inspired) and out (expired). The air tube extending from the nose is called the nasopharynx. The tube carrying air breathed in through the mouth is called the oropharynx. The nasopharynx and the oropharynx merge into the larynx. The oropharynx also carries swallowed substances, including food, water, and salivary secretion, which must pass into the esophagus and then the stomach. The larynx is protected by a

trap door called the epiglottis. The epiglottis prevents substances that have been swallowed, as well as substances that have been regurgitated (thrown up), from heading down into the larynx and toward the lungs.

A useful method of picturing the respiratory system is to imagine an upside-down tree. The larynx flows into the trachea, which is the tree trunk, and thus the broadest part of the respiratory tree. The trachea divides into two tree limbs, the right and left bronchi. Each one of these branches off into multiple smaller bronchi, which course through the tissue of the lung. Each bronchus divides into tubes of smaller and smaller diameter, finally ending in the terminal bronchioles. The air sacs of the lung, in which oxygen-carbon dioxide exchange actually takes place, are clustered at the ends of the bronchioles like the leaves of a tree. They are called alveoli.

The tissue of the lung which serves only a supportive role for the bronchi, bronchioles, and alveoli is called the lung stroma (or lung parenchyma).

Function of the respiratory system

The main function of the respiratory system is to provide oxygen, the most important energy source for the body's cells. Inspired air (the air we breath in) contains the oxygen, and travels down the respiratory tree to the alveoli. The oxygen moves out of the alveoli and is sent into circulation throughout the body as part of the red blood cells. The oxygen in the inspired air is exchanged within the alveoli for the waste product of human metabolism, carbon dioxide. The air we breathe out contains the gas called carbon dioxide. This gas leaves the alveoli during expiration. To restate this exchange of gases simply, we breathe in oxygen, we breathe out carbon dioxide.

Respiratory system defenses

The healthy human lung is sterile. There are no normally resident bacteria or viruses (unlike the upper respiratory system and parts of the gastrointestinal system, where bacteria dwell even in a healthy state). There are multiple safeguards along the path of the respiratory system. These are designed to keep invading organisms from leading to infection.

The first line of defense includes the hair in the nostrils, which serves as a filter for large particles. The epiglottis is a trap door of sorts, designed to prevent food and other swallowed substances from entering the larynx and then trachea. Sneezing and coughing, both provoked by the presence of irritants within the respiratory system, help to clear such irritants from the respiratory tract.

Mucus, produced through the respiratory system, also serves to trap dust and infectious organisms. Tiny hair like projections (cilia) from cells lining the respiratory tract beat constantly. They move debris trapped by mucus upwards and out of the respiratory tract. This mechanism of protection is referred to as the mucociliary escalator.

Cells lining the respiratory tract produce several types of immune substances which protect against various organisms. Other cells (called macrophages) along the respiratory tract actually ingest and kill invading organisms.

The organisms that cause pneumonia, then, are usually carefully kept from entering the lungs by virtue of these host defenses. However, when an individual encounters a large number of organisms at once, the usual defenses may be overwhelmed, and infection may occur. This can happen either by inhaling contaminated air droplets, or by aspiration of organisms inhabiting the upper airways.

Conditions predisposing to pneumonia

In addition to exposure to sufficient quantities of causative organisms, certain conditions may make an individual more likely to become ill with pneumonia. Various conditions are listed below.

Cigarette smoke, inhaled directly by a smoker or second-hand by a innocent bystander, interferes significantly with ciliary function, as well as inhibiting macrophage function, thus predisposing in individual to pneumonia.

Stroke, seizures, alcohol, and various drugs interfere with the function of the epiglottis. This leads to a leaky seal on the trap door, with possible contamination by swallowed substances and/or regurgitated stomach contents. Alcohol and drugs also interfere with the normal **cough** reflex. This further decreases the chance of clearing unwanted debris from the respiratory tract.

Viruses may interfere with ciliary function, allowing themselves or other microorganism invaders (such as bacteria) access to the lower respiratory tract. One of the most important viruses is HIV (human **immunodeficiency** virus), the causative virus in **AIDS** (acquired immunodeficiency syndrome). In recent years this virus has resulted in a huge increase in the incidence of pneumonia. Because AIDS results in a general decreased effectiveness of many aspects of the host's immune system, a patient with AIDS is susceptible to all kinds of pneumonia. This includes some previously rare parasitic types which would be unable

to cause illness in an individual possessing a normal immune system.

Pneumonia is sometimes a pulmonary condition affecting **cancer** patients, and may indicate that the cancer is progressing or that the patient has developed a new problem. Both cancer and the therapies used to treat it can injure the lungs or weaken the immune system in ways that make cancer patients especially susceptible to the bacteria, fungi, viruses, and other organisms that cause pneumonia. Tumors and infections can block the patient's airway or limit the lungs' ability to rid themselves of fluid and other accumulated secretions that make breathing difficult. Radiation treatment for **breast cancer** increases the risk of pneumonia in some patients by weakening lung tissue. Other factors that increase a cancer patient's risk of developing pneumonia include:

- radiation therapy
- chemotherapy
- surgery
- depressed white blood cell count (neutropenia)
- antibiotics
- steroids
- malnutrition
- limited mobility
- splenectomy-immune system deficits

Various chronic conditions predispose a person to infection with pneumonia. These include **asthma**, **cystic fibrosis**, and neuromuscular diseases which may interfere with the seal of the epiglottis. **Esophageal disorders** may result in stomach contents passing upwards into the esophagus. This increases the risk of aspiration into the lungs of those stomach contents with their resident bacteria. Diabetes, sickle cell anemia, lymphoma, leukemia, and **emphysema** also predispose a person to pneumonia.

Pneumonia is also one of the most frequent infectious complications of all types of surgery. Many drugs used during and after surgery may increase the risk of aspiration, impair the cough reflex, and cause a patient to under fill their lungs with air. **Pain** after surgery also discourages a patient from breathing deeply enough, and from coughing effectively.

Certain other conditions can increase the risk of pneumonia. These include the following:

- abnormal anatomical structure, particularly of the chest or lungs
- advanced age and associated immune system weakness

- esophageal disorders that may result in stomach contents passing upwards
- genetic factors and associated changes in DNA
- malnutrition

Pneumonia in children

Pneumonia can develop gradually in children after exposure to the causative organism, or it can develop quickly after another illness, reducing the lungs' ability to receive and distribute oxygen. It can be mild and easily cured with **antibiotics** and rest, or it can be severe and require hospitalization. The onset, duration, and severity of pneumonia depend upon the type of infective organism invading the body and the response of the child's immune system in fighting the infection. Respiratory distress represents 20% of all admissions of children to hospitals, and pneumonia is the underlying cause of most of these admissions.

Bacterial pneumonia develops after the child inhales or aspirates pathogens. Viral pneumonia stems primarily from inhaling infected droplets from the upper airway into the lungs. In neonates, pneumonia may result from colonization of the infant's nasopharynx by organisms that were in the birth canal at the time of delivery.

Pneumonia in the elderly

Pneumonia is one of the common and significant diseases of the elderly, especially those over the age of seventy. In general, the elderly are more susceptible to pneumonia than younger people. The elderly are also more likely to be hospitalized for pneumonia and need mechanical ventilation, resulting in a longer hospital stay than younger persons. In addition, many elderly people contract pneumonia while staying in a hospital for other conditions, because their immune systems are often compromised due to the condition that initially required treatment.

The elderly have a less effective mucociliary escalator, as well as changes in their immune system. This causes this age group to be more at risk for the development of pneumonia.

The intensity of symptoms and clinical manifestations of pneumonia are often less in the elderly than in younger patients, thus complicating diagnosis of the disease. The elderly may lose lung capacity as they age, making it harder for them to cough productively. They are also often used to feeling ill so may not recognize new symptoms of illness. Elderly people with pneumonia commonly exhibit acute confusion or **delirium** and deterioration of base metabolic functions.

Incidence

In the United States, pneumonia is the sixth most common disease leading to death; 2 million Americans develop pneumonia each year, and 40,000–70,000 die from it. Pneumonia is also the most common fatal infection acquired by already hospitalized patients. In developing countries, pneumonia ties with **diarrhea** as the most common cause of death. According to the Centers for Disease Control and Prevention (CDC), the number of deaths from pneumonia in the United States has declined slightly since 2001, however, even in nonfatal cases, pneumonia is a significant economic burden on the health care system. One study estimates that people in the American workforce who develop pneumonia cost employers five times as much in health care as the average worker.

The epidemic of HIV, has resulted in a huge increase in the incidence of pneumonia. Because AIDS results in immune system suppression, individuals with AIDS are highly susceptible to all kinds of pneumonia, including some previously rare parasitic types that would not cause illness in someone with a normal immune system.

Demographics

Every year in the United States, two million people of all ages develop pneumonia, including 4% of all the children in the country. It is the sixth most common disease leading to death and the fourth leading cause of death in the elderly; 40,000 to 70,000 people die from pneumonia each year. The incidence of pneumonia in children younger than one year of age is 35 to 40 per 1,000; 30 to 35 per 1,000 children ages two to four; and 15 per 1,000 children between ages five and nine. Fewer than 10 children in 1,000 over age nine are reported to develop pneumonia.

One sixth of the six million pneumonia cases that develop each year occur primarily in persons aged 65 years and older. Over 90% of all deaths from pneumonia occur in the older population. The incidence of development of pneumonia in the elderly is 20 to 40 illnesses per 1000 persons for pneumonia acquired in community settings, while the incidence rises to 100 to 250 per 1000 persons in cases acquired in long-term care facilities. An estimated 2.1% of elderly residents in long-term care facilities at any one time have pneumonia. About one billion dollars per year are spent on medical therapy to treat bacterial pneumonia in the elderly.

Causes

The list of organisms which can cause pneumonia is very large, and includes nearly every class of infecting organism: viruses, bacteria, bacteria-like organisms, fungi, and parasites. Some organisms are more frequently encountered by specific age groups. In addition, some characteristics of an individual may place him or her at greater risk for infection by particular types of organisms:

- Viruses cause the majority of pneumonias in young children (especially respiratory syncytial virus, parainfluenza and influenza viruses, and adenovirus).
- Adults are more frequently infected with bacteria (such as *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Staphylococcus aureus*).
- Pneumonia in older children and young adults is often caused by the bacteria-like *Mycoplasma pneumoniae* (often referred to as “walking” pneumonia).
- *Pneumocystis carinii pneumonia* (PCP) is an important cause of pneumonia in patients with immune problems (such as patients being treated for cancer with chemotherapy, or patients with AIDS). Classically considered a parasite, it appears to be more related to fungi.
- People who have reason to come into contact with bird droppings, such as poultry workers, are at risk for pneumonia caused by the organism *Chlamydia psittaci*.
- A very large, serious outbreak of pneumonia occurred in 1976, when individuals attending an American Legion convention were infected by a previously unknown organism. The outbreak caused twenty nine deaths among American Legion members who were staying at a Philadelphia hotel. Subsequently named *Legionella pneumophila*, it causes what is now called “Legionnaire’s disease.” The *Legionella* bacteria can live in water and can spread through air conditioning systems in hotels and hospitals. Susceptibility to the disease increases with increasing age.

Other bacteria that cause pneumonia, especially in institutional settings, include *Klebsiella*, *Pseudomonas aeruginosa*, *Enterobacter* species, *Proteus* species, ***Escherichia coli***, and other gram negative bacteria. Strains of anaerobic bacteria can be aspirated into the lungs by the elderly due to conditions associated with **aging** (such as sedative use or neurological conditions) and cause pneumonia. *Haemophilus influenzae* is a bacteria that causes pneumonia more frequently in patients with chronic **bronchitis**.

Pneumonia caused by *Mycoplasma pneumoniae* is a common cause of pneumonia that is usually not a significant threat to the health of the elderly, as it usually affects people younger than 40. Persons at highest risk for mycoplasma pneumonia are those living or working in crowded areas such as schools and homeless shelters, although many people who contract mycoplasma pneumonia have no identifiable risk factor. Symptoms typical of pneumonia are usually mild and appear over a period of one to three weeks. They may become more severe in some people.

PCP is caused by a fungus, *Pneumocystis jiroveci*. PCP develops in persons with weakened immune systems from causes such as cancer, chronic use of **corticosteroids** or other medications that affect the immune system, HIV/AIDS, or solid organ and/or bone marrow transplants. Symptoms of PCP include a mild and dry cough, **fever**, rapid breathing, and **shortness of breath**, especially upon **exercise** or activity. PCP was a rare disease before the AIDS disease developed. This type of pneumonia may also be referred to as **pneumocystis pneumonia**.

Chemical pneumonia is an unusual type of lung irritation. Although pneumonia usually is caused by a bacteria or virus, in chemical pneumonia, inflammation of lung tissue can be caused by many types of chemicals, including liquids, gases, and small particles, such as dust or fumes. Only a small percentage of pneumonias are caused by chemicals. Some chemicals only harm the lungs; however, some toxic chemicals may affect other organs in addition to the lungs and can result in serious organ damage or death. Aspiration pneumonia is another form of chemical pneumonia, where oral secretions or stomach contents are aspirated into the lungs. Inflammation develops from the toxic effects of stomach acid and enzymes on lung tissue. Symptoms of chemical pneumonia may include:

- burning of the nose, eyes, lips, mouth, and throat
- dry cough
- wet cough producing clear, yellow, or green mucus
- cough producing blood or frothy pink matter
- nausea or abdominal pain
- chest pain
- shortness of breath
- painful breathing or pleuritis (an inflammation of the outside covering of the lungs)
- headache
- flu symptoms

- weakness or a general ill feeling
- delirium or disorientation

Half of all pneumonia cases are caused by viruses, including the influenza virus, parainfluenza virus, adenovirus, rhinovirus, herpes simplex virus, respiratory syncytial virus, hantavirus, and cytomegalovirus. Many of these pneumonia infections are mild and may last only a short time. However pneumonia caused by the influenza virus may be severe and occasionally fatal. The symptoms of influenza pneumonia are similar to those of influenza, including fever, dry cough, **headache**, muscle pain, and weakness. However, within 12 to 36 hours, breathlessness develops, and the coughing increases, with a small amount of mucus produced. Patients have a high fever and may develop blueness of the lips. Eighty percent of deaths in recent influenza epidemics occurred in persons aged 65 and older, mostly due to development of complications such as **sepsis** or acute **respiratory distress syndrome**. Viral pneumonia can be further complicated by development of bacterial pneumonia.

Symptoms

Pneumonia is suspected in any patient who has fever, cough, chest pain, shortness of breath, and increased respirations (number of breaths per minute). Fever with a shaking chill is even more suspicious. Many patients cough up clumps of sputum, commonly known as spit. These secretions are produced in the alveoli during an infection or other inflammatory condition. They may appear streaked with pus or blood. Severe pneumonia results in the signs of oxygen deprivation. This includes blue appearance of the nail beds or lips (**cyanosis**).

The invading organism causes symptoms, in part, by provoking an overly-strong immune response in the lungs. In other words, the immune system, which should help fight off infections, kicks into such high gear, that it damages the lung tissue and makes it more susceptible to infection. The small blood vessels in the lungs (capillaries) become leaky, and protein-rich fluid seeps into the alveoli. This results in less functional area for oxygen-carbon dioxide exchange. The patient becomes relatively oxygen deprived, while retaining potentially damaging carbon dioxide. The patient breathes faster and faster, in an effort to bring in more oxygen and blow off more carbon dioxide.

Mucus production is increased, and the leaky capillaries may tinge the mucus with blood. Mucus

plugs actually further decrease the efficiency of gas exchange in the lung. The alveoli fill further with fluid and debris from the large number of white blood cells being produced to fight the infection.

Consolidation, a feature of bacterial pneumonias, occurs when the alveoli, which are normally hollow air spaces within the lung, instead, become solid due to quantities of fluid and debris.

Viral pneumonias and mycoplasma pneumonias do not result in consolidation. These types of pneumonia primarily infect the walls of the alveoli and the stroma of the lung.

Severe acute respiratory syndrome (SARS)

Severe acute respiratory syndrome, or SARS, is a contagious and potentially fatal disease that first appeared in the form of a multi-country outbreak in early February 2003. Later that month, the CDC began to work with the World Health Organization (WHO) to investigate the cause(s) of SARS and to develop guidelines for **infection control**. SARS has been described as an “atypical pneumonia of unknown etiology;” by the end of March 2003, the disease agent was identified as a previously unknown coronavirus.

The early symptoms of SARS include a high fever with chills, headache, **muscle cramps**, and weakness. This early phase is followed by respiratory symptoms, usually a dry cough and painful or difficult breathing. Some patients require mechanical ventilation. The mortality rate of SARS is thought to be about 3%.

Diagnosis

For the most part, diagnosis is based on the patient's report of symptoms, combined with examination of the chest. Listening with a stethoscope will reveal abnormal sounds, and tapping on the patient's back (which should yield a resonant sound due to air filling the alveoli) may instead yield a dull thump if the alveoli are filled with fluid and debris.

Laboratory diagnosis can be made of some bacterial pneumonias by obtaining a sputum specimen and staining the sputum with special chemicals and looking at it under a microscope. Identification of the specific type of bacteria may require culturing the sputum (using the sputum sample to grow greater numbers of the bacteria in a lab dish).

X-ray examination of the chest may reveal certain abnormal changes associated with pneumonia.

Localized shadows obscuring areas of the lung may indicate a bacterial pneumonia, while streaky or patchy appearing changes in the x-ray picture may indicate viral or mycoplasma pneumonia. These changes on x ray, however, are known to lag in time behind the patient's actual symptoms.

The doctor may do a **bronchoscopy** (visualizing inside the airway via a scope), or may remove a small piece of lung tissue (transbronchial biopsy) for microscopic examination and cultures. If the patient's condition continues to worsen, the doctor may remove additional lung tissue via thoracic needle biopsy or open **lung biopsy**, for microscopic analysis and cultures.

Treatment

Prior to the discovery of penicillin antibiotics, bacterial pneumonia was almost always fatal. Today, antibiotics, especially given early in the course of the disease, are very effective against bacterial causes of pneumonia. Erythromycin and tetracycline improve recovery time for symptoms of mycoplasma pneumonia. They do not, however, eradicate the organisms. Amantadine and acyclovir may be helpful against certain viral pneumonias.

A newer antibiotic named linezolid (Zyvox) is being used to treat penicillin-resistant organisms that cause pneumonia. Linezolid is the first of a new line of antibiotics known as oxazolidinones. Another new drug known as ertapenem (Invanz) is reported to be effective in treating bacterial pneumonia.

Patients may also be given fluids and possibly drug therapy to thin mucus secretions (mucolytic agents) or medication to open the airways of the lung (brochodilators). **Cough suppressants** may be given as well as pain medication and fever-reducing medication. Hospitalized patients often receive oxygen, respiratory therapy, and intravenous antibiotics and fluids.

Pneumonia in cancer patients must be treated promptly in order to speed recovery and prevent complications that could arise if the inflammation were allowed to linger. Treatment always includes bed rest and coughing to expel phlegm and other fluids from the lungs (productive cough). To determine which course of treatment would be most appropriate, a doctor considers when symptoms first appeared, what pattern the illness has followed, and whether cancer or its treatments have diminished the patient's infection-fighting ability (immune response).

KEY TERMS

Acute respiratory distress syndrome—A serious reaction to various forms of injuries to the lung, which is characterized by inflammation of the lung, leading to impaired gas exchange and release of inflammatory mediators causing inflammation and low blood oxygen and frequently resulting in multiple organ failure. This condition is life threatening and often lethal, usually requiring mechanical ventilation and admission to an intensive care unit.

Alveoli—The little air sacs clustered at the ends of the bronchioles, in which oxygen-carbon dioxide exchange takes place.

Aspiration—A situation in which solids or liquids which should be swallowed into the stomach are instead breathed into the respiratory system.

Bronchoscopy—The examination of the bronchi (the main airways of the lungs) using a flexible tube (bronchoscope). Bronchoscopy helps to evaluate and diagnose lung problems, assess blockages, obtain samples of tissue and/or fluid, and/or to help remove a foreign body.

CD4 count—A measure of the strength of the immune system. HIV continually kills CD4 cells. Over time, the body can not replace these lost CD4 cells and their number declines. As this happens, the body becomes more susceptible to infections. A normal CD4 count is 1000. The body starts to get more frequent common infections at around a count of

400. At around a CD4 count of 200, the body becomes susceptible to many unusual infections. It is best to start medications for HIV before the CD4 count drops below 200 to prevent these infections from developing.

Cilia—Hair-like projections from certain types of cells.

Consolidation—A condition in which lung tissue becomes firm and solid rather than elastic and air-filled because it has accumulated fluids and tissue debris.

Coronavirus—One of a family of RNA-containing viruses known to cause severe respiratory illnesses. In March 2003, a previously unknown coronavirus was identified as the causative agent of severe acute respiratory syndrome, or SARS.

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Sepsis—Presence of various pus-forming and other pathogenic organisms, or their toxins, in the blood or tissues.

Sputum—Material produced within the alveoli in response to an infectious or inflammatory process.

Stroma—A term used to describe the supportive tissue surrounding a particular structure. An example is that tissue which surrounds and supports the actually functional lung tissue.

Prognosis

Prognosis varies according to the type of organism causing the infection. Recovery following pneumonia with *Mycoplasma pneumoniae* is nearly 100%. *Staphylococcus pneumoniae* has a death rate of 30–40%. Similarly, infections with a number of gram negative bacteria (such as those in the gastrointestinal tract which can cause infection following aspiration) have a death rate of 25–50%. *Streptococcus pneumoniae*, (also referred to as **pneumococcal pneumonia**), the most common organism causing pneumonia, produces a death rate of about 5%. More complications occur in the very young or very old individuals who have multiple areas of the lung infected simultaneously. Individuals with other chronic illnesses (including **cirrhosis** of the liver, congestive **heart failure**, individuals without a functioning spleen, and individuals who have other diseases that result in a weakened

immune system, experience complications. Patients with immune disorders, various types of cancer, transplant patients, and AIDS patients also experience complications.

The chances of an early recovery (within two to three weeks) from pneumonia are enhanced if the pneumonia is detected early, if the patient has a strong immune system, if the infection has not spread throughout the body, and if the patient is not suffering from other diseases.

Prevention

Measures that can be taken to prevent pneumonia include frequent washing of hands, elimination of the use of tobacco (which damages the ability of the lungs to withstand infections), and wearing of masks in dusty or moldy areas. Since pneumonia often follows common respiratory infections such as the cold or flu,

an important preventive measure is to be alert to any symptoms of respiratory illness that last for more than a few days. The practice of deep breathing for patients recovering in the hospital from various diseases or surgeries is recommended to help prevent them from developing pneumonia.

Because many bacterial pneumonias occur in patients who are first infected with the influenza virus (the flu), yearly **vaccination** against influenza can decrease the risk of pneumonia for certain patients. This is particularly true of the elderly and people with chronic diseases (such as asthma, cystic fibrosis, other lung or heart diseases, **sickle cell disease**, diabetes, **kidney disease**, and forms of cancer).

A specific vaccine against *Streptococcus pneumoniae* is very protective, and should also be administered to patients with chronic illnesses.

Patients who have decreased immune resistance are at higher risk for infection with *Pneumocystis carinii*. They are frequently put on a regular drug regimen of trimethoprim sulfa and/or inhaled pentamidine to avoid pneumocystis pneumonia.

The flu vaccine helps prevent pneumonia caused by influenza viruses. This vaccine must be given yearly to protect against new viral strains.

Additional preventive therapy may be necessary for:

- AIDS patients with CD4 counts below 200
- People on chronic high-doses of corticosteroids
- People who have had previous episodes of PCP

Health care team roles

In most cases, a diagnosis of pneumonia is made in a physician's office, a general medical clinic, or emergency room by a primary care practitioner. Children and adolescents with pneumonia are most likely to be diagnosed by their primary care physician or pediatrician.

When patients are hospitalized for pneumonia, good nursing assessment and observation are primary requirements. These include monitoring vital signs, including oxygen saturation (the amount of oxygen circulating in the blood), encouraging the patient to move, breathe deeply, cough, and get out of bed with assistance (if indicated) to facilitate good lung expansion. The nurse should also provide education to the patient about the importance of coughing, breathing deeply, and taking in adequate fluid.

When at home, patient should be encouraged to drink fluids to loosen secretions and bring up phlegm.

Both patients and care givers should be made aware of potential **drug interactions** with other medications that the patient may be taking (for example, warfarin and antibiotics). Regular communication between the physician and the care giver is essential.

Resources

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ORGANIZATIONS

- American Lung Association, 1301 Pennsylvania Ave. NW, Washington, DC, 20004, (202) 785 3355 , (202) 452 1805, <http://www.lungusa.org>.
- American Thoracic Society, 61 Broadway, 6th Floor, New York, NY, 10006-2755, (212) 315-8600, <http://www.thoracic.org>.
- Centers for Disease Control and Prevention, 1600 Clifton Rd., NE, Atlanta, GA, 30333, (404) 639-3311, (800) 311-3435, <http://www.cdc.gov.org>.

National Heart, Lung, and Blood Institute (NHLBI), P. O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3255 (TTY), <http://www.nhlbi.nih.gov>.
 World Health Organization, Communicable Diseases, 20 Avenue Appia, 1211, Geneva 27, Switzerland, +4122791 4140, <http://www.who.int/gtb>.

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Pneumonitis see **Pneumonia**

Pneumothorax

Definition

Pneumothorax is a collection of air or gas in the chest or pleural space that causes part or all of a lung to collapse.

Description

Normally, the pressure in the lungs is greater than the pressure in the pleural space surrounding the lungs. However, if air enters the pleural space, the pressure in the pleura then becomes greater than the pressure in the lungs, causing the lung to collapse partially or completely. Pneumothorax can be either spontaneous or due to trauma.

If a pneumothorax occurs suddenly or for no known reason, it is called a spontaneous pneumothorax. This condition most often strikes tall, thin men between the ages of 20 to 40. In addition, people with lung disorders, such as **emphysema**, **cystic fibrosis**, and **tuberculosis**, are at higher risk for spontaneous pneumothorax. Traumatic pneumothorax is the result of accident or injury due to medical procedures performed to the chest cavity, such as **thoracentesis** or mechanical ventilation. Tension pneumothorax is a serious and potentially life-threatening condition that may be caused by traumatic injury, chronic lung disease, or as a complication of a medical procedure. In this type of pneumothorax, air enters the chest cavity, but cannot escape. This greatly increased pressure in the pleural space causes the lung to collapse completely, compresses the heart, and pushes the heart and associated blood vessels toward the unaffected side.



An x ray of a patient undergoing pneumothorax treatment. ECG electrodes attached to chest monitor heartbeat while endotracheal tube is inserted in windpipe. (Photo Researchers, Inc.)

Causes and symptoms

The symptoms of pneumothorax depend on how much air enters the chest, how much the lung collapses, and the extent of lung disease. Symptoms include the following, according to the cause of the pneumothorax:

- Spontaneous pneumothorax. Simple spontaneous pneumothorax is caused by a rupture of a small air sac or fluid-filled sac in the lung. It may be related to activity in otherwise healthy people or may occur during scuba diving or flying at high altitudes. Complicated spontaneous pneumothorax, also generally caused by rupture of a small sac in the lung, occurs in people with lung diseases. The symptoms of complicated spontaneous pneumothorax tend to be worse than those of simple pneumothorax, due to the underlying lung disease. Spontaneous pneumothorax is characterized by dull, sharp, or stabbing chest pain that begins suddenly and becomes worse with deep breathing or coughing. Other symptoms are shortness of breath, rapid breathing, abnormal breathing

movement (that is, little chest wall movement when breathing), and cough.

- **Tension pneumothorax.** Following trauma, air may enter the chest cavity. A penetrating chest wound allows outside air to enter the chest, causing the lung to collapse. Certain medical procedures performed in the chest cavity, such as thoracentesis, also may cause a lung to collapse. Tension pneumothorax may be the immediate result of an injury; the delayed complication of a hidden injury, such as a fractured rib, that punctures the lung; or the result of lung damage from asthma, chronic bronchitis, or emphysema. Symptoms of tension pneumothorax tend to be severe with sudden onset. There is marked anxiety, distended neck veins, weak pulse, decreased breath sounds on the affected side, and a shift of the mediastinum to the opposite side.

Diagnosis

To diagnose pneumothorax, it is necessary for the health care provider to listen to the chest (auscultation) during a **physical examination**. By using a stethoscope, the physician may note that one part of the chest does not transmit the normal sounds of breathing. A **chest x ray** will show the air pocket and the collapsed lung. An electrocardiogram (ECG) will be performed to record the electrical impulses that control the heart's activity. Blood samples may be taken to check for the level of arterial blood gases.

Treatment

A small pneumothorax may resolve on its own, but most require medical treatment. The object of treatment is to remove air from the chest and allow the lung to re-expand. This is done by inserting a needle and syringe (if the pneumothorax is small) or chest tube through the chest wall. This allows the air to escape without allowing any air back in. The lung will then re-expand itself within a few days. Surgery may be needed for repeat occurrences.

Prognosis

Most people recover fully from spontaneous pneumothorax. Up to half of patients with spontaneous pneumothorax experience recurrence. Recovery from a collapsed lung generally takes one to two weeks. Tension pneumothorax can cause **death** rapidly due to inadequate heart output or insufficient blood oxygen (hypoxemia), and must be treated as a medical emergency.

KEY TERMS

Electrocardiogram—A test that provides a typical record of normal heart action.

Mediastinum—The space between the right and left lung.

Pleural—Pleural refers to the pleura or membrane that enfolds the lungs.

Thoracentesis—Also called a pleural fluid tap, this procedure involves aspiration of fluid from the pleural space using a long, thin needle inserted between the ribs.

Prevention

Preventive measures for a non-injury related pneumothorax include stopping **smoking** and seeking medical attention for respiratory problems. If the pneumothorax occurs in both lungs or more than once in the same lung, surgery may be needed to prevent it from occurring again.

ORGANIZATIONS

American Association for Respiratory Care, 9425 N. MacArthur Blvd, Suite 100, Irving, TX, 75063-4706, (972) 243-2272, (972) 484-2720, info@aarc.org, http://www.aarc.org.

American Lung Association, 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20001, (202) 758-3355, (202) 452-1805, (800) 548-8252, info@lungusa.org, http://www.lungusa.org.

Lorraine Steefel, RN

Podiatry see **Foot care**

Poison ivy and poison oak

Definition

Poison ivy and poison oak are plants that cause an allergic skin reaction in most people who are exposed to them.

Demographics

An estimated 85 percent of the population is allergic to the urushiol oil found in poison ivy, oak, and sumac, according to the American Academy of Dermatology. Annually, up to 50 million Americans develop a



Poison oak plant. (© iStockPhoto/Joe Potato Photo.)

poison ivy, oak, or sumac rash. The chance of developing an allergic sensitivity to these poison plants decreases with age, and adults who have never been exposed to urushiol only have a 50 percent chance of developing **contact dermatitis** when exposed to poison ivy, oak, or sumac. It is possible for children who are highly reactive to urushiol to grow into adults who are barely sensitive to poison ivy, oak, or sumac, regardless of how many times they have been exposed to the plant oil.

Description

Poison ivy, which is generally thought of as a climbing vine, can also grow as a shrub or bush. It has leaves that are elliptical in shape and grow in groups of three on a stem. Poison ivy is common in the United States, except in the southwest, Alaska, and Hawaii. Poison oak, which grows as a shrub, has leaves that are shaped like oak leaves and also grow in groups of three to a stem. Poison oak is common in the United States, especially on the west coast from Mexico to Canada.

Not everyone is sensitive to poison ivy and poison oak; however, nine out of ten people who come in contact with either of the plants will have an allergic reaction to some degree. All parts of the plants are poisonous and the amount of time it takes for an allergic reaction to develop varies from person to person. The extent and severity of the reaction depends on the length of exposure, type of contact, and how sensitive the person is to the plants. If a person is going to have an allergic reaction, it will usually occur within one or two days of exposure. However, some people have a reaction within an

hour, whereas others don't experience a reaction until five days after the exposure.

Causes and symptoms

The substance that causes the allergic reaction is the same for both plants. It is an oily resin called urushiol. It only takes a small amount of the resin to cause a reaction. The resin can be transferred to the skin by directly touching the plant or indirectly by coming in contact with something that has touched the plant, such as tools, animals, or clothing. Although animals are rarely affected, they can carry the resin on their fur and transfer it to humans. According to the experts at the University of Maryland Medical Center, the "chemical [resin] can remain active for more than a year."

The symptoms for poison ivy and poison oak are the same. Usually the first symptoms to appear are itchiness and swelling in the areas of contact. The itchy rash that follows is made up of small pimple-like bumps (sometimes referred to as papules), as well as blisters that later break open, ooze, and crust over.

Diagnosis

A diagnosis is made based on the symptoms and a **physical examination** of the patient.

Treatment

Anyone who comes in contact with either plant should wash the exposed area with soap and water immediately. Taking a bath immediately after contact is not recommended, because that could spread the resin to other areas of the body. All clothing, including shoes and shoelaces, should be removed carefully and either washed separately or discarded.

For minor cases, hydrocortisone cream and Calamine lotion can provide relief until the symptoms disappear. Over-the-counter Benadryl capsules help with the **itching**. Some people find oatmeal or baking soda baths to be soothing as well. Oral **steroids**, such as prednisone, are available for more serious cases, especially those affecting the face, eyes, mouth, or genitals. If signs of infection develop, such as pus and a **fever**, patients should contact their doctors.

Patients should consult their physicians before they use any ointments that contain benzocaine or zirconium, because they can cause an allergic reaction that worsens the condition. Antihistamine



Poison ivy plant. (© iStockPhoto/norcon.)

ointments are not recommended for the same reason. The experts at the Alabama Cooperative Extension System caution that “some people have severe allergic reactions to these plants and can have swelling in the throat, breathing problems, weakness, **dizziness**, and bluish lips.” Emergency medical care should be sought if any serious reactions occur.

Prognosis

In most cases, the condition goes away in two weeks.

Prevention

The best prevention is know what the plants look like and to avoid them. A common saying should be kept in mind: Leaves of three, let them be.

People who plan to be in an area where poison ivy and poison oak might be found should wear protective clothing, such as long-sleeved shirts and long pants.

Eradication of the plants should be handled with care. As stated by the experts at the Alabama Cooperative Extension System, “burning can be dangerous and is not recommended for disposal or as a control measure, because the toxic oil from the plant can be carried in the smoke.” Instead they recommend spraying the plants with glyphosate, which is commonly known as the brands Roundup or Kleenup.

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Lee Ann Paradise,
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Poisoning

Definition

Poisoning occurs when any substance interferes with normal body functions after it is swallowed, inhaled, injected, or absorbed. The branch of medicine that deals with the detection and treatment of poisons is known as toxicology.

Demographics

Poisonings are a common occurrence and can be intentional or unintentional. About 10 million cases of poisoning occur in the United States each year. In 80% of the cases, the victim is a child under the age of five. About 50 children die each year from poisonings. Curiosity, inability to read warning labels, a desire to imitate adults, and inadequate supervision lead to childhood poisonings.

The elderly are the second most likely group to be poisoned. Mental confusion, poor eyesight, and the use of multiple drugs are the leading reasons that this group has a high rate of accidental poisoning. A substantial number of poisonings also occur as **suicide** attempts or drug overdoses.

According to the Centers for Disease Control (CDC), In 2008, unintentional poisoning caused about 732,316 emergency department (ED) visits, and 23% of those unintentional poisonings (166,015) resulted in hospitalization or transfer to another facility.

Common household, industrial, and agricultural products containing toxic substances

Alcohol (rubbing)	Dieffenbachia	Mercury
Antifreeze	Disinfectants/air fresheners	Metal primers
Arsenic	Drain openers	Metalworking materials
Art and craft supplies	English nightshade	Mothballs
Automotive fluids	Ethanol	Oven cleaners
Batteries, automotive	Flea collars/insect repellent	Paints, oil-based or alkyls
Batteries, household	Floor/furniture polish	Paints, water-based or latex
Building products	Foxglove	Paint strippers/thinners
Cleaning products	Gasoline	Pesticides
Cosmetics/personal care items	Glues/adhesives	Stains/finishes
Cyanide	Hemlock	Strychnine
Daffodil bulbs	Kerosene	Wood preservatives

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

Description

Poisons are common in the home and workplace, yet there are basically two major types. One group consists of products that were never meant to be ingested or inhaled, such as shampoo, paint thinner, pesticides, houseplant leaves, and carbon monoxide. The other group contains products that can be ingested in small quantities, but which are harmful if taken in large amounts, such as pharmaceuticals, medicinal herbs, or alcohol. Other types of poisons include the bacterial toxins that cause **food poisoning**, such as *Escherichia coli*; heavy metals, such as the lead found in the paint on older houses; and the venom found in the **bites and stings** of some animals and insects. The staff at a poison control center and emergency room doctors have the most experience diagnosing and treating poisoning cases.

Causes and symptoms

The effects of poisons are as varied as the poisons themselves. However, the exact mechanisms of only a few are understood. Some poisons interfere with metabolism. Others destroy the liver or kidneys. Some examples include heavy metals and some **pain** relief medications, including **acetaminophen** (Tylenol) and nonsteroidal anti-inflammatory drugs (Advil, Ibuprofen). A poison may severely depress the central nervous system, leading to **coma** and eventual respiratory and circulatory failure. Potential poisons in this category include anesthetics (e.g. ether and chloroform), opiates (e.g., morphine and codeine), and **barbiturates**. Some poisons directly affect the respiratory and circulatory systems. Carbon monoxide causes **death** by binding with hemoglobin that normally transports oxygen throughout the body. Certain corrosive vapors trigger the body to flood

the lungs with fluids, effectively drowning the person. Cyanide interferes with respiration at the cellular level. Another group of poisons interferes with the electrochemical impulses that travel between neurons in the nervous system. Yet another group, including **cocaine**, ergot, strychnine, and some snake venoms, causes potentially fatal seizures.

Severity of symptoms can range from **headache** and **nausea** to convulsions and death. The type of poison; the amount and time of exposure; and the age, size, and health of the victim are all factors that determine the severity of symptoms and the chances for recovery.

Plant poisoning

There are more than 700 species of poisonous plants in the United States. Plants are second only to medicines in causing serious poisoning in children under age five. There is no way to tell by looking at a plant if it is poisonous. Some plants, such as the yew shrub, are almost entirely toxic: needles, bark, seeds, and berries. In other plants, only certain parts are poisonous. The bulb of the hyacinth and daffodil are toxic, but the flowers are not, while the flowers of the jasmine plant are the poisonous part. Moreover, some plants are confusing because portions of them are eaten as food while other parts are poisonous. For example, the fleshy stem (tuber) of the potato plant is nutritious. However, its roots, sprouts, and vines are poisonous. The leaves of tomatoes are poisonous, while the fruit is not. Rhubarb stalks are good to eat, but the leaves are poisonous. Apricots, cherries, peaches, and apples all produce healthful fruit, but their seeds contain a form of cyanide that can kill a child if chewed in sufficient quantities. One hundred milligrams of moist, crushed apricot seeds can produce 217 mg of cyanide.

Common houseplants that contain some poisonous parts include:

- Aloe
- Amaryllis
- Cyclamen
- Dumb cane (also called Dieffenbachia)
- Philodendron

Common outdoor plants that contain some poisonous part include:

- Bird of paradise flower
- Buttercup
- Castor bean
- Chinaberry tree
- Daffodil
- English ivy
- Eucalyptus
- Foxglove
- Holly
- Horse chestnut
- Iris
- Jack-in-the-pulpit
- Jimsonweed (also called thornapple)
- Larkspur
- Lily-of-the-valley
- Morning glory
- Nightshade (several varieties)
- Oleander
- Potato
- Rhododendron
- Rhubarb
- Sweet pea
- Tomato
- Wisteria
- Yew

Symptoms of plant poisoning range from irritation of the skin or mucous membranes of the mouth and throat to nausea, **vomiting**, convulsions, irregular heartbeat, and even death. It is often difficult to tell if a person has eaten a poisonous plant because there are no tell-tale empty containers and no unusual lesions or odors around the mouth.

Many cases of plant poisoning involve plants that contain hallucinogens, such as peyote cactus buttons, certain types of mushrooms, and **marijuana**. A recent case of plant poisoning in France concerned *Datura*, or moonflower, a plant that has become popular with

young people trying to imitate Native American **puberty** rites.

Other cases of plant poisoning result from the use of herbal dietary supplements that have been contaminated by toxic substances. The Food and Drug Administration (FDA) has the authority to monitor herbal products on the market and issue warnings about accidental poisoning or other adverse affects associated with these products. For example, in 2002 a manufacturer of nettle capsules found to contain lead recalled the product following a warning from the FDA. Other dietary supplements have been found to contain small quantities of prescription medications or even toxic plants.

Household chemicals

Many products used daily in the home are poisonous if swallowed. These products often contain strong acids or strong bases (alkalis). Toxic household cleaning products include:

- ammonia
- bleach
- dishwashing liquids
- drain openers
- floor waxes and furniture polishes
- laundry detergents, spot cleaners, and fabric softeners
- mildew removers
- oven cleaners
- toilet bowl cleaners

Personal care products found in the home can also be poisonous. These include:

- deodorant
- hairspray
- hair straighteners
- nail polish and polish remover
- perfume
- shampoo

Signs that a person has swallowed one of these substances include evidence of an empty container nearby, nausea or **vomiting**, and **burns** on the lips and skin around the mouth if the substance was a strong acid or alkali. The chemicals in some of these products may leave a distinctive odor on the breath.

Pharmaceuticals

Both over-the-counter and prescription medicines can help the body heal if taken as directed.

However, when taken in large quantities, or with other drugs where there may be an adverse interaction, they can act as poisons. Drug overdoses, both accidental and intentional, are the leading cause of poisoning in adults. Medicinal herbs should be treated like pharmaceuticals and taken only in designated quantities under the supervision of a knowledgeable person. Herbs that have healing qualities when taken in small doses can be toxic in larger doses, or may interact with prescription medications in unpredictable ways.

Drug overdoses cause a range of symptoms, including excitability, sleepiness, confusion, unconsciousness, rapid heartbeat, convulsions, nausea, and changes in blood pressure. The best initial evidence of a **drug overdose** is the presence of an empty container near the victim.

Other causes of poisonings

People can be poisoned by fumes they inhale. Carbon monoxide is the most common form of inhaled poison. Other toxic substances that can be inhaled include:

- farm and garden insecticides and herbicides
- gasoline fumes
- insect repellent
- paint thinner fumes

Diagnosis

Initially, poisoning is suspected if the victim shows changes in behavior and signs or symptoms previously described. **Hallucinations** or other psychiatric symptoms may indicate poisoning by a hallucinogenic plant. Evidence of an empty container or information from the victim are helpful in determining exactly what substance has caused the poisoning. Some acids and alkalis leave burns on the mouth. Petroleum products, such as lighter fluid or kerosene, leave a distinctive odor on the breath. The vomit may be tested to determine the exact composition of the poison. Once hospitalized, the patient may be given blood and urine tests to determine his or her metabolic condition.

Treatment

Treatment for poisoning depends on the poison swallowed or inhaled. Contacting the poison control center or hospital emergency room is the first step in getting proper treatment. The poison control center's telephone number is often listed with emergency numbers on the inside cover of the telephone book, or it can be reached by dialing the operator. The poison control center will ask for specific information about

the victim and the poison, then give appropriate **first aid** instructions. If the patient is to be taken to a hospital, a sample of vomit and the poison container should be taken along, if they are available.

Most cases of plant poisoning are treated by inducing vomiting, if the patient is fully conscious. Vomiting can be induced by taking syrup of **ipecac**, an over-the-counter emetic available at any pharmacy.

For acid, alkali, or petroleum product poisonings, the patient should not vomit. Acids and alkalis can burn the esophagus if they are vomited, and petroleum products can be inhaled into the lungs during vomiting, resulting in **pneumonia**.

Once under medical care, doctors have the option of treating the patient with a specific remedy to counteract the poison (antidote) or with **activated charcoal** to absorb the substance inside the patient's digestive system. In some instances, pumping the stomach may be required. This technique, which is known as gastric lavage, involves introducing 20–30 mL of tap water or nine percent saline solution into the patient's digestive tract and removing the stomach contents with a siphon or syringe. The process is repeated until the washings are free of poison. Medical personnel will also provide supportive care as needed, such as intravenous fluids or mechanical ventilation.

If the doctor suspects that the poisoning was not accidental, he or she is required to notify law enforcement authorities. Most cases of malicious poisonings concern family members or acquaintances of the victim, but the number of intentional random poisonings of the general public has increased in recent years. A case reported in 2003 involved the use of nicotine to poison 1700 pounds of ground beef in a Michigan supermarket. Over a hundred persons fell ill after eating the poisoned beef.

Prognosis

The outcome of poisoning varies from complete recovery to death, and depends on the type and amount of the poison, the health of the victim, and the speed with which medical care is obtained.

Prevention

Most accidental poisonings are preventable. The number of deaths of children from poisoning has declined from about 450 per year in the 1960s to less than 50 each year since the 1990s. This decline has occurred mainly because of better packaging of toxic materials and better public education.

KEY TERMS

Antidote—A medication or remedy for counteracting the effects of a poison.

Emetic—A medication or substance given to induce vomiting.

Gastric lavage—A technique for washing poison out of the stomach by instilling water or saline solution through a tube, removing the stomach contents by suction, and repeating the process until the washings are free of poison. The procedure is also called stomach pumping.

Toxicology—The branch of medicine that deals with the effects, detection, and treatment of poisons.

Actions to prevent poisonings include:

- removing plants that are poisonous
- keeping medicines and household chemicals locked and in a place inaccessible to children
- keeping medications in child-resistant containers
- never referring to medicine as “candy”
- keeping cleaners and other poisons in their original containers
- disposing of outdated prescription medicines
- not purchasing over-the-counter medications with damaged protective seals or packaging
- avoiding the use of herbal preparations not made by a reputable manufacturer

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ORGANIZATIONS

American Association of Poison Control Centers (AAPCC), 3201 New Mexico Ave., Suite 330, Washington, DC, 20016, (202) 362–7217, POISONING EMERGENCIES: (800) 222–1222, <http://www.aapcc.org>.

National Toxicology Program (NTP) of the National Institute of Environmental Health Sciences (NIEHS), PO Box 12233, Research Triangle Park, NC, 27709, (919) 541–3419, <http://wwwntp-server.niehs.nih.gov>.

U. S. Food and Drug Administration (FDA), 5600 Fishers Ln., Rockville, MD, 20857, (888) 463–6332, <http://www.fda.gov>.

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Polarity therapy

Definition

Polarity therapy is a holistic, energy-based system that includes bodywork, diet, **exercise**, and lifestyle counseling for the purpose of restoring and maintaining proper energy flows throughout the body. The underlying concept of polarity therapy is that all energy within the human body is based in electromagnetic force and that disease results from improperly dissipated energy.

Purpose

Polarity therapy unblocks and recharges the flow of life energy and realigns unbalanced energy as a means of eliminating disease. Patients learn to release

KEY TERMS

Apana—Life sustaining energy centered in the larger intestine; the fifth of the five airs of Ayurvedic philosophy; the life force governing expulsion activity.

Ayurveda—(Sanskrit, *Ayur*, life, and *veda*, knowledge) Translated as “knowledge of life” or “science of longevity.” It became established as the traditional Hindu system of medicine.

Caduceus—The ancient and universal symbol of medicine consisting of the winged staff of Mercury and two intertwining serpents.

Primary energy pattern—A spiral motion that radiates from the umbilicus; the energy pattern associated with a child in the womb.

Prana—Life sustaining energy centered in the human brain; the first of the five airs of Ayurvedic philosophy; the life force governing inspiration and the conscious intellect.

QV—Quantum vacuum, a theory coined by physicists, which defines the interactions of energy that combine to form reality.

Reflexology—Belief that reflex areas in the feet correspond to every part of the body, including organs and glands, and that stimulating the correct reflex area can affect the body part.

Samana—Life sustaining energy of the smaller intestine; the fourth of the five airs of Ayurvedic philosophy; the life force governing side-to-side motion.

Tridosha—The combination of three basic principles of energy, or biological humor, that comprise life, according to Ayurvedic philosophy.

Udana—Life sustaining energy of the diaphragm, the third of the five airs of Ayurvedic philosophy, the life force governing upward motion.

Vyana—Life sustaining energy of the heart and lungs; the second of the five airs of Ayurvedic philosophy; the life force governing circular motion.

tension by addressing the source of the **stress** and by maintaining a healthy demeanor accordingly.

This treatment may be effective to promote health and healing to anyone willing to embrace the appropriate lifestyle. Polarity therapy is reportedly effective for anyone who has been exposed to toxic poisons. Likewise, HIV-positive individuals may find comfort in polarity therapy. Additionally this is an appropriate therapy for relieving general stress, back **pain**, stomach cramps, and other recurring maladies and conditions.

Description

Origins

Austrian-American chiropractor, osteopath, and naturopath Randolph Stone (1888–1981) developed polarity therapy as an integration of Eastern and Western principles and techniques of healing. Stone discovered the ancient principles of the Ayurvedic philosophy in the course of his travels during a sojourn in India. On a life-long quest to learn the fundamentals of human vitality, he also studied **reflexology** and **traditional Chinese medicine**.

Stone became committed to the principles of **Ayurvedic medicine**, which he interpreted in conjunction with his scientific and medical knowledge to define polarity therapy. According to the philosophy

of Ayurved, which is based in a set of principles called the tridosha—the energy of the human body is centered in five organs or regions (the brain; the cardio-pulmonary [heart and lungs] region, the diaphragm, the smaller intestine, and the larger intestine). One of five airs or energy forms controls each respective region: prana in the brain, vyana in the heart and lungs, udana in the diaphragm, samana in the smaller intestine, and apana in the larger intestine. The five airs control all directional motion in the body, with each air in command of a different type of movement. Stone established further that the prana, centered in the brain, ultimately controlled the combined forces of the body. Any impediment or restriction to the flow of prana in turn affects the health of the entire body. The prana force is nurtured through the flow of food and air into the body as well as through our interactions with other living beings and through the intake of the five sensory organs.

Stone devoted much of his life to defining an elaborately detailed cause and effect relationship between the human anatomy and illness, based on the energy flow of the prana. He further attributed electromagnetic energy as the basis of the energy forces. He used the medical symbol of the Caduceus to define the patterns of the flow and described the energy movement in detail in charts of the human body. Polarity therapy is based in charted energy

flows. The primary energy pattern is defined in a spiral motion that radiates from the umbilicus and defines the original energy flow of the fetus in the womb.

After determining the exact source of a patient's energy imbalance, the therapist begins the first of a series of bodywork sessions designed to rechannel and release the patient's misdirected prana. This therapy, akin to massage, is based in energetic pressure and involves circulating motions. In performing the regimen, the therapist pays strict attention to the pressure exerted at each location—even to which finger is used to apply pressure at any given point of the patient's anatomy. This technique, which comprises the central regimen or focal point of polarity therapy is very gentle and is unique to polarity therapy. It typically involves subtle rocking movements and cranial holds to stimulate body energy. Although firm, deep pushing touches are employed in conjunction with the massage technique, the polarity therapist never exerts a particularly forceful contact.

To support the bodywork, the therapist often prescribes a diet for the patient, to encourage cleansing and eliminate waste. The precepts of polarity therapy take into consideration specific interactions between different foods and the human energy fields.

Likewise, a series of exercises is frequently prescribed. These exercises, called polarity **yoga** include squats, stretches, rhythmic movements, deep breathing, and expression of sounds. They can be both energizing and relaxing. Counseling may be included whenever appropriate as a part of a patient's highly individual therapy regimen to promote balance.

Preparations

Therapists take a comprehensive case history from every patient prior to beginning treatment. This preliminary verbal examination often monopolizes the first therapy session. Depending upon circumstances, a therapist might have a need to assess the patient's physical structural balance through observation and **physical examination**.

Precautions

Polarity therapy is safe for virtually anyone, even the elderly and the most frail patients, because of the intrinsic gentleness of the **massage therapy**.

Side effects

Highly emotional releases of energy (laughter, tears, or a combination of both) are associated with this therapy.

Research and general acceptance

This is a complementary therapy of holistic, spiritually based treatment, which may be used in conjunction with a medical approach. Polarity therapy is practiced worldwide, but the majority of practitioners are based in the United States. Modern physicists employ concepts similar to Stone's basic theories of polarity in defining the quantum vacuum (QV) as a foundation of all reality. Still, this holistic regimen had not achieved the widespread acceptance anticipated by Stone before his death in 1981.

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ORGANIZATIONS

American Polarity Therapy Association, 122 N. Elm Street, Suite 512, Greensboro, NC, 27401, (336) 574-1121, (336) 574-1151, APTAoffices@polaritytherapy.org, <http://www.polaritytherapy.org>.

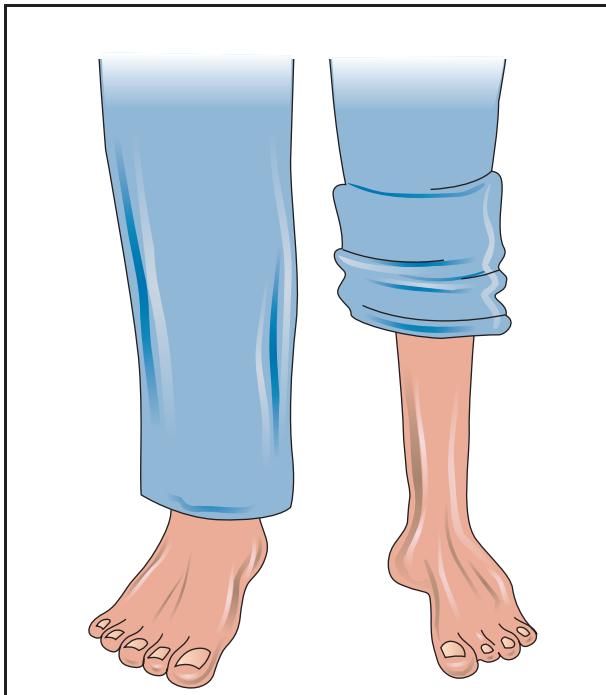
Trans-Hyperboreau Institute of Science, P.O. Box 2344, Sausalito, CA, 94966, (415) 331-0230, (415) 331-0231, (800) 485-8095.

Gloria Cooksey

Polio

Definition

Polio, or poliomyelitis, is an **infectious disease** caused by a virus that normally lives in the human digestive tract. About 90 percent of persons infected by the virus have no symptoms at all; in the other 10 percent, the polio virus causes an infection with symptoms ranging from a mild flu-like illness to **paralysis** of the lower limbs or **death** from paralysis of the muscles that control breathing. The term poliomyelitis comes from the Greek words *polio*, meaning gray, and *myelon*, referring to the spinal cord. The term is accurate, as an important consequence of the disease is the involvement of the spinal cord.



In its most severe form, polio causes paralysis of the muscles of the legs, arms, and respiratory system. All muscle tone is lost in the affected limb, and the muscle becomes flaccid and begins to atrophy, as shown in the illustration above.

(Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Demographics

Polio was widespread in the developed countries of Europe and North America in the first part of the twentieth century. The epidemics not only became more severe, but also affected adolescents and adults rather than mostly children. The older average age of patients was also marked by increased severity of symptoms. Since the introduction of effective vaccines, paralytic polio is almost unknown in the United States except among recent immigrants and other groups (such as the Amish) that do not routinely participate in community-wide **vaccination** programs. According to the Centers for Disease Prevention and Control (CDC), the incidence rate has been less than 0.01 cases per 100,000 people in the United States since 1965. The last case of wild-type polio in the country was reported in 1979. Only a few cases of paralytic poliomylitis are reported each year in the United States.

Worldwide, polio epidemics are most common in tropical countries during the months of July through September. Both sexes and all races are equally likely to get the disease if they are not protected by immunization.

There is hope that polio will soon follow **smallpox** as a disease that humankind has completely wiped out. In 1994 both North and South America were declared polio-free, followed by Australia, Japan, China and other countries around the Pacific Ocean in 2000, and Europe in 2002.

Description

There are three known types of polioviruses (called 1, 2, and 3), each causing a different strain of the disease. All are members of the viral family of enteroviruses (viruses that infect the gastrointestinal tract). Type 1 is the cause of epidemics and many cases of paralysis, which is the most severe manifestation of the infection. The virus is usually a harmless parasite of human beings.

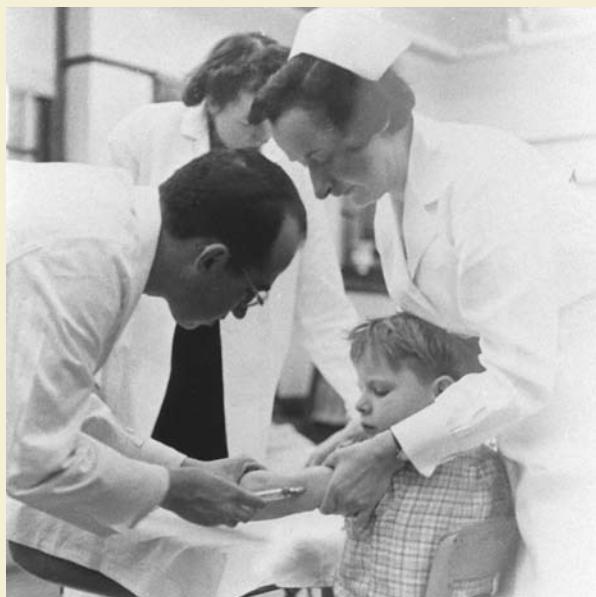
The most recent statistics indicate that of acute poliovirus infections, 4–8% lead only to nonspecific illness; 1–2% of infections finally result in neurologic symptoms. When the poliovirus does reach the central nervous system, inflammation and destruction of the spinal cord motor cells (anterior horn cells) occurs, which prevents them from sending out impulses to muscles. Loss of impulse transmission causes the muscles to become limp or soft and they cannot contract. This condition is referred to as flaccid paralysis and is the type found in polio. The extent of the paralysis depends on where the virus strikes and the number of cells that it destroys. Usually, some of the limb muscles are paralyzed; the abdominal muscles or muscles of the back may be paralyzed, affecting the person's posture. The neck muscles may become too weak for the head to be lifted. Paralysis of the face muscles may cause the mouth to twist or the eyelids to droop. Life may be threatened if paralysis of the throat or of the breathing muscles occurs.

Risk factors

Human beings are the only natural host of polioviruses; these viruses are not transmitted by animals. Some people are more likely than others to develop the paralytic form of the disease if they do become infected:

- Young children.
- Elderly adults.
- People who engage in hard physical labor or strenuous exercise.
- People who have recently had a tonsillectomy or dental surgery.
- Pregnant women.

DR. JONAS E. SALK (1914–1995)



(The Library of Congress.)

Jonas Salk was born in New York, New York, on October 28, 1914. He received his medical degree from New York University in 1939. In 1942, Salk began working for a former teacher, Thomas Francis, Jr., to produce

influenza vaccines, a project that continued until 1949. That year, as a research professor, Salk began a three-year project sponsored by the National Foundation for Infantile Paralysis, also known as the March of Dimes. Caused by the poliomyelitis virus, polio was also known as infantile paralysis. Periodic outbreaks of the disease, which attacks the nervous system, caused death or a lifetime of paralysis, especially in children. It was a difficult disease to study because sufficient viruses could not be obtained. Unlike bacteria, which can be grown in cultures, viruses need living tissue on which to grow. Once a method for preparing viruses was discovered and improved, sufficient viruses became available for research.

Salk first set out to confirm that there were three virus types responsible for polio and then began to experiment with ways to kill the virus and yet retain its ability to produce an immune response. By 1952, he had produced a dead virus vaccine that worked against the three virus types. He began testing. First the vaccine was tested on monkeys, then on children who had recovered from the disease, and finally on Salk's own family and children, none of whom had ever had the disease. Following large-scale trials in 1954, the vaccine was finally released for public use in 1955. The Salk vaccine was not the first vaccine against polio, but it was the first to be found safe and effective. By 1961, there was a 96 percent reduction in polio cases in the United States.

- People who travel frequently to areas where polio is still endemic. There are six such countries as of 2009, according to the World Health Organization (WHO): Afghanistan, Egypt, India, Niger, Nigeria, and Pakistan.
- An immune system weakened by HIV or certain types of cancer treatment.

Causes and symptoms

Polio is caused by a virus that enters the mouth through food or water that has been contaminated by fecal matter. It is an extremely contagious illness; anyone living with a recently infected person is likely to become infected too. Although people carrying the poliovirus are most contagious for 7–10 days before and after symptoms (if any) appear, they can spread the virus for weeks in their bowel movements.

Once inside the body, the polio virus takes between 6 and 20 days to incubate. It finds its way to the tissues lining the throat and the intestinal tract, where it multiplies rapidly. After about a week in the intestines, the virus travels to the tonsils and the lymph nodes, where it

multiplies further and then enters the bloodstream. It can remain within the blood and lymphatic system for as long as 17 weeks. In a minority of cases, the virus enters the central nervous system from the blood and lymph. It then multiplies in and destroys the nerve cells in the brain that control the movements of the muscles. These nerve cells are known as motor neurons. The location and severity of the paralytic polio that results when the motor neurons are damaged varies with the part of the central nervous system that is affected.

Minor forms of acute polio infection

Between 4 and 8 percent of acute polio infections are characterized by flu-like symptoms known as abortive poliomyelitis. People with this form of polio infection experience **sore throat** and **fever, nausea, vomiting**, abdominal **pain, constipation**, or **diarrhea**. Abortive polio is difficult to distinguish from the flu or other viral infections. Patients recover completely in about a week.

About 10% of people infected with poliovirus develop severe **headache** and pain and stiffness of

the neck and back. These symptoms are due to an inflammation of the meninges (tissues which cover the spinal cord and brain). This syndrome is called nonparalytic or aseptic **meningitis**. The term “aseptic” is used to differentiate this type of meningitis from those caused by bacteria. Patients with nonparalytic meningitis may experience a brief period of general illness followed by stiffness in the neck, back, or legs. They may also experience other abnormal sensations for a period of 2–10 days. As with abortive polio, patients with nonparalytic meningitis recover completely.

Paralytic polio

Between 1 and 2 percent of people infected with poliovirus develop the most severe form, paralytic polio. Some of these patients may have 2–3 symptom-free days between the minor illness and the major illness but the symptoms often appear without any previous minor illness. Symptoms again include headache and back and neck pain. The major symptoms, however, are due to invasion of the motor nerves, which are responsible for movement of the muscles.

Paralytic polio is usually divided into three types, depending on whether the paralysis affects the arms and legs (spinal polio; accounts for 79 percent of cases of paralytic polio); breathing, speaking, and swallowing (bulbar polio; 2 percent of cases); or the limbs as well as breathing and other functions (bulbospinal polio; 19 percent of cases). Bulbar polio is particularly likely to lead to death if the patient is not placed on a respirator because the virus affects the brain stem—the part of the brain that controls heartbeat as well as breathing and other vital functions.

The maximum state of paralysis in paralytic polio is usually reached within just a few days. The remaining unaffected nerves then begin the process of attempting to grow branches to compensate for the destroyed nerves. Fortunately, the nerve cells are not always completely destroyed. By the end of a month, the nerve impulses start to return to the apparently paralyzed muscle and by the end of six months, recovery is almost complete. If the nerve cells are completely destroyed, however, paralysis is permanent.

Diagnosis

The diagnosis of polio is based on a combination of the patient’s history and the type and location of symptoms—particularly such symptoms as a stiff neck, difficulty breathing, or abnormal reflexes. Fever and asymmetric flaccid paralysis without sensory loss in a child or young adult almost always indicate poliomyelitis. Nonparalytic poliomyelitis cannot be

KEY TERMS

Aseptic—Sterile; containing no microorganisms, especially no bacteria.

Asymptomatic—Having no symptoms of a disease even though the person may be infected by the organism that causes the disease.

Brainstem—The stalk of the brain that connects the two cerebral hemispheres to the spinal cord.

Endemic—A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.

Flaccid—Weak, soft, or floppy.

Gastrointestinal—Pertaining to the stomach and intestines.

Lymph/lymphatic—One of the three body fluids that is transparent and a slightly yellow liquid that is collected from the capillary walls into the tissues and circulates back to the blood supply.

Meningitis—Inflammation of the membranes that cover the brain and spinal cord.

Motor neuron—A type of cell in the central nervous system that controls the movement of muscles either directly or indirectly.

Neurologic—Pertaining to the nervous system.

Paralysis—The inability to voluntarily move.

distinguished clinically from aseptic meningitis due to other agents. Virus isolated from a throat swab and/or feces or blood tests demonstrating the rise in a specific antibody is required to confirm the diagnosis.

Examination

Tests

To confirm the diagnosis, samples of the patient’s stool, spinal fluid, or throat mucus may be collected and sent to a laboratory for analysis to see whether the sample contains the virus itself. A blood sample early in the infection may also be analyzed for evidence of antibodies to the poliovirus.

Procedures

A **lumbar puncture** is the procedure performed in order to obtain a sample of the patient’s spinal fluid. A long, thin needle is inserted into the lower back between the vertebrae to withdraw spinal fluid. This test can be used to reveal an increased number of white blood cells and no bacteria (aseptic meningitis).

Treatment

Traditional

There is no drug that can cure polio as of 2010.

Antibiotics are ineffective against any viral infection, including polio. Patients with abortive polio or non-paralytic meningitis do not usually need treatment other than resting at home.

Patients with paralytic polio may be placed on a respirator to help them breathe, particularly if they are diagnosed with bulbar polio. Other treatments include painkillers and hot packs for muscle aches, **physical therapy** to restore muscle strength, and occupational or **speech therapy** as needed. Physical therapy is the most important part of management of paralytic polio during recovery. Braces or special shoes may be recommended for some patients. A few patients may undergo surgery to restore limb function.

Prognosis

The overall prognosis for recovery from an acute attack of paralytic polio is generally good. Mortality is about 5–10 percent, mostly in elderly and very young patients; however, the death rate can reach 20–60 percent in cases of bulbar involvement. Half the patients with spinal polio recover fully; 25 percent have mild disabilities; and the remaining 25 percent are left with severe disabilities. Most patients recover from breathing problems, and only a small percentage of patients need long-term treatment on a respirator. Patients with muscle paralysis typically recover about 60 percent of their strength in the first 3–4 months of treatment.

About a quarter of patients who have recovered from paralytic polio develop a disorder called post-polio syndrome (PPS) between 10 and 40 years after the initial infection. PPS is not a reinfection although its cause is not completely understood as of 2009. PPS is marked by:

- Muscular weakness.
- Fatigue.
- Being easily exhausted after even small amounts of activity.
- Joint pain.
- Sleep disorders.
- Difficulty breathing or swallowing.
- Inability to tolerate cold temperatures.

PPS is treated with rest and such supportive measures as powered wheelchairs, pain relievers, and medications to help the patient sleep. Patients are also

encouraged to simplify their work habits and take frequent rest breaks.

Prevention

Polio can easily be prevented by administration of either the Salk vaccine, which contains an inactivated poliovirus, or the Sabin oral vaccine, which contains a weakened live virus. The Salk vaccine (also called the killed polio vaccine or inactivated polio vaccine) consists of a series of three shots that are given just under the skin as two doses 4–8 weeks apart followed by a third dose 6–12 months after the second dose. This immunization contains no live virus, just the components of the virus that provoke the recipient's immune system to react as if the recipient were actually infected with the poliovirus. The recipient thus becomes immune to infection with the poliovirus in the future. The Salk vaccine is the only polio vaccine that is given to people with weakened immune systems.

The Sabin vaccine (also called the oral polio vaccine or OPV) is given to infants by mouth at the same intervals as the diphtheria-pertussis-tetanus (DPT) immunization (three doses). It contains the live, but weakened, poliovirus, which make the recipient immune to future infections with poliovirus. It is given to adults in a single dose by mouth. It is not routinely given to people with weakened immune systems because it contains a live virus.

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ORGANIZATIONS

American Physical Therapy Association (APTA), 1111 North Fairfax Street, Alexandria, VA, 22314-1488, 703-684-APTA (2782), 800-999-APTA (2782), 703-684-7343, <http://www.apta.org//AM/Template.cfm?Section=Home>.

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, 800-232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

March of Dimes, 1275 Mamaroneck Avenue, White Plains, NY, 10605, 914-997-4488, <http://www.marchofdimes.com>.

U.S. National Institute of Allergy and Infectious Diseases (NIAID), 6610 Rockledge Drive, MSC 6612, Bethesda, MD, 20892-6612, 301-496-5717, 866-284-4107, 301-402-3573, <http://www3.niaid.nih.gov>.

World Health Organization (WHO), Avenue Appia 20, 1211 Geneva 27, Switzerland, + 41 22 791 21 11, + 41 22 791 31 11, info@who.int, <http://www.who.int/en>.

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Poliomyelitis see **Polio**

Polyangiitis overlap syndrome see **Vasculitis**

Polyarteritis nodosa see **Vasculitis**



A pair of human kidneys. The left is a polycystic kidney, and the right is a normal kidney. (A. Glauberman/Photo Researchers, Inc.)

over time. It was originally believed that the cysts eventually caused kidney failure by crowding out the healthy kidney tissue. It is now thought that the kidney damage seen in PKD is actually the result of the body's immune system. The immune system, in its attempts to rid the kidney of the cysts, instead progressively destroys the formerly healthy kidney tissue.

Demographics

One of the most common of all life-threatening genetic diseases, PKD affects more than 60,000 Americans. Over 12.5 million people worldwide are affected with PKD. Approximately one in every 400 to 1,000 people is affected with Autosomal Dominant Polycystic Kidney Disease (ADPKD). Another one in 10,000 is affected with Autosomal Recessive Polycystic Kidney Disease (ARPKD). PKD is observed in equal numbers in both males and females. PKD is also observed with equal frequency among ethnic groups.

Description

A healthy kidney is about the same size as a human fist. PKD cysts, which can be as small as the head of a pin or as large as a grapefruit, can expand the kidneys until each one is bigger than a football and weighs as much as 38lb (17 kg).

There are two types of PKD: infantile PKD, which generally shows symptoms prior to birth; and adult onset PKD. Individuals affected with infantile PKD are often stillborn. Among the liveborn individuals affected with infantile PKD, very few of these children survive to the age of two. The adult onset form of PKD is much more common. The time and degree of symptom onset in the adult form of PKD can vary widely, even within a single family with two or more affected individuals. Symptoms of this form of

Polycystic kidney disease

Definition

Polycystic kidney disease (PKD) is one of the most common of all life-threatening human genetic disorders. It is an incurable genetic disorder characterized by the formation of fluid-filled cysts in the kidneys of affected individuals. These cysts multiply

PKD usually start to appear between the ages of 20 and 50. Organ deterioration progresses more slowly in adult onset PKD than it does in the infantile form; but, if left untreated, adult onset PKD also eventually leads to kidney failure.

Causes and symptoms

Polycystic kidney disease is expressed as both a recessive and a dominant trait. A recessive genetic trait will not cause disease in a child unless it is inherited from both parents. A dominant genetic trait can be inherited from just one parent. Those people affected with autosomal dominant polycystic kidney disease (ADPKD) have the much more common adult onset form. Those with autosomal recessive polycystic kidney disease (ARPKD) have the infantile form.

There are mutations on at least three genes that cause adult onset PKD. Approximately 85% of these cases are known to arise from mutations in the PKD1 gene that has been mapped to a region on the short arm of chromosome 16 (16p13.3-p13.12). Another 10–15% of cases of adult onset PKD are thought to be caused by mutations in the PKD2 gene that has been mapped to a region on the long arm of chromosome 4 (4q21-q23). As of 2010, it is thought that the remainder of the cases of PKD are caused by mutations in the PKD3 gene.

Adult onset PKD is transmitted from parents to their offspring as a non-sex linked (autosomal) dominant trait. This means that if either parent carries this genetic mutation, there is a 50% chance their child will inherit this disease. In the case of two affected parents, there is a 75% probability that their children will be affected with adult onset PKD.

Infantile PKD is caused by a non-sex linked (autosomal) recessive genetic mutation that has been mapped to a region on the short arm of chromosome 6 (6p21). Both parents must be carriers of this mutation for their children to be affected with infantile PKD. In the case of two carrier parents, the probability is 25% that their child will be affected by infantile PKD.

A baby born with infantile PKD has floppy, low-set ears, a pointed nose, a small chin, and folds of skin surrounding the eyes (epicanthal folds). Large, rigid masses can be felt on the back of both thighs (flanks), and the baby usually has trouble breathing.

In the early stages of adult onset PKD, many people show no symptoms. Generally, the first symptoms to develop are high blood pressure (**hypertension**), general **fatigue**, **pain** in the lower back or the

backs of the thighs, headaches, and/or urinary tract infections accompanied by frequent urination.

As PKD becomes more advanced, the kidneys' inability to function properly becomes more pronounced. The cysts on the kidney may begin to rupture and the kidneys tend to be much larger than normal. Individuals affected with PKD have a much higher rate of **kidney stones** than the rest of the population at this, and later stages, of the disease. Approximately 60% of individuals affected with PKD develop cysts in the liver, while 10% develop cysts in the pancreas.

Because the kidneys are primarily responsible for cleaning the blood, individuals affected with PKD often have problems involving the circulatory system. These include an underproduction of red blood cells, which results in an insufficient supply of oxygen to the tissues and organs (anemia); an enlarged heart (cardiac hypertrophy) probably caused by long term hypertension; and a leakage of the valve between the left chambers (auricle and ventricle) of the heart (**mitral valve prolapse**). Less common (affecting approximately 5% of PKD patients) are brain aneurysms. An aneurysm is an abnormal and localized bulging of the wall of a blood vessel. If an aneurysm within the brain leaks or bursts, it may cause a **stroke** or even **death**.

Other health problems associated with adult onset PKD include chronic leg or back pain, frequent infections, and herniations of the groin and abdomen, including herniation of the colon (diverticular disease). A herniation, or **hernia**, is caused when a tissue, designed to hold the shape of an underlying tissue, becomes weakened at a particular spot. The underlying tissue pushes against this weakened area until the area is no longer able to hold back the underlying tissue and the area forms an abnormal bulge through which the underlying tissue projects. Diverticular disease is caused by a weakening of the muscles that hold the shape of the organs of the digestive tract. These muscles weaken allowing these organs, particularly one section of the colon, to form sac-like projections that can trap feces and become infected, or rupture.

In the final stages of PKD, the major symptom is kidney (renal) failure. Renal failure is indicated by an increase of nitrogen (in the form of urea) in the blood (uremia, or uremic poisoning). Uremia is a rapidly fatal condition without treatment.

Diagnosis

Many patients who have PKD do not have any symptoms. Their condition may not be discovered unless tests that detect it are performed for other reasons.

KEY TERMS

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes.

Cancer—A disease caused by uncontrolled growth of the body's cells.

Computed tomography (CT) scan—An imaging procedure that produces a three-dimensional picture of organs or structures inside the body, such as the brain.

Cyst—An abnormal sac or closed cavity filled with liquid or semisolid matter.

Diuretics—Medications that increase the excretion of urine.

Kidney—Either of two organs in the lumbar region that filter the blood, excreting the end products of the body's metabolism in the form of urine and

regulating the concentrations of hydrogen, sodium, potassium, phosphate and other ions in the body.

Magnetic resonance imaging (MRI)—A technique that employs magnetic fields and radio waves to create detailed images of internal body structures and organs, including the brain.

Ultrasonogram—A procedure where high-frequency sound waves that cannot be heard by human ears are bounced off internal organs and tissues. These sound waves produce a pattern of echoes, which are then used by the computer to create sonograms or pictures of areas inside the body.

Uremic poisoning—Accumulation of waste products in the body.

Examination

When symptoms of PKD are present, the diagnostic procedure begins with a family medical history and **physical examination** of the patient. If several family members have PKD, there is a strong likelihood that the patient has it too. If the disease is advanced, the doctor will be able to feel the patient's enlarged kidneys. Heart murmur, high blood pressure, and other signs of cardiac impairment can also be detected.

Tests

Urinalysis and a blood test called creatinine clearance can indicate how effectively the kidneys are functioning. Scanning procedures using intravenous dye reveal kidney enlargement or deformity and scarring caused by cysts. Ultrasound and **computed tomography scans** (CT scans) can reveal kidney enlargement and the cysts that caused it. CT scans can highlight cyst-damaged areas of the kidneys.

Procedures

A sampling of the kidney cells (biopsy) may be performed to verify the diagnosis.

Treatment

There is no way to prevent cysts from forming or becoming enlarged, or to prevent PKD from progressing to kidney failure. Treatment goals include

preserving healthy kidney tissue; controlling symptoms and, preventing infection and other complications.

Drugs

If adult PKD is diagnosed before symptoms become evident, urinalysis and other diagnostic tests are performed at six-week intervals to monitor the patient's health status. If results indicate the presence of infection or another PKD-related health problem, aggressive antibiotic therapy is initiated to prevent inflammation that can accelerate disease progression; iron supplements or infusion of red blood cells are used to treat anemia; and surgery may be needed to drain cysts that bleed, cause pain, have become infected, or interfere with normal kidney function.

Lowering high blood pressure can slow loss of kidney function. Blood-pressure control, which is the cornerstone of PKD treatment, is difficult to achieve. Therapy may include antihypertensive medications, diuretic medications, and/or a low-salt diet. As kidney function declines, some patients need dialysis and/or a kidney transplant.

Home remedies

There is no known way to prevent PKD, but certain lifestyle modifications can help control symptoms. People who have PKD should not drink heavily or smoke. They should not use **aspirin**, non-steroidal anti-inflammatory drugs (NSAIDs), or other prescription or over-the-counter medications that can impair

kidney function. Individuals affected with PKD should eat a balanced diet, **exercise** regularly, and maintain a weight appropriate for their height, age, and body type. Regular medical monitoring is also recommended.

Prognosis

There is no known cure for PKD. Those affected with infantile PKD generally die before the age of two. In adults, untreated disease can be rapidly fatal or continue to progress slowly, even after symptoms of kidney failure appear. About half of all adults with PKD also develop kidney failure. Unless the patient undergoes dialysis or has a kidney transplant, this condition usually leads to death within four years of diagnosis.

Although medical treatment can temporarily alleviate symptoms of PKD, the expanding cysts continue to increase pressure on the kidneys. Kidney failure and uremic poisoning (accumulation of waste products the body is unable to eliminate) generally cause death about 10 years after symptoms first appear.

Medications used to fight **cancer** and reduce elevated cholesterol levels have slowed the advance of PKD in laboratory animals. They may soon be used to treat adults and children who have the disease. Researchers are also evaluating the potential benefits of anti-inflammatory drugs, which may prevent the scarring that destroys kidney function.

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ORGANIZATIONS

- American Association of Kidney Patients, 3505 E. Frontage Rd., Suite 315, Tampa, FL, 33607, (800) 749-2257, info@aakp.org, http://www.aakp.org.
- American Kidney Fund (AKF), 6110 Executive Blvd., Suite 1010, Rockville, MD, 20852, (800) 638-8299, http://www.kidneyfund.org.
- American Society of Pediatric Nephrology, 3400 Research Forest Drive, Suite B-7, The Woodlands, TX, 77381, (281) 419-0052, info@aspneph.com, www.aspneoph.com.
- National Kidney and Urologic Disease Information Clearinghouse, 3 Information Way, Bethesda, MD, 20892, (301) 654-4415, www.niddk.nih.gov.
- National Kidney Foundation, 30 East 33rd St. , New York, NY, 10016, (800) 622-9010, http://www.kidney.org.
- National Organization for Rare Diseases (NORD), 55 Kenosia Ave. P.O. Box 1968, Danbury, CT, 06813, (203) 744-0100, orphan@rarediseases.org, http://www.rarediseases.org.
- Polycystic Kidney Disease Foundation, 8330 Ward Parkway, Suite 510, Kansas City, MO, 64114, (816) 931-2600, (800) PKD-CURE, pdkcure@pdkcure.org, http://www.pdkcure.org.

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Polycystic ovary syndrome

Definition

Polycystic ovary syndrome (PCOS) is a condition characterized by the accumulation of numerous cysts (fluid-filled sacs) on the ovaries associated with high male hormone levels, chronic anovulation (absent ovulation), and other metabolic disturbances. Classic symptoms include excess facial and body hair, **acne**, **obesity**, irregular menstrual cycles, and **infertility**.

Description

PCOS, also called Stein-Leventhal syndrome, is a group of symptoms caused by underlying hormonal and metabolic disturbances that affect about 6% of premenopausal women. PCOS symptoms appear as early as adolescence in the form of **amenorrhea** (missed periods), obesity, and **hirsutism**, the abnormal growth of body hair.

A disturbance in normal hormonal signals prevents ovulation in women with PCOS. Throughout

the cycle, estrogen levels remain steady, luteinizing hormone (LH) levels are high, and follicle stimulating hormone (FSH) and progesterone levels are low. Since eggs are rarely or never released from their follicles, multiple **ovarian cysts** develop over time.

One of the most important characteristics of PCOS is hyperandrogenism, the excessive production of male hormones (androgens), particularly testosterone, by the ovaries. This accounts for the male hair-growth patterns and acne in women with PCOS. Hyperandrogenism has been linked with **insulin resistance** (the inability of the body to respond to insulin) and hyperinsulinemia (high blood insulin levels), both of which are common in PCOS.

Causes and symptoms

While the exact cause of PCOS is unknown, it runs in families, so the tendency to develop the syndrome may be inherited. The interaction of hyperinsulinemia and hyperandrogenism is believed to play a role in chronic anovulation in susceptible women.

The numbers and types of PCOS symptoms that appear vary among women. These include:

- **Hirsutism.** Related to hyperandrogenism, this occurs in 70% of women.
- **Obesity.** Approximately 40–70% of persons with PCOS are overweight.
- **Anovulation and menstrual disturbances.** Anovulation appears as amenorrhea in 50% of women, and as heavy uterine bleeding in 30% of women. However, 20% of women with PCOS have normal menstruation.
- **Male-pattern hair loss.** Some women with PCOS develop bald spots.
- **Infertility.** Achieving pregnancy is difficult for many women with PCOS.
- **Polycystic ovaries.** Most, but not all, women with PCOS have multiple cysts on their ovaries.
- **Skin discoloration.** Some women with PCOS have dark patches on their skin.
- **Abnormal blood chemistry.** Women with PCOS have high levels of low-density lipoprotein (LDL or “bad”) cholesterol and triglycerides, and low levels of high-density lipoprotein (HDL or “good”) cholesterol.
- **Hyperinsulinemia.** Some women with PCOS have high blood insulin levels, particularly if they are overweight.

Diagnosis

PCOS is diagnosed when a woman visits her doctor for treatment of symptoms such as hirsutism, obesity, menstrual irregularities, or infertility. Women with PCOS are treated by a gynecologist, a doctor who treats diseases of the female reproductive organs, or a reproductive endocrinologist, a specialist who treats diseases of the body’s endocrine (hormones and glands) system and infertility.

PCOS can be difficult to diagnose because its symptoms are similar to those of many other diseases or conditions, and because all of its symptoms may not occur. A doctor takes a complete medical history, including questions about menstruation and reproduction, and weight gain. **Physical examination** includes a pelvic examination to determine the size of the ovaries, and visual inspection of the skin for hirsutism, acne, or other changes. Blood tests are performed to measure levels of luteinizing hormone, follicle stimulating hormone, estrogens, androgens, glucose, and insulin. A glucose-tolerance test may be administered. An ultrasound examination of the ovaries is performed to evaluate their size and shape. Most insurance plans cover the costs of diagnosing and treating PCOS and its related problems.

Treatment

PCOS treatment is aimed at correcting anovulation, restoring normal menstrual periods, improving fertility, eliminating hirsutism and acne, and preventing future complications related to high insulin and blood lipid (fat) levels. Treatment consists of weight loss, drugs or surgery, and hair removal, depending upon which symptoms are most bothersome, and whether a woman desires **pregnancy**.

Weight loss

In overweight women, weight loss (as little as 5%) through diet and **exercise** may correct hyperandrogenism, and restore normal ovulation and fertility. This is often tried first.

Drugs

HORMONAL DRUGS. Women who do not want to become pregnant and require **contraception** (spontaneous ovulation occurs occasionally among women with PCOS) are treated with low-dose oral contraceptive pills (OCPs). OCPs bring on regular menstrual periods and correct heavy uterine bleeding, as well as hirsutism, although improvement may not be seen for up to a year.

If an infertile woman desires to become pregnant, the first drug usually given to help induce ovulation is clomiphene citrate (Clomid), which results in pregnancy in about 70% of women but can cause multiple births. In the 20–25% of women who do not respond to clomiphene, other drugs that stimulate follicle development and induce ovulation, such as human menstrual gonadotropin (Pergonal) and human chorionic gonadotropin (HCG), are given. However, these drugs have a lower pregnancy rate (less than 30%), a higher rate of **multiple pregnancy** (from 5–30%, depending on the dose of the drug), and a higher risk of medical problems. Women with PCOS have a high rate of **miscarriage** (30%), and may be treated with the gonadotropin-releasing hormone agonist leuprolide (Lupron) to reduce this risk.

Since women with PCOS do not have regular endometrial shedding due to high estrogen levels, they are at increased risk for overgrowth of this tissue and **endometrial cancer**. The drug medroxyprogesterone acetate, when taken for the first 10 days of each month, causes regular shedding of the endometrium, and reduces the risk of **cancer**. However, in most cases, oral contraceptive pills are used instead to bring about regular menstruation.

OTHER DRUGS. Another drug that helps to trigger ovulation is the steroid hormone dexamethasone. This drug acts by reducing the production of androgens by the adrenal glands.

The antiandrogen spironolactone (Aldactazide), which is usually given with an oral contraceptive, improves hirsutism and male-pattern baldness by reducing androgen production, but has no effect on fertility. The drug causes abnormal uterine bleeding and is linked with **birth defects** if taken during pregnancy. Another antiandrogen used to treat hirsutism, flutamide (Eulexin), can cause liver abnormalities, **fatigue**, mood swings, and loss of sexual desire. A drug used to reduce insulin levels, metformin (Glucophage), has shown promising results in women with PCOS hirsutism, but its effects on infertility and other PCOS symptoms are unknown. Drug treatment of hirsutism is long-term, and improvement may not be seen for up to a year or longer.

Acne is treated with **antibiotics**, antiandrogens, and other drugs such as retinoic acids (vitamin A compounds).

Surgical treatment

Surgical treatment of PCOS may be performed if drug treatment fails, but it is not common. A wedge

resection, the surgical removal of part of the ovary and cysts through a laparoscope (an instrument inserted into the pelvis through a small incision), or an abdominal incision, reduces androgen production and restores ovulation. Although laparoscopic surgery is less likely to cause scar tissue formation than abdominal surgery, both are associated with the potential for scarring that may require additional surgery. Laparoscopic ovarian drilling is another type of laparoscopic surgery used to treat PCOS. The ovarian cysts are penetrated with a laser beam and some of the fluid is drained off. Between 50–65% of women may become pregnant after either type of surgery.

Some cases of severe hirsutism are treated by removal of the uterus (**hysterectomy**) and the ovaries (oophorectomy), followed by estrogen replacement therapy.

Other treatment

Hirsutism may be treated by hair removal techniques such as shaving, depilatories (chemicals that break down the structure of the hair), tweezing, waxing, electrolysis (destruction of the hair root by an electrical current), or the destruction of hair follicles by laser therapy. However, the treatments may have to be repeated.

Alternative treatment

PCOS can be addressed using many types of alternative treatment. The rebalancing of hormones is a primary focus of all these therapies. **Acupuncture** works on the body's energy flow according to the meridian system. Chinese herbs, such as *gui zhi fu ling wan*, can be effective. In **naturopathic medicine**, treatment focuses on helping the liver function more optimally in the horomonal balancing process. Dietary changes, including reducing animal products and fats, while increasing foods that nourish the liver such as carrots, dark green vegetables, lemons, and beets, can be beneficial. Essential fatty acids, including flax oil, evening primrose oil (*Oenothera biennis*), and black currant oil, act as anti-inflammatories and hormonal regulators. Western herbal medicine uses phytoestrogen and phytoprogesteronic herbs, such as blue cohosh (*Caulophyllum thalictroides*) and false unicorn root (*Chamaelirium luteum*), as well as liver herbs, like dandelion (*Taraxacum mongolicum*), to work toward hormonal balance. Supplementation with **antioxidants**, including zinc, and **vitamins** A, E, and C, is also recommended. Constitutional homeopathy can

KEY TERMS

Androgens—Male sex hormones produced by the adrenal glands and testes, the male sex glands.

Anovulation—The absence of ovulation.

Antiandrogens—Drugs that inhibit androgen production.

Estrogens—Hormones produced by the ovaries, the female sex glands.

Follicle stimulating hormone—A hormone that stimulates the growth and maturation of mature eggs in the ovary.

Gynecologist—A physician with specialized training in diseases and conditions of the female reproductive system.

Hirsutism—An abnormal growth of hair on the face and other parts of the body caused by an excess of androgens.

Hyperandrogenism—The excessive secretion of androgens.

Hyperinsulinemia—High blood insulin levels.

Insulin resistance—An inability to respond to insulin, a hormone produced by the pancreas that helps the body to use glucose.

Laparoscope—An instrument inserted into the pelvis through a small incision.

Luteinizing hormone—A hormone that stimulates the secretion of sex hormones by the ovary.

Ovarian follicles—Structures found within the ovary that produce eggs.

bring about a deep level of healing with the correct remedies.

Prognosis

With proper diagnosis and treatment, most PCOS symptoms can be adequately controlled or eliminated. Infertility can be corrected and pregnancy achieved in most women although, in some, hormonal disturbances and anovulation may recur. Women should be monitored for endometrial cancer. Because of the high rate of hyperinsulinemia seen in PCOS, women with the disorder should have their glucose levels checked regularly to watch for the development of diabetes. Blood pressure and cholesterol screening are also needed because these women also tend to have high levels of LDL cholesterol and

triglycerides, which put them at risk for developing heart disease.

Prevention

There is no known way to prevent PCOS, but if diagnosed and treated early, risks for complications such as and heart disease and diabetes may be minimized. Weight control through diet and exercise stabilizes hormones and lowers insulin levels.

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- Vanderbilt University School of Medicine. <http://www.mc.vanderbilt.edu/peds/pidl/adolesc/polcysov.htm>.
- Women's Health-UK. <http://www.womens-health.co.uk>.

ORGANIZATIONS

- American Academy of Family Physicians (AAFP), 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2680, (913) 906-6000, (913) 906-6075, 800 271-2237, <http://www.aafp.org>.
- American Medical Association, 515 N. State St., Chicago, IL, 60654, (800) 621-8335, <http://www.ama-assn.org>.
- Polycystic Ovarian Syndrome Association, P.O. Box 3403, Englewood, CO, 80155-3403, info@pcosupport.org, <http://www.pcosupport.org>.

L. Fleming Fallon, Jr., MD, DrPH

Polycythemia see **Secondary polycythemia**

Polycythemia rubra vera see **Polycythemia vera**

Polycythemia vera

Definition

Polycythemia vera (PV) is a chronic blood disorder marked by an abnormal increase in three types of blood cells produced by bone marrow: red blood cells (RBCs), white blood cells (WBCs), and platelets. PV is called a

myeloproliferative disorder, which means that the bone marrow is producing too many cells too quickly. Most of the symptoms of PV are related to the increased volume of the patient's blood and its greater thickness (high viscosity). PV sometimes evolves into a different myeloproliferative disorder or into acute leukemia.

Description

Polycythemia vera is a relatively common progressive disorder that develops over a course of 10–20 years. In the United States, PV affects about one person in every 200,000. PV has several other names, including splenomegalic polycythemia, Vaquez-Osler syndrome, erythremia, and primary polycythemia. Primary polycythemia means that the disorder is not caused or triggered by other illnesses. PV most commonly affects middle-aged adults. It is rarely seen in children or young adults and does not appear to run in families. The male/female ratio is 2:1.

Risk factors for polycythemia vera include:

- Caucasian race
- male sex
- age between 40 and 60

Causes and symptoms

The cause of PV remains uncertain. In general, the increased mass of red blood cells in the patient's blood causes both hemorrhage and abnormal formation of **blood clots** in the circulatory system (thrombosis). The reasons for these changes in clotting patterns are not yet fully understood.

Early symptoms

The symptoms of early PV may be minimal—it is not unusual for the disorder to be discovered during a routine blood test. More often, however, patients have symptoms that include headaches, ringing in the ears, tiring easily, memory problems, difficulty breathing, giddiness or lightheadedness, **hypertension**, visual problems, or **tingling** or burning sensations in their hands or feet. Another common symptom is **itching** (pruritus). Pruritus related to PV is often worse after the patient takes a warm bath or shower.

Some patients' early symptoms include unusually heavy bleeding from minor cuts, nosebleeds, stomach ulcers, or bone **pain**. In a few cases, the first symptom is the development of blood clots in an unusual part of the circulatory system (e.g., the liver).

Later symptoms and complications

As the disease progresses, patients with PV may have episodes of hemorrhage or thrombosis. Thrombosis is the most frequent cause of **death** from PV. Other complications include a high level of uric acid in the blood and an increased risk of peptic ulcer disease. About 10% of PV patients eventually develop **gout**; another 10% develop peptic ulcers.

Spent phase

The spent phase is a development in late PV that affects about 30% of patients. The bone marrow eventually fails and the patient becomes severely anemic, requiring repeated blood transfusions. The spleen and liver become greatly enlarged—in the later stages of PV, the patient's spleen may fill the entire left side of the abdomen.

Diagnosis

Physical examination

PV is often a diagnosis of exclusion, which means that the doctor will first rule out other possible causes of the patient's symptoms. The doctor can detect some signs of the disorder during a **physical examination**. Patients with PV will have an enlarged spleen (splenomegaly) in 75% of cases. About 50% will have a slightly enlarged liver. The doctor can feel these changes when he or she presses on (palpates) the patient's abdomen while the patient is lying flat. An **eye examination** will usually reveal swollen veins at the back of the eye. Patients with PV often have unusually red complexions; mottled red patches on their legs, feet, or hands; or swelling at the ends of the fingers.

Diagnostic criteria for PV

Accurate diagnosis of PV is critical because its treatment may require the use of drugs with the potential to cause leukemia. The results of the patient's blood tests are evaluated according to criteria worked out around 1970 by the Polycythemia Vera Study Group. The patient is considered to have PV if all three major criteria are met; or if the first two major criteria and any two minor criteria are met.

Major criteria:

- red blood cell mass greater than 36 mL/kg in males, greater than 32 mL/kg in females
- arterial oxygen level greater than 92%
- splenomegaly

Minor criteria:

- platelet count greater than 400,000/mm³

- WBC greater than 12,000/mm³ without fever or infection
- leukocyte alkaline phosphatase (LAP) score greater than 100 with increased blood serum levels of vitamin B₁₂

Laboratory testing

BLOOD TESTS. The diagnosis of PV depends on a set of findings from blood tests. The most important single measurement is the patient's red blood cell mass as a proportion of the total blood volume. This measurement is made by tagging RBCs with radioactive chromium (⁵¹Cr) in order to determine the patient's RBC volume. While a few patients with PV may have a red cell mass level within the normal range if they have had recent heavy bleeding, a high score may eliminate the need for some other tests. A score higher than 36 mL/kg for males and 32 mL/kg for females on the ⁵¹Cr test suggests PV. Measurements of the oxygen level in the patient's arterial blood, of the concentration of vitamin B₁₂ in the blood serum, and of leukocyte alkaline phosphatase (LAP) staining can be used to distinguish PV from certain types of leukemia or from other types of polycythemia. LAP staining measures the intensity of enzyme activity in a type of white blood cell called a neutrophil. In PV, the LAP score is higher than normal whereas in leukemia it is below normal.

BONE MARROW TESTS. Bone marrow testing can be used as part of the diagnostic process. A sample of marrow can be cultured to see if red blood cell colonies develop without the addition of a hormone that stimulates RBC production. The growth of a cell colony without added hormone indicates PV. Bone marrow testing is also important in monitoring the progress of the disease, particularly during the spent phase.

GENETIC TESTING. **Genetic testing** can be used to rule out the possibility of chronic myeloid leukemia. Patients with this disease have a characteristic chromosomal abnormality called the Philadelphia chromosome. The Philadelphia chromosome does *not* occur in patients with PV.

Imaging studies

Imaging studies are not necessary to make the diagnosis of PV. In some cases, however, imaging studies can detect enlargement of the spleen that the doctor may not be able to feel during the physical examination.

KEY TERMS

Anagrelide—An orphan drug that is approved for treating PV patients on an investigational basis. Anagrelide works by controlling the level of platelets in the blood.

Leukocyte alkaline phosphatase (LAP) test—A blood test that measures the level of enzyme activity in a type of white blood cell called neutrophils.

Myeloproliferative disorder—A disorder in which the bone marrow produces too many cells too rapidly.

Myelosuppressive therapy—Any form of treatment that is aimed at slowing down the rate of blood cell production.

Orphan drug—A drug that is known to be useful in treatment but lacks sufficient funding for further research and development.

Philadelphia chromosome—An abnormal chromosome that is found in patients with a chronic form of leukemia but not in PV patients.

Phlebotomy—Drawing blood from a patient's vein as part of diagnosis or therapy. Phlebotomy is sometimes called venesection. It is an important part of the treatment of PV.

Pruritus—An itching sensation or feeling. In PV the itching is not confined to a specific part of the body and is usually worse after a warm bath or shower.

Spent phase—A late development in PV leading to failure of the bone marrow and severe anemia.

Splenomegaly—Abnormal enlargement of the spleen. Splenomegaly is a major diagnostic criterion of PV.

Treatment

Treatment of PV is tailored to the individual patient according to his or her age, the severity of the symptoms and complications, and the stage of the disease.

Phlebotomy

Phlebotomy is the withdrawal of blood from a vein. It is the first line of treatment for patients with PV. Phlebotomy is used to bring down the ratio of red blood cells to fluid volume (the **hematocrit**) in the patient's blood to a level below 45%. In most cases the doctor will withdraw about 500 mL of blood (about 15 fluid ounces) once or twice a week until the hematocrit is low enough. Phlebotomy is considered the best course of treatment for patients younger than 60 and for women of childbearing age. Its drawback is that patients remain at some risk for either thrombosis or hemorrhage.

Myelosuppression

Myelosuppressive therapies are used to slow down the body's production of blood cells. They are given to patients who are older than 60 and at high risk for thrombosis. These therapies, however, increase the patient's risk of developing leukemia. The substances most frequently used include hydroxyurea (Hydrea), interferon alfa (Intron), or radioactive phosphorus (^{32}P). ^{32}P is used only in elderly patients with life expectancies of less than five years because it causes

leukemia in about 10% of patients. Interferon alfa is expensive and causes side effects resembling the symptoms of **influenza** but is an option for some younger PV patients.

Investigational treatment

The Food and Drug Administration (FDA) has approved the use of anagrelide, an orphan drug, for investigational use in the treatment of PV. Anagrelide has moderate side effects and controls the platelet level in over 90% of patients.

Treatment of complications

The itching caused by PV is often difficult to control. Patients with pruritus are given diphenhydramine (Benadryl) or another antihistamine. Patients with high levels of uric acid are usually given allopurinol (Lopurin, Zyloprim) by mouth. Supportive care includes advice about diet—splenomegaly often makes patients feel full after eating only a little food. This problem can be minimized by advising patients to eat small meals followed by rest periods.

Because of the clotting problems related to PV, patients should not undergo surgery until their blood counts are close to normal levels. Female patients of childbearing age should be warned about the dangers of **pregnancy** related to their clotting abnormalities.

Prognosis

The prognosis for untreated polycythemia vera is poor; 50% of patients die within 18 months after diagnosis. Death usually results from **heart failure**, leukemia, or hemorrhage. Patients being treated for PV can expect to live between 11 and 15 years on average after diagnosis.

ORGANIZATIONS

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Rebecca J. Frey, PhD



Polydactyly and syndactyly

Definition

Polydactyly and syndactyly are congenital irregularities of the hands and feet. Polydactyly is the occurrence of extra fingers or toes, and syndactyly is the webbing or fusing together of two or more fingers or toes.

Description

Polydactyly can vary from an unnoticeable rudimentary finger or toe to fully developed extra digits.

Syndactyly also exhibits a large degree of variation. Digits can be partially fused or fused along



Syndactyly is the webbing or fusing together of two or more fingers or toes. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)

Polydactyly is the occurrence of extra or partial fingers or toes. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)

their entire length. The fusion can be simple with the digits connected only by skin, or it can be complicated with shared bones, nerves, vessels, or nails.

Polydactyly and syndactyly can occur simultaneously when extra digits are fused. This condition is known as polysyndactyly.

Causes and symptoms

Polydactyly and syndactyly are due to errors in the process of fetal development. For example, syndactyly results from the failure of the programmed cell **death** that normally occurs between digits. Most often these errors are due to genetic defects.

Polydactyly and syndactyly can both occur by themselves as isolated conditions or in conjunction with other symptoms as one aspect of a multi-symptom disease. There are several forms of isolated syndactyly and several forms of isolated polydactyly; each of these, where the genetics is understood, is caused by an autosomal dominant gene. This means that since the gene is autosomal (not sex-linked), males and females are equally likely to inherit the trait. This also means that since the gene is dominant, children who have only one parent with the trait have a 50% chance

KEY TERMS

Autosomal chromosome—One of the non-X or non-Y chromosomes.

Congenital—A condition present at birth.

Digit—A finger or a toe.

Dominant trait—A genetic trait that will always express itself when present as one of a pair of genes (as opposed to a recessive trait where two copies of the gene are necessary to give the individual the trait).

Gene—A portion of a DNA molecule that either codes for a protein or RNA molecule or has a regulatory function.

Triploidy—The condition where an individual has three entire sets of chromosomes instead of the usual two.

Trisomy—An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

of inheriting it. However, people in the same family carrying the same gene can have different degrees of polydactyly or syndactyly.

Polydactyly and syndactyly are also possible outcomes of a large number of rare inherited and developmental disorders. One or both of them can be present in over 100 different disorders where they are minor features compared to other characteristics of these diseases.

For example, polydactyly is a characteristic of Meckel syndrome and Laurence-Moon-Biedl syndrome. Polydactyly may also be present in Patau's syndrome, asphyxiating thoracic dystrophy, hereditary spherocytic **hemolytic anemia**, Moebius syndrome, VACTERL association, and Klippel-Trenaunay syndrome.

Syndactyly is a characteristic of Apert syndrome, Poland syndrome, Jarcho-Levin syndrome, oral-facial-digital syndrome, Pfeiffer syndrome, and Edwards syndrome. Syndactyly may also occur with Gordon syndrome, Fraser syndrome, Greig cephalopolysyndactyly, **phenylketonuria**, Saethre-Chotzen syndrome, Russell-Silver syndrome, and triploidy.

In some isolated cases of polydactyly or syndactyly, it is not possible to determine the cause. Some of these cases might nevertheless be due to genetic defects; sometimes there is too little information to demonstrate a genetic cause. Some cases might be due to external factors like exposure to toxins or womb anomalies.

Diagnosis

Polydactyly and syndactyly can be diagnosed by external observation, x ray, and fetal sonogram.

Treatment

Polydactyly can be corrected by surgical removal of the extra digit or partial digit. Syndactyly can also be corrected surgically, usually with the addition of a skin graft from the groin.

Prognosis

The prognosis for isolated polydactyly and syndactyly is excellent. When polydactyly or syndactyly are part of a larger condition, the prognosis depends on the condition. Many of these conditions are quite serious, and early death may be the outcome.

Prevention

There is no known prevention for these conditions.

Resources

OTHER

OMIM Home Page, Online Mendelian Inheritance in Man.
<http://www.ncbi.nlm.nih.gov/Omim>.

ORGANIZATIONS

March of Dimes Birth Defects Foundation, 1275 Mamaroneck Ave., White Plains, NY, 10605, (914) 997-4488,
<http://www.modimes.org>.

National Institute of Child Health and Human Development, Bldg 31, Room 2A32, MSC 2425, 31 Center Drive, Bethesda, MD, 20892-2425, 8666 760-5947, (800) 370-2943, <http://www.nichd.nih.gov>.

Lorraine Lica, PhD

Polyendocrine deficiency syndromes see
Polyglandular deficiency syndromes

Polyglandular deficiency syndromes

Definition

Polyglandular deficiency syndromes are disorders characterized by the failure of more than one endocrine gland to make hormones in sufficient quantities for the body to function normally.

Description

The endocrine system is a diverse group of glands located all over the body that work together to regulate the body's metabolic activities. It includes:

- the pituitary gland, located deep in the brain, is considered the "master gland" that regulates many of the others
- the thyroid gland is located in the neck and sets the metabolic speed of many processes,
- the parathyroid glands, attached to the back of the thyroid, regulate calcium balance,
- the adrenal glands are located on top of the kidneys and make four separate kinds of hormones,
- the gonads (sex organs) produce sex hormones,
- the pancreas is responsible for the production of digestive juices, insulin, and glucagon.

There are over a dozen different syndromes that involve failure of more than one endocrine gland.

Causes and symptoms

The cause of polyglandular deficiency syndromes is usually an autoimmune response—a condition in which the body generates antibodies to its own tissues. The immune system may attack one or more glands; however, because of their interdependence, the destruction of one gland can often lead to the impairment of another. Other causes may include **infectious disease**; insufficient blood flow to the glands due to an obstruction such as a blood clot; or the presence of a tumor.

Doctors usually group polyglandular deficiency syndromes into three types:

- Type I occurs during childhood and is characterized by failure of the adrenals, parathyroids, thyroid, and gonads combined with hepatitis, hair loss, skin pigment changes, and inability of the bowel to absorb adequate nutrition. These children also get a persistent skin fungus infection called candidiasis.
- Type II occurs during adulthood and is characterized by failure of the adrenals, thyroid (Schmidt's syndrome), and gonads combined with similar nutritional

KEY TERMS

Antibody—A weapon in the body's immune defense arsenal that attacks a specific antigen.

Congenital—Present at birth.

Myasthenia gravis—A disease that causes muscle weakness.

Rubella—German measles.

Syndrome—A collection of abnormalities that occur often enough to suggest they have a common cause.

failures and hair and skin changes. These patients also have myasthenia gravis. This type of polyglandular deficiency syndrome often produces insulin-dependent diabetes mellitus (IDDM).

- Type III disease may produce diabetes or adrenal failure combined with thyroid problems. It may also include baldness (alopecia), anemia, and vitiligo (condition characterized by white patches on normally pigmented skin).

Not all symptoms of any syndrome appear at once or in the same patient.

Diagnosis

Because these diseases evolve over time, the final diagnosis may not appear for years. A family history is very helpful in knowing what to expect. Any single endocrine abnormality should heighten suspicion that there are others, since they so often occur together, both as underproduction and overproduction of hormones. Most hormone levels can be monitored through blood tests. Many of the antibodies that characterize these conditions can also be found by blood testing.

Treatment

Fortunately there are replacements available for all the missing hormones. Careful balancing of them all can provide a reasonably comfortable quality of life for these patients.

Resources

BOOKS

Fauci, Anthony S., et al., eds. *Harrison's Principles of Internal Medicine*. 17th ed. New York: McGraw-Hill Professional, 2008.

J. Ricker Polsdorfer, MD

Polyhydramnios and oligohydramnios

Definition

Polyhydramnios and oligohydramnios are amniotic fluid abnormalities. Polyhydramnios is an excess of amniotic fluid and oligohydramnios is a deficiency of amniotic fluid.

Description

Amniotic fluid is the liquid that surrounds the developing fetus during **pregnancy**. It is contained within the amniotic membrane that forms the amniotic sac (bag of waters). During the first three months after conception (first trimester), amniotic fluid is mainly derived from the blood plasma that diffuses through the thin tissues of the fetus into the surrounding space. After the fetal kidneys form and become functional at about 10–11 weeks, fetal urine becomes the main source of amniotic fluid and remains so for the rest of the pregnancy. In addition, the lungs also produce liquid that becomes part of the amniotic fluid. Other contributions come from fetal oral and nasal secretions and from the fetal surface of the placenta. Amniotic fluid removal is largely due to fetal swallowing and absorption into the fetal blood. Uptake also occurs across the placental surface. The volume of amniotic fluid normally increases throughout pregnancy, reaching a peak at about 32–33 weeks and remaining fairly constant or decreasing slightly thereafter. There is a wide range of normal fluid volumes with an average of 700–800 mL at 32–33 weeks. Through the processes of swallowing and urination, a fetus can recycle the entire volume in less than 24 hours. Because the normal values for amniotic fluid volume increase during pregnancy, the actual volume that constitutes polyhydramnios is dependent on the gestational age of the fetus. During the last two months of pregnancy, polyhydramnios usually refers to amniotic fluid volumes greater than 1,700–1,900 mL. Severe cases are associated with much greater fluid volume excesses. The range of fluid values diagnostic of oligohydramnios is not as wide as that for polyhydramnios. Less than 300 mL, or lower than the 5% percentile for gestational age, is usually considered the upper threshold.

Causes and symptoms

Polyhydramnios, also referred to as hydramnios, can have any one of a number of causes related either to an underlying maternal or fetal condition. Maternal diabetes, which is associated with a macrosomic (enlarged) fetus, is a common cause. The medication

lithium, used to treat depression, can also increase amniotic fluid levels. Twin gestations are prone to polyhydramnios. Infections passed from mother to fetus such as **rubella**, **cytomegalovirus**, and **toxoplasmosis**, can also result in damage to the fetus and elevated amniotic fluid levels. Fetal abnormalities, including many that are life-threatening or lead to a significant impairment in the quality of life, are found in up to a quarter of all patients. For this reason, the initial finding of excess amniotic fluid should be followed by thorough diagnostic studies to determine the cause and the prognosis.

Because fetal swallowing is a major factor in amniotic fluid removal, fetal abnormalities that prevent fluid uptake should be investigated. These include gastrointestinal obstructions such as **esophageal atresia** and duodenal atresia, as well as neurological conditions that affect swallowing including anencephaly. Certain cardiac abnormalities, kidney disorders, and genetic conditions such as **myotonic dystrophy** and alpha-thalassemia can also cause polyhydramnios. Fetal chromosome abnormalities are frequently associated with elevated amniotic fluid levels. The more severe the polyhydramnios the more likely it is that fetal abnormalities will be present. In addition, there are other, infrequent causes, and in a number of cases, no cause can be found. Polyhydramnios can lead to maternal abdominal discomfort and respiratory difficulties as well as preterm labor. When polyhydramnios is associated with fetal abnormalities, perinatal mortality is significantly increased.

Oligohydramnios is most commonly associated with abnormalities of the fetal kidneys. Since fetal urine is the main source of amniotic fluid in the latter two-thirds of pregnancy, any condition that interferes with fetal urine production can lead to oligohydramnios. Renal agenesis, cystic kidneys, and bladder outlet obstructions are common. Meckel-Gruber syndrome, a lethal autosomal recessive genetic disorder featuring brain and kidney abnormalities and extra digits is one specific cause. Placental insufficiency and fetal growth retardation can also result in oligohydramnios. **Premature rupture of membranes**, especially between 16 and 24 weeks is another cause and, because amniotic fluid is important in lung growth, it can lead to underdevelopment of the lungs (pulmonary hypoplasia). In general, regardless of the cause, oligohydramnios that arises early in a pregnancy, can cause hypoplastic lungs. It can also result in space limitations within the amniotic sac that cause fetal compression and orthopedic abnormalities such as clubbed feet in the newborn. In general, oligohydramnios that begins near the time of delivery is associated with a better outcome than cases that have an onset earlier in pregnancy.

KEY TERMS

Alpha-thalassemia—An inherited disorder that interferes with the normal production of hemoglobin.

Anencephaly—Congenital absence of the brain. Occurs during the first month of embryonic development.

Autosomal recessive—A pattern of inheritance in which both copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is one that is located on one of the autosomes or non-sex chromosomes. When both parents have one abnormal copy of the same gene, they have a 25% chance with each pregnancy that their offspring will have the disorder.

Congenital—Present at birth.

Duodenal atresia—Closure or blockage of the duodenum, the upper section of the small intestine.

Esophageal atresia—Blockage or closure of the esophagus, the tube leading from the mouth to the stomach.

Gestational age—The estimated age of a fetus expressed in weeks, calculated from the first day of the last normal menstrual period.

Myotonic dystrophy—A genetic defect resulting in abnormal muscle function.

Placenta—The flat, spongy structure that forms within the uterus during pregnancy and provides nourishment to the developing fetus.

Renal agenesis—Failure of the fetal kidneys to form. Oligohydramnios usually associated with absence of both kidneys.

Diagnosis

In current obstetrical practice, polyhydramnios and oligohydramnios are usually detected during a routine prenatal ultrasound. If the ultrasonographer suspects that excess or reduced fluid is present, it is customary to take measurements of pockets of fluid visualized around the fetus, calculate the amniotic fluid index (AFI), and compare it to AFI values found in standard tables. Subsequent ultrasound measurements can then be used to track the increase or decrease in fluid.

It is extremely important that the cause of an abnormal AFI be sought. Because of the high risk of fetal abnormalities, detailed ultrasound exams (targeted exams) should then be performed. The mother should be counseled about the possible complications and offered additional testing as necessary. For example, an **amniocentesis** for prenatal chromosome analysis may be important because of the high risk of fetal chromosome abnormalities. This test is usually indicated if fetal abnormalities are suspected on the basis of the ultrasound exam. An amniocentesis can also be used to check for fetal infections and some rare single gene defects.

Treatment

Effective treatments for polyhydramnios and oligohydramnios are limited. To relieve maternal discomfort, an excess fluid level can be reduced by inserting a needle into the amniotic sac and using a syringe to withdraw excess fluid. This can be done

repeatedly, if necessary. In oligohydramnios, the opposite approach of adding fluid either by increasing oral intake in the mother or by directly infusing saline into the amniotic sac has been tried in select cases. If the cause of oligohydramnios is a fetal bladder obstruction, it may be possible to place a small tube in the bladder to shunt the fluid into the amniotic sac.

Alternative treatment

In select cases where polyhydramnios is thought to be due to an increased output of fetal urine, the drug indomethacin has been used with some success, but there is concern about side effects, particularly on the fetus. Another similar drug, sulindac, is currently being investigated. If oligohydramnios is due to premature rupture of the membranes, a protocol to manage complications should be instituted.

Prognosis

The prognosis for both polyhydramnios and oligohydramnios depends on the cause. If excess or reduced amniotic fluid is the result of an underlying fetal abnormality, the nature of that abnormality will determine the prognosis. This is one reason why it is important to perform the necessary follow-up studies. A woman who has been diagnosed with polyhydramnios or oligohydramnios needs to be made fully aware of the types of testing available and carefully counseled about the diagnosis and its impact on the chance for a successful pregnancy outcome and a healthy infant.

Prevention

In order to prevent polyhydramnios or oligohydramnios, it would be necessary to prevent the underlying cause. Good control of maternal diabetes and the prevention of infections transmittable from mother to fetus are two approaches for a subset of cases, but, in general, prevention is not possible.

Resources

BOOKS

- Cunningham, F. Gary, et al. *Williams Obstetrics*. 23rd ed. New York: McGraw-Hill Medical, 2010.
 Rodeck, Charles H., and Martin J. Whittle. *Fetal Medicine: Basic Science and Clinical Practice*. 2nd ed. New York: Churchill Livingstone, 2009.

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KEY TERMS

Anemia—A condition in which the blood lacks enough red blood cells (hemoglobin).

Atrophy—Wasting away of a body part.

Frozen shoulder—A shoulder that becomes scarred and cannot move.

Giant cell arteritis—Also called temporal arteritis. A condition which causes the inflammation of temporal arteries. It can cause blindness when the inflammation effects the ophthalmic artery.

NSAIDs—Nonsteroidal anti-inflammatory drugs like aspirin, ibuprofen, and naproxen.

Syndrome—A collection of abnormalities that occur often enough to suggest they have a common cause.

Diagnosis

Symptoms are usually present for over a month by the time patients seek medical attention. A mild anemia is often present. One blood test, called an **erythrocyte sedimentation rate**, is very high, much more so than in most other diseases. The most important issue in evaluating polymyalgia rheumatica is to check for giant cell arteritis. Giant cell arteritis can lead to blindness if left untreated.

Treatment

Polymyalgia rheumatica responds dramatically to cortisone-like drugs in modest doses. In fact, one part of confirming the diagnosis is to observe the response to this treatment. It may also respond to **nonsteroidal anti-inflammatory drugs** (NSAIDs). Temporal arteritis is also treated with cortisone, but in higher doses.

Prognosis

The disease often remits after time, with no further treatment required.

Resources

BOOKS

- Fauci, Anthony S., et al., eds. *Harrison's Principles of Internal Medicine*. 17th ed. New York: McGraw-Hill Professional, 2008.

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Polymyositis

Definition

Polymyositis is an inflammatory muscle disease causing weakness and **pain**. **Dermatomyositis** is identical to polymyositis with the addition of a characteristic skin rash.

Description

Polymyositis (PM) is an inflammatory disorder in which muscle tissue becomes inflamed and deteriorates, causing weakness and pain. It is one of several types of inflammatory muscle disease, or myopathy. Others include dermatomyositis (DM) and inclusion body **myositis**. All three types are progressive conditions, usually beginning in adulthood. A fourth type, juvenile dermatomyositis, occurs in children. Although PM and DM can occur at any age, 60% of cases appear between the ages of 30 and 60. Females are affected twice as often as males.

Causes and symptoms

Causes

The cause of PM and DM is not known, but it is suspected that a variety of factors may play a role in the development of these diseases. PM and DM may be autoimmune diseases, caused by the immune system's attack on the body's own tissue. The reason for this attack is unknown, although some researchers believe that a combination of immune system susceptibility and an environmental trigger may explain at least some cases. Known environmental agents associated with PM and DM include infectious agents such as *Toxoplasma*, *Borrelia* (**Lyme disease** bacterium), and coxsackievirus. Most cases, however, have no obvious triggers (direct causative agents). There may also be a genetic component in the development of PM and DM.

Symptoms

The early symptoms of PM and DM are slowly progressing muscle weakness, usually symmetrical between the two sides of the body. PM and DM affect primarily the muscles of the trunk and those closest to the trunk, while the hands, feet, and face usually are not involved. Weakness may cause difficulty walking, standing, and lifting objects. Rarely, the muscles of breathing may be affected. Weakness of the muscles used for swallowing can cause difficulty with swallowing (dysphagia). Joint pain and/or swelling also may be present. Later in the course of these diseases, muscle

wasting or shortening (contracture) may develop in the arms or legs. Heart abnormalities, including electrocardiogram (ECG) changes and **arrhythmias**, develop at some time during the course of these diseases in about 30% of patients.

Dermatomyositis is marked by a skin rash. The rash is dusky, reddish, or lilac in color, and is most often seen on the eyelids, cheeks, bridge of the nose, and knuckles, as well as on the back, upper chest, knees, and elbows. The rash often appears before the muscle weakness.

Diagnosis

PM and DM are often difficult diseases to diagnose, because they are rare, because symptoms come on slowly, and because they can be mistaken for other diseases that cause muscle weakness, especially limb girdle **muscular dystrophy**.

Accurate diagnosis involves:

- A neurological exam.
- Blood tests to determine the level of the muscle enzyme creatine kinase, whose presence in the circulation indicates muscle damage.
- Electromyography, an electrical test of muscle function.
- Muscle biopsy, in which a small sample of affected muscle is surgically removed for microscopic analysis. A biopsy revealing muscle cells surrounded by immune system cells is a strong indicator of myositis.

Treatment

PM and DM respond to high doses of **immunosuppressant drugs** in most cases. The most common medication used is the corticosteroid prednisone. Prednisone therapy usually leads to improvement within two or three months, at which point the dose can be tapered to a lower level to avoid the significant side effects associated with high doses of prednisone. Unresponsive patients are often given a replacement or supplementary immunosuppressant, such as azathioprine, cyclosporine, or methotrexate. Intravenous immunoglobulin treatments may help some people who are unresponsive to other immunosuppressants.

Pain can usually be controlled with an over-the-counter analgesic, such as **aspirin**, ibuprofen, or naproxen. A speech-language therapist can help suggest exercises and tips to improve difficulty in swallowing. Avoiding weight gain helps prevent overtaxing weakened muscles.

KEY TERMS

Autoimmune disease—A disease in which the body's immune system, responsible for fighting off foreign invaders such as bacteria and viruses, begins to attack and damage a part of the body as if it were foreign.

Immunosuppressant—A drug that reduces the body's natural immunity by suppressing the natural functioning of the immune system.

Alternative treatment

As with all autoimmune conditions, food allergies or intolerances and environmental triggers may be contributing factors. For **food allergies** and intolerances, an elimination challenge diet can be used under the supervision of a trained practitioner, naturopath, or nutritionist, to identify trigger foods. These foods can then be eliminated from the person's diet. For environmental triggers, it is helpful to identify the source so that it can be avoided or eliminated. A thorough **detoxification** program can help alleviate symptoms and change the course of the disease. Dietary changes from processed foods to whole foods that do not include allergen triggers can have significant results. Nutrient supplements, especially the **antioxidants** zinc, selenium, and **vitamins** A, C, and E, can be beneficial. Constitutional homeopathic treatment can work at a deep level to rebalance the whole person. **Acupuncture** and Chinese herbs can be effective in symptom alleviation and deep healing. Visualization, **guided imagery**, and hypnosis for **pain management** are also useful.

Prognosis

The progression of PM and DM varies considerably from person to person. Immunosuppressants can improve strength, although not all patients respond, and relapses may occur. PM and DM can lead to increasing weakness and disability, although the life span usually is not significantly affected. About half of the patients recover and can discontinue treatment within five years of the onset of their symptoms. About 20% still have active disease requiring ongoing treatment after five years, and about 30% have inactive disease but some remaining muscle weakness.

Prevention

There is no known way to prevent myositis, except to avoid exposure to those environmental agents that may be associated with some cases.

ORGANIZATIONS

Muscular Dystrophy Association, 3300 East Sunrise Drive, Tucson, AZ, 85718, (800) 572-1717, <http://www.mdausa.org>.

The Myositis Association, 1737 King Street, Suite 600, Alexandria, VA, 22314, (800) 821-7356, TMA@myositis.org, <http://www.myositis.org>.

Myositis Support Group, 146 Newtown Road, Southampton, England, SO19 9HR, 44023 8044 9708, 44023 8039 6402, msg@myositis.org.uk, <http://www.myositis.org.uk>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), 1 AMS Circle, Bethesda, MD, 20892-3675, 301 495-4484, (301) 718-6366, (877) 226-4267, NIAMSinfo@mail.nih.gov, <http://www.niams.nih.gov>.

Richard Robinson

Polyneuritis see **Peripheral neuropathy**

Polysomnography

Definition

The word polysomnography, derived from the Greek roots “poly,” meaning many, “somno,” meaning sleep, and “graphy” meaning to write, refers to multiple tests performed on patients while they sleep. Polysomnography is an overnight test to evaluate **sleep disorders**. Polysomnography generally includes monitoring of the patient's airflow through the nose and mouth, blood pressure, electrocardiographic activity, blood oxygen level, brain wave pattern, eye movement, and the movement of respiratory muscle and limbs.

Purpose

Polysomnography is used to help diagnose and evaluate a number of sleep disorders. For instance, it can help diagnose **sleep apnea**, a common disorder in middle-aged and elderly obese men, in which the muscles of the soft palate in the back of the throat relax and close off the airway during sleep. This may cause the person to snore loudly and gasp for air at night, and to be excessively sleepy and doze off during the day. Another syndrome often evaluated by polysomnography is **narcolepsy**. In narcolepsy, people have sudden attacks of sleep and/or cataplexy (temporary loss of muscle tone caused by moments of emotion, such as fear, anger, or surprise, which causes people to slump or fall over), sleep **paralysis**

or **hallucinations** at the onset of sleep. Polysomnography is often used to evaluate parasomnias (abnormal behaviors or movements during sleep), such as sleep walking, talking in one's sleep, nightmares, and **bedwetting**. It can also be used to detect or evaluate seizures that occur in the middle of the night, when the patient and his or her family are unlikely to be aware of them.

Precautions

Polysomnography is extremely safe and no special precautions need to be taken.

Description

Polysomnography requires an overnight stay in a sleep laboratory. During this stay, while the patient sleeps, he or she is monitored in a number of ways that can provide very useful information.

One form of monitoring is **electroencephalography** (EEG), in which electrodes are attached to the patient's scalp in order to record his or her brain wave activity. The electroencephalograph records brain wave activity from different parts of the brain and charts them on a graph. The EEG not only helps doctors establish what stage of sleep the patient is in, but may also detect seizures.

Another form of monitoring is continuous electro-oculography (EOG), which records eye movement and is used to determine when the patient is going through a stage of sleep called rapid-eye-movement (REM) sleep. Both EEG and EOG can be helpful in determining sleep latency (the time that transpires between lights out and the onset of sleep), total sleep time, the time spent in each sleep stage, and the number of arousals from sleep.

The air flow through the patient's nose and mouth are measured by heat-sensitive devices called thermistors. This can help detect episodes of apnea (stopped breathing), or hypnoeaa (inadequate breathing). Another test called pulse oximetry measures the amount of oxygen in the blood, and can be used to assess the degree of oxygen **starvation** during episodes of hypnoeaa or apnea.

The electrical activity of the patient's heart is also measured on an electrocardiogram, or ECG. Electrodes are affixed to the patient's chest and they pick up electrical activity from various areas of the heart. They help detect cardiac arrhythmias (abnormal heart rhythms), which may occur during periods of sleep apnea. Blood pressure is also measured: sometimes

episodes of sleep apnea can dangerously elevate blood pressure.

In some cases, sleep laboratories monitor the movement of limbs during sleep. This can be helpful in detecting such sleep disorders as periodic limb movements.

Preparation

The patient may be asked to discontinue taking any medications used to help him/her sleep. Before the patient goes to sleep, the technician hooks him or her up to all of the monitors being used.

Aftercare

Once the test is over, the monitors are detached from the patient. No special measures need to be taken after polysomnography.

Normal results

A normal result in polysomnography shows normal results for all parameters (EEG, ECG, blood pressure, eye movement, air flow, pulse oximetry, etc.) monitored throughout all stages of sleep.

Abnormal results

Polysomnography may yield a number of abnormal results, indicating a number of potential disorders. For instance, abnormal transitions in and out of various stages of sleep, as documented by the EEG and the EOG, may be a sign of narcolepsy. Reduced air flow through the nose and mouth, along with a fall in oxygenation of the blood, may indicate apnea or hypopnea. If apnea is accompanied by abnormalities in ECG or elevations in blood pressure, this can indicate that sleep apnea may be particularly harmful. Frequent movement of limbs may indicate a sleep disorder called periodic limb movement.

ORGANIZATIONS

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

Robert Scott Dinsmoor

Pompe's disease see **Glycogen storage diseases**

Porphyrias

Definition

The porphyrias are disorders in which the body produces too much porphyrin and insufficient heme (an iron-containing nonprotein portion of the hemoglobin molecule). Porphyrin is a foundation structure for heme and certain enzymes. Excess porphyrins are excreted as waste in the urine and stool. Overproduction and overexcretion of porphyrins causes low, unhealthy levels of heme and certain important enzymes, creating various physical symptoms.

Description

Biosynthesis of heme is a multistep process that begins with simple molecules and ends with a large, complex heme molecule. Each step of the chemical pathway is directed by its own task-specific protein, called an enzyme. As a heme precursor molecule moves through each step, an enzyme modifies the precursor in some way. If a precursor molecule is not modified, it cannot proceed to the next step, causing a buildup of that specific precursor.

This situation is the main characteristic of the porphyrias. Owing to a defect in one of the enzymes of the heme biosynthesis pathway, protoporphyrins or porphyrins (heme precursors) are prevented from proceeding further along the pathway. These precursors accumulate at the stage of the enzyme defect causing an array of physical symptoms in an affected person. Specific symptoms depend on the point at which heme biosynthesis is blocked and which precursors accumulate. In general, the porphyrias primarily affect the skin and the nervous system. Symptoms can be debilitating or life threatening in some cases. Porphyria is most commonly an inherited condition. It can also, however, be acquired after exposure to poisonous substances.

Heme

Heme is produced in several tissues in the body, but its primary biosynthesis sites are the liver and the bone marrow. Heme synthesis for immature red blood cells, namely the erythroblasts and the reticulocytes, occurs in the bone marrow.

Although production is concentrated in the liver and bone marrow, heme is utilized in various capacities in virtually every tissue in the body. In most cells, heme is a key building block in the construction of factors that oversee metabolism and transport of oxygen and energy. In the liver, heme is a component of several vital enzymes, particularly cytochrome P450.

Cytochrome P450 is involved in the metabolism of chemicals, **vitamins**, fatty acids, and hormones; it is very important in transforming toxic substances into easily excretable materials. In immature red blood cells, heme is the featured component of hemoglobin. Hemoglobin is the red pigment that gives red blood cells their characteristic color and their essential ability to transport oxygen.

Heme biosynthesis

The heme molecule is composed of porphyrin and an iron atom. Much of the heme biosynthesis pathway is dedicated to constructing the porphyrin molecule. Porphyrin is a large molecule shaped like a four-leaf clover. An iron atom is placed at its center point in the last step of heme biosynthesis.

The production of heme may be compared to a factory assembly line. At the start of the line, raw materials are fed into the process. At specific points along the line, an addition or adjustment is made to further development. Once additions and adjustments are complete, the final product rolls off the end of the line.

The heme “assembly line” is an eight-step process, requiring eight different and properly functioning enzymes:

1. delta-aminolevulinic acid synthase
2. delta-aminolevulinic acid dehydratase
3. porphobilogen deaminase
4. uroporphyrinogen III cosynthase
5. uroporphyrinogen decarboxylase
6. coproporphyrinogen oxidase
7. protoporphyrinogen oxidase
8. ferrochelatase

The control of heme biosynthesis is complex. Various chemical signals can trigger increased or decreased production. These signals can affect the enzymes themselves or the production of these enzymes, starting at the genetic level. For example, one point at which heme biosynthesis may be controlled is at the first step. When heme levels are low, greater quantities of delta-aminolevulinic acid (ALA) synthase are produced. As a result, larger quantities of heme precursors are fed into the biosynthesis pathway to step up heme production.

Porphyrias

Under normal circumstances, when heme concentrations are at an appropriate level, precursor production decreases. However, a glitch in the biosynthesis pathway—represented by a defective enzyme—means that heme biosynthesis does not reach completion.

Because heme levels remain low, the synthesis pathway continues to churn out precursor molecules in an attempt to correct the heme deficit.

The net effect of this continued production is an abnormal accumulation of precursor molecules and development of some type of porphyria. Each type of porphyria corresponds with a specific enzyme defect and an accumulation of the associated precursor. Although there are eight steps in heme biosynthesis, there are only seven types of porphyrias; a defect in ALA synthase activity does not have a corresponding porphyria.

Enzymes involved in heme biosynthesis display subtle, tissue-specific variations; therefore, heme biosynthesis may be impeded in the liver, but normal in the immature red blood cells, or vice versa. Incidence of porphyria varies widely between types and occasionally by geographic location. Although certain porphyrias are more common than others, their greater frequency is only relative to other types. All porphyrias are considered to be rare disorders.

In the past, the porphyrias were divided into two general categories based on the location of the porphyrin production. Porphyrrias affecting heme biosynthesis in the liver were referred to as hepatic porphyrias. Porphyrrias that affect heme biosynthesis in immature red blood cells were referred to as erythropoietic porphyrias (erythropoiesis is the process through which red blood cells are produced). Porphyrrias were usually grouped into acute and non-acute types. Acute porphyrias produce severe attacks of pain and neurological effects. Non-acute porphyrias present as chronic diseases.

The acute porphyrias, and the heme biosynthesis steps at which enzyme defects occur, are:

- ALA dehydratase deficiency porphyria (step 2). This porphyria type is very rare. The inheritance pattern appears to be autosomal recessive. In autosomal recessively inherited disorders, a person must inherit two defective genes, one from each parent. A parent with only one gene for an autosomal recessive disorder does not display symptoms of the disease.
- Acute intermittent porphyria (step 3). Acute intermittent porphyria (AIP) is also known as Swedish porphyria, pyrroloporphyria, and intermittent acute porphyria. AIP is inherited as an autosomal dominant trait, which means that only one copy of the defective gene needs to be present for the disorder to occur. Simply inheriting this gene, however, does not necessarily mean that a person will develop the disease. Approximately five to 10 per 100,000 people in the United States carry a gene for AIP, but only 10% of these people ever develop symptoms of the disease.

• Hereditary coproporphyria (step 6). Hereditary coproporphyria (HCP) is inherited in an autosomal dominant manner. As with all porphyrias, it is an uncommon ailment. By 1977, only 111 cases of HCP were recorded; in Denmark, the estimated incidence is two in one million people.

• Variegate porphyria (step 7). Variegate porphyria (VP) is also known as porphyria variegata, protocoproporphyria, South African genetic porphyria, and Royal malady (supposedly King George III of England and Mary, Queen of Scots, suffered from VP). VP is inherited in an autosomal dominant manner and is especially prominent in South Africans of Dutch descent. Among that population, the incidence is approximately three in 1,000 persons. It is estimated that there are 10,000 cases of VP in South Africa. Interestingly, it appears that the affected South Africans are descendants of two Dutch settlers who came to South Africa in 1680. Among other populations, the incidence of VP is estimated to be one to two cases per 100,000 persons.

The non-acute porphyrias, and the steps of heme biosynthesis at which they occur, are:

- Congenital erythropoietic porphyria (step 4). Congenital erythropoietic porphyria (CEP) is also called Gunther's disease, erythropoietic porphyria, congenital porphyria, congenital hematoporphyria, and erythropoietic uroporphyrinia. CEP is inherited in an autosomal recessive manner. It is a rare disease, estimated to affect fewer than one in one million people. Onset of dramatic symptoms usually occurs in infancy, but may hold off until adulthood.
- Porphyria cutanea tarda (step 5). Porphyria cutanea tarda (PCT) is also called symptomatic porphyria, porphyria cutanea symptomatica, and idiosyncratic porphyria. PCT may be acquired, typically as a result of disease (especially hepatitis C), drug or alcohol use, or exposure to certain poisons. PCT may also be inherited as an autosomal dominant disorder, however most people remain latent—that is, symptoms never develop. PCT is the most common of the porphyrias, but the incidence of PCT is not well defined.
- Hepatoerythropoietic porphyria (step 5). Hepatoerythropoietic porphyria (HEP) affects heme biosynthesis in both the liver and the bone marrow. HEP results from a defect in uroporphyrinogen decarboxylase activity (step 5), and is caused by defects in the same gene as PCT. Disease symptoms, however, strongly resemble congenital erythropoietic porphyria. HEP seems to be inherited in an autosomal recessive manner.

- Erythropoietic protoporphyrria (step 8). Also known as protoporphyrria and erythrohepatic protoporphyrria, erythropoietic protoporphyrria (EPP) is more common than CEP; more than 300 cases have been reported. In these cases, onset of symptoms typically occurred in childhood.

Causes and symptoms

General characteristics

The underlying cause of all porphyrias is a defective enzyme important to the heme biosynthesis pathway. Porphyrias are inheritable conditions. In virtually all cases of porphyria an inherited factor causes the enzyme's defect. An environmental trigger—such as diet, drugs, or sun exposure—may be necessary before any symptoms develop. In many cases, symptoms do not develop. These asymptomatic individuals may be completely unaware that they have a gene for porphyria.

All of the hepatic porphyrias—except porphyria cutanea tarda—follow a pattern of acute attacks separated by periods during which no symptoms are present. For this reason, this group is often referred to as the acute porphyrias. The erythropoietic porphyrias and porphyria cutanea tarda do not follow this pattern and are considered to be chronic conditions.

The specific symptoms of each porphyria vary based on which enzyme is affected and whether that enzyme occurs in the liver or in the bone marrow. The severity of symptoms can vary widely, even within the same type of porphyria. If the porphyria becomes symptomatic, the common factor between all types is an abnormal accumulation of protoporphyrins or porphyrin.

ALA dehydratase porphyria (ADP)

ADP is characterized by a deficiency of ALA dehydratase. ADP is caused by mutations in the delta-aminolevulinate dehydratase gene (ALAD) at 9q34. Of the few cases on record, the prominent symptoms are **vomiting**, pain in the abdomen, arms, and legs, and neuropathy. (Neuropathy refers to nerve damage that can cause pain, **numbness**, or paralysis.) The nerve damage associated with ADP could cause breathing impairment or lead to weakness or **paralysis** of the arms and legs.

Acute intermittent porphyria (AIP)

AIP is caused by a deficiency of porphobilogen deaminase, which occurs due to mutations in the hydroxymethylbilane synthase gene (HMBS) located

at 11q23.3. Symptoms of AIP usually do not occur unless a person with the deficiency encounters a trigger substance. Trigger substances can include hormones (for example **oral contraceptives**, menstruation, **pregnancy**), drugs, and dietary factors. Most people with this deficiency never develop symptoms.

Attacks occur after **puberty** and commonly feature severe abdominal pain, **nausea**, **vomiting**, and **constipation**. Muscle weakness and pain in the back, arms, and legs are also typical symptoms. During an attack, the urine is a deep reddish color. The central nervous system may also be involved. Possible psychological symptoms include **hallucinations**, confusion, seizures, and mood changes.

Congenital erythropoietic porphyria (CEP)

CEP is caused by a deficiency of uroporphyrinogen III cosynthase due to mutations in the uroporphyrinogen III cosynthase gene (UROS) located at 10q25.2-q26.3. Symptoms are often apparent in infancy and include reddish urine and possibly an enlarged spleen. The skin is unusually sensitive to light and blisters easily if exposed to sunlight. (Sunlight induces protoporphyrin changes in the plasma and skin. These altered protoporphyrin molecules can cause skin damage.) Increased hair growth is common. Damage from recurrent blistering and associated skin infections can be severe. In some cases facial features and fingers may be lost to recurrent damage and infection. Deposits of protoporphyrins can sometimes lead to red staining of the teeth and bones.

Porphyria cutanea tarda (PCT)

PCT is caused by deficient uroporphyrinogen decarboxylase. PCT is caused by mutations in the uroporphyrinogen decarboxylase gene (UROD) located at 1p34. PCT may occur as an acquired or an inherited condition. The acquired form usually does not appear until adulthood. The inherited form may appear in childhood, but often demonstrates no symptoms. Early symptoms include blistering on the hands, face, and arms following minor injuries or exposure to sunlight. Lightening or darkening of the skin may occur along with increased hair growth or loss of hair. Liver function is abnormal but the signs are mild.

Hepatoerythropoietic porphyria (HEP)

HEP is linked to a deficiency of uroporphyrinogen decarboxylase in both the liver and the bone marrow. HEP is an autosomal recessive disease caused by mutations in the gene responsible for PCT, the uroporphyrinogen decarboxylase gene (UROD), located

at 1p34. The gene is shared, but the mutations, inheritance, and specific symptoms of these two diseases are different. The symptoms of HEP resemble those of CEP.

Hereditary coproporphyria (HCP)

HCP is similar to AIP, but the symptoms are typically milder. HCP is caused by a deficiency of coproporphyrinogen oxidase due to mutations in a gene by the same name at 3q12. The greatest difference between HCP and AIP is that people with HCP may have some skin sensitivity to sunlight. However, extensive damage to the skin is rarely seen.

Variegate porphyria (VP)

VP is caused by a deficiency of protoporphyrinogen oxidase. There is scientific evidence that VP is caused by mutation in the gene for protoporphyrinogen oxidase located at 1q22. Like AIP, symptoms of VP occur only during attacks. Major symptoms of this type of porphyria include neurological problems and sensitivity to light. Areas of the skin that are exposed to sunlight are susceptible to burning, blistering, and scarring.

Erythropoietic protoporphyrinia (EPP)

Owing to deficient ferrochelatase, the last step in the heme biosynthesis pathway—the insertion of an iron atom into a porphyrin molecule—cannot be completed. This enzyme deficiency is caused by mutations in the ferrochelatase gene (FECH) located at 18q21.3. The major symptoms of this disorder are related to sensitivity to light—including both artificial and natural light sources. Following exposure to light, a person with EPP experiences burning, **itching**, swelling, and reddening of the skin. Blistering and scarring may occur but are neither common nor severe. EPP is associated with increased risks for **gallstones** and liver complications. Symptoms can appear in childhood and tend to be more severe during the summer when exposure to sunlight is more likely.

Diagnosis

Depending on the array of symptoms an individual may exhibit, the possibility of porphyria may not immediately come to a physician's mind. In the absence of a family history of porphyria, non-specific symptoms, such as abdominal pain and vomiting, may be attributed to other disorders. Neurological symptoms, including confusion and hallucinations, can lead to an initial suspicion of psychiatric disease. Diagnosis is more easily accomplished in cases in which non-specific symptoms appear in combination with

symptoms more specific to porphyria, like neuropathy, sensitivity to sunlight, or certain other manifestations. Certain symptoms, such as urine the color of port wine, are hallmark signs very specific to porphyria. DNA analysis is not yet of routine diagnostic value.

A common initial test measures protoporphyrins in the urine. However, if skin sensitivity to light is a symptom, a blood plasma test is indicated. If these tests reveal abnormal levels of protoporphyrins, further tests are done to measure heme precursor levels in red blood cells and the stool. The presence and estimated quantity of porphyrin and protoporphyrins in biological samples are easily detected using spectrofluorometric testing. Spectrofluorometric testing uses a spectrofluorometer that directs light of a specific strength at a fluid sample. The porphyrins and protoporphyrins in the sample absorb the light energy and fluoresce, or glow. The spectrofluorometer detects and measures fluorescence, which indicates the amount of porphyrins and protoporphyrins in the sample.

Whether heme precursors occur in the blood, urine, or stool gives some indication of the type of porphyria, but more detailed biochemical testing is required to determine their exact identity. Making this determination yields a strong indicator of which enzyme in the heme biosynthesis pathway is defective; which, in turn, allows a diagnosis of the particular type of porphyria.

Biochemical tests rely on the color, chemical properties, and other unique features of each heme precursor. For example, a screening test for acute intermittent porphyria (AIP) is the Watson-Schwartz test. In this test, a special dye is added to a urine sample. If one of two heme precursors—porphobilinogen or urobilinogen—is present, the sample turns pink or red. Further testing is necessary to determine whether the precursor present is porphobilinogen or urobilinogen—only porphobilinogen is indicative of AIP.

Other biochemical tests rely on the fact that heme precursors become less soluble in water (able to be dissolved in water) as they progress further through the heme biosynthesis pathway. For example, to determine whether the Watson-Schwartz urine test is positive for porphobilinogen or urobilinogen, chloroform is added to the test tube. Chloroform is a water-insoluble substance. Even after vigorous mixing, the water and chloroform separate into two distinct layers. Urobilinogen is slightly insoluble in water, while porphobilinogen tends to be water soluble. The porphobilinogen mixes more readily in water than chloroform, so if the water layer is pink (from the dye added to the urine

sample), that indicates the presence of porphobilinogen, and a diagnosis of AIP is probable.

As a final test, measuring specific enzymes and their activities may be done for some types of porphyrias; however, such tests are not done as a screening method. Certain enzymes, such as porphobilinogen deaminase (the defective enzyme in AIP), can be easily extracted from red blood cells; other enzymes, however, are less readily collected or tested. Basically, an enzyme test involves adding a certain amount of the enzyme to a test tube that contains the precursor it is supposed to modify. Both the production of modified precursor and the rate at which it appears can be measured using laboratory equipment. If a modified precursor is produced, the test indicates that the enzyme is doing its job. The rate at which the modified precursor is produced can be compared to a standard to measure the efficiency of the enzyme.

Treatment

Treatment for porphyria revolves around avoiding acute attacks, limiting potential effects, and treating symptoms. Treatment options vary depending on the specific type of porphyria diagnosed. **Gene therapy** has been successful for both CEP and EPP. In the future, scientists expect development of gene therapy for the remaining porphyrias. Given the rarity of ALA dehydratase porphyria, definitive treatment guidelines for this rare type have not been developed.

Acute intermittent porphyria, hereditary coproporphyria, and variegate porphyria

Treatment for acute intermittent porphyria, hereditary coproporphyria, and variegate porphyria follows the same basic regime. A person who has been diagnosed with one of these porphyrias can prevent most attacks by avoiding precipitating factors, such as certain drugs that have been identified as triggers for acute porphyria attacks. Individuals must maintain adequate **nutrition**, particularly with respect to carbohydrates. In some cases, an attack can be stopped by increasing carbohydrate consumption or by receiving carbohydrates intravenously. In 2004, a report from Turkey revealed successful treatment of an acute intermittent porphyria attack with a drug called fluoxetine.

When attacks occur prompt medical attention is necessary. Pain is usually severe, and narcotic **analgesics** are the best option for relief. Phenothiazines can be used to counter nausea, vomiting, and **anxiety**, and chloral hydrate or diazepam is useful for **sedation** or to induce sleep. Hematin, a drug administered

intravenously, may be used to halt an attack. Hematin seems to work by signaling the pathway of heme biosynthesis to slow production of precursors. Women, who tend to develop symptoms more frequently than men owing to hormonal fluctuations, may find ovulation-inhibiting hormone therapy to be helpful.

Gene therapy is a possible future treatment for these porphyrias. An experimental animal model of AIP has been developed and research is in progress.

Congenital erythropoietic porphyria

The key points of congenital erythropoietic porphyria treatment are avoiding exposure to sunlight and prevention of skin trauma or skin infection. Liberal use of **sunscreens** and consumption of beta-carotene supplements can provide some protection from sun-induced damage. Medical treatments such as removing the spleen or administering transfusions of red blood cells can create short-term benefits, but these treatments do not offer a cure. Remission can sometimes be achieved after treatment with oral doses of **activated charcoal**. Severely affected patients may be offered **bone marrow transplantation** which appears to confer long-term benefit.

Porphyria cutanea tarda

As with other porphyrias, the first line of defense is avoidance of factors, especially alcohol, that could bring about symptoms. Regular blood withdrawal is a proven therapy for pushing symptoms into remission. If an individual is anemic or cannot have blood drawn for other reasons, chloroquine therapy may be used.

Erythropoietic protoporphyrina

Avoiding sunlight, using sunscreens, and taking beta-carotene supplements are typical treatment options for erythropoietic protoporphyrina. The drug cholestyramine may reduce the skin's sensitivity to sunlight as well as the accumulated heme precursors in the liver. **Liver transplantation** has been used in cases of liver failure. In 2004, a report in a medical journal told of one case of successful treatment of a 19-year-old patient with acute intermittent porphyria with liver transplantation. While she had only been studied for 1.5 years, the authors said her quality of life was good and they hoped that the procedure would offer cure for select patients with severe forms of the disease.

Alternative treatment

Acute porphyria attacks can be life-threatening events, so attempts at self-treatment can be dangerous.

KEY TERMS

Autosomal dominant—A pattern of genetic inheritance in which only one abnormal gene is needed to display the trait or disease.

Autosomal recessive—A pattern of genetic inheritance in which two abnormal genes are needed to display the trait or disease.

Biosynthesis—The manufacture of materials in a biological system.

Bone marrow—A spongy tissue located in the hollow centers of certain bones, such as the skull and hip bones. Bone marrow is the site of blood cell generation.

Enzyme—A protein that catalyzes a biochemical reaction or change without changing its own structure or function.

Erythropoiesis—The process through which new red blood cells are created; it begins in the bone marrow.

Erythropoietic—Referring to the creation of new red blood cells.

Gene—A building block of inheritance, which contains the instructions for the production of a

particular protein, and is made up of a molecular sequence found at a section of DNA. Each gene is found on a precise location on a chromosome.

Hematin—A drug administered intravenously to halt an acute porphyria attack. It causes heme biosynthesis to decrease, preventing the further accumulation of heme precursors.

Heme—The iron-containing molecule in hemoglobin that serves as the site for oxygen binding.

Hemoglobin—Protein-iron compound in the blood that carries oxygen to the cells and carries carbon dioxide away from the cells.

Hepatic—Referring to the liver.

Neuropathy—A condition caused by nerve damage. Major symptoms include weakness, numbness, paralysis, or pain in the affected area.

Porphyrin—A large molecule shaped like a four-leaf clover. Combined with an iron atom, it forms a heme molecule.

Protoporphyrin—A precursor molecule to the porphyrin molecule.

Alternative treatments can be useful adjuncts to conventional therapy. For example, some people may find relief for the pain associated with acute intermittent porphyria, hereditary coproporphyria, or variegate porphyria through **acupuncture** or hypnosis. Relaxation techniques, such as **yoga** or **meditation**, may also prove helpful in **pain management**.

Prognosis

Even when porphyria is inherited, symptom development depends on a variety of factors. In the majority of cases, a person remains asymptomatic throughout life. About one percent of acute attacks can be fatal. Other symptoms may be associated with temporarily debilitating or permanently disfiguring consequences. Measures to avoid these consequences are not always successful, regardless of how diligently they are pursued. Although pregnancy has been known to trigger porphyria attacks, dangers associated with pregnancy as not as great as was once thought.

Prevention

For the most part, the porphyrias are attributable to inherited genes; such inheritance cannot be prevented. However, symptoms can be limited or

prevented by avoiding factors that trigger symptom development.

People with a family history of an acute porphyria should be screened for the disease. Even if symptoms are absent, it is useful to know about the presence of the gene to assess the risks of developing the associated porphyria. This knowledge also reveals whether a person's offspring may be at risk. Prenatal testing for certain porphyrias is possible. Prenatal diagnosis of congenital erythropoietic porphyria has been successfully accomplished. Any prenatal tests, however, would not indicate whether a child would develop porphyria symptoms; only that the potential is there.

Resources

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ORGANIZATIONS

The American Porphyria Foundation, 4900 Woodway, Suite 780, Houston, TX, 77056-1837, (713) 266-9617, (713) 840-9552, (866) 273-3635, <http://www.porphyriafoundation.com>.

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Port-wine stain see **Birthmarks**

Portacaval shunting see **Portal vein bypass**

Portal-systemic encephalopathy see **Liver encephalopathy**

function, but also suffer from an enlarged spleen, **jaundice**, and damage to the vascular system brought on by years of **alcoholism**. They are likely to experience serious complications during surgery. Some patients are aggressively uncooperative with medical personnel. Under these conditions, half the patients may not survive the operation.

Description

A choice of portal vein bypasses is available. Portal vein bypass is usually performed as an emergency operation in a hospital under **general anesthesia**. The surgeon makes an abdominal incision and finds the portal vein. In portacaval shunting, blood from the portal vein is diverted into the inferior vena cava. This is the most common bypass. In splenorenal shunting, the splenic vein (a part of the portal vein), is connected to the renal vein. A mesocaval shunt connects the superior mesenteric vein (another part of the portal vein) to the inferior vena cava.

Portal pressure can also be reduced in a procedure called transvenous intrahepatic portosystemic shunt (TIPS). A catheter is threaded into the portal vein, and an expandable balloon or wire mesh is inserted to divert blood from the portal vein to the hepatic vein. The rate of serious complications in TIPS is only 1–2%. The operation cannot be performed at all hospitals, but is becoming the preferred treatment for reducing portal pressure.

Preparation

Standard preoperative blood and urine tests are performed, and liver function is evaluated. The heart and arterial blood pressure are monitored both during and after the operation.

Aftercare

The patient will be connected to a heart monitor and fed through a nasogastric tube. Vital functions are monitored through blood and urine tests. Patients receive **pain** medication and **antibiotics**. Once released from the hospital, patients are expected to abstain from alcohol and follow a diet and medication schedule designed to reduce the risks of re-bleeding.

Risks

Portal vein bypass surgery is high risk because it is performed on patients who are generally in poor health. Only half the patients survive, although the chances of survival are greater with TIPS surgery.

Purpose

The portal vein carries blood from the stomach and abdominal organs to the liver. It is a major vein that splits into many branches. High pressure in the portal vein causes swelling and bleeding from blood vessels in the esophagus. This situation occurs when the liver is damaged from **cirrhosis** of the liver, a condition usually caused by prolonged, excessive alcohol consumption.

Massive internal bleeding caused by high pressure in the portal vein occurs in about 40% of patients with cirrhosis. It is initially fatal in at least half of these patients. Patients who survive are likely to experience bleeding recurrence. Portal vein bypass, also called portacaval shunting, is performed on these surviving patients to control bleeding.

Precautions

Most patients who need portal vein bypass surgery not only have **liver disease** and poor liver

KEY TERMS

Cirrhosis—A chronic degenerative liver disease common among alcoholics.

Inferior vena cava—A large vein that returns blood from the legs, pelvis, and abdomen to the heart.

Portal vein—Formed by a fusion of small veins that end in a network of capillaries, the portal vein delivers blood to the liver.

Those patients who survive the operation still face the risk of **heart failure**, brain disease due to a decrease in the liver's conversion of waste products (**liver encephalopathy**), hemorrhage, lung complications, infection, **coma**, and **death**.

Normal results

The survival rate is directly related to the amount of liver damage patients have. The less damage, the more likely the patient is to recover. Cooperation with restrictions on alcohol and diet affect long-term survival.

Resources

BOOKS

McPhee, Stephen, and Maxine Papadakis. *Current Medical Diagnosis and Treatment*, 2010, 49th ed. New York: McGraw-Hill Medical, 2009.

Tish Davidson, A.M.

Positron emission tomography (PET)

Definition

Positron emission tomography (PET) is a non-invasive scanning technique that utilizes small amounts of radioactive positrons (positively charged particles) to visualize body function and metabolism.

Purpose

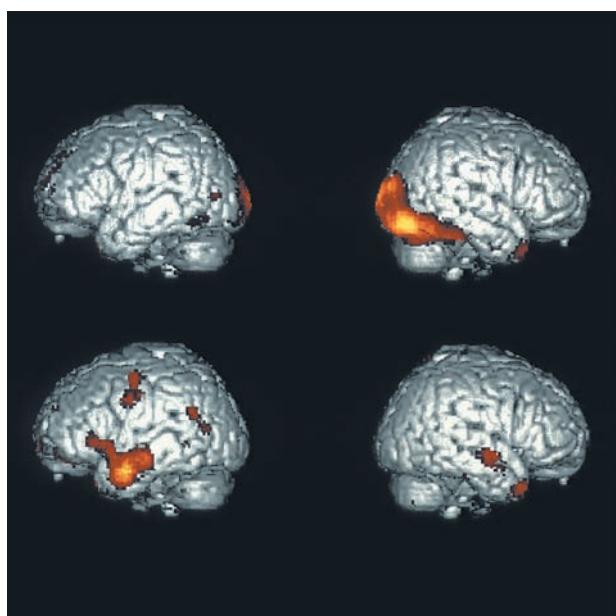
PET is the fastest growing nuclear medicine tool in terms of increasing acceptance and applications. It is useful in the diagnosis, staging, and treatment of **cancer** because it provides information that cannot be obtained by other techniques such as computed tomography (CT) and **magnetic resonance imaging** (MRI).

PET scans are performed at medical centers equipped with a small cyclotron. Smaller cyclotrons and increasing availability of certain radiopharmaceuticals are making PET a more widely used imaging modality.

Physicians first used PET to obtain information about brain function, and to study brain activity in various neurological diseases and disorders including **stroke**, **epilepsy**, **Alzheimer's disease**, Parkinson's disease, and Huntington's disease; and in psychiatric disorders such as **schizophrenia**, depression, **obsessive-compulsive disorder**, **attention deficit hyperactivity disorder (ADHD)**, and **Tourette syndrome**. PET is now used to evaluate patients for these cancers: head and neck, lymphoma, melanoma, lung, colorectal, breast, and esophageal. PET also is used to evaluate heart muscle function in patients with **coronary artery disease** or **cardiomyopathy**.

Description

PET involves injecting a patient with a radiopharmaceutical similar to glucose. An hour after injection of this tracer, a PET scanner images a specific metabolic function by measuring the concentration and distribution of the tracer throughout the body.



A PET scan showing brain activity while patient recognizes faces—left sides at left/right sides at right. Activity is prevalent in temporal lobe (bottom scans). (Photo Researchers, Inc.)

KEY TERMS

Electron—One of the small particles that make up an atom. An electron has the same mass and amount of charge as a positron, but the electron has a negative charge.

Gamma ray—A high-energy photon emitted by radioactive substances.

Half-life—The time required for half of the atoms in a radioactive substance to disintegrate.

Photon—A light particle.

Positron—One of the small particles that make up an atom. A positron has the same mass and amount of charge as an electron, but the positron has a positive charge.

When it enters the body, the tracer courses through the bloodstream to the target organ, where it emits positrons. The positively charged positrons collide with negatively charged electrons, producing gamma rays. The gamma rays are detected by photomultiplier-scintillator combinations positioned on opposite sides of the patient. These signals are processed by the computer and images are generated.

PET provides an advantage over CT and MRI because it can determine if a lesion is malignant. The two other modalities provide images of anatomical structures, but often cannot provide a determination of malignancy. CT and MRI show structure, while PET shows function. PET has been used in combination with CT and MRI to identify abnormalities with more precision and indicate areas of most active metabolism. This additional information allows for more accurate evaluation of cancer treatment and management.

Resources

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ORGANIZATIONS

American College of Physicians, 190 N. Independence Mall West, Philadelphia, PA, 19106-1572, (215) 351-2600, (800) 523-1546, <http://www.acponline.org>.

American Heart Association, 7272 Greenville Avenue, Dallas, TX, 75231, (214) 373-6300, (800) 242-8721, <http://www.americanheart.org>.

American Medical Association, 515 N. State Street, Chicago, IL, 60610, (312) 464-5000, <http://www.ama-assn.org>.

National Cancer Institute, Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD, 20892-2580, (800) 422-6237, <http://www.nci.nih.gov>.

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Post-concussion syndrome

Definition

Post-concussion syndrome (PCS) is a common but controversial disorder that presents with variety of symptoms including—but not limited to—headache, **dizziness**, **fatigue**, and personality changes.

Description

PCS occurs in approximately 23–93% of persons with mild to severe head injuries. It is estimated that a neurologist (a physician who specializes in nerve and brain disorders) sees five patients with PCS per month. There is no accurate correlation between the severity of injury and the development of PCS symptoms, since signs of the disorder can occur in someone who was just dazed by an injury. Some studies suggest that PCS symptoms occur at a higher rate in patients who were unconscious after trauma.

Causes and symptoms

PCS is most commonly caused by minor **head injury** called a **concussion**. The majority of patients with minor head injury characteristically develop PCS with distinct symptoms. Patients may report problems with concentration, recent memory, and abstract thinking. Additionally, patients may develop dizziness, irritability, fatigue, and personality changes. Elderly patients are particularly affected by disequilibrium and chronic dizziness even after minor trauma.

Diagnosis

There are no specific or reliable tests to diagnose PCS. A neuropsychologist can perform an in-depth neuropsychologic assessment that can determine

KEY TERMS

Disequilibrium—Difficulty with equilibrium that can mean a deficiency in balance and/or orientation.

Neuropsychologist—A clinical psychologist who specializes in assessing psychological status caused by a brain disorder.

presence or absence and extent of impairment. These tests may be performed for medical purposes.

Treatment

Treatment for PCS can be extensive. Medications for **headache** and **pain** may be indicated (**analgesics** and **muscle relaxants**). Antidepressants may be given to improve **insomnia**, irritability, or **anxiety**. Pain control could be achieved with **acupuncture**, nerve blocks, or transcutaneous **electrical nerve stimulation** (TENS, electrical stimulation of muscle groups). It is important for clinicians to educate caretakers and to provide referrals for **family therapy** and cognitive **rehabilitation** for the affected person.

Prognosis

The overall outcome is difficult to assess. Limited interpretation in literature is primarily due to the subjective nature of symptoms. Patient recovery is directed and evaluated by cognitive function changes, subjective symptoms, and return to work. Most cases of PCS can be a financial strain and threaten family stability. There may be compensation and litigation claims, which is often stressful and aggravates symptoms.

Resources

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Laith Farid Gulli, M.D.

Post-herpetic neuralgia see **Neuralgia**

Post-traumatic stress disorder

Definition

Post-traumatic **stress** disorder (PTSD) is a complex **anxiety** disorder that may occur when a person experiences or witnesses an event perceived as a threat and in which he or she experiences fear, terror, or helplessness. PTSD is sometimes summarized as “a normal reaction to abnormal events.” It was first defined as a distinctive disorder in 1980. Originally diagnosed in veterans of the Vietnam War, it is now recognized in civilian survivors of **rape** or other criminal assaults; natural disasters; plane crashes, train collisions, or industrial explosions; acts of terrorism; **child abuse**; or war.

Demographics

PTSD can develop in almost anyone in any age group exposed to a sufficiently terrifying event or chain of events. The National Institute of Mental Health (NIMH) estimated in 2007 that about 7.7 million adults in the United States have PTSD. One study found that 3.7 percent of a sample of teenage boys and 6.3 percent of adolescent girls had PTSD. It is estimated that a person’s risk of developing PTSD over the course of their life is between 8 and 10 percent. On average, 30 percent of soldiers who have been in a war zone develop PTSD. Women are at greater risk of PTSD following **sexual assault** or domestic violence, while men are at greater risk of developing PTSD following military combat.

Traumatic experiences are surprisingly common in the general North American population. More than 10% of the men and 6% of the women in one survey reported experiencing four or more types of trauma in their lives. The most frequently mentioned traumas are:

- witnessing someone being badly hurt or killed
- involvement in a fire, flood, earthquake, severe hurricane, or other natural disaster
- involvement in a life-threatening accident (workplace explosion or transportation accident)
- military combat

PTSD is more likely to develop in response to an intentional human act of violence or cruelty such as a rape or mugging than as a reaction to an impersonal catastrophe like a flood or hurricane. It is not surprising that the traumatic events most frequently mentioned by men diagnosed with PTSD are rape, combat exposure, childhood neglect, and childhood

physical abuse. For women diagnosed with PTSD, the most common traumas are rape, sexual molestation, physical attack, being threatened with a weapon, and childhood physical abuse.

PTSD can also develop in therapists, rescue workers, or witnesses of a frightening event as well as in those who were directly involved. This process is called vicarious traumatization.

Description

The experience of PTSD has sometimes been described as like being in a horror film that keeps replaying and can't be shut off. It is common for people with PTSD to feel intense fear and helplessness, and to relive the frightening event in nightmares or in their waking hours. Sometimes the memory is triggered by a sound, smell, or image that reminds the sufferer of the traumatic event. These reexperiences of the event are called flashbacks. A person with PTSD is also likely to be jumpy and easily startled or to go numb emotionally and lose interest in activities they used to enjoy. They may have problems with memory and with getting enough sleep. In some cases they may feel disconnected from the real world or have moments in which their own bodies seem unreal; these symptoms are indications of dissociation, a process in which the mind splits off certain memories or thoughts from conscious awareness. Many people with PTSD turn to alcohol or drugs in order to escape the flashbacks and other symptoms of the disorder, even if only for a few minutes.

Risk factors

Factors that influence the likelihood of a person's developing PTSD include:

- The nature, intensity, and duration of the traumatic experience. For example, someone who just barely escaped from the World Trade Center before the towers collapsed is at greater risk of PTSD than someone who saw the collapse from a distance or on television.
- The person's previous history. People who were abused as children, who were separated from their parents at an early age, or who have a previous history of anxiety or depression are at increased risk of PTSD.
- Genetic factors. Vulnerability to PTSD is known to run in families.
- The availability of social support after the event. People who have no family or friends are more likely to develop PTSD than those who do.

HIGH-RISK POPULATIONS. Some subpopulations in the United States are at greater risk of developing PTSD. The lifetime prevalence of PTSD among persons living in depressed urban areas or on Native American reservations is estimated at 23%. For victims of violent crimes, the estimated rate is 58%.

PTSD also appears to be more common in seniors than in younger people. Thirteen percent of the senior population reports they are affected by PTSD in comparison to 7–10% of the entire population. Reports of **elder abuse** crimes have gone up by 200% since 1986. Also, the incidence of PTSD is known to be higher in Holocaust survivors, war veterans, and **cancer** or heart surgery survivors, which accounts for a significant portion of older Americans. Of those seniors who are military veterans, there is an increasing number who are isolated and/or in poor health as a result of PTSD.

Children are also susceptible to PTSD and their risk is increased exponentially as their exposure to the event increases. Children experiencing abuse, the **death** of a parent, or those located in a community suffering a traumatic event can develop PTSD. Two years after the Oklahoma City bombing of 1995, 16% of children within a 100-mile radius of Oklahoma City with no direct exposure to the bombing had increased symptoms of PTSD. Weak parental response to the event, having a parent suffering from PTSD symptoms, and intensified exposure to the event via the media all increase the possibility of a child's developing PTSD symptoms. In addition, a developmentally inappropriate sexual experience for a child may be considered a traumatic event, even though it may not have actually involved violence or physical injury.

MILITARY VETERANS. Studies conducted between 2004 and 2006 with veteran participants from Operation Iraqi Freedom and Operation Enduring Freedom (Afghanistan) found a strong correlation between duration of combat exposure and PTSD. Veterans of combat in Iraq reported a higher rate of PTSD than those deployed to Afghanistan because of longer exposure to warfare.

Information about PTSD in veterans of the Vietnam era is derived from the National Vietnam Veterans Readjustment Survey (NVVRS), conducted between 1986 and 1988. The estimated lifetime prevalence of PTSD among American veterans of this war is 30.9% for men and 26.9% for women. An additional 22.5% of the men and 21.2% of the women have been diagnosed with partial PTSD at some point in their lives. The lifetime prevalence of PTSD among veterans of World War II and the Korean War is estimated at 20%.

CROSS-CULTURAL ISSUES. Further research needs to be done on the effects of ethnicity and culture on post-traumatic symptoms. As of the early 2000s, most PTSD research has been done by Western clinicians working with patients from a similar background. Researchers do not yet know whether persons from non-Western societies have the same psychological reactions to specific traumas or whether they develop the same symptom patterns.

Causes and symptoms

The causes of PTSD are not completely understood. One major question that has not been answered as of 2009 is why some people involved in a major disaster develop PTSD and other survivors of the same event do not. For example, a survey of 988 adults living close to the World Trade Center conducted in November 2001 found that only 7 percent had been diagnosed with PTSD following the events of September 11; the other 93 percent were anxious and upset, but they did not develop PTSD. Research into this question is ongoing.

Causes

When PTSD was first suggested as a diagnostic category for *DSM-III* in 1980, it was controversial precisely because of the central role of outside stressors as causes of the disorder. Psychiatry has generally emphasized the internal abnormalities of individuals as the source of mental disorders; prior to the 1970s, war veterans, rape victims, and other trauma survivors were often blamed for their symptoms and regarded as cowards, moral weaklings, or masochists. The high rate of psychiatric casualties among Vietnam veterans, however, led to studies conducted by the Veterans Administration. These studies helped to establish PTSD as a legitimate diagnostic entity with a complex set of causes.

BIOCHEMICAL/PHYSIOLOGICAL CAUSES. Present neurobiological research indicates that traumatic events cause lasting changes in the human nervous system, including abnormal levels of secretion of stress hormones. In addition, in PTSD patients, researchers have found changes in the amygdala and the hippocampus—the parts of the brain that form links between fear and memory. Experiments with ketamine, a drug that inactivates one of the neurotransmitters in the central nervous system, suggest that trauma works in a similar way to damage associative pathways in the brain. **Positron emission tomography (PET)** scans of PTSD patients suggest that trauma affects the parts of the brain that govern speech and language.

SOCIOCULTURAL CAUSES. Studies of specific populations of PTSD patients (combat veterans, survivors of rape or genocide, former political hostages or prisoners, etc.) have shed light on the social and cultural causes of PTSD. In general, societies that are highly authoritarian, glorify violence, or sexualize violence have high rates of PTSD even among civilians.

OCCUPATIONAL FACTORS. Persons whose work exposes them to traumatic events or who treat trauma survivors may develop secondary PTSD (also known as compassion **fatigue** or burnout). These occupations include specialists in emergency medicine, police officers, firefighters, search-and-rescue personnel, psychotherapists, disaster investigators, etc. The degree of risk for PTSD is related to three factors: the amount and intensity of exposure to the suffering of trauma victims, the worker's degree of empathy and sensitivity, and unresolved issues from the worker's personal history.

PERSONAL VARIABLES. Although the most important causal factor in PTSD is the traumatic event itself, individuals differ in the intensity of their cognitive and emotional responses to trauma; some persons appear to be more vulnerable than others. In some cases, this greater vulnerability is related to temperament or natural disposition, with shy or introverted people being at greater risk. In other cases, the person's vulnerability results from chronic illness, a physical disability, or previous traumatization—particularly abuse in childhood. As of 2009, researchers have not found any correlation between race or ethnicity and biological vulnerability to PTSD.

Symptoms

DSM-IV-TR specifies six diagnostic criteria for PTSD:

- Traumatic stressor: The patient has been exposed to a catastrophic event involving actual or threatened death or injury, or a threat to the physical integrity of the self or others. During exposure to the trauma, the person's emotional response was marked by intense fear, feelings of helplessness, or horror. In general, stressors caused intentionally by human beings (genocide, rape, torture, abuse, etc.) are experienced as more traumatic than accidents, natural disasters, or "acts of God."
- Intrusive symptoms: The patient experiences flashbacks, traumatic daydreams, or nightmares, in which he or she relives the trauma as if it were recurring in the present. Intrusive symptoms result from an abnormal process of memory formation. Traumatic memories have two distinctive characteristics: 1) they can be triggered by stimuli that remind the patient of

- the traumatic event; 2) they have a “frozen” or wordless quality, consisting of images and sensations rather than verbal descriptions.
- **Avoidant symptoms:** The patient attempts to reduce the possibility of exposure to anything that might trigger memories of the trauma, and to minimize his or her reactions to such memories. This cluster of symptoms includes feeling disconnected from other people, psychic numbing, and avoidance of places, persons, or things associated with the trauma. Patients with PTSD are at increased risk of substance abuse as a form of self-medication to numb painful memories.
 - **Hyperarousal:** Hyperarousal is a condition in which the patient’s nervous system is always on “red alert” for the return of danger. This symptom cluster includes hypervigilance, insomnia, difficulty concentrating, general irritability, and an extreme startle response. Some clinicians think that this abnormally intense startle response may be the most characteristic symptom of PTSD.
 - **Duration of symptoms:** The symptoms must persist for at least one month.
 - **Significance:** The patient suffers from significant social, interpersonal, or work-related problems as a result of the PTSD symptoms. A common social symptom of PTSD is a feeling of disconnection from other people (including loved ones), from the larger society, and from spiritual, religious, or other significant sources of meaning.

Diagnosis

The diagnosis of PTSD is based on the patient’s history, including the timing of the traumatic event and the duration of the patient’s symptoms.

Examination

Consultation with a mental health professional for diagnosis and a plan of treatment is always advised. Many of the responses to trauma, such as **shock**, terror, irritability, blame, guilt, grief, sadness, emotional numbing, and feelings of helplessness, are natural reactions. For most people, resilience is an overriding factor and trauma effects diminish within six to sixteen months. It is when these responses continue or become debilitating that PTSD is often diagnosed.

As outlined in DSM-IV, the exposure to a traumatic stressor means that an individual experienced, witnessed or was confronted by an event or events involving death or threat of death, serious injury or the threat of bodily harm to oneself or others. The individual’s response must involve intense fear, helplessness, or horror. A two-pronged approach to evaluation is

KEY TERMS

Benzodiazepines—A class of drugs that have a hypnotic and sedative action, used mainly as tranquilizers to control symptoms of anxiety.

Cognitive-behavioral therapy—A type of psychotherapy used to treat anxiety disorders (including PTSD) that emphasizes behavioral change as well as alteration of negative thought patterns.

Cortisol—A hormone produced by the adrenal glands near the kidneys in response to stress.

Dissociation—The splitting off of certain mental processes from conscious awareness. Many PTSD patients have dissociative symptoms.

Flashback—A temporary reliving of a traumatic event.

Hyperarousal—A state of increased emotional tension and anxiety, often including jitteriness and being easily startled.

Hypervigilance—A condition of abnormally intense watchfulness or wariness. Hypervigilance is one of the most common symptoms of PTSD.

Prevalence—The percentage of a population that is affected by a specific disease at a given time.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, raising the levels of serotonin. SSRIs include Prozac, Zoloft, and Paxil.

Trauma—A severe injury or shock to a person’s body or mind.

considered the best way to make a valid diagnosis because it can gauge under-reporting or over-reporting of symptoms. The two primary forms are structured interviews and self-report questionnaires. Spouses, partners and other family members may also be interviewed. Because the evaluation may involve subtle reminders of the trauma in order to gauge a patient’s reactions, individuals should ask for a full description of the evaluation process beforehand. Asking what results can be expected from the evaluation is also advised.

A number of structured interview forms have been devised to facilitate the diagnosis of post traumatic stress disorder:

- The Clinician Administered PTSD Scale (CAPS) developed by the National Center for PTSD
- The Structured Clinical Interview for DSM (SCID)

- Anxiety Disorders Interview Schedule-Revised (ADIS)
- PTSD-Interview
- Structured Interview for PTSD (SI-PTSD)
- PTSD Symptom Scale Interview (PSS-I)

Self-reporting checklists provide scores to represent the level of stress experienced. Some of the most commonly used checklists are:

- The PTSD Checklist (PCL), which has one list for civilians and one for military personnel and veterans
- Impact of Event Scale-Revised (IES-R)
- Keane PTSD Scale of the MMPI-2
- The Mississippi Scale for Combat Related PTSD and the Mississippi Scale for Civilians
- The Post Traumatic Diagnostic Scale (PDS)
- The Penn Inventory for Post-Traumatic Stress
- Los Angeles Symptom Checklist (LASC)

Tests

There are no laboratory or imaging tests that can detect PTSD, although the doctor may order imaging studies of the brain to rule out head injuries or other physical causes of the patient's symptoms.

Treatment

Traditional

Treatment for PTSD usually involves a combination of medications and **psychotherapy**. If the patient has started to abuse alcohol or drugs, they must be treated for the substance abuse before being treated for PTSD. If the patient is diagnosed with coexisting depression, treatment should focus on the PTSD because its course, biology, and treatment response are different from those associated with major depression. Patients with the disorder are usually treated as outpatients; they are not hospitalized unless they are threatening to commit **suicide** or harm other people.

Mainstream forms of psychotherapy used to treat patients who have already developed PTSD include:

- Cognitive-behavioral therapy. There are two treatment approaches to PTSD included under this heading: exposure therapy, which seeks to desensitize the patient to reminders of the trauma; and anxiety management training, which teaches the patient strategies for reducing anxiety. These strategies may include relaxation training, biofeedback, social skills training, distraction techniques, or cognitive restructuring.

- Psychodynamic psychotherapy. This approach helps the patient recover a sense of self and learn new coping strategies and ways to deal with intense emotions related to the trauma. Typically, it consists of three phases: 1) establishing a sense of safety for the patient; 2) exploring the trauma itself in depth; 3) helping the patient re-establish connections with family, friends, the wider society, and other sources of meaning.
- Discussion groups or peer-counseling groups. These groups are usually formed for survivors of specific traumas, such as combat, rape/incest, and natural or transportation disasters. They help patients to recognize that other survivors of the shared experience have had the same emotions and reacted to the trauma in similar ways. They appear to be especially beneficial for patients with guilt issues about their behavior during the trauma (e.g., submitting to rape to save one's life, or surviving the event when others did not).
- Family therapy. This form of treatment is recommended for PTSD patients whose family life has been affected by the PTSD symptoms.

Drugs

In general, medications are used most often in patients with severe PTSD to treat the intrusive symptoms of the disorder as well as feelings of anxiety and depression. These drugs are usually given as one part of a treatment plan that includes psychotherapy or **group therapy**. As of 2009, there is no single medication that appears to be a "magic bullet" for PTSD. The **selective serotonin reuptake inhibitors** (SSRIs) appear to help the core symptoms when given in higher doses for five to eight weeks, while the tricyclic antidepressants (TCAs) or the **monoamine oxidase inhibitors** (MAOIs) are most useful in treating anxiety and depression.

Sleep problems can be lessened with brief treatment with an anti-anxiety drug, such as a benzodiazepine like alprazolam (Xanax), but long-term usage can lead to disturbing side effects, such as increased anger, drug tolerance, dependency, and abuse. **Benzodiazepines** are also not given to PTSD patients diagnosed with coexisting drug or alcohol abuse.

Alternative

Relaxation training, which is sometimes called anxiety management training, includes breathing exercises and similar techniques intended to help the patient prevent hyperventilation and relieve the muscle tension associated with the fight-or-flight

reaction of anxiety. **Yoga**, aikido, t'ai chi, and dance therapy help patients work with the physical as well as the emotional tensions that either promote anxiety or are created by the anxiety.

Other alternative or complementary therapies are based on physiological and/or energetic understanding of how the trauma is imprinted in the body. These therapies affect a release of stored emotions and resolution of them by working with the body rather than merely talking through the experience. One example of such a therapy is Somatic Experiencing (SE), developed by Dr. Peter Levine. SE is a short-term, biological, body-oriented approach to PTSD or other trauma. This approach heals by emphasizing physiological and emotional responses, without re-traumatizing the person, without placing the person on medication, and without the long hours of conventional therapy.

When used in conjunction with therapies that address the underlying cause of PTSD, such relaxation therapies as **hydrotherapy**, **massage therapy**, and **aromatherapy** are useful to some patients in easing PTSD symptoms. Essential oils of lavender, chamomile, neroli, sweet marjoram, and ylang-ylang are commonly recommended by aromatherapists for stress relief and anxiety reduction.

Some patients benefit from spiritual or religious counseling. Because traumatic experiences often affect patients' spiritual views and beliefs, counseling with a trusted religious or spiritual advisor may be part of a treatment plan. A growing number of pastoral counselors in the major Christian and Jewish bodies in North America have advanced credentials in trauma therapy. Native Americans are often helped to recover from PTSD by participating in traditional tribal rituals for cleansing memories of war and other traumatic events. These rituals may include sweat lodges, prayers and chants, or consultation with a shaman or tribal healer.

Several controversial methods of treatment for PTSD have been introduced since the mid-1980s. Some have been developed by mainstream medical researchers while others are derived from various forms of alternative medicine. These methods are controversial because they do not offer any scientifically validated explanations for their effectiveness. They include:

- Eye Movement Desensitization and Reprocessing (EMDR). This is a technique in which the patient reimagines the trauma while focusing visually on movements of the therapist's finger. It is claimed that the movements of the patient's eyes reprogram the brain and allow emotional healing.

• Tapas Acupressure Technique (TAT). TAT was developed in 1993 by a licensed acupuncturist named Tapas Fleming. It is derived from traditional Chinese medicine (TCM), and its practitioners maintain that a large number of acupuncture meridians enter the brain at certain points on the face, especially around the eyes. Pressure on these points is thought to release traumatic stress.

- Thought Field Therapy. This therapy combines the acupuncture meridians of TCM with analysis of the patient's voice over the telephone. The therapist then provides an individualized treatment for the patient.
- Traumatic Incident Reduction. This is a technique in which the patient treats the trauma like a videotape and "runs through" it repeatedly with the therapist until all negative emotions have been discharged.
- Emotional Freedom Techniques (EFT). EFT is similar to TAT in that it uses the body's acupuncture meridians, but it emphasizes the body's entire "energy field" rather than just the face.
- Counting Technique. Developed by a physician, this treatment consists of a preparation phase, a counting phase in which the therapist counts from 1 to 100 while the patient reimagines the trauma, and a review phase. Like Traumatic Incident Reduction, it is intended to reduce the patient's hyperarousal.

Prognosis

The prognosis of PTSD is difficult to determine because patients' personalities and the experiences they undergo vary widely. A majority of patients get better, including some who do not receive treatment. One study reported that the average length of PTSD symptoms in patients who get treatment is 32 months, compared to 64 months in patients who are not treated.

Factors that improve a patient's chances for full recovery include prompt treatment, early and ongoing support from family and friends, a high level of functioning before the frightening event, and an absence of alcohol or substance abuse.

About 30 percent of people with PTSD never recover completely, however. A few commit suicide because their symptoms get worse rather than improving.

Health care team roles

It is essential for all treatment team members to know their roles and execute them properly throughout the treatment and recovery phases of this disorder. Depending on whether outpatient or inpatient treatment is being provided, the team leaders may include

psychiatrists, psychologists, nursing staff, behavior specialists, physical therapists, and other medical/behavioral staff. In some cases it may be appropriate to include the patient's religious or spiritual advisor as a member of the team.

Prevention

PTSD is impossible to prevent completely because natural disasters and human acts of violence will continue to occur. In addition, it is not possible to tell beforehand how any given individual will react to a specific type of trauma. Prompt treatment after a traumatic event may lower the survivor's risk of developing severe symptoms.

Resources

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- National Center for Posttraumatic Stress Disorder. *Hope for Recovery: Understanding PTSD*. [10-minute video] http://www.ncptsd.va.gov/ncmain/ncdocs/videos/env_hoperecovery_gpv.html
- National Center for Posttraumatic Stress Disorder Fact Sheet. *What Is PTSD?* http://www.ncptsd.va.gov/ncmain/ncdocs/fact_shts/fs_what_is_ptsd.html
- National Institute of Mental Health (NIMH). *Helping Children Cope with Violence and Disasters: What Parents Can Do*. <http://www.nimh.nih.gov/health/publications/helping-children-and-adolescents-cope-with-violence-and-disasters-what-parents-can-do/index.shtml>.

ORGANIZATIONS

- American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209-3901, 703-907-7300, apa@psych.org, <http://www.psych.org>.
- Anxiety Disorders Association of America (ADAA), 8730 Georgia Ave., Suite 600, Silver Spring, MD, 20910, 240-485-1001, 240-485-1035, information@adaa.org, <http://www.adaa.org>.
- International Society for Traumatic Stress Studies (ISTSS), 111 Deer Lake Road, Suite 100, DeerfieldIL, United States, 60015, 847-480-9028, 847-480-9282, istss@istss.org, <http://www.istss.org>.
- National Alliance on Mental Illness (NAMI), 2107 Wilson Blvd., Suite 300, Arlington, VA, 22201-3042, 703-524-7600, Hotline: 800-950-NAMI (6264), 703-524-9094, <http://www.nami.org/Hometemplate.cfm>.
- National Center for Posttraumatic Stress Disorder (NCPTSD), Information line: 802-296-6300, ncptsd@va.gov, <http://www.ncptsd.va.gov/ncmain/index.jsp>.
- National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD, 20892-9663, 301-443-4513, 866-615-6464, 301-443-4279, nimhinfo@nih.gov, <http://www.nimh.nih.gov/index.shtml>.

Rebecca J. Frey, PhD

Postmenopausal bleeding

Definition

Postmenopausal bleeding is bleeding from the reproductive system that occurs six months or more after menstrual periods have stopped due to menopause.

Description

Menopause, the end of ovulation and menstrual periods, naturally occurs for most women age 40–55 years. The process of ending ovulation and menstruation is gradual, spanning one to two years.

Postmenopausal bleeding is bleeding that occurs after menopause has been established for at least six months. It is different from infrequent, irregular periods (**oligomenorrhea**) that occur around the time of menopause.

Many women experience some postmenopausal bleeding. However, postmenopausal bleeding is not normal. Because it can be a symptom of a serious medical condition, any episodes of postmenopausal bleeding should be brought to the attention of a woman's doctor.

Women taking estrogen (called **hormone replacement therapy** or HRT) are more likely to experience postmenopausal bleeding. So are obese women, because fat cells transform male hormones (androgens) secreted by the adrenal gland into estrogen.

Causes and symptoms

Postmenopausal bleeding can originate in different parts of the reproductive system. Bleeding from the vagina may occur because when estrogen secretion stops, the vagina dries out and can diminish (atrophy). This is the most common cause of bleeding from the lower reproductive tract.

Lesions and cracks on the vulva may also bleed. Sometimes bleeding occurs after intercourse. Bleeding can occur with or without an associated infection.

Bleeding from the upper reproductive system can be caused by:

- hormone replacements
- endometrial cancer
- endometrial polyps
- cervical cancer
- cervical lesions
- uterine tumors
- ovarian cancer
- estrogen-secreting tumors in other parts of the body

The most common cause of postmenopausal bleeding is HRT. The estrogen in the replacement therapy eases the symptoms of menopause (like hot flashes), and decreases the risk of **osteoporosis**. Sometimes this supplemental estrogen stimulates the uterine lining to grow. When the lining is shed, postmenopausal bleeding occurs. Most women on HRT usually

take the hormone progesterone with the estrogen, and may have monthly withdrawal bleeding. This is a normal side effect.

About 5–10% of postmenopausal bleeding is due to **endometrial cancer** or its precursors. Uterine hyperplasia, the abnormal growth of uterine cells, can be a precursor to **cancer**.

Diagnosis

Diagnosis of postmenopausal bleeding begins with the patient. The doctor will ask for a detailed history of how long postmenopausal bleeding has occurred. A woman can assist the doctor by keeping a record of the time, frequency, length, and quantity of bleeding. She should also tell the doctor about any medications she is taking, especially any estrogens or **steroids**.

After taking the woman's history, the doctor does a pelvic examination and **Pap test**. The doctor will examine the vulva and vagina for signs of atrophy, and will feel for any sign of uterine polyps. Depending on the results of this examination, the doctor may want to do more extensive testing.

Invasive diagnostic procedures

Endometrial biopsy allows the doctor to sample small areas of the uterine lining, while cervical biopsy allows the cervix to be sampled. Tissues are then examined for any abnormalities. This is a simple office procedure.

Dilatation and curettage (D & C) is often necessary for definitive diagnosis. This is done under either general or **local anesthesia**. After examining the tissues collected by an endometrial biopsy or D & C, the doctor may order additional tests to determine if an estrogen-secreting tumor is present on the ovaries or in another part of the body.

Non-invasive diagnostic procedures

With concerns about the rising cost of health care, vaginal probe ultrasound is increasingly being used more than endometrial biopsy to evaluate women with postmenopausal bleeding. Vaginal ultrasound measures the thickness of the endometrium. When the endometrial stripe is less than 0.2 in (5 mm) thick, the chance of cancer is less than 1%. The disadvantage of vaginal ultrasound is that it often does not show polyps and fibroids in the uterus.

A refinement of vaginal probe ultrasound is saline infusion sonography (SIS). A salt water (saline) solution is injected into the uterus with a small tube (catheter) before the vaginal probe is inserted. The presence

KEY TERMS

Dilatation and curettage (D & C)—A procedure performed under anesthesia during which the cervix is opened more (or dilated) and tissue lining the uterus is scraped out with a metal, spoon-shaped instrument or a suction tube. The procedure can be used to diagnose a problem or to remove growths (polyps).

Endometrial biopsy—The removal of uterine tissue samples either by suction or scraping; the cervix is not dilated. The procedure has a lower rate of diagnostic accuracy than D & C, but can be done as an office procedure under local anesthesia.

Endometrium—The tissue lining the inside of the uterus.

Fibroid tumors—Non-cancerous (benign) growths in the uterus. These growths occur in 30–40% of women over age 40, and do not need to be removed unless they are causing symptoms that interfere with a woman's normal activities.

Osteoporosis—The excessive loss of calcium from the bones, causing the bones to become fragile and break easily. Postmenopausal women are especially vulnerable to this condition because estrogen, a hormone that protects bones against calcium loss, decreases drastically after menopause.

of liquid in the uterus helps make any structural abnormalities more distinct. These two non-invasive procedures cause less discomfort than endometrial biopsies and D & Cs, but D & C still remains the definitive test for diagnosing uterine cancer.

Treatment

It is common for women just beginning HRT to experience some bleeding. Most women who are on HRT also take progesterone with the estrogen and may have monthly withdrawal bleeding. Again, this is a normal side effect that usually does not require treatment.

Postmenopausal bleeding due to bleeding of the vagina or vulva can be treated with local application of estrogen or HRT.

When diagnosis indicates cancer, some form of surgery is required. The uterus, cervix, ovaries, and fallopian tubes may all be removed depending on the type and location of the cancer. If the problem is estrogen- or androgen-producing tumors elsewhere in the body, these must also be surgically removed.

Postmenopausal bleeding that is not due to cancer and cannot be controlled by any other treatment usually requires a **hysterectomy**.

Prognosis

Response to treatment for postmenopausal bleeding is highly individual and is not easy to predict. The outcome depends largely on the reason for the bleeding. Many women are successfully treated with hormones. As a last resort, hysterectomy removes the source of the problem by removing the uterus. However, this operation is not without risk and the possibility of complications. The prognosis for women who have various kinds of reproductive cancer varies with the type of cancer and the stage at which the cancer is diagnosed.

Prevention

Postmenopausal bleeding is not a preventable disorder. However, maintaining a healthy weight will decrease the chances of it occurring.

ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329, 800 227-2345, <http://www.cancer.org>.
National Cancer Institute (National Institutes of Health), NCI Office of Communications and Education, 6116 Executive Blvd. Suite 300, Bethesda, MD, 20892-8322, 800 4-CANCER (422-6237), cancergovstaff@mail.nih.gov, <http://www.cancer.gov>.

Tish Davidson, A.M.

Postpartum blues see **Postpartum depression**

Postpartum depression

Definition

Postpartum depression is a mood disorder that begins after **childbirth** and usually lasts at least six weeks.

Description

Postpartum depression, or PPD, affects approximately 15% of all childbearing women. The onset of postpartum depression tends to be gradual and may persist for many months or develop into a second bout following a subsequent **pregnancy**. Mild to moderate cases are sometimes unrecognized by women themselves. Many women feel ashamed and may conceal their difficulties. This is a serious problem that disrupts women's lives and can have effects on the baby,

KEY TERMS

Hyperemesis—Severe vomiting during pregnancy. Hyperemesis appears to increase a woman's risk of postpartum depression.

Postpartum—Following childbirth.

other children, partners, and other relationships. Levels of depression for fathers can also increase significantly.

Postpartum depression is often divided into two types: early onset and late onset. Early-onset PPD most often seems like the “blues,” a mild brief experience during the first days or weeks after birth. During the first week after the birth, up to 80% of mothers experience the “baby blues.” This period is usually a time of extra sensitivity; symptoms include tearfulness, irritability, **anxiety**, and mood changes, which tend to peak between three to five days after childbirth. The symptoms normally disappear within two weeks without requiring specific treatment apart from understanding, support, skills, and practice. In short, some depression, **fatigue**, and anxiety may fall within the “normal” range of reactions to giving birth.

Late-onset PPD appears several weeks after birth. It involves slowly growing feelings of sadness, depression, lack of energy, chronic fatigue, inability to sleep, change in appetite, significant weight loss or gain, and difficulty caring for the baby.

Causes and symptoms

The cause of postpartum depression has been extensively studied. Alterations of hormone levels of prolactin, progesterone, estrogen, and cortisol are not significantly different from those of patients who do not suffer from postpartum depression. However, some research indicates a change in a brain chemical that controls the release of cortisol.

Research seems to indicate that postpartum depression is unlikely to occur in a patient with an otherwise psychologically uncomplicated pregnancy and past history. There is no association of postpartum depression with marital status, social class, or the number of live children born to the mother. However, there seems to be an increased chance to develop this disorder after pregnancy loss.

Certain characteristics have been associated with increased risk of developing postpartum depression. These risk factors include:

- medical indigence—being in need of health care and not being able to receive it, possibly due to lack of medical insurance
- being younger than 20 years old at time of delivery
- being unmarried
- having been separated from one or both parents in childhood or adolescence
- receiving poor parental support and attention in childhood
- having had limited parental support in adulthood
- poor relationship with husband or boyfriend
- economic problem with housing or income
- dissatisfaction with amount of education
- low self-esteem
- past or current emotional problem(s)
- family history of depression

Experts cannot always say what causes postpartum depression. Most likely, it is caused by a combination of factors that vary from person to person. Some researchers think that women are vulnerable to depression at all major turning points in their reproductive cycle, childbirth being only one of these markers. Factors before the baby's birth that are associated with a higher risk of PPD include severe **vomiting** (hyperemesis), **premature labor** contractions, and psychiatric disorders in the mother. In addition, new mothers commonly experience some degree of depression during the first weeks after birth. Pregnancy and birth are accompanied by sudden hormonal changes that affect emotions. Additionally, the 24-hour responsibility for a newborn infant represents a major psychological and lifestyle adjustment for most mothers, even after the first child. These physical and emotional stresses are usually accompanied by inadequate rest until the baby's routine stabilizes, so fatigue and depression are not unusual.

In addition to hormonal changes and disrupted sleep, certain cultural expectations appear to place women from those cultures at increased risk of postpartum depression. For example, women who bear daughters in societies with a strong preference for sons (such as Communist China) are at increased risk of postpartum depression. In other cultures, a strained relationship with the husband's family is a risk factor. In Western countries, domestic violence is associated with a higher rate of PPD.

Experiences of PPD vary considerably but usually include several symptoms.

Feelings:

- persistent low mood
- inadequacy, failure, hopelessness, helplessness

- exhaustion, emptiness, sadness, tearfulness
- guilt, shame, worthlessness
- confusion, anxiety, and panic
- fear for the baby and of the baby
- fear of being alone or going out

Behaviors:

- lack of interest or pleasure in usual activities
- insomnia or excessive sleep, nightmares
- not eating or overeating
- decreased energy and motivation
- withdrawal from social contact
- poor self-care
- inability to cope with routine tasks

Thoughts:

- inability to think clearly and make decisions
- lack of concentration and poor memory
- running away from everything
- fear of being rejected by the partner
- worry about harm or death to partner or baby
- ideas about suicide

Some symptoms may not indicate a severe problem. However, persistent low mood or loss of interest or pleasure in activities, along with four other symptoms occurring together for a period of at least two weeks, indicate clinical depression and require adequate treatment.

There are several important risk factors for postpartum depression, including the following:

- stress
- lack of sleep
- poor nutrition
- lack of support from one's partner, family, or friends
- family history of depression
- labor/delivery complications for mother or baby
- premature or postmature delivery
- problems with the baby's health
- separation of mother and baby
- a difficult baby (temperament, feeding, sleeping problems)
- pre-existing neurosis or psychosis

Physical and emotional **stress** during delivery in conjunction with great demands for infant care may cause the patient to neglect other family members, increasing the woman's feelings of self-worthlessness, isolation, and being trapped. Patients may also feel as if they are inadequate mothers, causing them guilt and embarrassment.

Demographics

There is a 20% to 30% risk of postpartum depression for women who had a previous depressive episode that was not associated with pregnancy. Additionally, there is an increased risk of recurrence in subsequent pregnancies since more than half of patients will have more than one episode.

Diagnosis

Diagnosis of postpartum depression entails a clinical interview with the patient to assess symptoms. A doctor or other professional healthcare provider may ask the mother about thoughts and feelings, and take a detailed personal history. Clinical assessment may be conducted by a psychologist or psychiatrist, who can determine the risk factors and diagnose the condition. A comprehensive psychological assessment interview could reveal a previous depressive cycle or a family history of depression—important risk factors. The most widely used standard for diagnosis is the Edinburgh Postnatal Depression Scale (EPDS). This is a simple and short 10-question scale. A score of 12 or greater on the EPDS is considered high risk for postpartum depression.

Treatment

Several treatment options exist, including medication, **psychotherapy**, counseling, and group treatment and support strategies. Treatment should begin as soon as the diagnosis is established. One effective treatment combines antidepressant medication and psychotherapy. These types of medication are often effective when used for three to four weeks. Any medication use must be carefully considered if the woman is **breastfeeding**, but with some medications, continuing breastfeeding is safe. There are many classes of antidepressant medications. Two of the most commonly prescribed for PPD are **selective serotonin reuptake inhibitors** (SSRIs) such as citalopram (Celexa), escitalopram (Lexapro), fluoxetine (Prozac), paroxetine (Paxil, Pexeva), and sertraline (Zoloft), and tricyclines, such as amitriptyline (Elavil), desipramine (Norpramin), imipramine (Tofranil), and nortriptyline (Aventyl, Pamelor). Nevertheless, medication alone is never sufficient and should always be accompanied by counseling or other support services. Also, many women with postpartum depression feel isolated. It is important for these women to know that they are not alone in their feelings. There are various postpartum depression support groups available in local communities, often sponsored by non-profit

organizations or hospitals. For women who have thoughts of **suicide**, it is imperative to seek help immediately.

When medications are combined with psychological therapy, the rates for successful treatment are increased. Interpersonal therapy and **cognitive-behavioral therapy** have been found to be effective.

Adjunct therapies such as **Acupuncture**, **traditional Chinese medicine**, **yoga**, **meditation**, and herbs may be considered to help the mother suffering from postpartum depression.

Some strategies that may help new mothers cope with the stress of becoming a parent include:

- Valuing her role as a mother and trusting her own judgment.
- Making each day as simple as possible.
- Avoiding extra pressures or unnecessary tasks.
- Trying to involve her partner more in the care of the baby from the beginning.
- Discussing with her partner how both can share the household chores and responsibilities.
- Scheduling frequent outings, such as walks and short visits with friends.
- Sharing her feelings with her partner or a friend who is a good listener.
- Talking with other mothers to help keep problems in perspective.
- Trying to sleep or rest when the baby is sleeping.
- Taking care of her health and well being.

Exercise, including yoga, can help enhance a new mother's emotional wellbeing. New mothers should also try to cultivate good sleeping habits and learn to rest when they feel physically or emotionally tired. It is important for a woman to learn to recognize her own warning signs of fatigue and respond to them by taking a break.

Expected results

When a woman has supportive friends and family, mild postpartum depression usually disappears quickly. If depression becomes severe and a mother cannot care for herself and the baby, hospitalization may be necessary. Medication, counseling, and support from others usually resolve even severe depression in three to six months. The prognosis for postpartum depression is better if it is detected early during its clinical course and a combination

of SSRIs and psychotherapy is available and initiated.

Prevention

Mothers should be advised prior to hospital discharge that if the "maternity blues" last longer than two weeks or pose tough difficulties with family interactions, they should call the hospital where their baby was delivered and pursue a referral for a psychological evaluation. Education concerning risk factors and reduction of these is important. Prophylactic (preventive) use of SSRIs is indicated two to three weeks before delivery to prevent the disorder in a patient with a past history of depression, since recurrence rates are high if the mother had a previous depressive episode.

Resources

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ORGANIZATIONS

- Kristin Brooks Hope Center, 615 Seventh St. NE, Washington, DC, 20002, (202) 536-3200, (800) 442-4673, <http://www.hopeline.com>.
- National Institute of Mental Health, 6001 Executive Blvd., Room 8184, MSC 9663, Bethesda, MD, 20892, (866) 615-6464, <http://www.nimh.nih.gov>.
- Online PPD Support Group, P.O. Box 611, Issaquah, WA, 98027, <http://www.ppdsupportpage.com>.
- Postpartum Support International., PO Box 60931, Santa Barbara, CA, 93160, (805) 967-7636, (800) 944-4773, <http://www.postpartum.net>.

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Postpartum psychosis see **Postpartum depression**

Postpolio syndrome

Definition

Postpolio syndrome (PPS) is a condition that strikes survivors of the disease **polio**. PPS occurs about 20–30 years after the original bout with polio, and causes slow but progressive weakening of muscles.

Description

Polio is a disease caused by the poliovirus. It most commonly infects younger children, although it can

also infect older children and adults. About 90% of people infected by poliovirus develop only a mild case or no illness at all. However, infected people can continue to spread the virus to others. In its most severe form polio causes **paralysis** of the muscles of the legs, arms, and respiratory system.

About 1% of all people infected with poliovirus develop the actual disease known as polio. In these cases, the virus (which enters the person's body through the mouth) multiplies rapidly within the intestine. The viruses then invade the nearby lymphatic system. Eventually, poliovirus enters the bloodstream, which allows it to gain access to the central nervous system or CNS (the brain and spinal cord). The virus may actually infect a nerve elsewhere in the body, and then spread along that nerve to enter the brain.

The major illness associated with poliovirus often follows a mild illness, which has symptoms of **fever**, **nausea**, and **vomiting**. However, after a symptom-free interval of several days, the patient who is on the way to a major illness develops new symptoms such as **headache** and back and neck **pain**. These symptoms are due to invasion of the nervous system. The motor nerves (those nerves responsible for movement of the muscles) become inflamed, injured, and destroyed. The muscles, therefore, no longer receive any messages from the brain or spinal cord. The muscles become weak, floppy, and then totally paralyzed (unable to move). All muscle tone is lost in the affected limb, and the muscle begins to decrease in size (atrophy). The affected muscles are often only on one side (asymmetric paralysis) of the body. Sensation (the person's ability to feel) is not affected in these paralyzed limbs.

The maximum state of paralysis is usually reached within just a few days. The remaining, unaffected nerves then begin the process of attempting to grow branches to compensate (make up for) the destroyed nerves. This process continues for about six months. Whatever function has not been regained in this amount of time will usually be permanently lost.

Causes and symptoms

PPS occurs in about 25% of patients, several decades after their original infection with polio. However, long-term follow-up indicates that two thirds of polio survivors may experience new weakness. Several theories exist as to the cause of this syndrome.

One such theory has looked at the way function is regained by polio survivors. Three mechanisms seem to be at work:

- injured nerves recuperate and begin functioning again
- muscles that still have working nerve connections grow in size and strength, in order to take over for other paralyzed muscles
- working nerves begin to send small branches out to muscles whose original nerves were destroyed by polio

As a person ages, injured nerves that were able to regain function may fail again, as may muscles that have been over-worked for years in order to compensate for other paralyzed muscles. Even the uninjured nerves that provided new nerve twigs to the muscles may begin to falter after years of relative over-activity. This theory, then, suggests that the body's ability to compensate for destroyed nerves may eventually begin to fail. The compensating nerves and muscles grow older, and because they've been working so much harder over the years, they wear out relatively sooner than would be expected of normal nerves and muscles. Some researchers look at this situation as a form of premature **aging**, brought on by overuse.

Other researchers note that normal aging includes the loss of a fair number of motor nerves. When a patient has already lost motor nerves through polio, normal loss of motor nerves through aging may cause the number of remaining working nerves to drop low enough to cause symptoms of weakness.

Other theories of PPS include the possibility that particles of the original polioviruses remain in the body. These particles may exert a negative effect, decades later, or they may cause the body's immune system to produce substances originally intended to fight the invading virus, but which may accidentally set off a variety of reactions within the body that actually serve to interfere with the normal functioning of the nerves and muscles.

Still other researchers are looking at the possibility that polio patients have important spinal cord changes which, over time, affect the nerves responsible for movement.

The symptoms of PPS include generalized **fatigue**, low energy, progressively increasing muscle weakness, shrinking muscle size (atrophy), involuntary twitching of the muscle fibers (fasciculations), painful muscles and joints, difficulties with breathing and swallowing, and sleep problems.

Survivors of polio may also develop arthritis of the spine, shoulders, or arms, related to the long-term use of crutches or overcompensation for weak leg muscles.

KEY TERMS

Asymmetric—Not occurring equally on both sides of the body.

Atrophy—Shrinking, growing smaller in size.

Flaccid—Weak, soft, floppy.

Paralysis—The inability to voluntarily move.

Diagnosis

Diagnosis is primarily through history. When a patient who has recovered from polio some decades previously begins to experience muscle weakness, PPS must be strongly suspected.

Treatment

Just as there are no treatments available to reverse the original damage of polio, there are also no treatments available to reverse the damaging effects of postpolio syndrome. Attempts can be made to relieve some of the symptoms, however.

Pain and inflammation of the muscles and joints can be treated with anti-inflammatory medications, application of hot packs, stretching exercises, and **physical therapy**. Exercises to maintain/increase flexibility are particularly important. However, an **exercise** regimen must be carefully designed, so as not to strain already fatigued muscles and nerves.

Some patients will require new types of braces to provide support for weakening muscles. Others will need to use wheelchairs or motorized scooters to maintain mobility.

Sleep problems and respiratory difficulties may be related to each other. If breathing is labored during sleep, the blood's oxygen content may drop low enough to interfere with the quality of sleep. This may require oxygen supplementation, or even the use of a machine to aid in breathing.

Prognosis

Prognosis for patients with postpolio syndrome is relatively good. It is a very slow, gradually progressing syndrome. Only about 20% of all patients with PPS will need to rely on new aids for mobility or breathing. It appears that the PPS symptoms reach their most severe about 30–34 years after original diagnosis of polio.

Prevention

There is no way to prevent PPS. However, paying attention to what types of exertion worsen symptoms may slow the progression of the syndrome.

ORGANIZATIONS

March of Dimes Birth Defects Foundation, 1275 Mamaroneck Ave., White Plains, NY, 10605, (914) 997-4488, <http://www.modimes.org>.

Polio Survivors Association, 12720 Lareina Ave., Downey, CA, 90242, (562) 862-4508, info@polioassociation.org, <http://polioassociation.org>.

Post-Polio Health International (PHI), 4207 Lindell Blvd., Suite 110, St. Louis, MO, 63108-2930, (314) 534-0475, (314) 534-5070, info@post-polio.org, <http://www.post-polio.org>.

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Postpoliomyelitis muscular atrophy see
Postpolio syndrome

Postpoliomyelitis syndrome see **Postpolio syndrome**

Poststreptococcal glomerulonephritis see
Acute poststreptococcal glomerulonephritis

Postural drainage see **Chest physical therapy**

Postural hypotension see **Orthostatic hypotension**

Postviral thrombocytopenia see **Idiopathic thrombocytopenic purpura**

Potassium hydroxide test see **KOH test**

Potassium imbalance see **Hyperkalemia; Hypokalemia**

PPD skin test see **Tuberculin skin test**

PWS. This uncontrollable appetite can lead to health problems and behavior disturbances.

Description

The first patients with features of PWS were described by Dr. Prader, Dr. Willi, and Dr. Lambert in 1956. Since that time, the complex genetic basis of PWS has begun to be understood. Initially, scientists found that individuals with PWS have a portion of genetic material deleted (erased) from chromosome 15. In order to have PWS, the genetic material must be deleted from the chromosome 15 received from one's father. If the deletion is on the chromosome 15 inherited from one's mother a different syndrome develops. This was an important discovery. It demonstrated for the first time that the genes inherited from one's mother can be expressed differently than the genes inherited from one's father.

Over time, scientists realized that some individuals with PWS do not have genetic material deleted from chromosome 15. Further studies found that these patients inherit both copies of chromosome 15 from their mother. This is not typical. Normally, an individual receives one chromosome 15 from their father and one chromosome 15 from their mother. When a person receives both chromosomes from the same parent it is called "uniparental disomy." When a person receives both chromosomes from his or her mother it is called "maternal uniparental disomy."

Scientists are still discovering other causes of PWS. A small number of patients with PWS have a change (mutation) in the genetic material on the chromosome 15 inherited from their father. This mutation prevents certain genes on chromosome 15 from working properly. PWS develops when these genes do not work normally.

Newborns with PWS generally have poor muscle tone, (hypotonia) and do not feed well. This can lead to poor weight gain and **failure to thrive**. Genitalia can be smaller than normal. Hands and feet are also typically smaller than normal. Some patients with PWS have unique facial characteristics. These unique facial features are typically subtle and detectable only by physicians.

As children with PWS age, development is typically slower than normal. Developmental milestones, such as crawling, walking and talking occur later than usual. Developmental delay continues into adulthood for approximately 50% of individuals with PWS. At about one to two years of age, children with PWS

Prader-Willi syndrome

Definition

Prader-Willi syndrome (PWS) is a genetic condition caused by the absence of chromosomal material from chromosome 15. The genetic basis of PWS is complex. Characteristics of the syndrome include developmental delay, poor muscle tone, short stature, small hands and feet, incomplete sexual development, and unique facial features. Insatiable appetite is a classic feature of

develop an uncontrollable, insatiable appetite. Left to their own devices, individuals with PWS will eat until they suffer from life-threatening **obesity**. The desire to eat can lead to significant behavior problems.

The symptoms and features of PWS require life long support and care. If food intake is strictly monitored and various therapies provided, individuals with PWS have a normal life expectancy.

PWS affects approximately 1 in 10,000 to 25,000 live births. It is the most common genetic cause of life-threatening obesity. It affects both males and females. PWS can be seen in all races and ethnic groups.

Causes and symptoms

In order to comprehend the various causes of PWS, the nature of chromosomes and genes must be well understood. Human beings have 46 chromosomes in the cells of their body. Chromosomes contain genes, which regulate the function and development of the body. An individual's chromosomes are inherited from his/her parents. Each parent normally gives a child 23 chromosomes. A child receives 23 chromosomes from the egg and 23 chromosomes from the sperm.

The 46 chromosomes in the human body are divided into pairs based on their physical characteristics. Each pair is assigned a number or a letter. When viewed under a microscope, chromosomes within the same pair appear identical because they contain the same genes.

Most chromosomes have a constriction near the center called the centromere. The centromere separates the chromosome into long and short arms. The short arm of a chromosome is called the "p arm," and the long arm is called the "q arm."

Chromosomes in the same pair contain the same genes. However, some genes work differently depending on if they were inherited from the egg or the sperm. Sometimes, genes are silenced when inherited from the mother. Other times, genes are silenced when inherited from the father. When genes in a certain region on a chromosome are silenced, they are said to be "imprinted." Imprinting is a normal process that does not typically cause disease. If normal imprinting is disrupted a genetic disease can develop.

Individuals have two complete copies of chromosome 15. One chromosome 15 is inherited from the mother, or "maternal" in origin. The other chromosome 15 is inherited from the father, or is "paternal" in origin.

Chromosome 15 contains many different genes. There are several genes found on the q arm of chromosome 15 that are imprinted. A gene called "SNPRN" is an example of one of these genes. It is normally imprinted, or silenced, if inherited from the mother. The imprinting of this group of maternal genes does not typically cause disease. The genes in this region should not be imprinted if paternal in origin. Normal development depends on these paternal genes being present and active. If these genes are deleted, not inherited, or incorrectly imprinted PWS develops.

Seventy percent of the cases of PWS are caused when a piece of material is deleted, or erased, from the paternal chromosome 15. This deletion occurs in a specific region on the q arm of chromosome 15. The piece of chromosomal material that is deleted contains genes that must be present for normal development. These paternal genes must be working normally, because the same genes on the chromosome 15 inherited from the mother are imprinted. When these paternal genes are missing, the brain and other parts of the body do not develop as expected. This is what causes the symptoms associated with PWS.

In 99% of the cases of PWS the deletion is sporadic. This means that it happens randomly and there is not an apparent cause. It does not run in the family. If a child has PWS due to a sporadic deletion in the paternal chromosome 15, the chance the parents could have another child with PWS is less than 1%. In fewer than 1% of the cases of PWS there is a chromosomal rearrangement in the family which causes the deletion. This chromosomal rearrangement is called a "translocation." If a parent has a translocation the risk of having a child with PWS is higher than 1%.

PWS can also develop if a child receives both chromosome 15s from his/her mother. This is seen in approximately 25% of the cases of PWS. Maternal uniparental disomy for chromosome 15 leads to PWS because the genes on chromosome 15 that should have been inherited from the father are missing, and the genes on both the chromosome 15s inherited from the mother are imprinted.

PWS caused by maternal uniparental is sporadic. This means that it occurs randomly and there is not an apparent cause. If a child has PWS due to maternal uniparental disomy the chance the parents could have another child with PWS is less than 1%.

Approximately 3–4% of patients with PWS have a change (mutation) in a gene located on the q arm of chromosome 15. This mutation leads to incorrect

imprinting. This mutation causes genes inherited from the father to be imprinted or silenced, which should not normally be imprinted. If a child has PWS due to a mutation that changes imprinting, the chance the parents could have another child with PWS is approximately 5%.

Infants with PWS have weak muscle tone (hypotonia). This hypotonia causes problems with sucking and eating. Infants with PWS may have problems gaining weight. Some infants with PWS are diagnosed with "failure to thrive" due to slow growth and development. During infancy, babies with PWS may also sleep more than normal and have problems controlling their temperature.

Some of the unique physical features associated with PWS can be seen during infancy. Genitalia that is smaller than normal is common. This may be more evident in males with PWS. Hands and feet may also be smaller than average. The unique facial features seen in some patients with PWS may be difficult to detect in infancy. These facial features are very mild and do not cause physical problems.

As early as six months, but more commonly at one to two years a compulsive desire to eat develops. This uncontrollable appetite is a classic feature of PWS. Individuals with PWS lack the ability to feel full or satiated. This uncontrollable desire to eat is thought to be related to a difference in the brain, which controls hunger. Over-eating (hyperphagia), a lack of a desire to **exercise**, and a slow metabolism places individuals with PWS at high risk for severe obesity. Some individuals with PWS may also have a reduced ability to vomit.

Behavior problems are a common feature of PWS. Some behavior problems develop from the desire to eat. Other reported problems include obsessive-compulsive behaviors, depression, and temper tantrums. Individuals with PWS may also pick their own skin (skin picking). This unusual behavior may be due to a reduced **pain** threshold.

Developmental delay, learning disabilities, and **mental retardation** are associated with PWS. Approximately 50% of individuals with PWS have developmental delay. The remaining 50% are described as having mild mental retardation. The mental retardation can occasionally be more severe. Infants and children with PWS are often delayed in development.

Puberty may occur early or late, but it is usually incomplete. In addition to the effects on sexual development and fertility, individuals do not undergo the normal adolescent growth spurt and may be short as

adults. Muscles often remain underdeveloped and body fat is increased.

Diagnosis

During infancy the diagnosis of PWS may be suspected if poor muscle tone, feeding problems, small genitalia, or the unique facial features are present. If an infant has these features, testing for PWS should be performed. This testing should also be offered to children and adults who display features commonly seen in PWS (developmental delay, uncontrollable appetite, small genitalia, etc.). There are several different genetic tests that can detect PWS. All of these tests can be performed from a blood sample.

Methylation testing detects 99% of the cases of PWS. Methylation testing can detect the absence of the paternal genes that should be normally active on chromosome 15. Although methylation testing can accurately diagnose PWS, it can not determine if the PWS is caused by a deletion, maternal uniparental disomy, or a mutation that disrupts imprinting. This information is important for **genetic counseling**. Therefore, additional testing should be performed.

Chromosome analysis can determine if the PWS is the result of a deletion in the q arm of chromosome 15. Chromosome analysis, also called "karyotyping," involves staining the chromosomes and examining them under a microscope. In some cases the deletion of material from chromosome 15 can be easily seen. In other cases, further testing must be performed. FISH (fluorescence in-situ hybridization) is a special technique that detects small deletions that cause PWS.

More specialized DNA testing is required to detect maternal uniparental disomy or a mutation that disrupts imprinting. This DNA testing identifies unique DNA patterns in the mother and father. The unique DNA patterns are then compared with the DNA from the child with PWS.

PWS can be detected before birth if the mother undergoes **amniocentesis** testing or **chorionic villus sampling** (CVS). This testing is only recommended if the mother or father is known to have a chromosome rearrangement, or if they already have a child with PWS syndrome.

Treatment

There is currently not a cure for PWS. Treatment during infancy includes therapies to improve muscle tone. Some infants with PWS also require special nipples and feeding techniques to improve weight gain.

KEY TERMS

Amniocentesis—A procedure in which a needle is inserted through a pregnant woman's abdomen and into her uterus. Amniotic fluid is then removed from around the fetus and may be used for genetic testing.

Centromere—Major constriction in a chromosome.

Deletion—Removal of a piece of genetic material.

DNA—Deoxyribonucleic acid. Genes are made of sections of DNA.

FISH—(flourescence in-situ hybridization) Technique used to detect small deletions or rearrangements in chromosomes.

Gene—Segment of DNA that controls the development and function of the body. Genes are contained within chromosomes.

Hyperphagia—Over-eating.

Hypotonia—Low muscle tone.

Imprinting—Process that silences a gene or group of genes. The genes are silenced depending on if they are inherited through the egg or the sperm.

Maternal—From one's mother.

Maternal uniparental disomy—Chromosome abnormality in which both chromosomes in a pair are inherited from one's mother.

Methylation testing—DNA testing that detects if a gene is active or imprinted.

Mutation—A change in a gene.

Paternal—From one's father.

Translocation—Chromosome abnormality in which chromosomes are rearranged and placed together.

Uniparental disomy—Chromosome abnormality in which both chromosomes in a pair are inherited from the same parent.

improve the poor muscle tone and reduced height typically associated with PWS.

Special education may be helpful in treating developmental delays and behavior problems. Individuals with PWS typically excel in highly structured environments.

Prognosis

Life expectancy is normal and the prognosis good, if weight gain is well controlled.

Resources

BOOKS

Whittington, Joyce, and Tony Holland. *Prader-Willi Syndrome: Development and Manifestations*. Cambridge, UK: Cambridge University Press, 2010.

OTHER

Gene Clinics. <http://www.geneclinics.org/profiles/pws/details.html>.

OMIM. <http://www.ncbi.nlm.nih.gov/omim/176270>.

ORGANIZATIONS

Genetic Alliance, Inc., 4301 Connecticut Ave., NW, Suite 404, Washington, DC, 20008-2369, (202) 966-5557, (202) 966-8553, info@geneticalliance.org, <http://www.geneticalliance.org>.

International Prader-Willi Syndrome Organization, c/o BIRD Foundation Onlus, Via Bartolomeo Bizio I, 1-36023 Costozza (VI), Italy, <http://www.ipwso.org>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Prader-Willi Foundation, PO BOX 222, Baldwinsville, NY, 13027, (716) 276-2211, (800) 442-1655, alliance@prader-willi.org, <http://www.prader-willi.org>.

Prader-Willi Syndrome Association, 8588 Potter Park Drive, Suite 500, Sarasota, FL, 34238, (941) 312-0142, (800) 926-4797, pwsausa@pwsausa.org, <http://www.pwsausa.org>.

Holly Ann Ishmael, M.S.

Praziquantel see **Antihelminthic drugs**

Precocious puberty

Definition

Precocious **puberty** is defined as sexual development before the age of 7 in girls, and age 9 in boys. The American Academy of Pediatrics (AAP) identifies the specific signs of precocious puberty as the emergence of breast buds in girls of 6 or 7, or an increase in the

Treatment and management during childhood, adolescence, and adulthood is typically focused on weight control. Strict control of food intake is vital to prevent severe obesity. In many cases food must be made inaccessible. This may involve unconventional measures such as locking the refrigerator or kitchen cabinets. A lifelong restricted-calorie diet and regular exercise program are also suggested. Unfortunately, diet medications have not been shown to significantly prevent obesity in PWS. However, growth hormone therapy has been shown to

size of a boy's testicle before his ninth birthday. Another definition of precocious puberty is population-specific; it states that precocious puberty is puberty that occurs at an age 2.5 standard deviations below the mean in a given population under consideration. It is important to keep in mind, however, that puberty appears to be occurring earlier in children in the early twenty-first century than in previous generations, so that definitions of "early" puberty may change over time.

Precocious puberty is subdivided into two types, central precocious puberty and peripheral precocious puberty (also called precocious pseudopuberty). In central precocious puberty (CPP), the condition is caused by the early maturation of the hypothalamic-pituitary-gonadal (HPG) axis. The HPG axis is a term used by doctors to refer to the combined effects of the hypothalamus, the pituitary gland, and the gonads. This group of glands controls sexual development, reproduction, and **aging** in humans and other animals. Most cases (80%) of precocious puberty are classified as CPP.

In peripheral precocious puberty (or precocious pseudopuberty), the sex hormones producing early signs of sexual maturation in the child come from sources other than the HPG axis. The hormones may be released from tumors in the adrenal gland or the pituitary gland, or they may be present in the child's body as the result of consuming soy products or using creams or ointments containing estrogen or testosterone. Other causes of precocious pseudopuberty include **ovarian cysts** or tumors (in girls) or **germ cell tumors** or a rare genetic mutation (in boys).

Demographics

According to the AAP, precocious puberty affects about 1 in every 160 otherwise healthy children—the same proportion as the number of youngsters who experience belated puberty. Exact figures for precocious puberty are difficult to obtain, however, because the condition is defined as a departure from the "normal" age of puberty, and that age has dropped since statistics were first kept in the 1840s. The drop in the average age at puberty since the early 2000s has been most noticeable in China, southern Europe, and other countries with warm climates.

Girls are much more likely to develop central precocious puberty than boys, the sex ratio being variously given as 5:1 or 8:1. In the United States, 25% of African American girls are reported to develop CPP, compared to 8% for Hispanic and Caucasian girls. The reason for this difference is not known as of 2010.

About 80% of all cases of precocious puberty are central precocious puberty, the remaining 20% being precocious pseudopuberty.

Description

Not every child in North America reaches puberty at the same time, but in most cases it's safe to predict that sexual development will begin at about age 11 in girls and 12 or 13 in boys. Precocious puberty often begins before age 8 in girls, triggering the development of breasts and hair under the arms and in the genital region. The onset of ovulation and menstruation also may occur. In boys, the condition triggers the development of a large penis and testicles, with spontaneous erections and the production of sperm. Hair grows on the face, under arms and in the pubic area, and **acne** may become a complexion problem. Children with precocious puberty may also develop adult body odor when they perspire.

While the early onset of puberty may seem fairly benign, in fact it can cause problems when hormones trigger changes in the growth pattern, essentially halting growth before the child has reached normal adult height. Girls may never grow taller than 5 ft (152 cm) and boys often stop growing by about 5 ft 2 in (157 cm).

Abnormal growth patterns are not the only problem, however. Children with this condition look noticeably different from their peers, and may feel rejected by their friends and socially isolated. Adults may expect these children to act more maturely simply because they look so much older. As a result, many of these children—especially boys—are much more aggressive than others their own age, leading to behavior problems both at home and at school. Girls who mature early are at increased risk of **sexual abuse**.

Risk factors

Risk factors for precocious puberty include:

- Sex. Girls are five times more likely to develop CPP than boys.
- Race. African American girls are three times as likely to develop CPP as Caucasian girls. This racial difference, however, does not hold true for boys.
- Obesity, particularly in girls. One study found that obese girls had an 80% chance of developing breasts before their ninth birthday and starting menstruation before age 12.
- History of injury to the central nervous system caused by trauma, surgery, or radiation therapy.
- History of brain tumors or structural abnormalities in the brain.

- Exposure to products containing sex hormones, including skin or hair products, vitamins, or dietary supplements. Heavy consumption of soy products (including tofu) is a risk factor for precocious pseudopuberty in that soy contains phytoestrogens, substances found in certain plants that cause estrogen-like effects in humans.
- History of precocious puberty in other family members.

Causes and symptoms

Puberty begins when the part of the brain called the hypothalamus secretes a hormone (gonadotropin-releasing hormone or Gn-RH) that triggers the pituitary gland to release gonadotropins. These protein hormones in turn stimulate the gonads (ovaries or testes) to produce sex hormones. These sex hormones (especially estrogen in girls and testosterone in boys) are what causes the onset of sexual maturity.

The hormonal changes of central precocious puberty are normal—it is just that the whole process begins a few years too soon. In 90% of girls with precocious puberty, there is no underlying problem that causes the process of sexual maturation to begin too soon. About 50% of boys, however, do have an underlying condition. Some inherit the condition; the responsible gene may be passed directly from father to son, or inherited indirectly from the maternal grandfather through the mother, who does not begin early puberty herself. This condition is called familial male precocious puberty. This genetic condition in girls can be traced in only about 1% of cases.

Other conditions or disorders that may be associated with precocious puberty include:

- Tumors, cysts, or other abnormalities of the thyroid gland (in both sexes).
- History of radiation therapy for childhood cancer.
- McCune-Albright syndrome. This is a disorder that affects the bones, skin, and endocrine system. It is caused by a mutation on a single gene and cannot be passed down to the next generation. Girls with this syndrome may have their first menstrual period as early as two years of age.
- Disorders of the central nervous system, including cerebral palsy, neurofibromatosis, brain tumors, and tuberous sclerosis.
- Congenital adrenal hyperplasia (CAH). This is a genetic disorder in which the adrenal glands produce too much of the male sex hormone androgen.

Precocious puberty that cannot be traced to a specified cause is termed idiopathic or constitutional.

KEY TERMS

Central precocious puberty (CPP)—Precocious puberty resulting from the early maturation of the HPG axis.

Endocrine system—A system of ductless glands that secrete hormones that regulate a variety of body processes, including growth and sexual maturation. A doctor who specializes in diseases and disorders of these glands is called an endocrinologist.

Gonadotropins—Protein hormones secreted by the pituitary gland.

Hypothalamic-pituitary-gonadal (HPG) axis—A term used by doctors to refer to the combined effects of the hypothalamus, the pituitary gland, and the gonads. This group of glands controls sexual maturation in humans as well as other processes.

Idiopathic—Of unknown cause or spontaneous origin.

Peripheral precocious puberty—Precocious puberty resulting from the presence of sex steroids independent of the activation of the HPG axis. It is also called precocious pseudopuberty.

Precocious—Developing at an unusually early age.

Puberty—The process of maturation in which a child's body turns into an adult body capable of reproduction. The English word comes from a Latin word that means “age of maturity” or “adulthood.”

Diagnosis

Parents should consult their child's pediatrician if their child shows any of the signs or symptoms of precocious puberty. The pediatrician may refer the child to an endocrinologist (doctor who specializes in disorders of the glands that regulate growth and sexual maturation as well as other body processes), because it is important to distinguish between central precocious puberty and precocious pseudopuberty—the two conditions are treated very differently.

Examination

An office **physical examination** can reveal the development of sexual characteristics in a young child. The doctor will weigh the child and compare his or her development to charts of normal development for the child's sex and age. If the child has been referred to an endocrinologist, the specialist will take a

careful family history as well as a history of the child's development, including any surgeries or radiation treatments. **Bone x rays** can reveal bone age, and **pelvic ultrasound** may show an enlarged uterus and rule out ovarian or adrenal tumors. MRI or CAT scans should be considered to rule out intracranial tumors.

Tests

Blood tests can highlight higher-than-normal levels of hormones. To distinguish between CPP and precocious pseudopuberty, the doctor will administer an injection of Gn-RH hormone and then take a blood sample 30–60 minutes later. If the child has central precocious puberty, levels of two other hormones in the blood will rise. If the child has precocious pseudopuberty, the levels of these other hormones will remain the same.

The doctor may take another blood sample to test for levels of thyroid hormone if an abnormality of the thyroid gland is suspected.

Procedures

In the case of girls with precocious puberty, the doctor may perform a pelvic ultrasound to check for an ovarian cyst or tumor.

Treatment

Traditional

Treatment of precocious puberty aims to halt or reverse sexual development so as to stop the accompanying rapid growth that will limit a child's height. There are two possible approaches: either treat the underlying condition (such as an ovarian or intracranial tumor) or change the hormonal balance to stop sexual development. Tumors in the central nervous system can sometimes be successfully removed by surgery; however, it may not be possible to treat the underlying condition. Treatment of central precocious puberty is usually aimed at adjusting hormone levels by giving an analogue of Gn-RH, most commonly leuprolide (Lupron). Monthly injections of such medications slow down the HPG axis until the child reaches the normal age of puberty. Once the injections are stopped, puberty will begin again.

Drugs

There are several drugs that have been developed to treat precocious puberty:

- histrelin (Supprelin LA)
- nafarelin (Synarel)
- synthetic gonadotropin-releasing hormone agonist

- progestin (Depo-Provera)
- leuprolide (Lupron)

Prognosis

Drug treatments can slow growth to 2–3 in (5–7.5 cm) a year, allowing these children to reach normal adult height, although the long-term effects are not yet known. As of 2010, few prospective studies on these drugs have been performed; their psychosocial effects and their possible side effects of weight gain and reduced bone mineral density require further study. Children taking drugs to suppress early puberty should be seen for follow-up visits every 4–6 months to check on bone growth and (in girls) a decrease or at least no increase in breast size.

The prognosis of precocious pseudopuberty depends on its cause. Recovery from McCune-Albright syndrome depends on the extent of bone disease, but most girls have a good prognosis. The prognosis of CAH is excellent with proper treatment. The prognosis of girls with ovarian tumors depends on the stage at which the tumors were discovered.

Prevention

Some risk factors for precocious puberty, such as sex, race, or family history of early puberty, cannot be avoided. Parents can, however, keep children away from dietary supplements or adult medications containing sex hormones; limit the amount of soy products in the diet; and help their children maintain healthy weight levels.

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ORGANIZATIONS

- American Academy of Pediatrics (AAP), 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007, 847-434-4000, 847-434-8000, <http://www.aap.org>.
- National Institute of Child Health and Human Development (NICHD), Bldg 31, Room 2A32, MSC 2425, 31 Center Drive, Bethesda, MD, 20892, 800-370-2943, 866-760-5947, NICHDInformationResourceCenter@mail.nih.gov, <http://www.nichd.nih.gov>.

Carol A. Turkington
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Prednisone see **Corticosteroids**

Description

Blood pressure is a measurement of the pressure of blood on the walls of blood vessels called arteries. The arteries deliver blood from the heart to all of the tissues in the body. Blood pressure is reported as two numbers. For example, a normal blood pressure is reported as 110/70 mm Hg (read as 110 over 70 millimeters of mercury; or just 110 over 70). These two numbers represent two measurements, the systolic pressure and the diastolic pressure. The systolic pressure (the first number in the example; 110/70 mm Hg) measures the peak pressure of the blood against the artery walls. This higher pressure occurs as blood is being pumped out of the heart and into the circulatory system. The pumping chambers of the heart (ventricles) squeeze to force the blood out of the heart. The diastolic pressure (the second number in the example 110/70 mm Hg) measures the pressure, during the filling of the ventricles. At this point, the atria contract to fill the ventricles. Because the ventricles are relatively relaxed, and are not pumping blood into the arteries, pressure in the arteries is lower as well.

High blood pressure in pregnancy (**hypertension**) is a very serious complication. It puts both the mother and the fetus at risk for a number of problems. Hypertension can exist in several different forms:

- The preeclampsia-eclampsia continuum (also called pregnancy-induced hypertension or PIH). In this type of hypertension, high blood pressure is first noted sometime after week 20 of pregnancy and is accompanied by protein in the urine and swelling.
- Chronic hypertension. This type of hypertension usually exists before pregnancy or may develop before week 20 of pregnancy.
- Chronic hypertension with superimposed preeclampsia. This syndrome occurs when a woman with pre-existing chronic hypertension begins to have protein in the urine after week 20 of pregnancy.
- Late hypertension. This is a form of high blood pressure occurring after week 20 of pregnancy and is unaccompanied by protein in the urine and does not progress the way preeclampsia-eclampsia does.

Preeclampsia is most common among women who have never given birth to a baby (called nulliparas). About 7% of all nulliparas develop preeclampsia. The disease is most common in mothers under the age of 20, or over the age of 35. African-American women have higher rates of preeclampsia than do Caucasian women. Other risk factors include poverty, multiple pregnancies (twins, triplets, etc.), pre-existing chronic

Preeclampsia and eclampsia

Definition

Preeclampsia and eclampsia are complications of **pregnancy**. In preeclampsia, the woman has dangerously high blood pressure, swelling, and protein in the urine. If allowed to progress, this syndrome will lead to eclampsia.

hypertension or **kidney disease**, diabetes, excess amniotic fluid, and a condition of the fetus called nonimmune hydrops. The tendency to develop preeclampsia appears to run in families. The daughters and sisters of women who have had preeclampsia are more likely to develop the condition.

Causes and symptoms

Experts are still trying to understand the exact causes of preeclampsia and eclampsia. It is generally accepted that preeclampsia and eclampsia are problematic because these conditions cause blood vessels to leak. The effects are seen throughout the body.

- General body tissues. When blood vessels leak, they allow fluid to flow out into the tissues of the body. The result is swelling in the hands, feet, legs, arms, and face. While many pregnant women experience swelling in their feet, and sometimes in their hands, swelling of the upper limbs and face is a sign of a more serious problem. As fluid is retained in these tissues, the woman may experience significant weight gain (two or more pounds per week).
- Brain. Leaky vessels can cause damage within the brain, resulting in seizures or coma.
- Eyes. The woman may experience problems seeing, and may have blurry vision or may see spots. The retina may become detached.
- Lungs. Fluid may leak into the tissues of the lungs, resulting in shortness of breath.
- Liver. Leaky vessels within the liver may cause it to swell. The liver may be involved in a serious complication of preeclampsia, called the HELLP syndrome. In this syndrome, red blood cells are abnormally destroyed, chemicals called liver enzymes are abnormally high, and cells involved in the clotting of blood (platelets) are low.
- Kidneys. The small capillaries within the kidneys can leak. Normally, the filtration system within the kidney is too fine to allow protein (which is relatively large) to leave the bloodstream and enter the urine. In preeclampsia, however, the leaky capillaries allow protein to be dumped into the urine. The development of protein in the urine is very serious, and often results in a low birth weight baby. These babies have a higher risk of complications, including death.
- Blood pressure. In preeclampsia, the volume of circulating blood is lower than normal because fluid is leaking into other parts of the body. The heart tries to make up for this by pumping a larger quantity of blood with each contraction. Blood vessels usually expand in diameter (dilate) in this situation to decrease the work load on the heart. In preeclampsia,

however, the blood vessels are abnormally constricted, causing the heart to work even harder to pump against the small diameters of the vessels. This causes an increase in blood pressure.

The most serious consequences of preeclampsia and eclampsia include brain damage in the mother due to brain swelling and oxygen deprivation during seizures. Mothers can also experience blindness, kidney failure, liver rupture, and **placental abruption**. Babies born to preeclamptic mothers are often smaller than normal, which makes them more susceptible to complications during labor, delivery, and in early infancy. Babies of preeclamptic mothers are also at risk of being born prematurely, and can suffer the complications associated with **prematurity**.

Diagnosis

Diagnosing preeclampsia may be accomplished by noting painless swelling of the arms, legs, and/or face, in addition to abnormal weight gain. The patient's blood pressure is taken during every doctor's visit during pregnancy. An increase of 30 mm Hg in the systolic pressure, or 15 mm Hg in the diastolic pressure, or a blood pressure reading greater than 140/90 mm Hg is considered indicative of preeclampsia. A simple laboratory test in the doctor's office can indicate the presence of protein in a urine sample (a dipstick test). A more exact measurement of the amount of protein in the urine can be obtained by collecting urine for 24 hours, and then testing it in a laboratory to determine the actual quantity of protein present. A 24-hour urine specimen containing more than 500 mg of protein is considered indicative of preeclampsia.

Treatment

With mild preeclampsia, treatment may be limited to bed rest, with careful daily monitoring of weight, blood pressure, and urine protein via dipstick. This careful monitoring will be required throughout pregnancy, labor, delivery, and even for 2–4 days after the baby has been born. About 25% of all cases of eclampsia develop in the first few days after the baby's birth. If the diastolic pressure does not rise over 100 mm Hg prior to delivery, and no other symptoms develop, the woman can continue pregnancy until the fetus is mature enough to be delivered safely. Ultrasound tests can be performed to monitor the health and development of the fetus.

If the diastolic blood pressure continues to rise over 100 mm Hg, or if other symptoms like **headache**, vision problems, abdominal **pain**, or blood abnormalities

KEY TERMS

Capillary—The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules.

Diastolic—The phase of blood circulation in which the heart's pumping chambers (ventricles) are being filled with blood. During this phase, the ventricles are at their most relaxed, and the pressure against the walls of the arteries is at its lowest.

Placenta—The organ that provides oxygen and nutrition from the mother to the fetus during pregnancy. The placenta is attached to the wall of the uterus and leads to the fetus via the umbilical cord.

Placental abruption—An abnormal separation of the placenta from the uterus before the birth of the baby, with subsequent heavy uterine bleeding. Normally, the baby is born first and then the placenta is delivered within a half hour.

Systolic—The phase of blood circulation in which the heart's pumping chambers (ventricles) are actively pumping blood. The ventricles are squeezing (contracting) forcefully, and the pressure against the walls of the arteries is at its highest.

Urine dipstick test—A test using a small, chemically treated strip that is dipped into a urine sample; when testing for protein, an area on the strip changes color depending on the amount of protein (if any) in the urine.

Uterus—The muscular organ that contains the developing baby during pregnancy.

Ventricles—The two chambers of the heart that are involved in pumping blood. The right ventricle pumps blood into the lungs to receive oxygen. The left ventricle pumps blood into the circulation of the body to deliver oxygen to all of the body's organs and tissues.

develop, then the patient may require medications to prevent seizures. Magnesium sulfate is commonly given through a needle in a vein (intravenous, or IV). Medications that lower blood pressure (**antihypertensive drugs**) are reserved for patients with very high diastolic pressures (over 110 mm Hg), because lowering the blood pressure will decrease the amount of blood reaching the fetus. This places the fetus at risk for oxygen deprivation. If preeclampsia appears to be progressing toward true eclampsia, then medications may be given in order to start labor. Babies can usually be delivered

vaginally. After the baby is delivered, the woman's blood pressure and other vital signs will usually begin to return to normal quickly.

Prognosis

The prognosis in preeclampsia and eclampsia depends on how carefully a patient is monitored. Very careful, consistent monitoring allows quick decisions to be made, and improves the woman's prognosis. Still, the most common causes of **death** in pregnant women are related to high blood pressure.

About 33% of all patients with preeclampsia will have the condition again with later pregnancies. Eclampsia occurs in about 1 out of every 200 women with preeclampsia. If not treated, eclampsia is almost always fatal.

Prevention

More information on how preeclampsia and eclampsia develop is needed before recommendations can be made on how to prevent these conditions. Research is being done with patients in high risk groups to see if **calcium** supplementation, **aspirin**, or fish oil supplementation may help prevent preeclampsia. Most importantly, it is clear that careful monitoring during pregnancy is necessary to diagnose preeclampsia early. Although even carefully monitored patients may develop preeclampsia and eclampsia, close monitoring by practitioners will help decrease the complications of these conditions.

ORGANIZATIONS

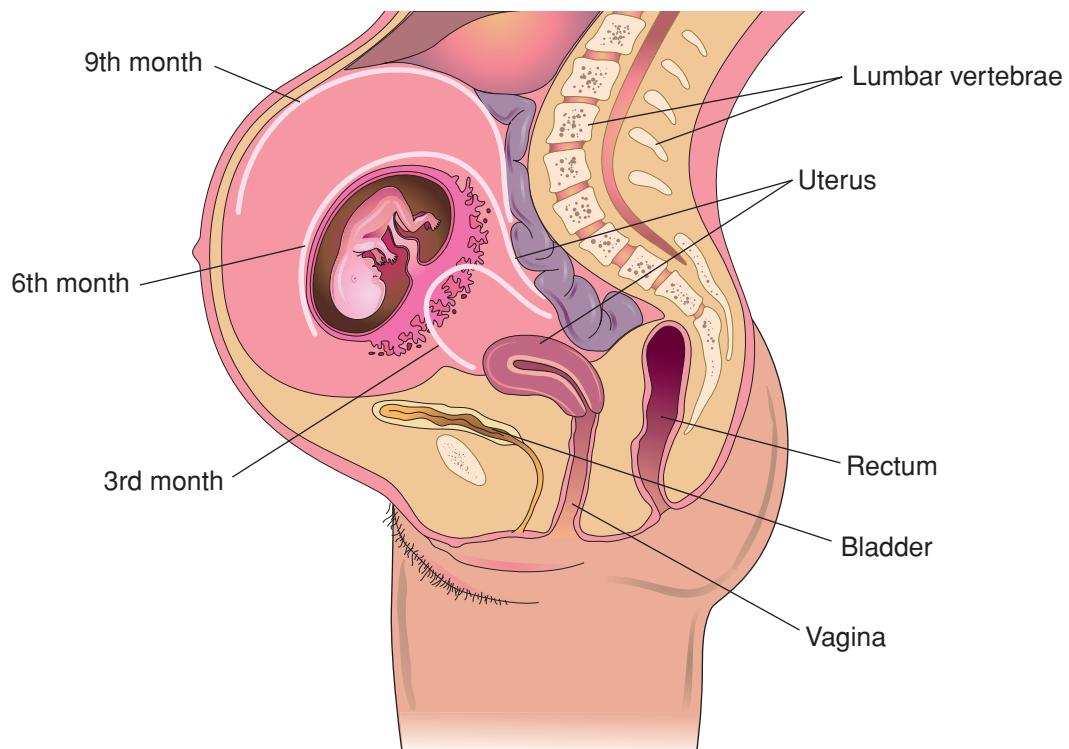
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Pregnancy

Definition

The period from conception to birth. After the egg is fertilized by a sperm and then implanted in the lining of the uterus, it develops into the placenta and embryo, and later into a fetus. Pregnancy usually lasts 40 weeks, beginning from the first day of the woman's last menstrual period, and is divided into three trimesters, each lasting three months.



Pregnancy usually lasts 40 weeks in humans, beginning from the first day of the woman's last menstrual period, and is divided into three trimesters. The illustration above depicts the position of the developing fetus during each trimester. ((Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Description

Pregnancy is a state in which a woman carries a fertilized egg inside her body. Due to technological advances, pregnancy is increasingly occurring among older women in the United States.

First month

At the end of the first month, the embryo is about a third of an inch long, and its head and trunk—plus the beginnings of arms and legs—have started to develop. The embryo receives nutrients and eliminates waste through the umbilical cord and placenta. By the end of the first month, the liver and digestive system begin to develop, and the heart starts to beat.

Second month

In this month, the heart starts to pump and the nervous system (including the brain and spinal cord) begins to develop. The 1 in (2.5 cm) long fetus has a complete cartilage skeleton, which is replaced by bone cells by month's end. Arms, legs and all of the major organs begin to appear. Facial features begin to form.

Third month

By now, the fetus has grown to 4 in (10 cm) and weighs a little more than an ounce (28 g). The major blood vessels and the roof of the mouth are almost completed, and the face starts to take on a more recognizably human appearance. Fingers and toes appear. All the major organs are now beginning to form; the kidneys are now functional and the four chambers of the heart are complete.

Fourth month

The fetus begins to kick and swallow, although most women still can not feel the baby move at this point. Now 4 oz (112 g), the fetus can hear and urinate, and has established sleep-wake cycles. All organs are now fully formed, although they will continue to grow for the next five months. The fetus has skin, eyebrows, and hair.

Fifth month

Now weighing up to a 1 lb (454 g) and measuring 8–12 in (20–30 cm), the fetus experiences rapid growth

as its internal organs continue to grow. At this point, the mother may feel her baby move, and she can hear the heartbeat with a stethoscope.

Sixth month

Even though its lungs are not fully developed, a fetus born during this month can survive with intensive care. Weighing 1–1.5 lbs (454–681 g), the fetus is red, wrinkly, and covered with fine hair all over its body. The fetus will grow very fast during this month as its organs continue to develop.

Seventh month

There is a better chance that a fetus born during this month will survive. The fetus continues to grow rapidly, and may weigh as much as 3 lb (1.3 kg) by now. Now the fetus can suck its thumb and look around its watery womb with open eyes.

Eighth month

Growth continues but slows down as the baby begins to take up most of the room inside the uterus. Now weighing 4–5 lbs (1.8–2.3 kg) and measuring 16–18 in (40–45 cm) long, the fetus may at this time prepare for delivery next month by moving into the head-down position.

Ninth month

Adding 0.5 lb (227 g) a week as the due date approaches, the fetus drops lower into the mother's abdomen and prepares for the onset of labor, which may begin any time between the 37th and 42nd week of gestation. Most healthy babies will weigh 6–9 lb (2.7–4 kg) at birth, and will be about 20 in. long.

Causes and symptoms

The first sign of pregnancy is usually a missed menstrual period, although some women bleed in the beginning. A woman's breasts swell and may become tender as the mammary glands prepare for eventual **breastfeeding**. Nipples begin to enlarge and the veins over the surface of the breasts become more noticeable.

Nausea and vomiting are very common symptoms and are usually worse in the morning and during the first trimester of pregnancy. They are usually caused by hormonal changes, in particular, increased levels of progesterone. Women may feel worse when their stomach is empty, so it is a good idea to eat several small meals throughout the day, and to keep things like crackers on hand to eat even before getting out of bed in the morning.

Many women also feel extremely tired during the early weeks. Frequent urination is common, and there may be a creamy white discharge from the vagina. Some women crave certain foods, and an extreme sensitivity to smell may worsen the **nausea**. Weight begins to increase.

In the second trimester (13–28 weeks) a woman begins to look noticeably pregnant and the enlarged uterus is easy to feel. The nipples get bigger and darker, skin may darken, and some women may feel flushed and warm. Appetite may increase. By the 22nd week, most women have felt the baby move. During the second trimester, nausea and **vomiting** often fade away, and the pregnant woman often feels much better and more energetic. Heart rate increases as does the volume of blood in the body.

By the third trimester (29–40 weeks), many women begin to experience a range of common symptoms. Stretch marks may develop on abdomen, breasts, and thighs, and a dark line may appear from the navel to pubic hair. A thin fluid may be discharged from the nipples. Many women feel hot, sweat easily and often find it hard to get comfortable. Kicks from an active baby may cause sharp pains, and lower backaches are common. More rest is needed as the woman copes with the added **stress** of extra weight. Braxton Hicks contractions may get stronger.

At about the 36th week in a first pregnancy (later in repeat pregnancies), the baby's head drops down low into the pelvis. This may relieve pressure on the upper abdomen and the lungs, allowing a woman to breathe more easily. However, the new position places more pressure on the bladder.

A healthy weight gain for most women is between 25 and 35 pounds. Women who are overweight should gain less; and women who are underweight should gain more. On average, pregnant women need an additional 300 calories a day. Generally, women will gain three to five pounds in the first three months, and then add one to two pounds a week until the baby is born. An average, healthy full-term baby at birth weighs 7.5 lb (3.4 kg), and the placenta and fluid together weigh another 3.5 lb. The remaining weight that a woman gains during pregnancy is mostly due to water retention and fat stores. Her breasts, for instance, gain about 2 lb. in weight, and she gains another 4 lb due to increased blood volume.

In addition to the typical, common symptoms of pregnancy, some women experience other problems that may be annoying, but which usually disappear after delivery. **Constipation** may develop as a result of food passing more slowly through the intestine.

Hemorrhoids and **heartburn** are fairly common during late pregnancy. Gums may become more sensitive and bleed more easily; eyes may dry out, making **contact lenses** feel painful. **Pica** (a craving to eat substances other than food) may occur. Swollen ankles and **varicose veins** may be a problem in the second half of pregnancy, and chloasma may appear on the face.

Chloasma, also known as the “mask of pregnancy” or melasma, is caused by hormonal changes that result in blotches of pale brown skin appearing on the forehead, cheeks, and nose. These blotches may merge into one dark mask. It usually fades gradually after pregnancy, but it may become permanent or recur with subsequent pregnancies. Some women also find that the line running from the top to the bottom of their abdomen darkens. This is called the linea nigra.

While the above symptoms are all considered to be normal, there are some symptoms that could be a sign of a more dangerous underlying problem. A pregnant woman with any of the following signs should contact her doctor immediately:

- abdominal pain
- rupture of the amniotic sac or leaking of fluid from the vagina
- bleeding from the vagina
- no fetal movement for 24 hours (after the fifth month)
- continuous headaches
- marked, sudden swelling of eyelids, hands, or face during the last three months
- dim or blurry vision during last three months
- persistent vomiting

Diagnosis

Many women first discover they are pregnant after a positive home pregnancy test. Pregnancy urine tests check for the presence of human chorionic gonadotropin (hCG), which is produced by a placenta. The newest home tests can detect pregnancy on the day of the missed menstrual period.

Home pregnancy tests are more than 97% accurate if the result is positive, and about 80% accurate if the result is negative. If the result is negative and there is no menstrual period within another week, the pregnancy test should be repeated. While home pregnancy tests are very accurate, they are less accurate than a pregnancy test conducted at a lab. For this reason, women may want to consider having a second pregnancy test conducted at their doctor’s office to be sure of the accuracy of the result.

Blood tests to determine pregnancy are usually used only when a very early diagnosis of pregnancy is needed. This more expensive test, which also looks for hCG, can produce a result within nine to 12 days after conception.

Once pregnancy has been confirmed, there are a range of screening tests that can be done to screen for **birth defects**, which affect about 3% of unborn children. Two tests are recommended for all pregnant women: alpha-fetoprotein (AFP) and the triple marker test.

Other tests are recommended for women at higher risk for having a child with a birth defect. This includes women over age 35, who had another child or a close relative with a birth defect, or who have been exposed to certain drugs or high levels of radiation. Women with any of these risk factors may want to consider **amniocentesis**, **chorionic villus sampling (CVS)** or ultrasound.

Other prenatal tests

There are a range of other prenatal tests that are routinely performed, including:

- PAP test
- gestational diabetes screening test at 24–28 weeks
- tests for sexually transmitted diseases
- urinalysis
- blood tests for anemia or blood type
- screening for immunity to various diseases, such as German measles

Treatment

Prenatal care is vitally important for the health of the unborn baby. A pregnant woman should be sure to eat a balanced, nutritious diet of frequent, small meals. Women should begin taking 400 mcg of **folic acid** several months before becoming pregnant, as folic acid has been shown to reduce the risk of spinal cord defects, such as **spina bifida**.

No medication (not even a nonprescription drug) should be taken except under medical supervision, since it could pass from the mother through the placenta to the developing baby. Some drugs, called teratogens, have been proven harmful to a fetus, but no drug should be considered completely safe (especially during early pregnancy). Drugs taken during the first three months of a pregnancy may interfere with the normal formation of the baby’s organs, leading to birth defects. Drugs taken later on in pregnancy may slow the baby’s growth rate, or they may damage specific fetal tissue (such as the developing teeth), or cause preterm birth. Herbal supplements and other “natural” remedies can also be extremely harmful to

KEY TERMS

Alpha-fetoprotein—A substance produced by a fetus' liver that can be found in the amniotic fluid and in the mother's blood. Abnormally high levels of this substance suggests there may be defects in the fetal neural tube, a structure that will include the brain and spinal cord when completely developed. Abnormally low levels suggest the possibility of Down's syndrome.

Braxton Hicks' contractions—Short, fairly painless uterine contractions during pregnancy that may be mistaken for labor pains. They allow the uterus to grow and help circulate blood through the uterine blood vessels.

Chloasma—A skin discoloration common during pregnancy, also known as the "mask of pregnancy" or melasma, in which blotches of pale brown skin appear on the face. It is usually caused by hormonal changes. The blotches may appear in the forehead, cheeks, and nose, and may merge into one dark mask. It usually fades gradually after pregnancy, but it may become permanent or recur with subsequent pregnancies. Some women may also find that the line

running from the top to the bottom of their abdomen darkens. This is called the linea nigra.

Embryo—An unborn child during the first eight weeks of development following conception (fertilization with sperm). For the rest of pregnancy, the embryo is known as a fetus.

Fetus—An unborn child from the end of the eighth week after fertilization until birth.

Human chorionic gonadotropin (hCG)—A hormone produced by the placenta during pregnancy.

Placenta—The organ that develops in the uterus during pregnancy that links the blood supplies of the mother and baby.

Rhythm method—The oldest method of contraception with a very high failure rate, in which partners periodically refrain from having sex during ovulation. Ovulation is predicted on the basis of a woman's previous menstrual cycle.

Spina bifida—A congenital defect in which part of the vertebrae fail to develop completely, leaving a portion of the spinal cord exposed.

an unborn baby and should not be taken during pregnancy without close supervision by a physician.

To have the best chance of having a healthy baby, a pregnant woman should avoid:

- smoking
- alcohol
- street drugs
- large amounts of caffeine
- artificial sweeteners

Nutrition

Women should begin following a healthy diet even before they become pregnant. This means cutting back on high-calorie, high-fat, high-sugar snacks, and increasing the amount of fruits, vegetables, and whole grains in her diet. Once she becomes pregnant, she should make sure to eat at least six to 11 servings of breads and other whole grains, three to five servings of vegetables, two to four servings of fruits, four to six servings of milk and milk products, three to four servings of meat and protein foods, and to drink six to eight glasses of water each day. She should limit **caffeine** to no more than one soft drink or cup of coffee per day.

Prognosis

Pregnancy is a natural condition that usually causes little discomfort provided the woman takes care of herself and gets adequate prenatal care. **Childbirth** education classes for the woman and her partner help prepare the couple for labor and delivery.

Prevention

There are many ways to avoid pregnancy. A woman has a choice of many methods of **contraception** which will prevent pregnancy, including (in order of least to most effective):

- spermicide alone
- natural (rhythm) method
- diaphragm or cap alone
- condom alone
- diaphragm with spermicide
- condom with spermicide
- intrauterine device (IUD)
- contraceptive pill
- sterilization (either a man or woman)
- avoiding intercourse

Resources

BOOKS

Kitzinger, Sheila. *The Complete Book of Pregnancy and Childbirth*. New York: Alfred A. Knopf, 2004.

OTHER

Doulas of North America. <http://www.dona.com>.
Planned Parenthood. <http://www.plannedparenthood.org>.
Pregnancy Information. <http://www.childbirth.org>.

ORGANIZATIONS

National Healthy Mothers, Healthy Babies Coalition, 2000 N. Beauregard Street, 6th Floor, Alexandria, VA, 22311, (703) 837-4792, (703) 684-5968, info@hmhb.org, <http://www.hmhb.org>.

National Institute of Child Health and Human Development, Bldg 31, Room 2A32, MSC 2425, 31 Center Drive, Bethesda, MD, 20892-2425, (800) 370-2943, <http://www.nichd.nih.gov>.

Debra Gordon

Pregnancy-induced high blood pressure see
Preeclampsia and eclampsia

Pregnancy test see **Human chorionic gonadotropin pregnancy test**

Preleukemia see **Myelodysplastic syndrome**

Premature atrial contractions see **Atrial ectopic beats**

Premature birth see **Prematurity**

Causes and symptoms

There are several reasons why a man may ejaculate prematurely. For some men, the cause is due to an innate reflex or psychological predisposition of the nervous system. Sometimes it can be caused by certain drugs, such as non-prescription cold medications. Psychological factors, such as **stress**, fear, or guilt can also play a role. Examples of psychological factors include guilt that the sexual activity is wrong or sinful, fear of getting caught, or stress from problems at work or home.

In general, symptoms are when a male reaches climax in less than two minutes or when it occurs before the male or couple want it to occur.

Diagnosis

There are no tests used to diagnose premature ejaculation. It is usually determined by the male involved based on his belief that he reached orgasm too quickly. General guidelines for premature ejaculation is if it occurs in two minutes or less, or prior to about 15 thrusts during sexual intercourse.

Treatment

In 1966, William H. Masters and Virginia E. Johnson published *Human Sexual Response*, in which they broke the first ground in approaching this topic from a new perspective. Their method was devised by Dr. James Seman and has been modified subsequently by Dr. Helen Singer Kaplan and others.

A competent and orthodox sex therapist will spend much more time focusing on the personal than the sexual relationship between the two people who come for treatment. Without emotional intimacy, sexual relations are superficial and sexual problems such as premature ejaculation are not always overcome.

With that foremost in mind, a careful plan is outlined that requires dedication, patience, and commitment by both partners. It necessarily begins by prohibiting intercourse for an extended period of time—at least a week, often a month. This is very important to the man because “performance anxiety” is the greatest enemy of performance. If he knows he cannot have intercourse he is able to relax and focus on the exercises. The first stage is called “sensate focus” and involves his concentration on the process of sexual arousal and climax. He should learn to recognize each step in the process, most particularly the moment just before the “point of no return.” Ideally, this stage of treatment requires the man’s partner to be devoted to his sensations. In order to regain equality,

Premature ejaculation

Definition

Premature ejaculation occurs when male sexual climax (orgasm) occurs before a man wishes it or too quickly during intercourse to satisfy his partner.

Description

Premature ejaculation is the most commonly reported sexual complaint of men and couples. The highest number of complaints is among teenage, young adult, and sexually inexperienced males. Increased risk is associated with sexual inexperience and lack of knowledge of normal male sexual responses.

he should in turn spend separate time stimulating and pleasing his mate, without intercourse.

At this point the techniques diverge. The original “squeeze technique” requires that the partner become expert at squeezing the head of the penis at intervals to prevent orgasm. The modified procedure, described by Dr. Ruth Westheimer, calls upon the man to instruct the partner when to stop stimulating him to give him a chance to draw back. A series of stages follows, each offering greater stimulation as the couple gains greater control over his arousal. This whole process has been called “outercourse.” After a period of weeks, they will have together retrained his response and gained satisfactory control over it. In addition, they will each have learned much about the other’s unique sexuality and ways to increase each other’s pleasure.

With either technique, the emphasis is on the mutual goal of satisfactory sexual relations for both partners.

However, the 1990s ushered in a new era in the treatment of premature ejaculation when physicians discovered that certain antidepression drugs had a side effect of delaying ejaculation. Clinical studies have shown that a class of antidepressants called selective serotonin reuptake inhibitors (SSRIs) can be very effective in prolonging the time to ejaculation. The individual drugs and the average amount of time they delay ejaculation are fluoxetine (Prozac), one to two minutes with doses of 20–40 milligrams per day (mg/day) and eight minutes with 60 mg/day; paroxetine (Paxil), three to 10 minutes with doses of 20–40 mg/day; and sertraline (Zoloft), two to five minutes with doses of 50–200 mg/day.

Alternative treatment

There are several alternative products, usually found in health food and **nutrition** stores, designed to be sprayed or rubbed on the penis to delay ejaculation. Although the products promise results, there are no valid clinical studies to support the claims. A device called a testicular restraint, sold through erotic mail-order magazines, sometimes helps men delay ejaculation. The Velcro-like device restrains the testicles from their natural tendency to move during sex. Testicular movement can cause premature ejaculation.

Prognosis

The “squeeze technique” has elicited a 95% success rate, whereby the patient is able to control ejaculation. Treatment with SSRIs is effective in 85–90% of cases. However, the effectiveness begins to decrease after five weeks of daily administration. Although

more studies are needed, this suggests the SSRIs are more effective when used on an as-needed basis.

Prevention

The best prevention is obtaining adequate information on normal sexual responses of males before having sex. It is also helpful to have sex in a comfortable, relaxed, private setting, free of guilt, stress, and fear.

ORGANIZATIONS

American Association for Marriage and Family Therapy,
112 South Alfred Street, Alexandria, VA, 22314-3061,
(703) 838-9808, (703) 838-9805, <http://www.aamft.org>.

American Association of Sex Educators, Counselors, and Therapists, 1444 I Street NW, Suite 700, Washington, DC, 20005, (202) 449-1099, (202) 216-9646, <http://www.aasect.org>.

Sexuality Information and Education Council of the U.S.,
90 John St. Suite 402, New York, NY, (212) 819-9770,
(212) 819-9770, (212) 819-9776, pmalone@siecus.org,
<http://www.siecus.org>.

Ken R. Wells

Premature labor

Definition

Premature labor is the term to describe contractions of the uterus that begin at weeks 20–36 of a pregnancy.

Description

The usual length of a human pregnancy is 38–42 weeks after the first day of the last menstrual period. Labor is a natural series of events that indicate that the birth process is starting. Premature labor is defined as contractions that occur after 20 weeks and before 37 weeks during the term of pregnancy. The baby is more likely to survive and be healthy if it remains in the uterus for the full term of the pregnancy. It is estimated that around 10% of births in the United States occur during the premature period. Premature birth is the greatest cause of newborn illness and **death**. In the United States, prematurity has a greater impact on African Americans.

Causes and symptoms

The causes of premature labor cannot always be determined. Some research suggests that infection of the urinary or reproductive tract may stimulate

premature labor and premature births. Multiple pregnancies (twins, triplets, etc.) are more likely to result in premature labor. **Smoking**, alcohol use, drug **abuse**, and poor **nutrition** can increase the risk of premature labor and birth. Adolescent mothers are also at higher risk for premature delivery. Women whose mothers took diethylstilbestrol (DES) when they carried them are more likely to deliver prematurely, as are women who have had previous surgery on the cervix.

The symptoms of premature labor can include contractions of the uterus or tightening of the abdomen, which occurs every ten minutes or more frequently. These contractions usually increase in frequency, duration, and intensity, and may or may not be painful. Other symptoms associated with premature labor can include menstrual-like cramps, abdominal cramping with or without **diarrhea**, pressure or pain in the pelvic region, low backache, or a change in the color or amount of vaginal discharge. As labor progresses, the cervix or opening of the uterus will open (dilate) and the tissue around it will become thinner (efface). **Premature rupture of membranes** (when the water breaks) may also occur.

An occasional contraction can occur anytime during the pregnancy and does not necessarily indicate that labor is starting. Premature contractions are sometimes confused with Braxton Hicks contractions, which can occur throughout the pregnancy. Braxton Hicks contractions do not cause the cervix to open or efface, and are considered “false labor.”

Diagnosis

The health care provider will conduct a **physical examination** and ask about the timing and intensity of the contractions. A vaginal examination is the only way to determine if the cervix has started to dilate or efface. Urine and blood samples may be collected to screen for infection. A vaginal culture (a cotton-tipped swab is used to collect some fluid and cells from the vagina) may be done to look for a vaginal infection. A fetal heart monitor may be placed on the mother’s abdomen to record the heartbeat of the fetus and to time the contractions. A fetal ultrasound may be performed to determine the age and weight of the fetus, the condition of the placenta, and to see if there is more than one fetus present. **Amniocentesis** will sometimes be performed. This is a procedure where a needle-like tube is inserted through the mother’s abdomen to draw out some of the fluid surrounding the fetus. Analysis of the amniotic fluid can determine if the baby’s lungs are mature. A baby with mature lungs is much more likely to survive outside the uterus.

KEY TERMS

Braxton Hicks contractions—Tightening of the uterus or abdomen that can occur throughout pregnancy. These contractions do not cause changes to the cervix and are sometimes called false labor or practice contractions.

Cervix—The opening at the bottom of the uterus, which dilates or opens in order for the fetus to pass into the vagina or birth canal during the delivery process.

Contraction—A tightening of the uterus during pregnancy. Contractions may or may not be painful and may or may not indicate labor.

Treatment

The goal of treatment is to stop the premature labor and prevent the fetus from being delivered before it is full term. A first recommendation may be for the woman with premature contractions to lie down with feet elevated and to drink juice or other fluids. If contractions continue or increase, medical attention should be sought. In addition to bed rest, medical care may include intravenous fluids. Sometimes, this extra fluid is enough to stop contractions. In some cases, oral or injectable drugs like terbutaline sulfate, ritodrine, magnesium sulfate, or nifedipine must be given to stop the contractions. These are generally very effective; however, as with any drug therapy, there are risks of side effects. Some women may need to continue on medication for the duration of the pregnancy. **Antibiotics** may be prescribed if a vaginal or **urinary tract infection** is detected. If the membranes have already ruptured, it may be difficult or impossible to stop premature labor. If infection of the membranes that cover the fetus (chorioamnionitis) develops, the baby must be delivered.

Prognosis

If premature labor is managed successfully, the pregnancy may continue normally for the delivery of a healthy infant. Once symptoms of preterm labor occur during the pregnancy, the mother and fetus need to be monitored regularly since it is likely that premature labor will occur again. If the preterm labor cannot be stopped or controlled, the infant will be delivered prematurely. Infants that are born prematurely have an increased risk of health problems including **birth defects**, lung problems, **mental retardation**, blindness, deafness, and developmental disabilities. If the infant is

born too early, its body systems may not be mature enough for it to survive. Evaluating the infant's lung maturity is one of the keys to determining the baby's chance of survival. Fetuses delivered further into pregnancy and those with more mature lungs are more likely to survive.

Prevention

Smoking, poor nutrition, and drug or alcohol abuse can increase the risk of premature labor and early delivery. Smoking and drug or alcohol use should be stopped. A healthy diet and prenatal vitamin supplements (prescribed by the health care provider) are important for the growth of the fetus and the health of the mother. Pregnant women are advised to see a health care provider early in the pregnancy and receive regular prenatal examinations throughout the pregnancy. The health care provider should be informed of any medications that the mother is receiving and any health conditions that exist before and during the pregnancy.

Resources

OTHER

"Am I in Labor?" *The Virtual Hospital Page*. University of Iowa. <http://www.vh.org>.

ORGANIZATIONS

March of Dimes Birth Defects Foundation, 1275 Mamaroneck Ave., White Plains, NY, 10605, (914) 997-4488, <http://www.modimes.org>.

Altha Roberts Edgren

Premature menopause

Definition

The average age at which American women go through **menopause** is 51 years. If menopause (hormonal changes at the end of the female reproductive years) occurs before age 40, it is said to be premature menopause. Possible causes include autoimmune problems and common **cancer** treatments.

Description

About half of all women will go through menopause before age 51 and the rest will go through it after. Most women will finish menopause between the ages of 42 and 58. A small number of women will find that their periods stop prematurely, before age 40.

Causes and symptoms

There are many possible causes of premature menopause. Women who have premature menopause often have **autoimmune disorders** like thyroid disease or **diabetes mellitus**. In these diseases, the body produces antibodies to one or more of its own organs. These antibodies interfere with the normal function of the organ. Just as antibodies might attack the thyroid or the pancreas (causing thyroid disease or diabetes), antibodies may attack the ovaries and stop the production of female hormones.

Cancer treatments like **chemotherapy** or radiation can cause premature menopause. The risk depends on the type and length of treatment and the age of the woman when she first begins radiation or chemotherapy.

If the ovaries are surgically removed (during a **hysterectomy**, for example) menopause will occur within a few days, no matter how old the woman is.

The symptoms of premature menopause are similar to those of regular menopause. Menstrual periods stop and women may notice hot flashes, vaginal dryness, mood swings, and sleep problems. Sometimes the first symptom of premature menopause is **infertility**. A woman may find that she cannot become pregnant because she is not ovulating (producing eggs) anymore.

When menopause occurs after the ovaries are surgically removed, the symptoms begin within several days after surgery and tend to be more severe. This happens because the drop in the level of estrogen is dramatic, unlike the gradual drop that usually occurs.

Diagnosis

Premature menopause can be confirmed by blood tests to measure the levels of follicle-stimulating hormone (FSH) and luteinizing hormone (LH). The levels of these hormones will be higher if menopause has occurred.

Because premature menopause is often associated with other hormonal problems, women who have premature menopause should be screened for diabetes, thyroid disease, and similar diseases.

Treatment

There is no treatment to reverse premature menopause. **Hormone replacement therapy** (HRT) can prevent the common symptoms of menopause and lower the long-term risk of **osteoporosis**. Women who have premature menopause should take HRT. Estrogen relieves the unpleasant symptoms of menopause, including the hot flashes and the vaginal dryness. Estrogen is especially important for women who go through

KEY TERMS

Autoimmune diseases—Diseases in which the body creates antibodies that attack one of its own organs.

Follicle-stimulating hormone (FSH)—A female hormone that regulates ovulation and menstruation.

Hormone replacement therapy (HRT)—Replacement of estrogen and progesterone lost by women who have gone through menopause. Hormone replacement therapy has been shown to lower the risk of osteoporosis and heart disease in elderly women.

Luteinizing hormone (LH)—A female hormone that regulates ovulation and menstruation.

Menopause—The end of a woman's reproductive years. The hormonal changes that accompany menopause include hot flashes, vaginal dryness, mood swings, sleep problems, and the end of menstrual periods.

premature menopause. The long-term health risks of menopause (osteoporosis and increased risk of heart disease) are even more likely to occur after premature menopause. However, women who have certain medical conditions (like **liver disease**, uterine cancer, or **breast cancer**) may not be candidates for estrogen.

If a woman still has her uterus after premature menopause, she will also need to take progesterone along with the estrogen. If her uterus has been removed, estrogen alone will be enough.

Women who wish to become pregnant after premature menopause now have the option of fertility treatments using donor eggs. This is similar to **in vitro fertilization**, but the eggs come from a donor instead of the woman who is trying to become pregnant.

Prevention

Premature menopause cannot be prevented.

Resources

BOOKS

Hackley, Barbara, Jan M. Kribs, and Mary Ellen Rousseau. *Primary Care of Women: A Guide for Midwives and Women's Health Providers*. Sudbury, MA: Jones and Bartlett Publishers, 2007.

Amy B. Tuteur, MD

Premature rupture of membranes

Definition

Premature rupture of membranes (PROM) is an event that occurs during **pregnancy** when the sac containing the developing baby (fetus) and the amniotic fluid bursts or develops a hole prior to the start of labor.

Description

During pregnancy, the unborn baby (fetus) is surrounded and cushioned by a liquid called amniotic fluid. This fluid, along with the fetus and the placenta, is enclosed within a sac called the amniotic membrane. The amniotic fluid is important for several reasons. It cushions and protects the fetus, allowing the fetus to move freely. The amniotic fluid also allows the umbilical cord to float, preventing it from being compressed and cutting off the fetus's supply of oxygen and nutrients. The amniotic membrane contains the amniotic fluid and protects the fetal environment from the outside world. This barrier protects the fetus from organisms (like bacteria or viruses) that could travel up the vagina and potentially cause infection.

Although the fetus is almost always mature at 36–40 weeks and can be born during that period without complication, a normal pregnancy lasts an average of 40 weeks. At the end of 40 weeks, the pregnancy is referred to as being "term." At term, labor usually begins. During labor, the muscles of the uterus contract repeatedly. This allows the cervix to begin to grow thinner (called effacement) and more open (dilatation). Eventually, the cervix will become completely effaced and dilated. In the most common sequence of events (about 90% of all deliveries), the amniotic membrane breaks (ruptures) around this time. The baby then leaves the uterus and enters the birth canal. Ultimately, the baby will be delivered out of the mother's vagina. In the 30 minutes after the birth of the baby, the placenta should separate from the wall of the uterus and be delivered out of the vagina.

Sometimes the membranes burst before the start of labor, and this is called premature rupture of membranes (PROM). There are two types of PROM. One occurs at a point in pregnancy before normal labor and delivery should take place. This is called preterm

PROM. The other type of PROM occurs at 36–40 weeks of pregnancy.

PROM occurs in about 10% of all pregnancies. Only about 20% of these cases are preterm PROM. Preterm PROM is responsible for about 34% of all premature births.

Causes and symptoms

The causes of PROM have not been clearly identified. Some risk factors include **smoking**, multiple pregnancies (twins, triplets, etc.), and excess amniotic fluid (**polyhydramnios**). Certain procedures carry an increased risk of PROM, including **amniocentesis** (a diagnostic test involving extraction and examination of amniotic fluid) and cervical cerclage (a procedure in which the uterus is sewn shut to avoid **premature labor**). A condition called **placental abruption** is also associated with PROM, although it is not known which condition occurs first. In some cases of preterm PROM, it is believed that bacterial infection of the amniotic membrane causes it to weaken and then break. However, most cases of PROM and infection occur in the opposite order, with PROM occurring first followed by an infection.

The main symptom of PROM is fluid leaking from the vagina. It may be a sudden, large gush of fluid, or it may be a slow, constant trickle of fluid. The complications that may follow PROM include premature labor and delivery of the fetus, infections of the mother and/or the fetus, and compression of the umbilical cord (leading to oxygen deprivation in the fetus).

Labor almost always follows PROM, although the delay between PROM and the onset of labor varies. When PROM occurs at term, labor almost always begins within 24 hours. Earlier in pregnancy, labor can be delayed up to a week or more after PROM. The chance of infection increases as the time between PROM and labor increases. While this may cause doctors to encourage labor in the patient who has reached term, the risk of complications in a premature infant may cause doctors to try delaying labor and delivery in the case of preterm PROM.

The types of infections that can complicate PROM include amnionitis and endometritis. Amnionitis is an infection of the amniotic membrane. Endometritis is an infection of the innermost lining of the uterus. Amnionitis occurs in 0.5%–1% of all pregnancies. In the case of PROM at term, amnionitis complicates about 3%–15% of pregnancies. About 15%–23% of all cases of preterm PROM will be complicated

by amnionitis. The presence of amnionitis puts the fetus at great risk of developing an overwhelming infection (**sepsis**) circulating throughout its bloodstream. Preterm babies are the most susceptible to this life-threatening infection. One type of bacteria responsible for overwhelming infections in newborn babies is called group B streptococci.

Diagnosis

Depending on the amount of amniotic fluid leaking from the vagina, diagnosing PROM may be easy. Some doctors note that amniotic fluid has a very characteristic musty smell. A **pelvic exam** using a sterile medical instrument (speculum) may reveal a trickle of amniotic fluid leaving the cervix, or a pool of amniotic fluid collected behind the cervix. One of two easy tests can be performed to confirm that the liquid is amniotic fluid. A drop of the fluid can be placed on nitrazine paper. Nitrazine paper is made so that it turns from yellowish green to dark blue when it comes in contact with amniotic fluid. Another test involves smearing a little of the fluid on a slide, allowing it to dry, and then viewing it under a microscope. When viewed under the microscope, dried amniotic fluid will be easy to identify because it will look “feathery” like a fern.

Once PROM has been diagnosed, efforts are made to accurately determine the age of the fetus and the maturity of its lungs. Premature babies are at great risk if they have immature lungs. These evaluations can be made using amniocentesis and ultrasound measurements of the fetus’s size. Amniocentesis also allows the practitioner to check for infection. Other indications of infection include a **fever** in the mother, increased heart rate of the mother and/or the fetus, high **white blood cell count** in the mother, foul smelling or pus-filled discharge from the vagina, and a tender uterus.

Treatment

Treatment of PROM depends on the stage of the patient’s pregnancy. In PROM occurring at term, the mother and baby will be watched closely for the first 24 hours to see if labor will begin naturally. If no labor begins after 24 hours, most doctors will use medications to start labor. This is called inducing labor. Labor is induced to avoid a prolonged gap between PROM and delivery because of the increased risk of infection.

Preterm PROM presents more difficult treatment decisions. The younger the fetus, the more likely it may die or suffer serious permanent damage if

KEY TERMS

Amniocentesis—A medical procedure during which a long, thin needle is inserted through the abdominal and uterine walls, and into the amniotic sac. A sample of amniotic fluid is withdrawn through the needle for examination.

Amniotic fluid—The fluid within the amniotic sac; the fluid surrounds, cushions, and protects the fetus.

Amniotic membrane—The thin tissue that creates the walls of the amniotic sac.

Cervical cerclage—A procedure in which the cervix is sewn closed; used in cases when the cervix starts to dilate too early in a pregnancy to allow the birth of a healthy baby.

Placenta—The organ that provides oxygen and nutrition from the mother to the fetus during pregnancy. The placenta is attached to the wall of the uterus, and leads to the fetus via the umbilical cord.

delivered prematurely. Yet the risk of infection to the mother and/or the fetus increases as the length of time from PROM to delivery increases. Depending on the age of the fetus and signs of infection, the doctor must decide either to try to prevent labor and delivery until the fetus is more mature, or to induce labor and prepare to treat the complications of **prematurity**. However, the baby will need to be delivered to avoid serious risks to both it and the mother if infection is present, regardless of the risks of prematurity.

A variety of medications may be used in PROM:

- Medication to induce labor (oxytocin) may be used, either in the case of PROM occurring at term or in the case of preterm PROM and infection.
- Tocolytics may be given to halt or prevent the start of labor. These may be used in the case of preterm PROM, when there are no signs of infection. Delaying the start of labor may give the fetus time to develop more mature lungs.
- Steroids may be used to help the fetus's lungs mature early. Steroids may be given in preterm PROM if the fetus must be delivered early because of infection or labor that cannot be stopped.
- Antibiotics can be given to fight infections. Research is being done to determine whether antibiotics should be given prior to any symptoms of infection to avoid the development of infection.

Prognosis

The prognosis in PROM varies. It depends in large part on the maturity of the fetus and the development of infection.

Prevention

The only controllable factor associated with PROM is smoking. Cigarette smoking should always be discontinued during a pregnancy.

ORGANIZATIONS

American College of Obstetricians and Gynecologists (ACOG), PO Box 96920, Washington, DC, 20090-6920, (202) 638-5577, <http://www.acog.org>.

Rosalyne Carson-DeWitt, MD

Premature ventricular contractions see
Ventricular ectopic beats

Prematurity

Definition

The length of a normal **pregnancy** or gestation is considered to be 40 weeks (280 days) from the date of conception. Infants born before 37 weeks gestation are considered premature and may be at risk for complications.

Description

More than one out of every ten infants born in the United States is born prematurely. Advances in medical technology have made it possible for infants born as young as 23 weeks gestational age (17 weeks premature) to survive. These premature infants, however, are at higher risk for **death** or serious complications, which include heart defects, respiratory problems, blindness, and brain damage.

Causes and symptoms

The birth of a premature baby can be brought on by several different factors, including **premature labor**; **placental abruption**, in which the placenta detaches from the uterus; **placenta previa**, in which the placenta grows too low in the uterus; **premature rupture of membranes**, in which the amniotic sac is torn, causing the amniotic fluid to leak out; **incompetent cervix**, in which the opening to the uterus opens too soon; and maternal toxemia, or blood

poisoning. While one of these conditions is often the immediate reason for a premature birth, the underlying cause is usually unknown. Prematurity is much more common in **multiple pregnancy** and for mothers who have a history of miscarriages or who have given birth to a premature infant in the past. One of the few, and most important, identifiable causes of prematurity is drug **abuse**, particularly **cocaine**, by the mother.

Infants born prematurely may experience major complications due to their low birth weight and the immaturity of their body systems. Some of the common problems among premature infants are **jaundice** (yellow discoloration of the skin and whites of the eyes), apnea (a long pause in breathing), and inability to breast- or bottle-feed. Body temperature, blood pressure, and heart rate may be difficult to regulate in premature infants. The lungs, digestive system, and nervous system (including the brain) are underdeveloped in premature babies, and are particularly vulnerable to complications. Some of the more common risks and complications of prematurity are described below.

Respiratory distress syndrome (RDS) is the most common problem seen in premature infants. Babies born too soon have immature lungs that have not developed **surfactant**, a protective film that helps air sacs in the lungs stay open. With RDS, breathing is rapid and the center of the chest and rib cage pull inward with each breath. Extra oxygen can be supplied to the infant through tubes that fit into the nostrils of the nose, or by placing the baby under an oxygen hood. In more serious cases, the baby may have to have a breathing tube inserted and receive air from a respirator or ventilator. A surfactant drug can be given in some cases to coat the lung tissue. Extra oxygen may be needed for a few days or weeks, depending on how small and premature the baby was at birth. Bronchopulmonary dysplasia is the development of scar tissue in the lungs, and can occur in severe cases of RDS.

Necrotizing enterocolitis (NEC) is a further complication of prematurity. In this condition, part of the baby's intestines are destroyed as a result of bacterial infection. In cases where only the innermost lining of the bowel dies, the infant's body can regenerate it over time; however, if the full thickness of a portion dies, it must be removed surgically and an opening (ostomy) must be made for the passage of wastes until the infant is healthy enough for the remaining ends to be sewn together. Because NEC is potentially fatal, doctors are

quick to respond to its symptoms, which include lethargy, **vomiting**, a swollen and/or red abdomen, **fever**, and blood in the stool. Measures include taking the infant off mouth feedings and feeding him or her intravenously; administering **antibiotics**; and removing air and fluids from the digestive tract via a nasal tube. Approximately 70% of NEC cases can be successfully treated without surgery.

Intraventricular hemorrhage (IVH) is another serious complication of prematurity. It is a condition in which immature and fragile blood vessels within the brain burst and bleed into the hollow chambers (ventricles) normally reserved for cerebrospinal fluid and into the tissue surrounding them. Physicians grade the severity of IVH according to a scale of I–IV, with I being bleeding confined to a small area around the burst vessels and IV being an extensive collection of blood not only in the ventricles, but in the brain tissue itself. Grades I and II are not uncommon, and the baby's body usually reabsorbs the blood with no ill effects. However, more severe IVH can result in **hydrocephalus**, a potentially fatal condition in which too much fluid collects in the ventricles, exerting increased pressure on the brain and causing the baby's head to expand abnormally. To drain fluid and relieve pressure on the brain, doctors will either perform lumbar punctures, a procedure in which a needle is inserted into the spinal canal to drain fluids; install a reservoir, a tube that drains fluid from a ventricle and into an artificial chamber under or on top of the scalp; or install a **ventricular shunt**, a tube that drains fluid from the ventricles and into the abdomen, where it is reabsorbed by the body. Infants who are at high risk for IVH usually have an ultrasound taken of their brain in the first week after birth, followed by others if bleeding is detected. IVH cannot be prevented; however, close monitoring can ensure that procedures to reduce fluid in the brain are implemented quickly to minimize possible damage.

Apnea of prematurity is a condition in which the infant stops breathing for periods lasting up to 20 seconds. It is often associated with a slowing of the heart rate. The baby may become pale, or the skin color may change to a blue or purplish hue. Apnea occurs most commonly when the infant is asleep. Infants with serious apnea may need medications to stimulate breathing or oxygen through a tube inserted in the nose. Some infants may be placed on a ventilator or respirator with a breathing tube inserted into the airway. As the baby gets older, and the lungs and brain tissues mature, the breathing usually becomes more

regular. A group of researchers in Cleveland reported in 2003, however, that children who are born prematurely are 3–5 times more likely to develop sleep-disordered breathing by age 10 than children who were full-term babies.

As the fetus develops, it receives the oxygen it needs from the mother's blood system. Most of the blood in the infant's system bypasses the lungs. Once the baby is born, its own blood must start pumping through the lungs to get oxygen. Normally, this bypass duct closes within the first few hours or days after birth. If it does not close, the baby may have trouble getting enough oxygen on its own. **Patent ductus arteriosus** is a condition in which the duct that channels blood between two main arteries does not close after the baby is born. In some cases, a drug, indomethacin, can be given to close the duct. Surgery may be required if the duct does not close on its own as the baby develops.

Retinopathy of prematurity is a condition in which the blood vessels in the baby's eyes do not develop normally, which can, in some cases, result in blindness. Premature infants are also more susceptible to infections. They are born with fewer antibodies, which are necessary to fight off infections.

Diagnosis

Many of the problems associated with prematurity depend on how early the baby is born and how much it weighs at birth. The most accurate way of determining the gestational age of an infant in utero is calculating from a known date of conception or using ultrasound imaging to observe development. When a baby is born, doctors can use the Dubowitz exam to estimate gestational age. This standardized test scores responses to 33 specific neurological stimuli to estimate the infant's neural development. Once the baby's gestational age and weight are determined, further tests and **electronic fetal monitoring** may need to be used to diagnose problems or to track the baby's condition. A blood pressure monitor may be wrapped around the arm or leg. Several types of monitors can be taped to the skin. A heart monitor or cardiorespiratory monitor may be attached to the baby's chest, abdomen, arms, or legs with adhesive patches to monitor breathing and heart rate. A thermometer probe may be taped on the skin to monitor body temperature. Blood samples may be taken from a vein or artery. X rays or ultrasound imaging may be used to examine the heart, lungs, and other internal organs.

Treatment

Treatment depends on the types of complications that are present. It is not unusual for a premature infant to be placed in a heat-controlled unit (an incubator) to maintain its body temperature. Infants that are having trouble breathing on their own may need oxygen either pumped into the incubator, administered through small tubes placed in their nostrils, or through a respirator or ventilator, which pumps air into a breathing tube inserted into the airway. The infant may require fluids and nutrients to be administered through an intravenous line in which a small needle is inserted into a vein in the hand, foot, arm, leg, or scalp. If the baby needs drugs or medications, they may also be administered through the intravenous line. Another type of line may be inserted into the baby's umbilical cord. This can be used to draw blood samples or to administer medications or nutrients. If heart rate is irregular, the baby may have heart monitor leads taped to the chest. Many premature infants require time and support with breathing and feeding until they mature enough to breathe and eat unassisted. Depending on the complications, the baby may require drugs or surgery.

A form of treatment that is being recommended by many mainstream practitioners is **massage therapy**. Research has shown that the risks of massaging preterm infants are minimal, and that the infants benefit from improved developmental scores, more rapid weight gain, and earlier discharge from the hospital. An additional benefit of massage therapy is closer bonding between the parents and their newborn child.

Prognosis

Advances in medical care have made it possible for many premature infants to survive and develop normally. However, whether or not a premature infant will survive is still intimately tied to his or her gestational age:

- 21 weeks or less: 0% survival rate
- 22 weeks: 0%–10% survival rate
- 23 weeks: 10%–35% survival rate
- 24 weeks: 40%–70% survival rate
- 25 weeks: 50%–80% survival rate
- 26 weeks: 80%–90% survival rate
- 27 weeks: greater than 90% survival rate

Physicians cannot predict long-term complications of prematurity; some consequences may not become evident until the child is school-aged. Minor disabilities like learning problems, poor coordination,

KEY TERMS

Apnea—A long pause in breathing.

Dubowitz exam—A standardized test that scores responses to 33 specific neurological stimuli to estimate an infant's neural development and gestational age.

Intraventricular hemorrhage (IVH)—A condition in which blood vessels within the brain burst and bleed into the hollow chambers (ventricles) normally reserved for cerebrospinal fluid and into the tissue surrounding them.

Jaundice—Yellow discoloration of skin and whites of the eyes that results from excess bilirubin in the body's system.

Necrotizing enterocolitis (NEC)—A condition in which part of the intestines are destroyed as a result of bacterial infection.

Respiratory distress syndrome (RDS)—Condition in which a premature infant with immature lungs does not develop surfactant. RDS is the most common problem seen in premature infants.

Retinopathy of prematurity—A condition in which the blood vessels in a premature infant's eyes do not develop normally, which can, in some cases, result in blindness.

Surfactant—A protective film that helps air sacs in the lungs stay open. Premature infants may not have developed this protective layer before birth and are more susceptible to respiratory problems without it. Some surfactant drugs are available. These can be given through a respirator and will coat the lungs when the baby breathes the drug in.

or short attention span may be the result of premature birth, but can be overcome with early intervention. The risks of serious long-term complications depend on many factors, including how premature the infant was at birth, weight at birth, and the presence or absence of breathing problems. Gender is a definite factor: a Swedish study published in 2003 found that boys are at greater risk of death or serious long-term consequences of prematurity than girls; for example, 60% of boys born at 24 weeks' gestation die, compared to 38% mortality for girls. The development of infection or the presence of a birth defect can also affect long-term prognosis. Infections in premature and very low birth weight infants are a risk factor for later disorders of the nervous system; a study done at Johns Hopkins reported that 77 out of a group of 213 premature infants developed neurologic disorders. Severe disabilities like brain damage, blindness, and chronic lung problems are possible and may require ongoing care.

Prevention

Some of the risks and complications of premature delivery can be reduced if the mother receives good prenatal care, follows a healthy diet, avoids alcohol or drug consumption, and refrains from cigarette **smoking**. In some cases of premature labor, the mother may be placed on bed rest or given drugs that can stop labor contractions for days or weeks, giving the developing infant more time to develop before delivery. The physician may prescribe a steroid medication to be given to the mother before the delivery to help speed

up the baby's lung development. The availability of a neonatal intensive care unit, a special hospital unit equipped and trained to deal with premature infants, can also increase the chances of survival.

A new medication may help to prevent spontaneous premature births. Researchers at Wake Forest University reported in June 2003 that a drug known as 17 alpha-hydroxyprogesterone caproate not only reduced the number of premature births in a group of women who received weekly injections of the drug compared to a placebo group, but also lowered the rates of necrotizing enterocolitis, intraventricular hemorrhage, and need for supplemental oxygen in their infants. However, research is still ongoing.

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ORGANIZATIONS

American Academy of Pediatrics (AAP), 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007-1098, (847) 434-4000, (847) 424-8000, kidsdocs@aap.org, <http://www.aap.org>.

National Institute of Child Health and Human Development, Bldg 31, Room 2A32, MSC 2425, 31 Center Drive, Bethesda, MD, 20892-2425, (866) 760-5947, (800) 370-2943, <http://www.nichd.nih.gov/>.

Altha Roberts Edgren
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Premenstrual dysphoric disorder

Definition

Premenstrual syndrome (PMS) is a collection of physical and emotional symptoms that occurs 5 to 11 days before a woman's period begins and goes away once menstruation starts. The most severe form of PMS is premenstrual dysphoric disorder (PMDD).

Description

PMS is estimated to affect 70%–90% of women of childbearing age. The more severe form of the disorder, PMDD, affects 3%–5% of women of childbearing age. Up to 40% of women have PMDD symptoms that are so severe they interfere with their daily activities. It is more common in women in their late 20s and early 40s, who have at least one child and a history of depression, anxiety/tension, affective lability, or irritability/anger.

Causes and symptoms

Although the actual cause of PMDD is not known, it is believed to be related to hormonal

changes that occur before menstruation. There are more than 150 signs and symptoms attributed to PMDD, and every woman experiences different ones at different times. There seem to be socioeconomic and genetic factors that precipitate PMDD. Twin studies have demonstrated a positive correlation with heritability and PMDD symptoms. Antianxiety medications have been shown to help improve symptoms associated with PMDD. The most common symptoms include **headache**; swelling of ankles, feet, and hands; backache; abdominal cramps, heaviness or **pain**; bloating and/or gas; **muscle spasms**; breast tenderness; weight gain; recurrent **cold sores**; **acne**; **nausea**; **constipation** or **diarrhea**; food cravings; anxiety or panic; confusion; difficulty concentrating; forgetfulness; poor judgment; and depression.

Diagnosis

PMDD is diagnosed when symptoms occur during the second half of the menstrual cycle (14 days or more after the first day of a woman's period), are absent for about seven days after the period ends, increase in severity as the cycle progresses, go away when the menstrual flow begins or shortly thereafter, and occur for at least three consecutive menstrual cycles. There are no tests to diagnose it. The diagnosis of PMDD emphasizes and requires psychologically important mood symptoms.

Treatment

The first prescription drug approved by the U.S. Food and Drug Administration for the treatment of PMDD was Sarafem (fluoxetine). Other prescription drugs approved for the treatment of PMDD include Paxil CR (paroxetine), Zoloft (sertraline), and the oral contraceptive Yaz (a combination of drospirenone and ethinyl estradiol). **Nonsteroidal anti-inflammatory drugs**, such as ibuprofen and **aspirin**, may help with bloating and pain.

Alternative treatment

Non-pharmaceutical treatments include a variety of lifestyle changes, such as following a healthy diet, **exercise**, **stress** relief therapies, and even such alternative therapies as **aromatherapy**. Certain **vitamins** and supplements may also help, such as vitamin B6, **calcium**, magnesium, and vitamin E. Certain herbs may also help with symptom relief, including vitex, black cohosh, valerian, kava kava, and **St. John's wort**.

KEY TERMS

Antidepressant—A medication used to relieve the symptoms of clinical depression.

Beta blockers—Class of drug, including Corgard (nadolol) and Lanoxin (digoxin), that primarily works by blunting the action of adrenaline, the body's natural fight-or-flight chemical.

Nonsteroidal anti-inflammatory drugs—This class of drugs includes aspirin and ibuprofen, and primarily works by interfering with the formation of prostaglandins, enzymes implicated in pain and inflammation.

Prognosis

The prognosis varies for each woman, and is largely dependent on how much work she is willing to do in terms of lifestyle changes. Additionally, planning for PMDD symptoms, joining a support group, and communicating with family members can help minimize the negative effects of PMDD and its impact on a woman's home and work environments.

Prevention

Some women may find their PMDD disappears periodically. Diet and **nutritional supplements** can have the greatest impact in preventing PMDD.

ORGANIZATIONS

National Association for Premenstrual Syndrome, 41 Old Road East Peckham, Kent, England, TN12 5AP, 4408815-7311, <http://www.pms.org.uk>.

Society for Women's Health Research, 1025 Connecticut Ave. NW, Suite 701, Washington, DC, 20036, (202) 223-8224, (202) 833-3472, info@swhr.org, <http://www.womenshealthresearch.org>.

Description

Approximately 75% of all menstruating women experience some symptoms that occur before or during menstruation. PMS encompasses symptoms severe enough to interfere with daily life. About 3%–8% of women experience the more severe PMDD. These symptoms can last 4–10 days and can have a substantial impact on a woman's life.

The reason some women get severe PMS while others have none is not understood. PMS symptoms usually begin when a woman is in her 20s. The disease may run in families and is also more prone to occur in women with a history of psychological problems. Overall, however, it is difficult to predict who is most at risk for PMS.

Causes and symptoms

Because PMS is restricted to the second half of a woman's menstrual cycle (after ovulation), it is thought that hormones play a role. During a woman's monthly menstrual cycle, which lasts 24–35 days, hormone levels change. The hormone estrogen gradually rises during the first half of a woman's cycle (the preovulatory phase) and falls dramatically at ovulation. After ovulation (the postovulatory phase), progesterone levels gradually increase until menstruation occurs. Both estrogen and progesterone are secreted by the ovaries, which are responsible for producing the eggs. The main role of these hormones is to cause thickening of the lining of the uterus (endometrium). However, estrogen and progesterone also affect other parts of the body, including the brain. In the brain and nervous system, estrogen can affect the levels of neurotransmitters, such as serotonin. Serotonin has long been known to have an effect on emotions, as well as eating behavior. It is thought that when estrogen levels go down during the postovulatory phase of the menstrual cycle, decreases in serotonin levels follow. Whether these changes in estrogen, progesterone, and serotonin are responsible for the emotional aspects of PMS is not known with certainty. However, most researchers agree that the chemical transmission of signals in the brain and nervous system is in some way related to PMS. This is supported by the fact that the times following **childbirth** and **menopause** are also associated with both depression and low estrogen levels.

Symptoms for PMS are varied and many, including both physical and emotional aspects that range from mild to severe. The physical symptoms include bloating, **headache**, food cravings, abdominal cramps,

Premenstrual syndrome

Definition

Premenstrual syndrome (PMS) refers to a set of physical and psychological symptoms that occur between ovulation and the onset of menstruation. Severe forms of this syndrome are referred to as **premenstrual dysphoric disorder** (PMDD). These symptoms may be related to hormones and emotional disorders.

KEY TERMS

- Antidepressant**—A drug used to control depression.
- Estrogen**—A female hormone important in the menstrual cycle.
- Neurotransmitter**—A chemical messenger used to transmit an impulse from one nerve to the next.
- Phytoestrogens**—Compounds found in plants that can mimic the effects of estrogen in the body.
- Progesterone**—A female hormone important in the menstrual cycle.
- Serotonin**—A neurotransmitter important in regulating mood.

back **pain**, tension, and breast tenderness. Psychological or emotional aspects include mood swings, irritability, anxiety, and depression.

Diagnosis

The best way to diagnose PMS is to track a woman's symptoms for several months. PMS is diagnosed by the presence of physical, psychological, and behavioral symptoms that are cyclic and occur in association with the premenstrual period of time. PMDD, which is far less common, was officially recognized as a disease in 1987. Its diagnosis depends on the presence of at least five symptoms related to mood that disappear within the first few days of menstruation. These symptoms must interfere with normal functions and activities of the individual. The diagnosis of PMDD has caused controversy in fear that it may be used against women, labeling them as being impaired by their menstrual cycles.

Treatment

There are many treatments for PMS and PMDD depending on the symptoms and their severity. For mild cases, treatment includes **vitamins**, **diuretics**, and pain relievers. Vitamins E and B₆ may decrease breast tenderness and help with **fatigue** and mood swings in some women. Diuretics that remove excess fluid from the body seem to work for some women. For more severe cases and for PMDD, treatments available include **antidepressant drugs**, hormone treatment, or (only in extreme cases) surgery to remove the ovaries. Hormone treatment usually involves **oral contraceptives**. This treatment, as well as removal of the ovaries, is used to prevent ovulation and the changes in hormones that accompany ovulation. Recent studies,

however, indicate that hormone treatment has little effect over placebo.

Antidepressants

The most progress in the treatment of PMS and PMDD has been through the use of antidepressant drugs. The most effective of these include sertraline (Zoloft), fluoxetine (Prozac), and paroxetine (Paxil). They are termed **selective serotonin reuptake inhibitors** (SSRIs) and act by indirectly increasing the brain's serotonin levels, thus stabilizing emotions. Some doctors prescribe antidepressant treatment for PMS throughout the cycle, while others direct patients to take the drug only during the latter half of the cycle. Antidepressants should be avoided by women wanting to become pregnant. A recent clinical study found that women who took sertraline had a significant improvement in productivity, social activities, and relationships compared with a placebo group. Side effects of sertraline were found to include **nausea**, **diarrhea**, and decreased libido.

Alternative treatment

There are alternative treatments that can both affect serotonin and hormone responses, as well as some of the physical symptoms of PMS.

Vitamins and minerals

Some women find relief with the use of vitamin and mineral supplements. Magnesium can reduce the fluid retention that causes bloating, while **calcium** may decrease both irritability and bloating. Magnesium and calcium also help relax smooth muscles and this may reduce cramping. Vitamin E may reduce breast tenderness, nervous tension, fatigue, and **insomnia**. Vitamin B₆ may decrease fluid retention, fatigue, irritability, and mood swings. Vitamin B₅ supports the adrenal glands and may help reduce fatigue.

Phytoestrogens and natural progesterone

The Mexican wild yam (*Dioscorea villosa*) contains a substance that may be converted to progesterone in the body. Because this substance is readily absorbed through the skin, it can be found as an ingredient in many skin creams. (Some products also have natural progesterone added to them.) Some herbalists believe that these products can have a progesterone-like effect on the body and decrease some of the symptoms of PMS.

The most important way to alter hormone levels may be by eating more phytoestrogens. These plant-derived compounds have an effect similar to estrogen in the body. One of the richest sources of phytoestrogens is soy products, such as tofu. Additionally, many supplements can be found that contain black cohosh (*Cimicifugaracemosa*) or dong quai (*Angelica sinensis*), which are herbs high in phytoestrogens. Red clover (*Trifolium pratense*), alfalfa (*Medicago sativa*), licorice (*Glycyrrhiza glabra*), hops (*Humulus lupulus*), and legumes are also high in phytoestrogens. Increasing the consumption of phytoestrogens is also associated with decreased risks of **osteoporosis**, **cancer**, and heart disease.

Antidepressant alternatives

Many antidepressants act by increasing serotonin levels. An alternative means of achieving this is to eat more carbohydrates. For instance, two cups of cereal or a cup of pasta has enough carbohydrates to effectively increase serotonin levels. An herb known as **St. John's wort** (*Hypericum perforatum*) has stood up to scientific trials as an effective antidepressant. As with the standard antidepressants, however, it must be taken continuously and does not show an effect until used for 46 weeks. There are also herbs, such as skullcap (*Scutellaria lateriflora*) and kava (*Piper methysticum*), that can relieve the anxiety and irritability that often accompany depression. An advantage of these herbs is that they can be taken when symptoms occur rather than continually. Chaste tree (*Vitex agnus-castus*), in addition to helping rebalance estrogen and progesterone in the body, also may relieve the anxiety and depression associated with PMS.

Prognosis

The prognosis for women with both PMS and PMDD is good. Most women who are treated for these disorders do well.

Prevention

Maintaining a good diet, one low in sugars and fats and high in phytoestrogens and complex carbohydrates, may prevent some of the symptoms of PMS. Women should try to **exercise** three times a week and keep in generally good health. Avoidance of **caffeine** and/or alcohol may help some women. Because PMS is often associated with **stress**, avoidance of stress or developing better means to deal with stress can be important.

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ORGANIZATIONS

- Advancement of Women's Health Research, 1828 L Street, N.W., Suite 625, Washington, DC, 20036, (202) 223-8224, <http://www.womens-health.org>.
- National Association for Premenstrual Syndrome, 7 Swift's Court, High Street, Seal, Kent, England, TN15 0EG, + 4401732 760011, <http://www.PMDD.org.uk>.

Cindy L. A. Jones, PhD

Prenatal surgery

Definition

Prenatal surgery is a surgical procedure performed on a fetus prior to birth.

Purpose

In most cases prenatal surgery is performed only when the fetus is not expected to survive delivery or live long after birth without prenatal intervention. The most common prenatal surgeries are for conditions in which the newborn will not be able to breathe on its own.

Most prenatal surgeries are performed for:

- Urinary tract obstructions in males, usually caused by a narrowing of the urinary tract, in which urine backs up and injures the kidneys. About 10% of fetal urinary tract obstructions require prenatal surgery to prevent multiple abnormalities and depleted amniotic fluid.
- Congenital diaphragmatic hernia (CDH), a condition in which the diaphragm—the muscle that separates the chest and the abdomen—does not form completely. Without surgery about 50% of fetuses with CDH do not survive after birth because of underdeveloped lungs.
- Congenital cystic adenomatoid malformation (CCAM), a condition in which one or more lobes

of the lungs become fluid-filled sacs called cysts. Large CCAMs may prevent lung development, cause heart failure, or prevent the fetus from ingesting amniotic fluid.

- Sacrococcygeal teratoma (SCT), tumors at the base of the tailbone. The most common tumor in newborns, occurring in one out of every 35,000–40,000 births, some prenatal SCTs are very large, hard, and full of blood vessels, and can stress the heart.
- Twin-to-twin transfusion syndrome (TTTS), a condition in which, because of abnormal blood-vessel connections in the placenta, one twin pumps the circulating blood for both twins. Affecting up to 15% of twins sharing a placenta (monochorionic), TTTS can lead to a variety of problems including heart failure.
- Twin reversed arterial perfusion (TRAP) sequence, a condition in which one twin lacks a heart. Occurring in about 1% of monochorionic twins, the healthy twin pumps all of the blood and, if untreated, 50–75% of these normal twins die.

Other conditions that may be treated by prenatal surgery include:

- various congenital defects that block air passages and will prevent the newborn from breathing on its own
- various lung malformations
- omphalocele, a birth defect in which portions of the stomach, liver, and intestines protrude through an opening in the abdominal wall
- fetal gastroschisis—a birth defect in which the stomach and intestines protrude through improperly formed abdominal wall muscles and float in the amniotic fluid
- bowel obstructions, usually caused by a narrowing in the small intestine
- hypoplastic left heart syndrome, in which the blood flow through the left side of the heart is obstructed
- X-linked severe combined immunodeficiency syndrome
- spina bifida (myelomeningocele)—the second most common birth defect in the United States, affecting one out of every 2,000 newborns. It is a lesion or hole where the nerves of the spinal cord are not completely enclosed and is not considered to be life-threatening.

Precautions

Prenatal surgery involves:

- serious risks for the mother and fetus
- travel to a hospital that performs the procedure

- possibly having to stay near the hospital until delivery
- extended postoperative bed rest, sometimes until delivery
- a significant financial commitment

Description

Prenatal surgery may be referred to as fetal surgery, antenatal surgery, or maternal-fetal surgery. There are only about 600 candidates for prenatal surgery in the United States each year. Of these, only about 10% actually undergo the procedure. Most prenatal surgeries are performed between 18 and 26 weeks of gestation. Some surgeries may not be covered by insurance.

Prenatal surgery usually requires a general anesthetic, although sometimes an epidural anesthetic to numb the abdominal region may be used. The fetus receives the anesthetic via the mother's blood. An anesthesiologist and a perinatologist monitor the heart rates of the mother and fetus during the procedure.

Prenatal surgeries include:

- inserting a device into the fetal bladder to drain urine into the amniotic sac for treating urinary tract obstruction
- draining or removing CCAMs
- destroying blood vessels leading to a large SCT
- amniotherapy for TTTS, in which a syringe through the mother's abdomen is used to remove fluid from the overfilled amniotic sac and replace it in the depleted sac of the twin pumping the blood
- destroying abnormal blood vessel connections in the placenta of TTTS twins
- severing the connections between TRAP sequence twins
- experimental hematopoietic-stem-cell transplants for X-linked severe combined immunodeficiency syndrome
- closing the lesion in spina bifida

Open surgeries

In open prenatal surgeries incisions are made through the mother's abdominal wall and the fetus is partially removed from the uterus or the entire uterus is removed through the mother's abdomen. Using ultrasound as a guide, the surgeon feels for the affected fetal part. The surgeon may knead and push on the uterus to move or flip the fetus away from the placenta, the disk-shaped organ within the uterus that

supplies the fetal blood. A narrow tube is placed through a tiny hole in the uterine wall to drain and collect the amniotic fluid. Opening the uterus is the riskiest part of prenatal surgery. The first incision is made at a point away from the placenta to prevent damaging it. Following the procedure the fetus is replaced in the uterus and the incision is stitched. Prior to the final stitch the amniotic fluid is re-injected into the uterus. The uterus is repositioned in the mother's body cavity and her abdominal wall is closed.

The first successful open fetal surgery was performed in 1981 for a urinary tract obstruction. The first successful open fetal surgery for CDH was performed in 1989.

Prenatal open surgery for CCAM requires opening the fetus's chest. If a large cyst does not have a hard component, procedures called thoracoamniotic shunting or catheter decompression may be used to drain it. Otherwise the surgeon must remove part or all of the cyst. The first successful resection (removal) of a CCAM from a fetal lung was performed in 1990. The first resectioning of a fetal SCT was performed in 1992.

In prenatal surgery for **spina bifida**, an incision the size of a small fist is made in the uterus. The surgeon loosens and lifts the tissues of the spinal-canal lesion and stitches them closed. Between 1997 and 2004, more than 200 open surgeries were performed for spina bifida. The surgery was available only as part of a prospective randomized clinical trial.

Less invasive procedures

For urinary tract obstructions, a needle may be used to insert a catheter through the mother's abdomen and uterus and into the fetal bladder where it drains the urine into the amniotic fluid. The catheter may have a wire mesh that expands in the bladder to prevent it from plugging up or dislodging.

The first successful fetoscopic temporary tracheal occlusion for CDH was performed in 1996. Small openings are made in the uterus and a tiny fiber-optic fetoscope is inserted to guide the operation. A needle-like instrument is used to place a balloon in the fetus's trachea to prevent lung fluid from escaping through the mouth, enabling the lungs to expand, grow, and push the abdominal organs out of the chest. The balloon is removed at birth.

Hypoplastic left heart syndrome is treated by passing a needle, guided by ultrasound, through the mother's abdominal wall, into the uterus, and into the fetal heart. A catheter is passed through the needle

across the fetus's aortic valve. A balloon is inflated, opening the valve and allowing blood to flow through the left side of the heart.

RADIOFREQUENCY ABLATION. Radiofrequency ablation (RFA) sometimes is used for SCT. Guided by ultrasound, a needle is inserted through the mother's abdomen and uterus and into the tumor. Radio-frequency waves sent through the needle destroy the blood supply to the tumor with heat. This slows the tumor's growth and may enable the fetus to survive until delivery. The first RFA of an SCT was performed in 1998.

TRAP sequence also may be treated by RFA. A 3-mm needle targets the exact point where the blood enters the twin without a heart. Using an echocardiographic device, RFA is applied until the blood vessels and surrounding tissue are destroyed and the blood flow is halted. This procedure has eliminated the need for open surgery to treat TRAP sequence.

LASER TREATMENT. If TTTS does not respond to amnioreduction, laser treatment to halt the abnormal blood circulation may be attempted. A thin fetoscope is inserted through the mother's abdominal and uterine walls and into the amniotic cavity of the recipient twin to examine the surface placental blood vessels. The abnormal blood vessel connections are located and eliminated with a laser beam. The first successful fetoscopic laser treatment for TTTS was performed in 1999.

EXIT. Ex utero intrapartum treatment (EXIT) is a surgery performed for a congenital defect that blocks a fetal airway. The fetus is removed from the womb by **cesarean section** but the umbilical cord is left intact so that the mother's placenta continues to sustain the fetus. After the air passage is cleared, the umbilical cord is cut and the newborn can breathe on its own. The EXIT procedure is used for various types of airway obstruction including CCAM.

Preparation

The decision to perform prenatal surgery is made on the basis of detailed ultrasound imaging of the fetus—including echocardiograms that use ultrasound to obtain images of the heart—as well as other diagnostic tools. Consultations include a perinatologist, a neonatologist, a pediatric surgeon, a clinical nurse specialist, and a social worker. Since additional congenital defects preclude prenatal surgery, **amniocentesis** or chorionic villi sampling (CVS) are used to check for chromosomal abnormalities in the fetus.

Prior to surgery the mother must:

- arrange for postoperative bed rest to prevent preterm labor
- prepare for the possibility of remaining near the hospital until delivery
- receive betamethasone, a steroid, in two intramuscular injections 12–24 hours apart to accelerate fetal lung maturation
- wear a fetal/uterine monitor

The mother usually receives medications called tocolytics to prevent contractions and labor during and after surgery. These include:

- terbutaline
- indocin suppositories before surgery and up to 48 hours after surgery
- magnesium sulfate for one to two days after surgery with careful monitoring
- nifedipine every four to six hours as the indocin is decreased, continuing until 37 weeks of gestation or delivery

Aftercare

In addition to usual postsurgical care, the mother:

- usually remains in the hospital for four to seven days
- lies on her side to help prevent contractions and ensure the best possible fetal circulation
- has a transparent dressing over the abdominal incision for fetal monitoring
- has continuous electronic fetal/uterine monitoring to check the fetal heart, the uterine response to tocolytics, and to watch for signs of preterm labor

After discharge from the hospital the mother is on modified bed rest, lying on her side, until 37 weeks of gestation. This increases blood flow to the fetus and reduces pressure on the cervix to help prevent uterine contractions. She sees a perinatologist once a week and has at least one ultrasound per week.

Risks

Most prenatal surgeries are high risk and may be considered experimental. The greatest risk is that the placenta will be nicked during surgery, causing blood hemorrhaging, uterine contractions, and birth of a premature infant who may not survive. Preterm labor is the most common complication of prenatal surgery. Fetoscopic surgeries are less dangerous and traumatic than open fetal surgery and reduce the risk of **premature labor**. Subsequent children of a mother who has undergone prenatal surgery usually are delivered by cesarean section because of uterine scarring.

Maternal risks

Risks to the mother include:

- extensive blood loss
- complications from general anesthesia
- side effects—potentially fatal—from medications to control premature labor
- rupture of the uterine incision
- infection of the wound or uterus
- psychological stress
- inability to have additional children
- death

Fetal risks

All fetuses that undergo surgery are born prematurely. Those born even six weeks early are at risk for walking and talking delays and learning disabilities. Infants born at 30 weeks of gestation or less are at risk for blindness, **cerebral palsy**, and brain hemorrhages.

About 25% of women undergoing prenatal surgery lose some amniotic fluid, often because of leakage at the uterine incision. Amniotic fluid is essential for lung development and protects the fetus from injury and infection. If all of the amniotic fluid is lost, the fetal lungs may not develop properly. Without the fluid cushion in which the fetus floats, the umbilical cord may be compressed, causing **death**.

Other risks to the fetus include:

- birth during surgery
- separation of the tissues surrounding the amniotic fluid sac and the uterus, causing early delivery or interference with blood flow to some fetal body part such as an arm or leg
- intrauterine infection requiring immediate birth of the fetus
- further damage to the spinal cord and nerves during surgery to treat spina bifida
- brain damage
- physical deformities
- death

Normal results

Although fetal surgeries heal without scarring, it is difficult to predict their outcome because relatively few have been performed. Results for specific conditions include:

- Fetal surgery for CDH lessens the severity of the condition so that the fetus usually survives delivery and lives long enough to undergo corrective surgery.

KEY TERMS

Amniocentesis—Withdrawal of amniotic fluid through the mother's abdominal wall, using a needle and syringe, to test for fetal disorders.

Amniotic fluid—The watery fluid within the amniotic sac that surrounds the fetus.

Cesarean section—C-section; incision through the abdominal and uterine walls to deliver a baby.

Chorion—The outermost membrane of the sac enclosing the fetus.

Chorionic villus sampling (CVS)—The removal of fetal cells from the chorion for the diagnosis of genetic disorders.

Congenital cystic adenomatoid malformation (CCAM)—A condition in which one or more lobes of the fetal lungs develop into fluid-filled sacs called cysts.

Congenital diaphragmatic hernia (CDH)—A condition in which the fetal diaphragm—the muscle dividing the chest and abdominal cavity—does not close completely.

Echocardiography—Ultrasonic examination of the heart.

Ex utero intrapartum treatment (EXIT)—A cesarean section in which the infant is removed from the uterus but the umbilical cord is not cut until after surgery; treats congenital defects that block an air passage.

Fetoscope—A fiber-optic instrument for viewing the fetus inside the uterus.

Monochorionic twins—Twins that share a single placenta.

Omphalocele—A congenital hernia in which a small portion of the fetal abdominal contents, covered by a membrane sac, protrudes into the base of the umbilical cord.

Placenta—The organ within the uterus that provides nourishment to the fetus.

Radiofrequency ablation (RFA)—A procedure in which radiofrequency waves are used to destroy blood vessels and tissues.

Sacrococcygeal teratoma (SCT)—A tumor occurring at the base of the fetus's tailbone.

Spina bifida—Myelomeningocele; a congenital defect in which the fetal backbone and spinal canal do not close completely, allowing the spinal cord and its surrounding membranes to protrude.

Tocolytic—A medication that inhibits uterine contractions.

Twin reversed arterial perfusion (TRAP) sequence—A condition in which one fetus lacks a heart and the other fetus pumps the blood for both.

Twin-to-twin transfusion syndrome (TTTS)—A condition in monochorionic twins in which there is a connection between the two circulatory systems so that the donor twin pumps the blood to the recipient twin without a return of blood to the donor.

Ultrasound—A procedure that uses high-frequency sound waves to image a fetus.

- Thoracoamniotic shunting for CCAM usually results in infant survival.
- The infant survival rate following prenatal removal of solid CCAMs is about 50%.
- RFA to slow the growth of a tumor usually enables the fetus to survive delivery, after which the tumor can be removed.
- The infant survival rate following prenatal treatment for TTTS is about 70%. Since TTTS is a progressive disorder, early intervention may prevent later complications.

Spina bifida arises during the first month of fetal development. Fluid leaking from the spinal cord and exposure of the cord to amniotic fluid causes damage throughout gestation. Lesions higher up in the spinal

cord can cause severe deformities, **paralysis**, and **mental retardation**. Prenatal surgery may reduce the abnormalities, although it does not cure the condition. Babies who survive prenatal surgery appear to be 33%–50% less likely to have **hydrocephalus**, a condition that requires surgically implanted tubes or shunts to remove fluid from the ventricles (cavities) of the brain. The surgery also appears to reverse hindbrain herniation, in which the back of the brain slips down into the spinal canal, causing breathing and swallowing problems and death in 15% of affected children. Children who had prenatal surgery to treat spina bifida appear to have better brain function than those who did not. However prenatal surgery does not prevent two of the most serious conditions associated with spina bifida: leg movement and bladder and bowel control.

The long-term prognosis for these children is not known.

Resources

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ORGANIZATIONS

Fetal Treatment Center, University of California at San Francisco Children's Hospital, 505 Parnassus Ave., San Francisco, CA, 94143, (888) 689-8273, referral. center@ucsfmedctr.org, <https://www.ucsfbenioffchildrens.org>.

Management of Myelomeningocele Study (MOMS), the GWU Biostatistics Center, 6110 Executive Blvd., Suite 750, Rockville, MD, 20852, (866) 458-4621, (866) 275-6667, MOMS@biostat.bsc.gwu.edu, <http://www.spinabifidamoms.com/english/index.html>.

Margaret Alic, Ph.D.

Preparing for surgery

Definition

Preparing for a planned surgery includes selecting a surgery center and surgeon to perform the procedure, scheduling the surgery, undergoing pre-surgical testing, meeting with health-care professionals and the surgical team, receiving education about the procedure, receiving and following all of the appropriate preoperative instructions, and signing a consent form.

Purpose

Preparing for surgery helps the patient understand what to expect before surgery and ensures the patient is physically and psychologically ready for the surgery.

Description

Most patients go to the surgery center or hospital the same day as the scheduled surgery; thus, many of the steps involved in preparing for surgery will take place from one to four weeks before the scheduled surgery. Many surgeries are performed on an outpatient basis, which means that the patient goes home the same day as the surgery.

Selecting a surgeon and surgery center

SURGEON. A surgeon, along with a multi-disciplinary team of surgical specialists, will perform the surgery. The surgeon should be board certified by the American Board of Surgery, as well as certified by the medical specialty board or boards related to the type of surgery performed. Certification from a medical specialty board means that the surgeon has completed an approved educational training program (including three to seven years of full-time training in an accredited residency program). Certification also includes examinations that assess the surgeon's knowledge, skills, and experience.

There are 24 certifying boards recognized by the American Board of Member Specialties (ABMS) and the American Medical Association (AMA). Most of the ABMS boards issue time-limited certificates, valid for six to 10 years. This requires physicians to become re-certified to maintain their board certification—a process that includes a credential review, continuing education in the specialty, and additional examinations. Even though board certification is not required for an individual physician to practice medicine, most hospitals require that a certain percentage of their staff be board certified.

The letters FACS (Fellow of the American College of Surgeons) after a surgeon's name are a further indication of a surgeon's qualifications. Those who become Fellows of the American College of Surgeons have passed a comprehensive evaluation of their surgical training and skills; they also have demonstrated their commitment to high standards of ethical conduct. This evaluation is conducted according to national standards that were

KEY TERMS

Case manager—A health-care professional who can provide assistance with a patient's needs beyond the hospital.

Discharge planner—A health-care professional who helps patients arrange for health and home care needs after they go home from the hospital.

Electrocardiogram (ECG, EKG)—A test that records the electrical activity of the heart using small electrode patches attached to the skin on the chest.

Infectious disease team—A team of physicians who help control the hospital environment to protect patients against harmful sources of infection.

Informed consent—An educational process between health-care providers and patients intended to instruct the patient about the nature and purpose of the procedure or treatment, the risks and benefits of the procedure, and alternatives, including the option of not proceeding with the test or treatment.

Inpatient surgery—Surgery that requires an overnight stay of one or more days in the hospital.

NPO—A term that means nothing by mouth. NPO refers to the time after which the patient is not allowed to eat or drink prior to a procedure or treatment.

Outpatient surgery—Also called same-day or ambulatory surgery. The patient arrives for surgery and returns home on the same day.

established to ensure that patients receive the best possible surgical care.

A surgeon's membership in professional societies is also an important consideration. Professional societies provide an independent forum for medical specialists to discuss issues of national interest and mutual concern. Examples of professional societies include the Society of Thoracic Surgeons (STS) and the American College of Physicians—American Society of Internal Medicine (ACP-ASIM).

To find information about a surgeon's qualifications, the patient can call a state or county medical association for assistance. A reference book is also available: *The Official ABMS Directory of Board Certified Medical Specialists*, which lists all surgeons who are certified by approved boards. This publication also

contains brief information about each surgeon's medical education and training, and it can be found in many libraries.

SURGERY CENTER. The surgeon will arrange for the procedure to be performed in a hospital where he or she has staff privileges. The patient should make sure the hospital has been accredited by the Joint Commission on Accreditation of Healthcare Organizations, a professionally sponsored program that stimulates a high quality of patient care in health-care facilities. Joint Commission accreditation means the hospital voluntarily sought accreditation and met national health and safety standards. There is also an accreditation option that is available for ambulatory surgery centers.

Selecting a surgery center that has a multi-disciplinary team of specialists is important. The surgery team should include surgeons, **infectious disease** specialists, pharmacologists, and advanced care registered nurses. Other surgical team members may include fellows and residents, clinical coordinators, physical therapists, respiratory therapists, registered dietitians, social workers, and financial counselors.

Choosing a surgery center with experience is important. Some questions to consider when choosing a surgery center or hospital include:

- How many surgeries are performed annually and what are the outcomes/survival rates of those surgeries?
- How do the surgery center's outcomes compare with the national average?
- Does the surgery center offer treatment for a patient's specific condition? How experienced is the staff in treating that condition?
- What is the center's success record in providing the specific medical treatment or procedure?
- Does the surgery center have experience treating patients the same age as the inquiring patient?
- Does the surgery center explain the patient's rights and responsibilities?
- Does the surgery center have a written description of its services and fees?
- How much does the patient's type of treatment cost at this surgery center?
- Is financial help available?
- Who will be responsible for the patient's specific care plan while he or she is in the hospital?
- If the center is far from the patient's home, will accommodations be provided for caregivers?

- What type of services are available during the patient's hospital stay?
- Will a discharge plan be developed before the patient goes home from the hospital?
- Does the hospital provide training to help the patient care for his or her condition at home?

Scheduling the surgery

Depending on the nature of the surgery, it may be scheduled within days or weeks after the surgery is determined to be the appropriate treatment option for the patient. The patient's surgery time may not be determined until the business day before the scheduled surgery. The patient may be instructed to call the surgical center to find out the time of the scheduled surgery.

The time the patient is told to report to the surgery center (arrival time) is not the time when the surgery will take place. Patients are told to arrive at the surgery center far enough in advance (usually about two hours prior to the scheduled surgery time) so they can be properly prepared for surgery. In some cases, the patient's surgery may need to be rescheduled if another patient requires emergency surgery at the patient's scheduled time.

The patient should ask the health-care providers if the scheduled surgery will be performed on an outpatient or inpatient basis. Outpatient means the patient goes home the same day as the surgery; inpatient means a hospital stay is required.

Presurgical testing

Presurgical testing, also called preoperative testing or surgical consultation, includes a review of the patient's medical history, a complete **physical examination**, a variety of tests, patient education, and meetings with the health-care team. The review of the patient's medical history includes an evaluation of the patient's previous and current medical conditions, surgeries and procedures, medications, and any other health conditions such as **allergies** that may impact the surgery. Presurgical testing is generally scheduled one week before the surgery.

The patient may find it helpful to bring along a family member or friend to the presurgical testing appointments. This caregiver can help the patient remember important details to prepare for surgery.

After attending the surgical consultation, the patient may desire to seek a second opinion to confirm the first doctor's treatment recommendations. The patient should check with his or her insurance

provider to determine if the second opinion consultation is covered.

Meeting with the surgical team

During the surgical consultation, the patient meets with the surgeon or a member of the surgeon's health-care team to discuss the surgery and other potential treatment options for the patient's medical condition. At some time before the surgery, the patient will meet with other health-care providers, including the anesthesiologist, nurse clinicians, and sometimes a dietitian, social worker, or **rehabilitation** specialist.

Patient education

The surgical team will ensure that the patient understands the potential benefits and risks of the procedure as well as what to expect before the procedure and during the recovery. Patient education may include one-on-one instruction from a health-care provider, educational sessions in a group setting, or self-guided learning videos or modules. Informative and instructional handouts are usually provided to explain specific presurgical requirements.

Some surgery centers offer services such as **guided imagery** and relaxation tapes, **massage therapy**, **aromatherapy**, or other complementary techniques to reduce a patient's level of **stress** and **anxiety** before a surgical procedure. Guided imagery is a form of focused relaxation that coaches the patient to visualize calm, peaceful images. Several research studies have proven that guided imagery can significantly reduce stress and anxiety before and after surgical and medical procedures and help the patient recover more rapidly. Guided imagery and relaxation tapes are available at many major bookstores and from some surgery centers. The patient may be able to listen to the tapes during the procedure, depending on the type of procedure being performed.

Preoperative instructions

Preoperative instructions include information about reserving blood products for surgery, taking or discontinuing medications before the surgery, eating and drinking before surgery, quitting **smoking**, limiting activities before surgery, and preparing items to bring to the hospital the day of surgery.

BLOOD TRANSFUSIONS AND BLOOD DONATION

Blood transfusions may be necessary during surgery. A blood **transfusion** is the delivery of whole blood or blood components to replace blood lost through trauma, surgery, or disease. About one in three hospitalized patients will require a blood transfusion. The

surgeon can provide an estimate of how much blood the patient's procedure may require.

To decrease the risk of infection and immunologic complications, some surgery centers offer a preoperative **blood donation** program. Autologous blood (from the patient) is the safest blood available for transfusion, since there is no risk of disease transmission. Methods of autologous donation or collection include:

- Intraoperative blood collection: the blood lost during surgery is processed, and the red blood cells are re-infused during or immediately after surgery.
- Preoperative donation: the patient donates blood once a week for one to three weeks before surgery. The blood is separated and the blood components needed are re-infused during surgery.
- Immediate preoperative hemodilution: the patient donates blood immediately before surgery to decrease the loss of red blood cells during surgery. Immediately after donating, the patient receives fluids to compensate for the amount of blood removed. Since the blood is diluted, fewer red blood cells are lost from bleeding during surgery.
- Postoperative blood collection: blood lost from the surgical site right after surgery is collected and re-infused after the surgical site has been closed.

The surgeon determines what type of blood collection process, if any, is appropriate.

MEDICATION GUIDELINES. Depending on the type of surgery scheduled, certain medications may be prescribed or restricted before the surgery. The health-care team will provide specific guidelines. If certain medications need to be restricted before surgery, the patient will receive a complete list of the medications (including prescription, over-the-counter, and herbal medications) to avoid taking before the scheduled surgery.

If the physician advises the patient to take prescribed medication within 12 hours before surgery, it should be taken with small sips of water.

The patient should not bring any medications to the hospital; all necessary medications, as ordered by the doctor, will be provided in the hospital.

EATING AND DRINKING BEFORE SURGERY. Before most surgeries, the patient is advised not to eat or drink anything after midnight the evening before the surgery. This includes no smoking and no gum chewing. The patient should not drink any alcoholic beverages for at least 24 hours before surgery, unless instructed otherwise. If the patient has diabetes or if the surgery is to be performed on a child, the patient

should ask the health-care team for specific guidelines about eating and drinking before surgery.

Smoking cessation

Patients who will undergo any surgical procedure are encouraged to quit smoking and stop using tobacco products at least two weeks before the procedure, and to make a commitment to be a nonsmoker after the procedure. Ideally, the patient should quit smoking at least eight weeks prior to surgery. Quitting smoking before surgery helps the patient recover more quickly from surgery. There are several smoking cessation programs available in the community. The patient should ask a health-care provider for more information if he or she needs help quitting smoking.

Activity before surgery

The patient should eat right, rest, and **exercise** as normal before surgery, unless given other instructions. The patient should try to get enough sleep to build up energy for the surgery. The health-care team may advise the patient to scrub the planned surgical site with a special disinfecting soap the evening before the surgery.

MAKING PLANS FOR HOME AND WORK. The patient should make arrangements ahead of time for someone to care for children and take care of any other necessary activities at home such as getting the mail or newspapers. The patient should inform family members about the scheduled surgery in advance, so they can provide help and support before, during, and after surgery.

The patient should ask the health-care team what supplies may be needed after surgery during recovery at home so these items can be purchased or rented ahead of time. Some supplies that may be needed include an adaptive chair for the toilet or bathtub, or supplies for changing the wound dressing at home. Ask the health care providers if home care assistance (in which a visiting nurse visits the home to provide medical care) will be needed after surgery.

Items to bring to the hospital

The patient should bring a list of current medications, allergies, and appropriate medical records upon admission to the surgery center. The patient should also bring a prepared list of questions to ask.

The patient should not bring valuables such as jewelry, credit cards, or other items. A small amount of cash (no more than \$20) may be packed to purchase items such as newspapers or magazines.

Women should not wear nail polish or makeup the day of surgery.

If a hospital stay is expected after surgery, the patient should only pack what is needed. Some essential items include a toothbrush, toothpaste, comb or brush, deodorant, razor, eyeglasses (if applicable), slippers, robe, pajamas, and one change of comfortable clothes to wear when going home. The patient should also bring a list of family members' names and phone numbers to contact in an emergency.

Transportation

The patient should arrange for transportation home, since the effects of anesthesia and other medications given before surgery make it unsafe to drive.

Preoperative preparation

Upon arriving at the hospital or surgery center, the patient will be required to complete paperwork and show an insurance identification card, if insured. An identification bracelet that includes the patient's name and doctor's name will be placed on the patient's wrist.

INFORMED CONSENT. The health-care provider will review the informed consent form and ask the patient to sign it. Informed consent is an educational process between health-care providers and patients. Before any procedure is performed, the patient is asked to sign a consent form. Before signing the form, the patient should understand the nature and purpose of the procedure or treatment, the risks and benefits of the procedure, and alternatives, including the option of not proceeding with the procedure. Signing the informed consent form indicates that the patient permits the surgery or procedure to be performed. During the discussion about the procedure, the health-care providers are available to answer the patient's questions about the consent form or procedure.

ADVANCED DIRECTIVES. The health-care provider will ask the patient if he or she has any advance directives to be included in the patient's file. Advance directives are legal documents that increase a patient's control over medical decisions. A patient may decide medical treatment in advance, in the event that he or she becomes physically or mentally unable to communicate his or her wishes. Advance directives either state what kind of treatment the patient wants to receive (living will), or authorize another person to make medical decisions for the patient when he or she is unable to do so (durable power of attorney). Advance directives are not required and may be changed or

canceled at any time. Any change should be written, signed and dated in accordance with state law, and copies should be given to the physician and to others who received original copies. Advance directives can be revoked either in writing or by destroying the document. Advance directives include do-not-resuscitate (DNR) orders. A DNR order indicates that a person—usually with a terminal illness or other serious medical condition—has decided not to have **cardiopulmonary resuscitation** (CPR) performed in the event that his or her heart or breathing stops.

TESTS AND PREOPERATIVE EVALUATION. Some routine tests will be performed, including blood pressure, temperature, pulse, and weight checks; blood tests; **urinalysis**; **chest x ray**; and electrocardiogram (ECG). A brief physical exam will be performed. In some cases, an enema may be required. The health-care team will ask several questions to evaluate the patient's condition and to complete the final preparations for surgery. The patient should inform the health-care team if he or she drinks alcohol on a daily basis so precautions can be taken to avoid complications during and after surgery.

FINAL SURGICAL PREPARATION. Preoperative preparation generally includes these steps:

- The patient changes into a hospital gown.
- The patient removes (as applicable) contact lenses and glasses, dentures, hearing aids, nail polish, and jewelry.
- The patient empties his or her bladder.
- The health-care providers clean and possibly shave the area on the body where the surgery will be performed.
- The patient may receive medication to aid relaxation.
- An intravenous catheter will be placed in a vein in the patient's arm to deliver fluids, medications, or blood during surgery.
- In some hospitals, the patient may wait in an area called a holding area until the operating room and surgical team are ready. Depending on the hospital's policy, one or two of the patient's family members may wait with the patient.
- The patient is taken to the operating room in a wheelchair or on a bed (also called a gurney) where monitors are placed to evaluate the patient's condition during surgery.
- Anesthesia is administered; the type of anesthesia administered will depend upon the procedure, the patient's general health, and medications.
- A catheter may be placed in the patient's bladder to drain urine.

- The patient's vital signs, including the blood oxygen level, electrical activity of the heart, blood pressure, pulse, temperature, breathing, mental status, and level of consciousness, are continuously monitored during and after the surgery.

Information for families

While the patient is in surgery, the family members wait in a designated waiting area. Some hospitals or surgery centers offer a pager to the patient's family so they can be contacted for updates about the progress of the surgery. It may be helpful for the patient to select a spokesperson from the family to communicate with the health-care providers. This may improve communication with the health-care providers as well as to other family members. The patient should also communicate his or her wishes regarding the spokesperson's telephone communications to other family members.

Educational classes may be available for family members to learn more about the patient's surgery and what to expect during the recovery.

When the surgery is complete, the surgeon usually contacts the family members to provide information about the surgery. If a problem or complication occurs during surgery, the family members are notified immediately.

Normal results

Patients who receive proper preparation for surgery, including physical and psychological preparation, experience less anxiety and are more likely to make a quicker recovery at home, with fewer complications. Patients who perceive their surgical and post-operative experiences as positive report that they had minimal pain and nausea, were relaxed, had confidence in the skills of their health-care team, felt they had some control over their care, and returned to their normal activities within the expected timeframe.

Resources

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- Lichtenberg, Maggie. *The Open Heart Companion: Preparation and Guidance for Open-Heart Surgery Recovery*. Sante Fe, NM: Open Heart Pub., 2006.

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- Callery, Peter. "Preparing Children for Surgery." *Pediatric Nursing* 17.3 (April 2005): 12–13.
- Larson, Heather. "Pre-Op Jitters: Preparing for Surgery." *Whole Life Times* (Jan 2002): 22–24.

Lucas, Brian. "Preparing Patients for Hip and Knee Replacement Surgery." *Nursing Standard* 22.2 (Sept 19, 2007): 50–58.

ORGANIZATIONS

- Agency for Health Care Policy and Research (AHCPR), Publications Clearinghouse, P.O. Box 8547, Silver Spring, MD, 20907, (800) 358-9295.
- American Association of Nurse Anesthetists (AANA), 222 South Prospect Avenue, Park Ridge, IL, 60068-4001, (847) 692-7050, <http://www.aana.com>.
- American Board of Surgery, 1617 John F. Kennedy Boulevard, Suite 860, Philadelphia, PA, 19103, (215) 568-4000, <http://www.absurgery.org>.
- American College of Surgeons, 633 N. Saint Clair Street, Chicago, IL, 60611-3211, (312) 202-5000, <http://www.facs.org>.
- American Society of Anesthesiologists (ASA), 520 North Northwest Highway, Park Ridge, IL, 60068-2573, (847) 825-5586, <http://www.asahq.org>.
- National Heart, Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 251-2222, <http://www.nhlbi.nih.gov>.

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Prepregnancy counseling

Definition

Prepregnancy counseling is advice supplied by an obstetrician, nurse, certified nurse-midwife, or **childbirth** educator about those steps a mother-to-be and father-to-be can take in preparation for pregnancy. Basically, it is a checklist for people to see if they are living lives that are most accommodating to having a healthy pregnancy. Prepregnancy counseling gives time for one to make changes before pregnancy.

Purpose

The purpose and goal of prepregnancy counseling is to help patients have full-term, healthy pregnancies and babies. The counseling and education are important because lifestyle habits such as **smoking** or alcohol usage can be hazardous to a developing fetus.

Precautions

Women who have diabetes should take special precautions before pregnancy. This counseling, usually provided by a team of professionals including a registered dietitian, diabetes educators, an

obstetrician, and others, helps to prevent early pregnancy loss and congenital malformations in infants of diabetic mothers.

Women who have a history of genetic disease can opt to have **genetic testing**. Prepregnancy counseling can include referrals to those specialists.

Women who are over 40 have higher **cesarean section** rates. They are also more likely than younger women to have conditions such as high blood pressure, and are more likely to have babies with genetic problems, such as **Down syndrome**.

Women who are considering pregnancy should avoid exposure to hazards such as chemicals, illicit drugs, alcohol, and smoking. They should reduce their **caffeine** intake and be careful not to let their body temperatures rise to dangerous levels.

Description

Prepregnancy counseling involves communicating important aspects about **nutrition**, medication use, and lifestyle months in advance of getting pregnant. Issues include diet, nutrition, **exercise**, smoking, alcohol, drugs, emotional health, and referral to **genetic counseling** if a patient knows of a history of inherited disease.

Preparation

The mother-to-be should stop using birth control pills to allow for at least two regular menstrual cycles before conception. This requires that she stop taking birth control pills several months before getting pregnant.

Other steps to prepare for pregnancy include:

Being at optimal weight. Women should not go on prepregnancy weight loss **diets** unless they are under the care of a physician; abrupt weight loss can affect the mother's menstrual cycle and reduce fertility.

Eating a balanced diet. This is achieved by taking a prenatal vitamin provided by a health care provider and focusing on nutrients that are important for a developing fetus. These include folate, or **folic acid**, which is important for the development of the baby's brain and spinal cord. Folate can be found in fortified cereals, citrus fruits, and green leafy vegetables. **Calcium** is important for baby and mother. It helps the baby's bones to develop normally and keeps the mother from suffering a calcium deficiency during pregnancy. Iron keeps the mother from developing anemia during pregnancy. Good sources of iron are green leafy vegetables, red meat, beans, and fortified cereal. Fiber helps mothers avoid **constipation**, a

KEY TERMS

Preeclampsia—Also called toxemia, preeclampsia is a condition during pregnancy that results in high blood pressure, swelling that doesn't go away, and large amounts of protein in the urine. Without treatment, it can progress to a dangerous condition called eclampsia, in which a woman goes into convulsions.

common occurrence during pregnancy. Good sources of fiber include beans, fruits, and vegetables.

Exercising on a regular basis. Exercise promotes general overall health.

Undergoing routine physical and dental exams. These include having a physical and breast examination and **Pap test**. Other tests might be recommended according to a woman's health and genetic history. They should also report any prescription drugs, over-the-counter medications, or natural **vitamins** and herbs they are taking. This is the time for a woman to make sure she is up to date on her immunizations. A dental exam, with x rays, can eliminate the need to have x rays while pregnant.

Getting psychological support. Mental support is also important in the prepregnancy stage. This can help a woman to relax and better prepare mentally and physically for what lies ahead.

Risks

About 10%–15% of couples in the United States experience **infertility**. When couples should seek medical evaluation and an infertility workup depends on their ages. Generally, it takes longer for older couples to conceive. Prepregnancy counseling might include a referral to a fertility specialist. While infertility is often treatable, treatment can be expensive, emotionally difficult, and time consuming. About 10% of the time, doctors cannot detect a reason for the infertility.

There always is the risk that a pregnancy goes awry or a baby is born with a medical condition, regardless of whether or not a person has had prepregnancy counseling.

Normal results

Prepregnancy counseling can provide guidelines for people so that they can maximize their chances to

have emotionally and physically healthy pregnancies and healthy babies.

Abnormal results

Many abnormal results, such as genetic conditions, **miscarriage**, **preeclampsia** (also known as toxemia), and preterm births, cannot be avoided even with prepregnancy counseling. Still, some abnormal results, such as miscarriages and preterm births, may occur when mothers and fathers lead unhealthy lifestyles despite their counseling.

Resources

OTHER

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Lisette Hilton

Presbyopia

Definition

The term presbyopia means "old eye" and is a vision condition involving the loss of the eye's ability to focus on close objects.

Description

Presbyopia is a condition that occurs as a part of normal **aging** and is not considered to be an eye disease. The process occurs gradually over a number of years. Symptoms are usually noticeable by age 40–45 and continue to develop until the process stabilizes some 10–20 years later. Presbyopia occurs without regard to other eye conditions.

Causes and symptoms

In the eye, the crystalline lens is located just behind the iris and the pupil. Tiny ciliary muscles pull and push the lens, adjusting its curvature and thereby adjusting the eye's focal power to bring objects into focus. As individuals age, the lens becomes less flexible and elastic, and the muscles become less powerful. Because these changes result in inadequate adjustment of the lens of the eye for

various distances, objects that are close will appear blurry. The major cause of presbyopia is loss of elasticity of the lens of the eye. Loss of ciliary muscle power, however, is also believed to contribute to the problem.

Symptoms of presbyopia result in the inability to focus on objects close at hand. As the lens hardens, it is unable to focus the rays of light that come from nearby objects. Individuals typically have difficulty reading small print, such as that in telephone directories and newspaper advertisements, and may need to hold reading materials at arm's length. Symptoms include **headache** and eyestrain when doing close work, blurry vision, and eye **fatigue**. Symptoms may be worse early in the morning or when individuals are fatigued. Dim lighting may also aggravate the problem.

Diagnosis

Presbyopia is officially diagnosed during an **eye examination** conducted by eye specialists, such as optometrists or ophthalmologists. After completing optometric college, doctors of optometry screen patients for eye problems and prescribe glasses and **contact lenses**. In contrast, ophthalmologists are medical doctors who specialize in eye diseases. They perform eye surgery, treat eye diseases, and also prescribe glasses and contact lenses.

A comprehensive eye examination requires at least 30 minutes. Part of the examination will assess vision while reading by using various strength lenses. If the pupils are dilated with drugs to permit a thorough examination of the retina, an additional hour is required. The cost of eye examinations can range from \$40 to \$250 depending on the complexity and site of the examination and the qualifications and reputation of the examiner. Some insurers cover the cost of routine eye examinations, while others do not. A thorough eye examination is recommended at regular intervals during the adult and aging years to monitor and diagnose eye conditions. However, individuals frequently self-diagnose presbyopia by trying on inexpensive mass-produced reading glasses until they find a pair that permits reading without strain.

Treatment

Presbyopia cannot be cured, but individuals can compensate for it by wearing reading, bifocal, or trifocal eyeglasses. A convex lens is used to make up for the lost automatic focusing power of the eye.

KEY TERMS

Accommodation—The ability of the eye to change its focus from near to distant objects.

Binocular vision—Using both eyes at the same time to see an image.

Ciliary muscles—The small muscles that permit the lens to change its shape in order to focus on near or distant objects.

Lens (or crystalline lens)—The eye structure behind the iris and pupil that helps focus light on the retina.

Visual acuity—Sharpness or clearness of vision.

Half-glasses can be worn, which leave the top open and uncorrected for distance vision. Bifocals achieve the same goal by allowing correction of other refractive errors (improper focusing of images on the retina of the eye).

In addition to glasses, contact lenses have also been found to be useful in the treatment of presbyopia. The two common types of contact lenses prescribed for this condition are bifocal and monovision contact lenses. Bifocal contact lenses are similar to bifocal glasses. The top portion of the lens serves as the distance lens while the lower serves as the near vision lens. To prevent rotation while in the eye, bifocal contacts use a specially manufactured type of lens. Good candidates for bifocal lenses are those patients who have a good tear film (moist eyes), good binocular vision (ability to focus both eyes together) and visual acuity in each eye, and no disease or abnormalities in the eyelids. The bifocal contact lens wearer must be motivated to invest the time it requires to maintain contact lenses and be involved in occupations that do not impose high visual demands. Further, bifocal contact lenses may limit binocular vision. Bifocal contact lenses are relatively expensive, in part due to the time it takes the patient to be accurately fitted.

An alternative to wearing eyeglasses or bifocal contact lenses is monovision contact lenses. Monovision fitting provides one contact lens that corrects for near vision and a second contact lens for the alternate eye that corrects for distance vision. If distance vision is normal, the individual wears only a single contact lens for near vision. Monovision works by having one eye focus for distant objects while the other eye becomes the reading eye. The brain learns to adapt to this and will automatically use the correct eye

depending on the location of material in view. Advantages of monovision are patient acceptability, convenience, and lower cost.

Several problems exist with the use of contact lenses in the treatment of presbyopia. Some individuals experience headache and fatigue during the adjustment period or find the slight decrease in visual acuity unacceptable. Monovision contact lenses usually result in a small reduction in high-contrast visual acuity when compared with bifocal contact lenses.

Prognosis

The changes in vision due to aging usually start in a person's early 40s and continue for several decades. At some point, there is no further development of presbyopia, as the ability to accommodate is virtually gone.

Prevention

There is no known way to prevent presbyopia.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, (415) 561-8500, <http://www.aoa.org>.

American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, (314) 991-4100, (314) 991-4101, (800) 365-2219, <http://www.aoa.org/>.

Lighthouse International, 111 East 59th Street, New York, NY, 10022-120, (212) 821-9200, (212) 821-9707, (800) 829-0500, info@lighthouse.org, <http://www.lighthouse.org>.

National Eye Institute, 2020 Vision Place, Bethesda, MD, (301) 496-5248, <http://www.nei.nih.gov/>.

Elaine Souder, PhD

Presenile dementia see **Alzheimer's disease**

Pressure sores see **Bedsores**

Preterm labor see **Premature labor**

Priapism

Definition

Priapism is a persistent, unwanted, and usually painful penile erection that is not caused by sexual stimulation or arousal and is not relieved by orgasm.

Demographics

Priapism is uncommon, with a worldwide overall incidence of 1.5 cases per 100,000 men each year, increasing to 2.9 cases per 100,000 in men over age 40. Priapism can occur in males of any age, from newborns to the elderly, but is most common in boys between the ages of 5 and 10 and in men aged 20–50.

In younger males, priapism is more often associated with **sickle cell disease** (SCD) and is a common problem among African American males with SCD. The incidence of priapism is as high as 27% in male children with SCD. Estimates of the occurrence of priapism in adult males with SCD range from 10%–89% and the incidence is highest in men aged 19–21. Among older men, priapism is more often associated with drugs. Between 0.05% and 6% of men treating **erectile dysfunction** with drugs experience priapism.

Description

During a normal erection, the spongy tissues in the penis fill with blood as the blood vessels relax and expand in response to physical or psychological stimulation. The blood is trapped in the penis by the contraction of blood vessels that drain the blood. Following sexual activity or withdrawal of stimulation, the blood drains from the penis and it becomes non-rigid or flaccid. Priapism occurs when the blood in the penile shaft does not drain properly after sexual stimulation and the shaft remains hard. Priapism persisting for more than four hours is a medical emergency. It can permanently damage the tissues of the penis and lead to erectile dysfunction (ED)—the inability to achieve or maintain a normal erection.

There are three types of priapism:

- Ischemic or low-flow priapism is the most common type. It occurs when the blood cannot exit the penis after an erection.
- Stuttering or recurring priapism is a form of ischemic priapism that occurs intermittently.
- Nonischemic or high-flow priapism results from too much blood flowing into the penis.

Risk factors

The major risk factors for priapism are sickle cell disease and drugs used to counteract erectile dysfunction. At least 25% of men who inject ED drugs for more than three months develop priapism.

Causes and symptoms

Priapism is caused by abnormalities in the blood, blood vessels, or nerves that interfere with normal blood flow into or out of the penis. Ischemic priapism

is most often caused by diseases of the blood such as SCD, leukemia, or **malaria** or by medications for treating ED. About two-thirds of all pediatric patients with priapism have SCD. The abnormally shaped red blood cells of SCD can clump, preventing them from flowing out of the penis. In developed countries, ED drugs are the most common cause of adult priapism.

Priapism—usually ischemic priapism—can be a side effect of a variety of drugs including:

- oral ED medications, such as sildenafil (Viagra) and vardenafil (Levitra)
- recreational use of ED drugs
- overdoses of vasodilators, such as papaverine, which are injected directly into the penis to induce an immediate erection in men with ED
- antidepressants, such as trazadone (Desyrel), fluoxetine (Prozac), and bupropion (Wellbutrin)
- antipsychotics, such as thorazine, risperidone (Risperdal), and olanzapine (Zyprexa)
- anti-anxiety medications, such as diazepam (Valium)
- blood thinners, such as warfarin (Coumadin) and heparin
- some blood pressure medications
- cocaine, marijuana, and ecstasy
- excessive alcohol consumption

Nonischemic priapism is usually the result of a ruptured artery or other injury or trauma to the genitals, perineum (the area between the scrotum and anus), or pelvis that interferes with normal blood circulation in the penis.

Other factors that can contribute to the occurrence of priapism include:

- spinal cord injury
- anesthesia
- nervous system diseases, such as multiple sclerosis
- metabolic diseases, such as diabetes
- blood clots
- poisonous venom from scorpions or black widow spiders
- carbon monoxide poisoning
- rarely, cancers affecting the penis

Sometimes the cause of priapism cannot be determined.

The major symptom of priapism is an unwanted erection lasting more than four hours that is not associated with sexual stimulation or that persists after stimulation is completed; however, **stuttering** priapism usually lasts less than three hours. Ischemic or stuttering priapism is usually painful. At the very

KEY TERMS

Antineoplastic—A drug used to inhibit the growth and spread of cancerous cells.

Doppler ultrasound—An imaging technique that can detect moving fluids.

Erectile dysfunction (ED)—The consistent inability to achieve or maintain a penile erection.

Infarction—Death of tissue due to inadequate blood supply.

Nuclear scanning—Use of injected radioactive elements to analyze blood flow.

Sickle cell disease (SCD)—A hereditary abnormality causing deformed red blood cells that can plug up small blood vessels.

least it causes penile tenderness. With these types of priapism the penile shaft is rigid but the tip (glans) is usually soft. Nonischemic priapism is usually painless and, although the penile shaft is erect, it is not rigid.

Diagnosis

Examination

An ischemic erection lasting more than four hours requires immediate emergency room treatment. The physician may be able to determine the type of priapism based on the rigidity and sensitivity of the penis. A persistent erection that resolves in less than four hours or recurring or stuttering priapism also requires diagnosis to prevent further episodes. A family physician or general practitioner may refer the patient to a urologist. Diagnosis includes a **physical examination** of the genitals, perineum, rectum, and abdomen. It also includes a medical and sexual history, a list of medications and other drug use, and symptoms. The physician will look for signs of injury or tumors that could be causing priapism.

Tests

Laboratory tests are used to determine causes underlying priapism:

- Laboratory blood gas measurements are performed on blood removed from the penis with a tiny needle. The visible appearance of the blood can indicate the type of priapism—dark blood indicates oxygen deprivation and ischemic priapism and bright red blood indicates nonischemic priapism.

- Red blood cell and platelet counts and other blood tests can indicate SCD, other blood disorders, or certain cancers.
- Toxicology tests on blood or urine samples can screen for illicit or prescription drug use.

Procedures

Nuclear scanning, Doppler ultrasound, or color duplex ultrasonography may be used to evaluate penile blood flow and distinguish between ischemic and nonischemic priapism. They may also reveal an injury, tumor, or other abnormality that may be an underlying cause of the priapism.

Treatment

Traditional

Aspiration is the emergency treatment for ischemic priapism that does not respond to the injection of medications. Under local anesthetic the excess blood is drained from the penis with a small needle and syringe. The penile veins may also be flushed with saline solution. This relieves **pain**, clears the tissues of dangerous oxygen-depleted blood, and may relieve the erection. However the procedure may have to be repeated frequently over several hours to completely end the erection. A urinary catheter may be inserted to drain the bladder. Patients with SCD also may receive supplemental oxygen, a blood **transfusion**, intravenous fluids, or other treatments for sickle-cell crises. Surgical procedures are used as a last resort for treating ischemic priapism. One procedure blocks much of the blood supply to the penis, enabling it to relax.

Nonischemic priapism often does not require treatment. If the priapism is caused by a ruptured artery, surgical ligation may be used to tie off the artery and restore normal blood flow. Sometimes surgery is used to insert material that temporarily blocks blood flow to the penis. Surgery may also be necessary to repair arteries or tissue damage resulting from an injury.

Both ischemic and nonischemic priapism are sometimes treated with a surgical shunt implanted in the penis to reroute the blood and restore normal circulation.

Drugs

Initial treatment of ischemic priapism usually involves the injection of an alpha-agonist or alpha-adrenergic sympathomimetic drug, such as phenylephrine, into the spongy tissue of the penis (intracavernous injection). This constricts blood vessels coming into the penis, limiting the inflow of blood and enabling the outgoing blood vessels to dilate and

the blood to flow out. Sometimes these injections must be repeated often over a period of several hours.

Home remedies

Placing ice and pressure on the perineum can sometimes help end an erection. Cold packs applied to the penis may alleviate nonischemic priapism.

Prognosis

If priapism is relieved within the first 12–24 hours, there is usually no residual damage to the penis. However, untreated priapism can last for several days. The blood that is trapped in the penis is deprived of oxygen and begins to damage or destroy tissues (infarction). This can lead to permanent ED or disfigurement of the penis. A recent study found that 92% of men with priapism lasting less than 24 hours retained erectile function, compared with only 22% of those with priapism lasting more than seven days.

Prevention

Measures for preventing priapism include:

- treatment of any underlying medical or substance abuse problems
- avoiding triggers such as alcohol or illicit drugs
- changing a prescription medication that may be causing priapism
- hormone treatment for adult men
- a prescription muscle relaxant, such as baclofen (Lioresal)
- self-injection of phenylephrine to halt a prolonged erection
- an antineoplastic drug called hydroxyurea for patients with sickle cell disease
- insertion of a penile prosthesis for permanent prevention of ischemic priapism

Resources

BOOKS

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ORGANIZATIONS

American Urological Association, 1000 Corporate Boulevard, Linthicum, MD, 21090, (410) 689-3700, (866) RING-AUA (746-4282), (410) 689-3800, aua@AUAAnet.org, <http://www.auanet.org>.

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Margaret Alic, PhD

Prickly heat

Definition

Also known as sweat retention syndrome or miliaria rubra, prickly heat is a common disorder of the sweat glands.

Description

The skin contains two types of glands: one produces oil and the other produces sweat. Sweat glands are coil-shaped and extend deep into the skin. They are capable of plugging up at several different depths, producing four distinct skin **rashes**.

- Miliaria crystallina is the most superficial of the occlusions. At this level, only the thin upper layer of skin is affected. Little blisters of sweat that cannot escape to the surface form. A bad sunburn as it just starts to blister can look exactly like this.
- Deeper plugging causes miliaria rubra as the sweat seeps into the living layers of skin, where it irritates and itches.
- Miliaria pustulosa (a complication of miliaria rubra) occurs when the sweat is infected with pyogenic bacteria and turns to pus.
- Deeper still is miliaria profunda. The skin is dry, and goose bumps may or may not appear.

There are two requirements for each of these phases of sweat retention: hot enough weather to

induce sweating, and failure of the sweat to reach the surface.

Causes and symptoms

Best evidence suggests that bacteria form the plugs in the sweat glands. These bacteria are probably normal inhabitants of the skin, and why they suddenly interfere with sweat flow is still not known.

Infants are more likely to get miliaria rubra than adults. All the sweat retention rashes are also more likely to occur in hot, humid weather.

Besides **itching**, these conditions prevent sweat from cooling the body, which it is supposed to do by evaporating from the skin surface. Sweating is the most important cooling mechanism available in hot environments. If it does not work effectively, the body can rapidly become too hot, with severe and even lethal consequences. Before entering this phase of heat **stroke**, there will be a period of heat exhaustion symptoms—dizziness, thirst, weakness—when the body is still effectively maintaining its temperature. Then the temperature rises, often rapidly, to 104°F (40°C) and beyond. This is an emergency of the first order, necessitating immediate and rapid cooling. The best method is immersion in ice water.

Diagnosis

Rash and dry skin in hot weather are usually sufficient to diagnose these conditions.

Treatment

The rash itself may be treated with topical anti-pruritics (itch relievers). Preparations containing aloe, menthol, camphor, eucalyptus oil, and similar ingredients are available commercially. Even more effective, particularly for widespread itching in hot weather, are cool baths mixed with corn starch and/or oatmeal (about 0.5 lb [224 g] of each).

Dermatologists can peel off the upper layers of skin using a special ultraviolet light. This will remove the plugs and restore sweating, but is not necessary in most cases.

The primary concern of prickly heat is that the body cannot cool itself adequately without sweating. Careful monitoring for symptoms of heat disease is important. If they appear, some decrease in the ambient temperature must be achieved by moving to the shade, taking a cool bath or shower, or turning up the air conditioner.

KEY TERMS

Ambient—Surrounding.

Pyogenic—Capable of generating pus. *Streptococcus*, *Staphylococcus*, and bowel bacteria are the primary pyogenic organisms.

Syndrome—A collection of abnormalities that occur together often enough to suggest they have a common cause.

Prognosis

The rash disappears in a day with cooler temperatures, but the skin may not recover its ability to sweat for two weeks—the time needed to replace the top layers of skin with new growth from below.

Prevention

Experimental application of topical **antiseptics** like hexachlorophene almost completely prevented these rashes.

Resources

BOOKS

McPhee, Stephen, and Maxine Papadakis. *Current Medical Diagnosis and Treatment*, 2010, 49th ed. New York: McGraw-Hill Medical, 2009.

J. Ricker Polsdorfer, MD

Primaquine see **Antimalarial drugs**

Primary biliary cirrhosis

Definition

Primary biliary **cirrhosis** is the gradual destruction of the biliary system for unknown reasons.

Description

Although the cause of this serious condition is not known, it has many features to suggest that it is an autoimmune disease. Autoimmunity describes the process whereby the body's defense mechanisms are turned against itself. The immune system is supposed to recognize and attack only dangerous foreign invaders like germs, but many times it attacks, for no apparent reason, the cells of the body itself.



A close-up image indicating biliary cirrhosis of the liver.
(Custom Medical Stock Photo, Inc. Reproduced by permission.)

Autoimmune reactions occur in many different tissues of the body, creating a great variety of diseases.

Primary biliary cirrhosis progressively destroys the system that drains bile from the liver into the intestines. Bile is a collection of waste products excreted by the liver. As the disease progresses it also scars the liver, leading to cirrhosis. In some patients, the disease destroys the liver in as little as five years. In others, it may lie dormant for a decade or more.

Causes and symptoms

Ninety percent of patients with this disease are women between the ages of 35 and 60. The first sign of primary biliary cirrhosis may be an abnormal blood test on routine examination. **Itching** is a common early symptom, caused by a buildup of bile in the skin. **Fatigue** is also common in the early stages of the disease. Later symptoms include **jaundice** from the accumulation of bile and signs of specific nutritional deficiencies—bruising from **vitamin K deficiency**, bone pain from **vitamin D deficiency**, night blindness from **vitamin A deficiency**, and skin **rashes**, possibly from vitamin E or essential fatty acid deficiency. All these vitamin problems are related to the absence of bile to assist in the absorption of nutrients from the intestines.

Diagnosis

Blood tests strongly suggest the correct diagnosis, but a **liver biopsy** is needed for confirmation. It is also usually necessary to x ray the biliary system to look for other causes of obstruction.

KEY TERMS

Biopsy—Surgical removal of tissue for examination.

Cirrhosis—Scarring, usually referring to the liver.

Immunosuppression—Techniques to prevent transplant graft rejection by the body's immune system.

Treatment

Of the many medicines tried to relieve the symptoms and slow the progress of this disease, only one has had consistently positive results. Ursodeoxycholic acid, a chemical that dissolves gallstones, provides substantial symptomatic relief. It is still unclear if it slows liver damage.

Primary biliary cirrhosis is a major reason for **liver transplantation**. Patients do so well that this is becoming the treatment of choice. As experience, technique, and immunosuppression progressively improve, patients with this disease will come to **transplant surgery** earlier and earlier in their disease course.

Prognosis

So far, this disease has not returned in a transplanted liver.

ORGANIZATIONS

American Liver Foundation, 75 Maiden Lane, Suite 603, New York, NY, 10038, (212) 668-1000, (212) 483-8179, <http://www.liverfoundation.org/>.

J. Ricker Polsdorfer, MD

Primary degenerative dementia see
Alzheimer's disease

Primary polycythemia see **Polycythemia vera**

Primary pulmonary hypertension see
Pulmonary hypertension

PRK see **Photorefractive keratectomy and laser-assisted in-situ keratomileusis**

Pro time see **Prothrombin time**

Probenecid see **Gout drugs**

Procainamide see **Antiarrhythmic drugs**

Prochlorperazine see **Antinausea drugs**

Proctitis

Definition

Proctitis is an inflammation of the rectum.

Description

Proctitis affects mainly adolescents and adults. It is most common in men around age 30. Proctitis is caused by several different **sexually transmitted diseases**. Male homosexuals and people who practice anal intercourse are more likely to suffer from proctitis. Patients who have **AIDS** or who are immunocompromised are also more at risk.

Causes and symptoms

Proctitis is caused most often by sexually transmitted diseases, including **gonorrhea**, **syphilis**, herpes simplex (**genital herpes**), **candidiasis**, and chlamydia. It can also be caused by inflammatory bowel diseases, such as **Crohn's disease**, or ulcerative **colitis**, a chronic recurrent ulceration in the colon. Occasionally it is caused by an amoeba that causes **dysentery**.

Discharge of blood and mucus and intense **pain** in the area of the rectum and anus are all signs of proctitis. Patients feel the urge to have frequent bowel movements even when there is nothing present to eliminate. They may also have **constipation**, **diarrhea**, **fever**, and open sores around the anus. Other symptoms include cramping, lower back pain, difficulty urinating, and **impotence**.

Diagnosis

Proctitis is diagnosed by a patient history and **physical examination**. It is confirmed by a proctoscopy (examination of the rectum with an endoscope inserted through the anus). Proctoscopy usually shows a red, sore, inflamed lining of the rectum. Biopsies, smears, and lab cultures of rectal material are used to determine the exact cause of the inflammation so that the underlying cause can be treated appropriately.

Since the two problems often occur together, in the presence of proctitis, the large bowel should be examined for ulcerative colitis.

Treatment

Once the underlying cause of the inflammation is diagnosed, appropriate treatment begins. **Antibiotics** are given for bacterial infections. There is no cure for genital herpes, but the antiviral drug acyclovir is often

KEY TERMS

Candidiasis—A common fungal infection caused by yeast that thrives in moist, warm areas of the body.

Chlamydia—A gonorrhea-like bacterial infection.

Proctoscopy—A procedure in which a thin tube containing a camera and a light is inserted into the rectum so that the doctor can visually inspect it.

Rectum—The final section of the large intestine.

Ulcerative colitis—Chronic ulceration of the colon and rectum.

prescribed to reduce symptoms. Corticosteroid suppositories or ointments such as hydrocortisone are used to lessen discomfort, and the patient is encouraged to take warm baths to ease painful symptoms. Ulcerative proctitis often responds well to corticosteroid **enemas** or foam, or to sulfasalazine and related drugs.

Alternative treatment

Depending on the cause of proctitis, alternative medicine has several types of treatments available. If proctitis is related to gonorrhea, syphilis, or chlamydia, appropriate antibiotic treatment is recommended. Supplementation with *Lactobacillus acidophilus* is also recommended during and following antibiotic therapy to help rebuild normal gut flora that is destroyed by antibiotics. If proctitis is herpes related, antiviral herbs taken internally, as well as applied topically, can be helpful. Sitz baths and compresses of herbal infusions (herbs steeped in hot water) and decoctions (herbal extracts prepared by boiling the herb in water) can be very effective. Among the herbs recommended are calendula (*Calendula officinalis*), comfrey (*Symphytum officinale*), and plantain (*Plantago major*). Proctitis related to candidiasis requires dietary alterations, especially elimination of sugar from the diet. Any immunocompromised person needs close medical attention. If proctitis is related to inflammatory bowel diseases, the resolution of the underlying condition should contribute to resolution of the proctitis. **Acupuncture** and homeopathic treatment can be very useful in resolving inflammatory bowel diseases.

Prognosis

Proctitis caused by bacteria is curable with antibiotics. Genital herpes is not curable. Although

symptoms can be suppressed, proctitis may reoccur. Patients with AIDS are especially susceptible to candidiasis infections, which may be hard to control. Recovering from proctitis caused by inflammatory bowel diseases is variable and depends on successful management of those diseases. Severe proctitis can result in permanent narrowing of the anus.

Prevention

Proctitis is best prevented by using **condoms** and practicing safer sex to prevent acquiring sexually transmitted diseases. Avoiding anal intercourse also helps prevent damage to the rectum.

Resources

OTHER

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Tish Davidson, A.M.

Proctosigmoidoscopy see **Sigmoidoscopy**

Progesterone assay see **Sex hormones tests**

KEY TERMS

Multifocal—Having many focal points. In progressive multifocal leukoencephalopathy, it means that damage caused by the disease occurs at multiple sites.

Opportunistic infection—An illness caused by infecting organisms that would not be able to produce disease in a person with a healthy immune system but are able to take advantage of an impaired immune response.

Causes and symptoms

Although at least 80% of the adults in the United States have been exposed to JC virus (as evidenced by the presence of antibodies to this virus), very few will develop PML. Little is certain about what causes JCV to produce active disease, but the virus persists in the kidneys of otherwise healthy people without making them ill. Recent evidence suggests that after prolonged compromise of the immune system, the virus changes into a form that can reach brain tissue and cause disease. In PML, the JCV infects and kills the cells (oligodendrocytes) that produce myelin, which is needed to form the sheath that surrounds and protects nerves.

About 45% of people with PML experience vision problems, most often a blindness affecting half of the visual field of each eye. Mental impairment affects about 38% of people with PML. Eventually, about 75% experience extreme weakness. Other symptoms include lack of coordination, **paralysis** on one side of the body (hemiparesis), and problems in speaking or using language.

Diagnosis

Diagnosis is difficult but usually relies on a neurologist and radiologist assessing the white matter of the brain on a computed tomography or **magnetic resonance imaging** (MRI) scan. Tests of the cerebrospinal fluid can help distinguish between PML and other diseases, such as **multiple sclerosis** and acute hemorrhagic leukoencephalopathy. The rapid clinical progression in immunocompromised patients is another distinguishing factor.

Treatment

Currently, there is no known cure for PML, although it sometimes responds to treatment in patients with AIDS who are taking anti-HIV drugs

Progressive multifocal leukoencephalopathy

Definition

Progressive multifocal leukoencephalopathy (PML) is a rapidly progressive neuromuscular disease caused by opportunistic infection of brain cells (oligodendrocytes and astrocytes) by the JC virus (JCV).

Description

PML is an opportunistic infection associated with **AIDS** and certain cancers. It occurs in people with inadequate immune response and carries a poor prognosis. The incidence of PML, once quite rare, is rising as the numbers of people living with persistently compromised immune systems rises. An estimated 2%–7% of people with HIV disease will develop PML. The infection also occurs among people undergoing long-term **chemotherapy** for **cancer**. PML is not considered a contagious disease. According to the Centers for Disease Control definition of AIDS, PML in the presence of HIV infection is sufficient to form a diagnosis of AIDS.

(such as AZT, alpha-interferon, and peptide T). Although several agents have shown some potential in the last few years, such as the highly toxic cancer drug cytarabine, none are safe enough or sufficiently effective to be approved for PML.

Prognosis

PML is usually a very aggressive disease. The time between the onset of symptoms and **death** can be as little as one to six months. However, some patients infected with HIV have improved without receiving treatment specifically for PML.

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“NINDS Progressive Multifocal Leukoencephalopathy Information Page.” National Institute of Neurological Disorders and Stroke. <http://www.ninds.nih.gov/disorders/pml/pml.htm> (accessed January 5, 2011).

Jill S. Lasker

Progressive supranuclear ophthalmoplegia
see **Progressive supranuclear palsy**

Progressive supranuclear palsy

Definition

Progressive supranuclear palsy (PSP; also known as Steele-Richardson-Olszewski syndrome) is a rare disease that gradually destroys nerve cells in the parts of the brain that control eye movements, breathing, and muscle coordination. The loss of nerve cells causes palsy, or **paralysis**, that slowly gets worse as the disease progresses. The palsy affects ability to move the eyes, relax the muscles, and control balance.

Description

Progressive supranuclear palsy is a disease of middle age. Symptoms usually begin in the 60s, rarely before age 45 or after age 75. Men develop PSP more often than women do. It affects three to four people per million each year.

Causes and symptoms

PSP affects the brainstem, the basal ganglia, and the cerebellum. The brainstem is located at the top of the spinal cord. It controls the most basic functions needed for survival—the involuntary (unwilled)

movements such as breathing, blood pressure, and heart rate. The brainstem has three parts: the medulla oblongata, the pons, and the midbrain. The parts affected by PSP are the pons, which controls facial nerves and the muscles that turn the eye outward, and the midbrain, the visual center. The basal ganglia are islands of nerve cells located deep within the brain. They are involved in the initiation of voluntary (willed) movement and control of emotion. Damage to the basal ganglia causes muscle stiffness (spasticity) and **tremors**. The cerebellum is located at the base of the skull. It controls balance and muscle coordination.

Vision is controlled by groups of cells called *nuclei* in the brainstem. In PSP, the nuclei continue to function, but the mechanisms that control the nuclei are destroyed. The term *supranuclear* means that the damage is done above (*supra*) the nuclei. Patients with PSP have difficulty with voluntary (willed) eye movement. At first, the difficulty only occurs in trying to look down. As the disease progresses, ability to move the eyes right and left is also affected. However, reflex or unwilling eye movements remain normal. Thus, when the patient’s head is tilted upwards, the eyes move to look down. These reflex movements remain normal until late in the course of the disease. The upper eyelids may be pulled back, the eyebrows raised, and the brow wrinkled, causing a typical wide-eyed stare. Rate of blinking may decrease from the normal 20–30 per minute to three to five per minute. It becomes difficult to walk downstairs, to maintain eye contact during conversation, or to move the eyes up and down to read.

The earliest symptoms of PSP may be frequent falls or stiff, slow movements of the arms and legs. These symptoms may appear as much as five years before the characteristic vision problems. Walking becomes increasingly awkward, and some patients tend to lean and fall backward. Facial muscles may be weak, causing slurred speech and difficulty swallowing. Sleep may be disturbed and thought processes slowed. Although memory remains intact, the slowed speech and thought patterns and the rigid facial expression may be mistaken for senile **dementia** or **Alzheimer’s disease**. Emotional responses may become exaggerated and inappropriate, and the patient may experience **anxiety**, depression, and agitation.

The cause of PSP is not known. Most people who develop PSP come from families with no history of the disease, so it does not seem to be inherited, except in certain rare instances. People who have PSP seem to lack the neurotransmitters dopamine and homovanillic acid in the basal ganglia. Neurotransmitters are chemicals that help carry electrical impulses along the nervous system. Transmitting structures in brain cells called neurofibrils become disorganized (neurofibrillary

tangles). Neurofibrillary tangles are also found in Alzheimer's disease, but the pattern is somewhat different.

Diagnosis

PSP is sometimes mistaken for **Parkinson's disease**, which is also associated with stiffness, frequent falls, slurred speech, difficulty swallowing, and decreased spontaneous movement. The facial expression in Parkinson's, however, is blank or mask-like, whereas in PSP it is a grimace and wide-eyed stare. PSP does not cause the uncontrolled shaking (tremor) in muscles at rest that is associated with Parkinson's disease. Posture is stooped in Parkinson's disease, but erect in PSP. Speech is of low volume in both diseases, but is more slurred and irregular in rhythm in PSP.

Multiple strokes or abnormal accumulations of fluid within the skull (**hydrocephalus**) can also cause balance problems similar to PSP. **Magnetic resonance imaging (MRI)** scans of the brain may be needed to rule out these conditions. In advanced cases, MRI shows characteristic abnormalities in the brainstem described as "mouse ears."

Treatment

PSP cannot be cured. Drugs are sometimes given to relieve symptoms, but drug treatment is usually disappointing. Dopaminergic medications used in Parkinson's disease, such as levodopa (Sinemet), sometimes decrease stiffness and ease spontaneous movement. Anticholinergic medications, such as trihexyphenidyl (Artane), which restore function to neurotransmitters, or tricyclic drugs, such as amitriptyline (Elavil), may improve speech, walking, and inappropriate emotional responses.

Speech therapy may help manage the swallowing and speech difficulty in PSP. As the disease progresses, the difficulty in swallowing may cause the patient to choke and get small amounts of food in the lungs. This condition can cause aspiration **pneumonia**. The patient may also lose too much weight. In these cases, a feeding tube may be needed. The home environment should be modified to decrease potential injury from falls. Walkers can be weighted in front, to prevent backward falls and handrails can be installed in the bathroom. Because the patient cannot look down, low objects like throw rugs and coffee tables should be removed. Dry eyes from infrequent blinking can be treated with drops or ointments.

Prognosis

The patient's condition gradually deteriorates. After about seven years, balance problems and

KEY TERMS

Basal ganglia—Brain structure at the base of the cerebral hemispheres, involved in controlling movement.

Brainstem—Brain structure closest to the spinal cord, involved in controlling vital functions, movement, sensation, and nerves supplying the head and neck.

Cerebellum—The part of the brain involved in coordination of movement, walking, and balance.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct images of internal structures.

Parkinson's disease—A slowly progressive disease that destroys nerve cells. Parkinson's is characterized by shaking in resting muscles, a stooping posture, slurred speech, muscular stiffness, and weakness.

stiffness make it nearly impossible for the patient to walk. Persons with PSP become more and more immobile and unable to care for themselves. **Death** is not caused by the PSP itself. It is usually caused by pneumonia related to **choking** on secretions or by **starvation** related to swallowing difficulty. It usually occurs within 10 years, but if good general health and **nutrition** are maintained, the patient may survive longer.

Prevention

PSP cannot be prevented.

ORGANIZATIONS

American Academy of Neurology, 1080 Montreal Ave., St. Paul, MN, 55116, (651) 695-2717, (651) 695-2791, (800) 879-1960, memberservices@aan.com, <http://www.aan.com/>.

Society for Progressive Supranuclear Palsy, Inc., Suite #5065 Johns Hopkins Outpatient Center, 601 N. Caroline St., Baltimore, MD, 21287, (800) 457-4777, <http://www.psp.org>.

Laurie Barclay, MD

Progressive systemic sclerosis see
Scleroderma

Prolactin test

Definition

Prolactin is a hormone secreted by the anterior portion of the pituitary gland (sometimes called the “master gland”). Its role in the male has not been demonstrated, but in females, prolactin promotes **lactation**, or milk production, after **childbirth**.

Purpose

The prolactin test is used to diagnose pituitary dysfunction that might be caused by a tumor called an adenoma. In some circumstances, the test is also used to evaluate absence of menstrual periods (**amenorrhea**), or spontaneous production of milk (**galactorrhea**) by a woman who is not pregnant or lactating.

Precautions

Stress from trauma, illness, surgery, or even nervousness about a blood test can elevate prolactin levels. Drugs that may increase prolactin include phenothiazines, **oral contraceptives**, opiates, histamine antagonists, **monoamine oxidase inhibitors** (MAO inhibitors), estrogen, and **antihistamines**. Drugs that can decrease values include levodopa and dopamine.

Description

Prolactin is also known as the lactogenic hormone or lactogen. It is essential for the development of the mammary glands for lactation during **pregnancy**, and for stimulating and maintaining lactation after childbirth. Like the human growth hormone, prolactin acts directly on tissues, and its levels rise in response to sleep and to physical or emotional stress. During sleep, prolactin levels can increase to the circulating levels found in pregnant women (as high as ten to twenty times the normal level).

Prolactin secretion is controlled by prolactin-releasing and prolactin-inhibiting chemicals (factors) secreted by an area of the brain called the hypothalamus. Another hormone, thyroid-releasing hormone, or TRH, can also stimulate prolactin.

Tumors of the pituitary, called adenomas, are the most common cause of excessive levels of prolactin. Depending on the type of cell involved, these tumors are also called prolactin-secreting pituitary acidophilic or chromophobic adenomas. Moderately high prolactin levels are found to a lesser extent in women with secondary amenorrhea, galactorrhea, low thyroid,

KEY TERMS

Adenoma—A benign tumor.

Amenorrhea—The absence or abnormal stoppage of menstrual periods.

Factor—Any of several substances necessary to produce a result or activity in the body. The term is used when the chemical nature of the substance is unknown. In endocrinology, when the chemical nature is known, factors are renamed hormones.

Galactorrhea—Excessive or spontaneous flow of milk.

Pituitary gland—A gland located at the base of the brain and controlled by the hypothalamus. It controls most endocrine functions and is responsible for things such as kidney function, lactation, and growth and development.

anorexia, and a disorder known as **polycystic ovary syndrome**, a disease whose cause is not well known.

Because high prolactin levels are more likely due to pituitary adenoma than other causes, the prolactin level is used to diagnose and monitor this type of tumor. Several stimulation and suppression tests, with TRH or levodopa, respectively, have been designed to differentiate pituitary adenoma from other causes of prolactin overproduction.

Preparation

This test requires a blood sample that should be drawn in the morning at least two hours after the patient wakes (samples drawn earlier may show sleep-induced peak levels). The patient need not restrict food or fluids nor limit physical activity, but should relax for approximately 30 minutes before the test.

Risks

Risks posed by this test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or lightheadedness after venipuncture, or **hematoma** (blood accumulating under the puncture site).

Normal results

Reference ranges vary from laboratory to laboratory but are generally within the following values:

- adult male: 0–20 ng/mL
- adult female: 0–20 ng/mL
- pregnant female: 20–400 ng/mL

Abnormal results

Increased prolactin levels are found in galactorrhea, amenorrhea, prolactin-secreting pituitary tumor, infiltrative diseases of the hypothalamus, and metastatic **cancer** of the pituitary gland. Higher levels than normal are seen in stress related to **anorexia nervosa**, surgery, strenuous **exercise**, trauma, and renal (kidney) failure.

Decreased prolactin levels are seen in Sheehan's syndrome, a condition of severe hemorrhage after obstetric delivery that causes decreased blood supply to the pituitary.

Resources

BOOKS

Pagana, Kathleen Deska, and Timothy J. Pagana. *Mosby's Manual of Diagnostic and Laboratory Tests*. 4th ed. St. Louis: Mosby, 2009.

Janis O. Flores

Prolactinoma see **Galactorrhea**

Prolapsed disk see **Herniated disk**

Prolonged QT syndrome

Definition

Prolonged QT syndrome, also known as long QT syndrome (LQTS), refers to a group of disorders that increase the risk for sudden **death** due to an abnormal heartbeat.

Description

Abnormal heartbeats (cardiac **arrhythmias**) are a primary cause of sudden death, especially in the young population. In the United States, an estimated 1 in 300,000 individuals per year die suddenly due to irregular heart rhythms. One of the better understood causes of these arrhythmias is LQTS.

The QT of LQTS refers to an interval between two points (Q and T) on the common electrocardiogram (ECG, EKG) used to record the electrical activity of the heart. This electrical activity, in turn, is the result of small molecules (ions such as **sodium** and potassium) passing in and out of channels in the membranes surrounding heart cells. A prolonged QT interval indicates an abnormality in electrical activity that leads to irregularities in heart muscle contraction. One of these

irregularities is a specific pattern of very rapid contractions (tachycardia) of the lower chambers of the heart called torsade de pointes, a type of **ventricular tachycardia**. The rapid contractions, which are not effective in pumping blood to the body, result in a decreased flow of oxygen-rich blood to the brain. This can result in a sudden loss of consciousness (syncope) and death.

Causes and symptoms

Both inherited and acquired forms of LQTS have been identified. Most acquired forms are thought to be due to certain drugs including adrenaline (epinephrine), several **antihistamines** and **antibiotics**, specific heart medications, **diuretics**, and others. It has been proposed, but not yet documented, that individuals who experience LQTS after using one of these medications may actually have a genetic defect that increases their tendency to cardiac arrhythmias. Severe weight loss such as is associated with **anorexia nervosa** can also disrupt ion balances in the heart and result in prolongation of the QT interval.

Four inherited forms of LQTS have been described to date. Jervell and Lange-Neilsen syndrome, named for the physicians who described the condition in 1957, is associated with congenital deafness and is inherited as an autosomal recessive trait. Romano-Ward syndrome, the most common inherited form of LQTS, was first described in the 1960s. It is inherited in an autosomal dominant pattern and is not associated with other physical impairments such as deafness. The remaining two forms are Timothy syndrome and Andersen syndrome.

At least 10 different genes have been associated with the inherited forms of LQTS. The genes involved in LQTS play important roles in the formation of ion channels in the cell membrane, and, thus, mutations in these genes disrupt normal cardiac rhythms.

LQTS usually presents with symptoms that constitute a life-threatening emergency. Sudden loss of consciousness or cardiac arrest can be brought on by emotional or physical **stress** in young and otherwise healthy individuals, both female and male. Fright, anger, surprise, sudden awakening as a result of loud sounds (alarm clock, telephone), and physical activities, especially swimming, have all been reported to precipitate an episode of cardiac arrhythmia in susceptible individuals. Sudden death often occurs. Although the information is preliminary,

KEY TERMS

Anorexia nervosa—Eating disorder marked by malnutrition and weight loss commonly occurring in young women.

Autosomal dominant—A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50% chance of passing it to each of their offspring.

Autosomal recessive—A pattern of inheritance in which both copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes.

When both parents have one abnormal copy of the same gene, they have a 25% chance with each pregnancy that their offspring will have the disorder.

Diuretic—An agent that increases the production of urine.

Electrocardiogram—A record of the electrical activity of the heart showing certain waves called P, Q, R, S, and T waves. The Q, R, S, T waves are associated with contraction of the ventricles, the lower two chambers of the heart.

Sympathetic nervous system—A division of the autonomic nervous system, the portion of the nervous system that controls involuntary bodily functions such as heart rate.

Syndactyly—A fusion of two or more toes or fingers.

recent research has also suggested that a small number of SIDS (**sudden infant death syndrome**) cases may be due to mutations in one or more of the genes associated with LQTS.

Diagnosis

Problems exist in diagnosing LQTS. Although the method of diagnosis is the electrocardiogram, most young, healthy people do not routinely undergo this test, and thus their first, and possibly fatal, episode of LQTS comes without warning. In some cases, a non-fatal episode is mistakenly treated as a seizure, and so the follow-up assessment does not include an electrocardiogram. In addition, some cases of LQTS cannot be diagnosed by a routine electrocardiogram. That is, the QT interval is not found to be prolonged in routine testing. If LQTS is suspected either because of a previous episode of syncope or because of a family member with LQTS, an **exercise** electrocardiogram should be performed. In all instances where an individual is diagnosed with LQTS, family members should be thoroughly evaluated, and a detailed family history should be taken noting any individuals with episodes of sudden loss of consciousness and any cases of unexplained sudden death. Because many of the genes involved in LQTS have been identified, **genetic testing** can offer a more reliable means of diagnosis of other family members at risk. The first step in determining if this type of testing is appropriate in any particular situation is to consult a genetic counselor or medical geneticist.

Treatment

A conventional treatment is the oral administration of beta blockers, medications that decrease the input from the sympathetic nervous system to the heart. Although beta blockers do not correct the abnormalities in the ion channels of the heart cells, they do appear to decrease the occurrence of cardiac arrhythmias. However, these medications are not helpful in all cases, and are actually contraindicated in some individuals. Potassium supplementation is also being explored as a treatment in certain cases. As the genetics of LQTS becomes better understood, it should be possible to tailor treatments that will be effective for each of the various gene mutations.

Alternative treatment

In some patients, severing of the sympathetic nerve to the heart has decreased the occurrence of arrhythmias. **Pacemakers** and defibrillators appear to hold promise as new forms of treatment. As devices of this type are developed that are smaller in size, they may come into more widespread use, either alone or in conjunction with specific medications.

Prognosis

LQTS is a lifelong condition. Individuals who are not diagnosed and treated are at an increased risk of syncope and sudden death. Adequate treatment can

decrease this risk. There is no cure. Individuals with one of the inherited forms of LQTS are at risk of passing the mutation and the disease to their offspring.

Prevention

The risk of cardiac arrhythmias due to acquired forms of LQTS can be decreased by avoiding the medications and situations that trigger episodes. At present there is no genetic therapy to correct the gene mutations present in the inherited forms of LQTS, but individuals who are known to have an inherited form may also be able to lessen the risk of a life-threatening episode by avoiding such environmental triggers and by taking the appropriate medications.

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ORGANIZATIONS

National Organization for Rare Disorders (NORD), 55
Kenosia Avenue, PO Box 1968, Danbury, CT 06813-1968, (203) 744-0100, (800) 999-6673, TTD: (203) 797-9590, <http://www.rarediseases.org>.

Sudden Arrhythmia Death Syndromes Foundation, 508 E. South Temple, Suite #20, Salt Lake City, UT, 84102, (800) 786-7723, <http://www.sads.org>.

Sallie Boineau Freeman, PhD

PROM see **Premature rupture of membranes**

Promethaz see **Antihistamines**

Prophylaxis

Definition

A prophylaxis is a measure taken to maintain health and prevent the spread of disease. Antibiotic prophylaxis is the focus of this article and refers to the use of **antibiotics** to prevent infections.

Purpose

Antibiotics are well known for their ability to treat infections. But some antibiotics also are prescribed to *prevent* infections. This usually is done only in certain situations or for people with particular medical problems. For example, people with abnormal heart valves have a high risk of developing heart valve infections after even minor surgery. This happens because bacteria from other parts of the body get into the bloodstream during surgery and travel to the heart valves. To prevent these infections, people with heart valve problems often take antibiotics before having any kind of surgery, including dental surgery.

Antibiotics also may be prescribed to prevent infections in people with weakened immune systems, such as people with **AIDS** or people who are having **chemotherapy** treatments for **cancer**. But even healthy people with strong immune systems may occasionally be given preventive antibiotics—if they are having certain kinds of surgery that carry a high risk of infection, or if they are traveling to parts of the world where they are likely to get an infection that causes **diarrhea**, for example.

In all of these situations, a physician should be the one to decide whether antibiotics are necessary. Unless a physician says to do so, it is not a good idea to take antibiotics to prevent ordinary infections.

Because the overuse of antibiotics can lead to resistance, drugs taken to prevent infection should be used only for a short time.

Description

Among the drugs used for antibiotic prophylaxis are amoxicillin (a type of penicillin) and **fluoroquinolones** such as ciprofloxacin (Cipro) and trovafloxacin (Trovan). These drugs are available only with a physician's prescription and come in tablet, capsule, liquid, and injectable forms.

Recommended dosage

The recommended dosage depends on the type of antibiotic prescribed and the reason it is being used. For the correct dosage, check with the physician or dentist who prescribed the medicine or the pharmacist who filled the prescription. Be sure to take the medicine exactly as prescribed. Do not take more or less than directed, and take the medicine only for as long as the physician or dentist says to take it.

KEY TERMS

AIDS—Acquired immunodeficiency syndrome. A disease caused by infection with the human immunodeficiency virus (HIV). In people with this disease, the immune system breaks down, opening the door to other infections and some types of cancer.

Antibiotic—A medicine used to treat infections.

Chemotherapy—Treatment of an illness with chemical agents. The term is usually used to describe the treatment of cancer with drugs.

Immune system—The body's natural defenses against disease and infection.

Precautions

If the medicine causes **nausea**, **vomiting**, or diarrhea, check with the physician or dentist who prescribed it as soon as possible. Patients who are taking antibiotics before surgery should not wait until the day of the surgery to report problems with the medicine. The physician or dentist needs to know right away if problems occur.

For other specific precautions, see the entry on the type of drug prescribed such as **penicillins** or fluoroquinolones.

Side effects

Antibiotics may cause a number of side effects. For details, see entries on specific types of antibiotics. Anyone who has unusual or disturbing symptoms after taking antibiotics should get in touch with his or her physician.

Interactions

Whether used to treat or to prevent infection, antibiotics may interact with other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. Anyone who takes antibiotics for any reason should inform the physician about all the other medicines he or she is taking and should ask whether any possible interactions may interfere with drugs' effects. For details of **drug interactions**, see entries on specific types of antibiotics.

Nancy Ross-Flanigan

Proportionate dwarfism see **Pituitary dwarfism**

Proptosis see **Exophthalmos**

Prostaglandins see **Drugs used in labor**

Prostate biopsy

Definition

Prostate biopsy is a surgical procedure that involves removing a small piece of prostate tissue for microscopic examination.

Purpose

This test is usually done to determine whether the patient has **prostate cancer**. Occasionally, it may also be used to diagnose a condition called benign prostatic hyperplasia that causes enlargement of the prostate. In the United States, prostate **cancer** is the most common cancer among men over 50, and is the second leading cause of cancer deaths. According to statistics released by the American Cancer Society in 2010, African American men in the United States have 1.6 times the risk of developing prostate cancer after age 50 than Caucasian or Asian American men. On the other hand, the rate of deaths from prostate cancer has declined further among African American men than among men of other races since 2000.

Prostate biopsy is recommended when a **digital rectal examination** (a routine screening test for prostate diseases) reveals a lump or some other abnormality in the prostate. In addition, if blood tests reveal that the levels of certain markers, such as PSA, are higher than normal, the doctor may order a biopsy.

Description

The prostate gland is one of the three male sex glands and lies just below the urinary bladder, in the area behind the penis and in front of the rectum. It secretes semen, the liquid portion of the ejaculate. The urethra carries the urine from the urinary bladder and the semen from the sex glands to the outside of the body.

Prostate biopsies can be performed in three different ways. They can be performed by inserting a needle through the perineum (the area between the base of the penis and the rectum), by inserting a needle through the wall of the rectum, or by cystoscopy. Before the procedure is performed, the patient may be given a sedative to help him relax. Patients undergoing cystoscopy may

be given either **general anesthesia** or **local anesthesia**. The doctor will ask the patient to have an enema before carrying out the biopsy. The patient is also given **antibiotics** to prevent any possible infection.

Needle biopsy via the perineum

The patient lies either on one side or on his back with his knees up. The skin of the perineum is thoroughly cleansed with an iodine solution. A local anesthetic is injected at the site where the biopsy is performed. Once the area is numb, the doctor makes a small (1 in.) incision in the perineum. The doctor places one finger in the rectum to guide the placement of the needle. The needle is then inserted into the prostate, a small amount of tissue is collected, and the needle is withdrawn. The needle is then re-inserted into another part of the prostate. Tissue may be taken from several areas. Pressure is then applied at the biopsy site to stop the bleeding. The procedure generally takes 15–30 minutes and is usually done in a physician's office or in a hospital operating room. Although it sounds painful, it typically causes only slight discomfort.

Needle biopsy via the rectum

This procedure is also done in the physician's office or in the hospital operating room, and is usually done without any anesthetic, although some doctors prefer to inject a local anesthetic, usually lidocaine. The patient is asked to lie on his side or on his back with his legs in stirrups. The doctor attaches a curved needle guide to his finger and then inserts the finger into the rectum. After firmly placing the needle guide in the rectum, the biopsy needle is pushed along the guide, through the wall of the rectum and into the prostate. The needle is rotated gently, prostate tissue samples are collected and the needle is withdrawn. When an ultrasound probe is used to guide the needle, the procedure is called a transrectal ultrasound-guided biopsy, or TRUS.

Cytoscopy

For this procedure, the patient is given either a general or a local anesthetic. An instrument called a cytoscope (a thin-lighted tube with telescopic lenses) is passed through the urethra. By looking through the cytoscope, the doctor can see if there is any blockage in the urethra and remove it. Tissue samples from the urinary bladder or the prostate can be collected for microscopic examination.

This test is generally performed in an operating room or in a physician's office. An hour before the

procedure, the patient is given a sedative to help him relax. An intravenous (IV) line will be placed in a vein in the arm to give medications and fluids if necessary. The patient is asked to lie on a special table with his knees apart and stirrups are used to support his feet and thighs. The genital area is cleansed with an anti-septic solution. If general anesthesia is being used, the patient is given the medication through the IV tube or inhaled gases or both. If a local anesthetic is being used, the anesthetic solution is gently instilled into the urethra.

After the area is numb, a cytoscope is inserted into the urethra and slowly pushed into the prostate. Tiny forceps or scissors are inserted through the cytoscope to collect small pieces of tissue that are used for biopsy. The cytoscope is then withdrawn. The entire procedure may take 30–45 minutes. Sometimes a catheter (tube) is left in the urinary bladder to help the urine drain out, until the swelling in the urethra has subsided.

Alternate procedures

Many different tests can be performed to diagnose prostate diseases and cancer. A routine screening test called digital **rectal examination** (DRE) can identify any lumps or abnormality with the prostate. Blood tests that measure the levels of certain protein markers, such as PSA, can indicate the presence of prostate cancer cells. X rays and other imaging techniques (such as **computed tomography scans**, **magnetic resonance imaging** [MRI], and ultrasonograms), where detailed pictures of areas inside the body are put together by a computer, can also be used to determine the extent and spread of the disease. However, a prostate biopsy and examination of the cells under a microscope remains the most definitive test for diagnosing and grading prostate cancer as of 2010.

Preparation

Before scheduling the biopsy, the doctor should be informed of all the medications that the patient is taking; whether the patient is allergic to any medication; and whether he has any bleeding problems. The patient may be given an antibiotic shortly before the test to reduce the risk of any infection afterwards. If the biopsy is done through the perineum, there are no special preparations. If it is being done through the rectum, the patient is asked to take an enema and is instructed on how to do it.

If a cytoscopy is being performed, the patient is asked to sign a consent form. The patient is also asked to take antibiotics before and for several days after the

KEY TERMS

Benign prostatic hyperplasia (BPH)—A noncancerous condition of the prostate that causes overgrowth of the prostate tissue, thus enlarging the prostate and obstructing urination.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes.

Computed tomography (CT) scan—A medical procedure in which a series of x rays are taken and put together by a computer in order to form detailed pictures of areas inside the body.

Digital rectal examination—A routine screening test that is used to detect any lumps in the prostate gland or any hardening or other abnormality of the prostate tissue. The doctor inserts a gloved and lubricated finger (digit) into the patient's rectum, which lies just behind the prostate. Typically, since a majority of tumors develop in the posterior

region of the prostate, they can be detected through the rectum.

Magnetic resonance imaging (MRI)—A medical procedure used for diagnostic purposes where pictures of areas inside the body are created using a magnet linked to a computer.

Pathologist—A doctor who specializes in the diagnosis of disease by studying cells and tissues under a microscope.

Ultrasonogram—A procedure in which high-frequency sound waves that cannot be heard by human ears are bounced off internal organs and tissues. These sound waves produce a pattern of echoes that are then used by the computer to create sonograms or pictures of areas inside the body.

Urethra—The tube that carries the urine from the urinary bladder and (in males) the semen from the sex glands to the outside of the body.

test to prevent infection due to insertion of the instruments. If a general anesthetic is going to be used, food and liquids will be restricted for at least eight hours before the test.

Aftercare

Following a needle biopsy, the patient may experience some **pain** and discomfort. He should avoid strenuous activities for the rest of the day. He may also notice some blood in his urine for two to three days after the test and some amount of rectal bleeding. If there is persistent bleeding, pain, or **fever**, and if the patient is unable to urinate for 24 hours, the doctor should be notified immediately.

When a cystoscopy is performed under a local anesthetic, the patient is asked to lie down for 30 minutes after the test and is then allowed to go. If general anesthesia is used, the patient is taken to the recovery room and kept there until he wakes up and is able to walk. He is allowed food and liquids after he wakes up. After general anesthesia, the patient may experience some tiredness and aching of the muscles throughout the body. If local anesthesia was administered, there is a brief burning sensation and a strong urge to urinate when the cystoscope is removed.

After the procedure, it is common to experience frequent urination with a burning sensation for a few

days. Drinking a lot of fluids will help reduce the burning sensation and the chances of an infection. There may also be some blood in the urine. However, if **blood clots** are seen, or if the patient is unable to pass urine eight hours after the cystoscopy, the doctor should be notified. In addition, if the patient develops a high fever, and complains of chills or abdominal pain after the procedure, he should see the doctor right away. Although serious infections are rare, a few patients develop such severe illnesses as **meningitis** following a prostate biopsy.

Risks

Prostate biopsy performed with a needle is a low-risk procedure. The possible complications include some bleeding into the urethra, bleeding from the rectum, an infection, a temporarily lowered sperm count, or an inability to urinate. These complications are treatable and the doctor should be notified of them.

Cystoscopy is generally a very safe procedure. The most common complication is an inability to urinate due to a swelling of the urethra. A catheter (tube) may have to be inserted to help drain out the urine. If there is an infection after the procedure, antibiotics are given to treat it. In very rare instances, the urethra or the urinary bladder may be perforated because of the insertion of the instrument. If this complication occurs, surgery may be needed to repair the damage.

Normal results

If the prostate tissue samples show no sign of inflammation, and if no cancerous cells are detected, the results are normal.

Abnormal results

Analysis of the prostate tissue under the microscope reveals any abnormalities. In addition, the presence of cancerous cells can be detected. If a tumor is present, the pathologist “grades” the tumor, in order to estimate how aggressive the tumor is. The most commonly used grading system is called the “Gleason system.”

Normal prostate tissue has certain characteristic features that the cancerous tissue lacks. In the Gleason system, prostate cancers are graded by how closely they resemble normal prostate tissue. The system assigns a grade ranging from one to five. The grades assigned to two areas of cancer are added up for a combined score that is between two and ten. A score between two and four is considered low and implies that the cancer is a slow-growing one. A Gleason score of eight to ten is high and indicates that the cancer is aggressive. The higher the Gleason score, the more likely it is that the cancer is fast-growing and may have already grown out of the prostate and spread to other areas (metastasized).

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ORGANIZATIONS

- American Cancer Society, 1599 Clifton Rd., NE, Atlanta, GA, 30329-4251, (800) 227-2345, <http://www.cancer.org>.
- American Urological Association, 1000 Corporate Blvd., Linthicum, MD, 21090, 866 RING-AUA.
- New Prostate Cancer InfoLink, <http://prostatecancerinfolink.net>.
- ZERO: The Project to End Prostate Cancer, 10 G Street NE, Suite 601, Washington, DC, 20002, (888) 245-9455, <http://www.zerocancer.org/index.html>.

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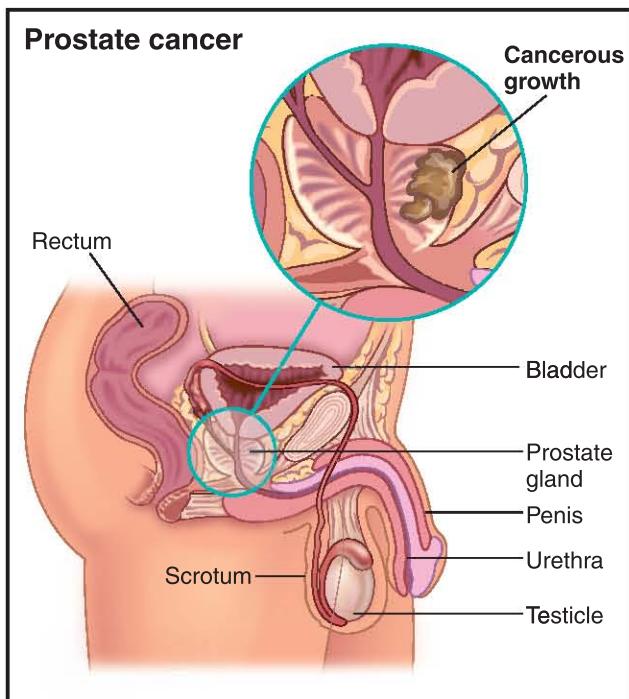
Prostate cancer

Definition

Prostate **cancer** is a disease in which cells in the prostate gland become abnormal and start to grow uncontrollably, forming tumors.

Demographics

Prostate cancer is the most commonly diagnosed malignancy among adult males in Western countries. Although prostate cancer is often very slow growing, it can be aggressive, especially in younger men. Given its slow growing nature, many men with the disease die of other causes rather than from the cancer itself.



This illustration shows the anatomy of the prostate and surrounding organs, with a cancerous growth on the prostate. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

About 190,000 new cases of prostate cancer are diagnosed in the United States every year. It is projected that one in every six American men will develop prostate cancer at some point during a lifetime. Prostate cancer is second only to lung cancer as a leading cause of death from cancer in American men. Almost 10% of deaths from cancer are caused by prostate cancer.

Description

Risk factors

The precise cause of prostate cancer is not known. However, there are several known risk factors for disease including age over 50, African American heritage, a family history of the disease, and possibly diets high in fat and red meat.

Prostate cancer is a malignancy of one of the major male sex glands. Along with the testicles and the seminal vesicles, the prostate secretes the fluid that makes up semen. The prostate is about the size of a walnut and lies just behind the urinary bladder. A tumor in the prostate interferes with proper control of the bladder and normal sexual functioning. Often the first symptom of prostate cancer is difficulty in urinating. However, because a very common, non-cancerous condition of the prostate, benign

prostatic hyperplasia (BPH), also causes the same problem, difficulty in urination is not necessarily due to cancer.

Cancerous cells within the prostate itself are generally not deadly on their own. However, as the tumor grows, some of the cells break off and spread to other parts of the body through the lymph or the blood, a process known as metastasis. The most common sites for prostate cancer to metastasize are the seminal vesicles, the lymph nodes, the lungs, and various bones around the hips and the pelvic region. The effects of these new tumors are what can cause death.

Prostate cancer affects African American men twice as often as Caucasian men; the mortality rate among African Americans is also two times as high. African Americans have the highest rate of prostate cancer of any world population group.

Causes and symptoms

Frequently, prostate cancer has no symptoms and the disease is diagnosed when the patient goes for a routine screening examination. However, when the tumor is big or the cancer has spread to the nearby tissues, the following symptoms may be seen:

- weak or interrupted flow of the urine
- frequent urination (especially at night)
- difficulty starting urination
- inability to urinate
- pain or burning sensation when urinating
- blood in the urine
- persistent pain in lower back, hips, or thighs (bone pain)
- impotence (trouble achieving an erection)
- painful ejaculation

Diagnosis

Prostate cancer is curable when detected early. However, the early stages of prostate cancer are often asymptomatic, so the disease often goes undetected until the patient has a routine **physical examination**. Diagnosis of prostate cancer can be made using some or all of the following tests.

Digital rectal examination (DRE)

In order to perform this test, the doctor puts a gloved and lubricated finger (digit) into the rectum to feel for any lumps in the prostate. The rectum lies just behind the prostate gland, and a majority of prostate tumors begin in the posterior region of the prostate. If

KEY TERMS

Antiandrogen—A substance that blocks the action of androgens, the hormones responsible for male characteristics. Used to treat prostate cancers that require male hormones for growth.

Benign prostate hyperplasia (BPH)—A noncancerous swelling of the prostate.

Brachytherapy—A method of treating cancers, such as prostate cancer, involving the implantation near the tumor of radioactive seeds.

Gleason Grading System—A method of predicting the tendency of a tumor in the prostate to metastasize based on how similar the tumor is to normal prostate tissue.

Granulocyte/macrophage colony stimulating factor (GM-CSF)—Also known as sargramostim, a substance produced by cells of the immune system that stimulates an attack upon foreign cells. Used to treat prostate cancers as a genetically engineered component of a vaccine that stimulates the body to attack prostate tissue.

Histopathology—The study of diseased tissues at a minute (microscopic) level.

Luteinizing hormone-releasing hormone (LH-RH) agonist—A substance that blocks the action of LHRH, a hormone that stimulates the production of testosterone (a male hormone) in men. Used to treat prostate cancers that require testosterone for growth.

Orchiectomy—Surgical removal of the testes as a way of treating prostate cancer by eliminating the production of testosterone.

Prostate-specific antigen—A protein made by the cells of the prostate that is increased by both BPH and prostate cancer.

Radical prostatectomy—Surgical removal of the entire prostate, a common method of treating prostate cancer.

Transurethral resection of the prostate (TURP)—Surgical removal of a portion of the prostate through the urethra, a method of treating the symptoms of an enlarged prostate, whether from BPH or cancer.

If the doctor does detect an abnormality, he or she may order more tests in order to confirm these findings. DRE is less effective than the prostate specific antigen (PSA) test in detecting prostate cancer, but use of DRE may facilitate detection of prostate cancer in men whose PSA levels are within normal limits.

Blood tests

Blood tests are used to measure the amounts of certain protein markers, such as PSA, found circulating in the blood. The cells lining the prostate generally make this protein, and a small amount can be detected normally in the bloodstream. In contrast, prostate cancers often produce a lot of this protein, significantly raising the circulating levels. A finding of a high PSA may indicate that cancer is present.

Transrectal ultrasound (TRUS)

A small probe is placed in the rectum and sound waves are released from the probe. These sound waves bounce off the prostate tissue and an image is created. Though the insertion of the probe into the rectum may be slightly uncomfortable, the procedure is generally painless and only takes 20 minutes. TRUS is not used as a screening test because it may not be sensitive

enough to detect cancer in its earliest stages. TRUS is most commonly used during a biopsy of the prostate.

Prostate biopsy

If cancer is suspected from the results of any of the above tests, the doctor will remove a small piece of prostate tissue with a hollow needle, a procedure known as core needle biopsy. Most urologists will take multiple samples, sometimes as many as 18, of the suspicious lesion. These samples are then checked under the microscope for the presence of cancerous cells. **Prostate biopsy** is the most definitive diagnostic tool for prostate cancer, and this procedure is done quickly and with little **pain** or discomfort.

X rays and imaging techniques

A chest x ray may be ordered to determine whether the cancer has spread to the lungs. Imaging techniques (such as **computer tomography scans** [CT scans] and **magnetic resonance imaging** [MRI]), where a computer is used to generate a detailed picture of the prostate and areas nearby, may be done to get a clearer view of the internal organs. A **bone scan** may be used to check whether the cancer has spread to the bone.

Treatment

Once cancer is detected during the microscopic examination of the prostate tissue during a biopsy, doctors will determine two different numerical scores that will help define the patient's treatment and prognosis.

Tumor grading

Initially, the pathologist will grade the tumor based on his or her examination of the biopsy tissue. The pathologist scores the appearance of the biopsy sample using the Gleason system. This system uses a scale of one to five based on the sample's similarity or dissimilarity to normal prostate tissue. If the tissue is very similar to normal tissue, it is still well differentiated and given a low grading number, such as one or two. As the tissue becomes more and more abnormal (less and less differentiated), the grading number increases, up to five. Less differentiated tissue is considered more aggressive and more likely to be the source of metastases.

The Gleason grading system is best predictive of the prognosis of a patient if the pathologist gives two scores to a particular sample: a primary and a secondary pattern. The two numbers are then added together and that is the Gleason score reported to the patient. Thus, the lowest Gleason score available is two (a primary and secondary pattern score of one each). A typical Gleason score is five (which can be a primary score of two and a secondary score of three or vice versa). The highest score available is ten, with a pure pattern of very undifferentiated tissue—that is, of grade five. The higher the score, the more abnormal behavior of the tissue, the greater the chance for metastases, and the more serious the prognosis after surgical treatment. A study found that the 10-year cancer survival rate without evidence of disease for grade two, three, and four cancers is 94% of patients. The rate is 91% for grade five cancers, 78% for grade six, 46% for grade seven, and 23% for grade eight, nine, and ten cancers.

Cancer staging

The second numeric score determined by the doctor will be the stage of the cancer (Stages I–IV), which takes into account the grade of the tumor determined by the pathologist. Based on the recommendations of the American Joint Committee on Cancer (AJCC), two kinds of data are used for staging prostate cancer. Clinical data is based on the external symptoms of the cancer, while histopathological data is based on surgical removal of the prostate and examination of its tissues. Clinical data is most useful to make treatment decisions,

while pathological data is the best predictor of prognosis. For this reason, the staging of prostate cancer takes into account both clinical and histopathologic information. Specifically, doctors look at tumor size, lymph node involvement, the presence of visceral (internal organ) involvement, and the grade of the tumor.

Treatment options

The doctor and the patient will decide on the treatment mode after considering many factors. For example, the patient's age, the stage of the disease, his general health, and the presence of any co-existing illnesses have to be considered. In addition, the patient's personal preferences and the risks and benefits of each treatment protocol are also taken into account before any decision is made.

SURGERY. For stage I and stage II prostate cancer, surgery is the most common method of treatment because it theoretically offers the chance of completely removing the cancer from the body. Radical **prostatectomy** involves complete removal of the prostate. The surgery can be done using a perineal approach, where the incision is made between the scrotum and the anus, or using a retropubic approach, where the incision is made in the lower abdomen. Perineal approach is also known as nerve-sparing prostatectomy, as it is thought to reduce the effect on the nerves and thus reduce the side effects of **impotence** and incontinence. However, the retropubic approach allows for the simultaneous removal of the pelvic lymph nodes, which can give important pathological information about the tumor's spread.

The drawback to surgical treatment for early prostate cancer is the significant risk of side effects that impact the quality of life of the patient. Studies by the National Cancer Institute (NCI) found that, even when using nerve-sparing techniques, 60% to 80% of men treated with radical prostatectomy reported themselves as impotent (unable to achieve an erection sufficient for sexual intercourse) two years after surgery. This side effect can be sometimes countered by prescribing **sildenafil citrate** (Viagra). Furthermore, 8% to 10% of patients were incontinent in that time span. Despite the side effects, the majority of men were reported as satisfied with their treatment choice. Additionally, there is some evidence that the skill and the experience of the surgeon are central factors in the occurrence and severity of side effects.

Newer surgical options used to treat prostate cancer include laparoscopic radical prostatectomy (LRP) and robotic-assisted LRP. LRP uses smaller incisions,

which typically results in decreased blood loss, less pain, shorter hospital stays, and faster recovery times as compared to more traditional surgical approaches. Robotic LRP involves the surgeon using robotic arms (using a device known as the da Vinci System) to perform the operation through small incisions made in the patient's abdomen. The advantage of using this method is enhanced maneuverability and precision. The device is expensive, however, and may not be available in many communities. Patients deciding on these newer approaches are advised to seek out surgeons with significant experience in LRP techniques.

Another treatment for prostate cancer is cryosurgery, or **cryotherapy**. Guided by ultrasound, surgeons insert up to eight cryoprobes through the skin and into close proximity with the tumor. Liquid nitrogen (temperature of -321 degrees F, or -196°C) is circulated through the probe, freezing the tumor tissue. In prostate surgery, a warming tube is also used to keep the urethra from freezing. Patients currently spend a day or two in the hospital following the surgery, but it could be an outpatient procedure in the near future. Recovery time is about one week. Side effects have been reduced in recent years, although impotence still affects almost all who have had cryosurgery for prostate cancer. Cryosurgery is considered a good alternative for those too old or sick to have traditional surgery or radiation treatments or when these more traditional treatments are unsuccessful. There is limited information about the long-term efficacy of this treatment for prostate cancer. Cryosurgery is not used as a first-line treatment for prostate cancer.

RADIATION THERAPY. **Radiation therapy** involves the use of high-energy x rays to kill cancer cells or to shrink tumors. It can be used instead of surgery for stage I and II cancer. The radiation can either be administered from a machine outside the body (external beam radiation), or small radioactive pellets can be implanted in the prostate gland in the area surrounding the tumor, called brachytherapy or interstitial implantation. Pellets containing radioactive iodine ($I-125$) or palladium ($Pd-103$) can be implanted on an outpatient basis, where they remain permanently. The radioactive effect of the seeds lasts only about a year.

A newer technique is temporary or high-dose brachytherapy, in which radioactive needles are inserted into implanted catheters in prostate tissue. Treatments typically take 5 to 15 minutes, after which the radioactive source is removed. Once therapy is completed, the catheters are removed.

Other newer radiation techniques that are delivered from outside of the body include three-dimensional radiation therapy (3D-CRT), intensity modulated radiation therapy (IMRT), conformal proton beam radiation therapy, and stereotactic radiosurgery (also known by names such as Gamma Knife, Novalis, and Cyberknife).

The side effects of radiation depend on the method of delivery and can include inflammation of the bladder, rectum, and small intestine as well as disorders of blood clotting (coagulopathies). Impotence and incontinence are often delayed side effects of the treatment. A study indicated that bowel control problems were more likely after radiation therapy when compared to surgery, but impotence and incontinence were more likely after surgical treatment. Long-term results with radiation therapy are dependent on stage.

HORMONE THERAPY. Hormone therapy, also called androgen deprivation therapy or androgen suppression therapy, is commonly used when the cancer is in an advanced stage and has spread to other parts of the body, such as stage III or stage IV. Prostate cells need the male hormone testosterone to grow. Decreasing the levels of this hormone or inhibiting its activity will cause the cancer to shrink. Hormone levels can be decreased in several ways.

Orchiectomy is a surgical procedure that involves complete removal of the testicles, leading to a decrease in the levels of testosterone. Drugs that may be given to decrease the amount of testosterone made by the testicles include luteinizing hormone-releasing hormone (LHRH) analogs or agonists, such as leuprolide (Lupron, Viadur, Eligard), goserelin (Zoladex), triptorelin (Trelstar), and histrelin (Vantas), and luteinizing hormone-releasing hormone antagonists, such as degarelix (Firmagon). Anti-androgens, which block the body's ability to use androgens (such as testosterone), may be prescribed in combination with orchiectomy or with LHRH analogs as first-line hormone therapy. Anti-androgens include flutamide (Eulexin), bicalutamide (Casodex), and nilutamide (Nilandron).

There are some serious and unpleasant side effects to hormone therapy. Men may have "hot flashes," enlargement and tenderness of the breasts, or impotence and loss of sexual desire. Another side effect is **osteoporosis**, or loss of bone mass leading to brittle and easily fractured bones.

EXPECTANT MANAGEMENT (WATCHFUL WAITING AND ACTIVE SURVEILLANCE). Watchful waiting means

no immediate treatment is recommended, but doctors keep the patient under careful observation. This is often done using regular PSA testing. This option is generally used in older patients when the tumor is not very aggressive and the patients have other, more life-threatening illnesses. Prostate cancer in older men tends to be slow growing. Therefore, the risk of the patient dying from prostate cancer, rather than from other causes, is relatively small.

Active surveillance, as the name implies, takes a more aggressive approach. The cancer is more closely monitored, typically every three to six months.

Prognosis

The prognosis for cancers at Stages I and II is very good. For men treated with stage I or stage II disease, almost 100% are alive after five years. Although the cancers of Stage III are more advanced, the five-year prognosis is still good, with 70% of men diagnosed at this stage still living.

Once the cancer has spread outside of the prostate to distant organs, the cancer cannot be cured by current treatment options. Median survival for patients in this advanced stage is typically one to three years, although some men with slower growing tumors may live for much longer.

The spread of the cancer into the pelvis, lymph system, or distant locations are very significant events, as the five-year survival rate drops to about 31% for prostate cancers in Stage IV at time of diagnosis.

Prevention

Because the cause of the cancer is not known, there is no definite way to prevent prostate cancer. However, mandatory screening for prostate cancer is controversial. Because the cancer is so slow growing, and the side effects of the treatment can have significant impact on patient quality of life, some medical organizations question the wisdom of yearly exams. Nevertheless, the National Cancer Institute reports that aggressive screening methods have achieved a reduction in the death rate of prostate cancer of about 2.3% for African Americans and about 4.6% for Caucasians since the mid-1990s, with a 20% increase in overall survival rate during that period.

Current recommendations for men who choose to be screened for prostate cancer are directed to men ages 50 and over whose life expectancy is 10 years or longer, men at high risk (African American men and men with a first-degree relative such as a father or a brother who were diagnosed with prostate cancer prior to age 65)

who are aged 45 or older, and men considered to be at highest risk (age of 40 to 45 with several first-degree relatives diagnosed with prostate cancer prior to age 65). For men who choose to be screened, testing with PSA and DRE is recommended. If the PSA level result is 2.5 ng/mL, the man may only need to be retested every two years. If the PSA level is 1.5 ng/mL, the current recommendation is for yearly retesting.

A low-fat diet may slow the progression of prostate cancer. To reduce the risk or progression of prostate cancer, the American Cancer Society recommends a diet rich in fruits, vegetables and dietary fiber, and low in red meat and saturated fats.

Current clinical studies are researching the effects of isoflavones (soy proteins) on prostate cancer risk. The outcomes of this research are not yet available.

Two recent clinical trials, the Prostate Cancer Prevention Trial and the Reduction by Dutasteride of Prostate Cancer Events (REDUCE) study, looked at the effects of administering drugs known as 5 alpha-reductase inhibitors (**finasteride** or dutasteride) for the prevention of prostate cancer. Men who took these drugs as part of participation in the trials developed less prostate cancers than men who did not receive the drugs. However, some men in the REDUCE trial who did develop prostate cancer developed cancers with higher Gleason scores than men who developed prostate cancer and did not take dutasteride. The results of these trials are still being analyzed.

Resources

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ORGANIZATIONS

- National Cancer Institute, 6116 Executive Blvd., Suite 300, Bethesda, MD, 20892–8322, (800) 4–CANCER (422–6237), cancergovstaff@mail.nih.gov, <http://www.cancer.gov>.
- The Prostate Cancer Foundation, 1250 Fourth St., Santa Monica, CA, 90401, (800) 757–CURE (2873), info@pcf.org, <http://www.capture.org>.

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- Prostate gland removal see **Prostatectomy**
Prostate sonogram see **Prostate ultrasound**

Prostate ultrasound

Definition

A prostate ultrasound is a diagnostic test used to detect potential problems with a man's prostate. An ultrasound test uses very high frequency sound waves that are passed through the body. The pattern of reflected sound waves, or "echoes," shows the outline of the prostate. This test can show whether the prostate is enlarged, and whether an abnormal growth that might be **cancer** is present.

Purpose

The prostate is a chestnut-shaped organ surrounding the beginning of the urethra in men. It produces a milky fluid that is part of the seminal fluid discharged during ejaculation. An ultrasound can see if the prostate has become enlarged, which occurs most in men over age 50. Cancer of the prostate also tends to affect older men.

During a **physical examination**, a doctor may perform a **digital rectal examination**. In this examination, the doctor uses a gloved and lubricated finger inserted in the rectum to feel for any abnormalities. If this examination shows that the prostate is enlarged or

a hard lump is present, an ultrasound may be done. Another reason a doctor might perform an ultrasound is if a blood test shows abnormal levels of a substance called prostate-specific antigen (PSA). Abnormal levels of PSA may indicate the presence of cancer.

If there is a suspicious lump, the doctor will want to take a sample of some of the tissue (**prostate biopsy**) to test it to see whether it is in fact cancer. Doing an ultrasound first will show the doctor what part of the prostate should be taken as a sample. Ultrasound can also show whether cancerous tissue is still only within the prostate or whether it has begun to spread to other locations. If **prostate cancer** is present and the doctor decides to treat it with a surgical freezing procedure, ultrasound is used as an aid in the procedure.

An ultrasound can reveal other types of prostate disease as well. For example, it can show if there is inflammation of the prostate (**prostatitis**). Sometimes it is used to determine why a man is unable to father children (**infertility**).

Precautions

A prostate ultrasound study is generally not performed on men who have recently had surgery on their lower bowel. This is because the test requires placing an ultrasound probe about the size of a finger into the rectum.

Description

Prostate ultrasound is generally done using a technique called the transrectal method. This procedure can be done in an outpatient clinic. The cylinder-shaped ultrasound probe is gently placed in the rectum as the patient lies on his left side with the knees bent. The probe is rocked back and forth to obtain images of the entire prostate. The procedure takes about 15–25 minutes to perform. After the test, the patient's doctor can be notified right away, and usually he or she will have a written report within 36 hours.

Preparation

To prepare for a prostate ultrasound, an enema is taken two to four hours before the exam. The patient should not urinate for one hour before the test. If biopsies may be done, the doctor will prescribe an antibiotic that usually is taken in four doses starting the night before the biopsy, the morning of the test, that evening, and the following morning.

KEY TERMS

Benign prostatic hypertrophy (BPH)—Benign prostatic hypertrophy is an enlargement of the prostate that is not cancerous. However, it may cause problems with urinating or other symptoms.

Prostate-specific antigen (PSA)—A substance that often is produced by cancers of the prostate. It can be detected in a blood test.

Urethra—The tube through which urine passes from the bladder and is excreted to outside the body.

Aftercare

There is some discomfort, but less than most patients expect. In fact, worrying ahead of time is usually the hardest part. Generally, the patient is allowed to leave after a radiologist or urologist has reviewed the results. There may be some mucus or a small amount of bleeding from the rectum after the ultrasound. Some patients notice a small amount of blood in the urine for up to two days after the test. Blood may also be present in the semen. As long as the amount of blood is small, there is no cause for concern.

Risks

There are no serious risks from a prostate ultrasound study. Infection is rare and probably is a result of biopsy rather than the sonogram itself. If the ultrasound probe is moved too vigorously, some bleeding may continue for a few days.

Normal results

Modern ultrasound techniques can display both the smooth-surfaced outer shell of the prostate and the core tissues surrounding the urethra. The entire volume of the prostate should be less than 20 milliliters, and its outline should appear as a smooth, echo-reflecting (echogenic) rim. Some irregularities within the substance of the gland and **calcium** deposits are normal findings.

Abnormal results

An **enlarged prostate** with dimmed echoes may indicate either prostatitis or benign enlargement of the gland, called benign prostatic hypertrophy (BPH). A distinct lump of tissue more likely means cancer.

Cancer also often appears as an irregular area within the gland that distorts the normal pattern of echoes. In either case, a biopsy should clarify the diagnosis.

ORGANIZATIONS

American Urological Association Foundation, 1000

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Prostatectomy

Definition

Prostatectomy refers to the surgical removal of part of the prostate gland (transurethral resection, a procedure performed to relieve urinary symptoms caused by benign enlargement), or all of the prostate (radical prostatectomy, the curative surgery most often used to treat **prostate cancer**).

Purpose

Benign disease

When men reach their mid-40s, the prostate gland begins to enlarge. This condition, benign prostatic hyperplasia (BPH), is present in more than half of men in their 60s and as many as 90% of those over 90. Because the prostate surrounds the urethra, the tube leading urine from the bladder out of the body, the enlarging prostate narrows this passage and makes urination difficult. The bladder does not empty completely each time a man urinates, and, as a result, he must urinate with greater frequency, night and day. In time, the bladder can overfill, and urine escapes from the urethra, resulting in incontinence. An operation called transurethral resection of the prostate (TURP) relieves symptoms of BPH by removing the prostate tissue that is blocking the urethra. No incision is needed. Instead a tube (retroscope) is passed through the penis to the level of the prostate, and tissue is either removed or destroyed so that urine can freely pass from the body.

Malignant disease

Prostate **cancer** is the single most common form of non-skin cancer in the United States and the most common cancer in men over 50. Half of men over 70 and almost all men over the age of 90 have prostate

cancer, and the American Cancer Society estimates that almost 218,000 new cases will be diagnosed in 2010. This condition does not always require surgery. In fact, many elderly men adopt a policy of “watchful waiting,” especially if their cancer is growing slowly. Younger men often elect to have their prostate gland totally removed along with the cancer it contains—an operation called radical prostatectomy. The two main types of this surgery, radical retropubic prostatectomy and radical perineal prostatectomy, are performed only on patients whose cancer is limited to the prostate. If cancer has broken out of the capsule surrounding the prostate gland and spread in the area or to distant sites, removing the prostate will not prevent the remaining cancer from growing and spreading throughout the body.

Precautions

Potential complications of TURP include bleeding, infection, and reactions to general or **local anesthesia**. About one man in five will need to have the operation again within 10 years.

Open (incisional) prostatectomy for cancer should not be done if the cancer has spread beyond the prostate, as serious side effects may occur without the benefit of removing all the cancer. If the bladder is retaining urine, it is necessary to insert a catheter before starting surgery. Patients should be in the best possible general condition before radical prostatectomy. Before surgery, the bladder is inspected using an instrument called a cystoscope to help determine the best surgical technique to use, and to rule out other local problems.

Description

TURP

This procedure does not require an abdominal incision. With the patient under either general or spinal anesthesia, a cutting instrument or heated wire loop is inserted to remove as much prostate tissue as possible and seal blood vessels. The excised tissue is washed into the bladder, then flushed out at the end of the operation. A catheter is left in the bladder for one to five days to drain urine and blood. Advanced laser technology enables surgeons to safely and effectively burn off excess prostate tissue blocking the bladder opening with fewer of the early and late complications associated with other forms of prostate surgery. This procedure can be performed on an outpatient basis, but urinary symptoms do not improve until swelling subsides several weeks after surgery.

Radical prostatectomy

RADICAL RETROPUBLIC PROSTATECTOMY. This is a useful approach if the prostate is very large or cancer is suspected. With the patient under general or spinal anesthesia or an epidural, a horizontal incision is made in the center of the lower abdomen. Some surgeons begin the operation by removing pelvic lymph nodes to determine whether cancer has invaded them, but recent findings suggest there is no need to sample them in patients whose likelihood of lymph node metastases is less than 18%. A doctor who removes the lymph nodes for examination will not continue the operation if they contain cancer cells, because the surgery will not cure the patient. Other surgeons remove the prostate gland before examining the lymph nodes. A tube (catheter) inserted into the penis to drain fluid from the body is left in place for 14–21 days.

Originally, this operation also removed a thin rim of bladder tissue in the area of the urethral sphincter—a muscular structure that keeps urine from escaping from the bladder. In addition, the nerves supplying the penis often were damaged, and many men found themselves impotent (unable to achieve erections) after prostatectomy. A newer surgical method called potency-sparing radical prostatectomy preserves sexual potency in 75% of patients and fewer than 5% become incontinent following this procedure.

RADICAL PERINEAL PROSTATECTOMY. This procedure is just as curative as radical retropubic prostatectomy but is performed less often because it does not allow the surgeon to spare the nerves associated with erection or, because the incision is made above the rectum and below the scrotum, to remove lymph nodes. Radical perineal prostatectomy is sometimes used when the cancer is limited to the prostate and there is no need to spare nerves or when the patient's health might be compromised by the longer procedure. The perineal operation is less invasive than retropubic prostatectomy. Some parts of the prostate can be seen better, and blood loss is limited. The absence of an abdominal incision allows patients to recover more rapidly. Many urologic surgeons have not been trained to perform this procedure. Radical prostatectomy procedures last one to four hours, with radical perineal prostatectomy taking less time than radical retropubic prostatectomy. The patient remains in the hospital three to five days following surgery and can return to work in three to five weeks. Ongoing research indicates that laparoscopic radical prostatectomy may be as effective as open surgery in treatment of early-stage disease.

Cryosurgery

Also called **cryotherapy** or cryoablation, this minimally invasive procedure uses very low temperatures to freeze and destroy cancer cells in and around the prostate gland. A catheter circulates warm fluid through the urethra to protect it from the cold. When used in connection with ultrasound imaging, cryosurgery permits very precise tissue destruction. Traditionally used only in patients whose cancer had not responded to radiation, but now approved by Medicare as a primary treatment for prostate cancer, cryosurgery can safely be performed on older men, on patients who are not in good enough general health to undergo radical prostatectomy, or to treat recurrent disease. Recent studies have shown that total cryosurgery, which destroys the prostate, is at least as effective as radical prostatectomy without the trauma of major surgery.

Preparation

As with any type of major surgery done under **general anesthesia**, the patient should be in optimal condition. Most patients having prostatectomy are in the age range when cardiovascular problems are frequent, making it especially important to be sure that the heart is beating strongly, and that the patient is not retaining too much fluid. Because long-standing prostate disease may cause kidney problems from urine “backing up,” it also is necessary to be sure that the kidneys are working properly. If not, a period of catheter drainage may be necessary before doing the surgery.

Aftercare

Following TURP, a catheter is placed in the bladder to drain urine and remains in place for two to three days. A solution is used to irrigate the bladder and urethra until the urine is clear of blood, usually within 48 hours after surgery. Whether **antibiotics** should be routinely given remains an open question. Catheter drainage also is used after open prostatectomy. The bladder is irrigated only if **blood clots** block the flow of urine through the catheter. Patients are given intravenous fluids for the first 24 hours, to ensure good urine flow. Patients resting in bed for long periods are prone to blood clots in their legs (which can pass to the lungs and cause serious breathing problems). This can be prevented by elastic stockings and by periodically exercising the patient’s legs. The patient remains in the hospital one to two days following surgery and can return to work in one to two weeks.

KEY TERMS

BPH—Benign prostatic hypertrophy, a very common noncancerous cause of prostatic enlargement in older men.

Catheter—A tube that is placed through the urethra into the bladder in order to provide free drainage of urine and blood following either TURP or open prostatectomy.

Cryosurgery—In prostatectomy, the use of a very low-temperature probe to freeze and thereby destroy prostatic tissue.

Impotence—The inability to achieve and sustain penile erections.

Incontinence—The inability to retain urine in the bladder until a person is ready to urinate voluntarily.

Prostate gland—The gland surrounding the male urethra just below the base of the bladder. It secretes a fluid that constitutes a major portion of the semen.

Urethra—The tube running from the bladder to the tip of the penis that provides a passage for eliminating urine from the body.

Risks

The complications and side effects that may occur during and after prostatectomy include:

- Excessive bleeding, which in rare cases may require blood transfusion.
- Incontinence caused by damage to the sphincter (the muscular valve that keeps urine in the bladder) during retropubic prostatectomy.
- Impotence, occurring when nerves to the penis are injured during the retropubic operation. The “nerve-sparing” technique has drastically cut down on this problem.
- Some patients who receive a large volume of irrigating fluid after TURP develop high blood pressure, vomiting, trouble with their vision, and mental confusion. This condition is caused by a low salt level in the blood, and is reversed by giving salt solution.
- A permanent narrowing of the urethra called a stricture occasionally develops when the urethra is damaged during TURP.
- There is about a 34% chance that the cancer will recur within 10 years of the procedure. In addition, about 25% of patients experience what is known as

biochemical recurrence, which means that the level of prostate-specific antigen (PSA) in the patient's blood serum begins to rise rapidly. Recurrence of the tumor or biochemical recurrence can be treated with radiation therapy or androgen deprivation therapy.

Normal results

In patients with BPH who have the TURP operation, urination should become much easier and less frequent, and dribbling or incontinence should cease. In patients having radical prostatectomy for cancer, a successful operation will remove the tumor and prevent its spread to other areas of the body (metastasis). If examination of lymph nodes shows that cancer has spread beyond the prostate at the time of surgery, other measures are available to control the tumor.

Resources

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ORGANIZATIONS

- American Urological Association Foundation, 1000 Corporate Blvd., Linthicum, MD, 21090, (410) 689-3700, (410) 689-3800, (866) 746-4282, aufoundation@auafoundation.org, <http://www.urologyhealth.org>.

Cancer Research Institute (National Headquarters), One Exchange Plaza, 55 Broadway, Suite 1802, New York, NY, 10006, (212) 688-7515, (212) 832-9376, (800) 992-2623, <http://www.cancerresearch.org>.

National Prostate Cancer Coalition, 1154 Fifteenth Street, NW, Washington, DC, 20005, (202) 463-9455, (202) 463-9456, (888) 245-9455, info@fightprostatecancer.org, <http://www.zerocancer.org>.

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Prostate-specific antigen test

Definition

Prostate-specific antigen, or PSA, is a protein produced by the prostate gland that may be found in elevated levels in the blood when a person develops certain diseases of the prostate, notably **prostate cancer**. PSA is *specific*, because it is present only in prostate tissue. It is not specific for prostate *cancer*, however, as it may also be elevated in men with benign enlargement of this organ.

Purpose

The blood test for PSA is used to screen men to detect prostate **cancer** at an early stage, and also to monitor its response to treatment. After lung cancer, prostate cancer is the most common form of cancer in men in the United States. Any routine physical exam of a man aged 50 and older should include a **digital rectal examination** (DRE), in which the doctor's finger probes the surface of the prostate gland to detect any suspicious area of hardness or a tumor mass. PSA testing may be ordered with or without DRE. If the PSA test is positive, a sample of prostate tissue (biopsy) may be taken to confirm that cancer is present. If negative, the test may be repeated immediately to confirm the diagnosis, or repeated the next year. Many physicians today routinely do both a DRE and a PSA test each year on their older male patients, so that if cancer does develop, it will be found at an early stage, making it easier to treat. These tests may be ordered in men younger than age 50 if they are considered to be at high risk for the development of prostate cancer. The combination of a DRE and a PSA test can detect approximately 80% of all prostate cancers.

At present, the PSA test is widely accepted as a way of telling whether a patient with cancer is

KEY TERMS

Antibody—A substance formed in the body in reaction to some foreign material invading the body, or sometimes to diseased body tissue such as prostate cancer. An antibody also may be prepared in the laboratory and used to measure the amount of antigen in the blood.

Antigen—Either a foreign substance such as a virus or bacterium, or a protein produced by diseased or injured body tissue.

Biopsy—A procedure using a hollow needle to obtain a small sample of tissue, such as from the prostate. Often done to determine whether cancer is present.

BPH—Benign prostatic hyperplasia, a noncancerous disorder that causes the prostate to enlarge.

responding to treatment. Because only the prostate produces PSA, its presence in the blood following complete removal of the prostate (radical **prostatectomy**) indicates that some cancer has been left behind.

Description

The PSA test is a radioimmunoassay. Any antigen causes the body to produce antibodies in an attempt to neutralize or eliminate the antigen, often a substance that harms body tissues. In the laboratory, a sample of the patient's blood is exposed to the antibody against PSA, so that the amount of antigen (PSA) can be measured. The results generally are available the next day.

Preparation

No special measures are needed when doing a PSA test other than taking the usual precautions to prevent infection at the needle puncture site.

Aftercare

There are no specific aftercare requirements related to this test.

Risks

There are no specific risks associated with the PSA test.

Although the level of PSA usually is elevated in men with prostate cancer, it also may be abnormally high (though usually not *as* high) in men with non cancerous enlargement of the prostate (benign

prostatic hyperplasia or BPH). If thousands of men have the PSA test routinely each year, many of them will have unnecessary tests (such as biopsy or an ultrasound study) to confirm cancer. If a "false-positive" result is obtained, where the PSA level seems high but really is not, some men may even be treated for prostate cancer when no cancer is present.

Results

Normal results

Each laboratory has its own normal range for PSA. In fact, laboratories may redefine the normal range whenever starting to use a new batch of test chemicals. A PSA level of 4.0 ng/mL or greater in men who are not considered to be at high risk for the development of prostate cancer is a common level that may trigger a need for additional testing to rule out or confirm the presence of prostate cancer. A PSA level that falls between 2.5 and 4.0 ng/mL, especially in men who are considered to be at high risk for the development of prostate cancer, may indicate a need for additional testing in this high risk group.

Abnormal results

Some experts believe that more than 90% of men with prostate cancer will have an elevated PSA level. Others claim that as many as one-third of cancers will be missed. The amount of PSA in the blood drops when cancer is successfully treated, but rises again if the tumor recurs, especially if it spreads to other parts of the body. A new variation of the PSA test shows how much of the material is bound to other protein in the blood and how much is "free." This procedure may be more accurate and could well indicate whether either prostate cancer or BPH is present.

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Prostatic acid phosphatase test see **Acid phosphatase test**

Prostatitis

Definition

Prostatitis is an inflammation of the male prostate gland. There are three types of prostatitis:

- Acute bacterial prostatitis develops rapidly.
- Chronic bacterial prostatitis develops gradually, lasts longer, and usually has less severe symptoms.
- Chronic nonbacterial or type III prostatitis, which used to be called prostodynia and is now referred to as chronic pelvic pain syndrome, is the most common type of prostatitis.

Demographics

Prostatitis is one of the most common urologic diseases, accounting for about 40% of visits to a urologist in the United States. As many as 50% of men may experience prostatitis at some point in their lives. It is estimated that chronic prostatitis accounts for up to two million physician visits each year in the United States, primarily for the nonbacterial form. Although prostatitis is rare in young boys, bacterial prostatitis can affect males of any age. Chronic bacterial prostatitis is most common in young and middle-aged men. Acute bacterial prostatitis is the least common type.

Description

The prostate is a walnut-shaped male reproductive gland that surrounds the urethra at the neck of the bladder and supplies fluid for semen. The urethra is the tube that carries urine from the bladder to the outside of the body. Inflammation of the prostate from prostatitis results in blockages in its tiny ducts, causing the build up of secretions and swelling of the prostate. Prostatitis is one of the three major disorders of the prostate. The others are prostate enlargement and **cancer**.

Risk factors

Risk factors for bacterial prostatitis include:

- multiple sexual partners
- anal intercourse, especially without a condom
- recent catheterization or insertion of another instrument into the urethra
- recent urinary tract infection
- abnormality of the urinary tract

Additional risk factors for chronic bacterial prostatitis include:

- age over 30
- excessive alcohol use

- injury to the perineum—the area between the scrotum and anus

Possible risk factors for nonbacterial prostatitis include:

- infection from bacteria, viruses, fungi, or parasites called trichomonads
- irritation from urine backing up into the prostate
- exposure to chemicals
- a nerve disorder affecting the lower urinary tract
- problems with toilet training
- sexual abuse
- stress

Causes and symptoms

Bacterial prostatitis is caused by bacteria from infected urine flowing backward from the urethra into the prostate ducts. Acute bacterial prostatitis can be caused by any bacterium that causes a **urinary tract infection**, including:

- *Escherichia coli* (*E. coli*)
- *Staphylococcus aureus*
- *Pseudomonas aeruginosa*
- *Klebsiella pneumonia*
- *Proteus mirabilis*
- *Enterococci*

Prostatitis caused by *E. coli* or other bacteria can occur spontaneously or in association with:

- anal intercourse
- urinary tract infections
- epididymitis—an inflammation of the ducts from the testes
- urethritis—inflammation of the urethra
- obstruction of the bladder outlet
- catheterization
- cystoscopy
- prostate biopsy
- trauma
- abnormality of the foreskin
- transurethral surgery

Some sexually transmitted infections (STIs) can cause acute prostatitis, typically in men younger than age 35. These STIs include:

- gonorrhea
- chlamydia
- trichomonas
- *Ureaplasma urealyticum*

Chronic bacterial prostatitis is most often caused by:

- *E. coli*
- *Klebsiella pneumonia*
- *Enterobacter cloacae*
- *Proteus* species

Chronic bacterial prostatitis often develops from an acute prostatitis infection or is associated with:

- recurrent urinary tract infections
- epididymitis
- urethritis

The exact cause of chronic nonbacterial prostatitis/chronic pelvic **pain** syndrome is unknown. Possible causes include:

- persistent infection with a bacteria, virus, or yeast
- pelvic inflammation or nerve or muscle abnormalities or spasms
- an autoimmune disorder or other inappropriate immune system response
- a uric acid disorder
- prostate stones
- constriction of the urethra
- benign prostatic hyperplasia (BPH)—a noncancerous growth
- prostate cancer
- food allergy
- stress
- irregular sexual activity

Symptoms of prostatitis vary greatly. Inflammation of the prostate can also occur without symptoms. Acute bacterial prostatitis usually develops rapidly with urinary symptoms, pain in the perineum or lower back, and **fever** and chills. Chronic prostatitis symptoms usually develop more slowly and are less severe. Some patients have ongoing mild symptoms or symptomatic episodes.

Signs and symptoms of prostatitis may include:

- Difficulties with urination. Most urinary problems are caused by the swollen prostate blocking the urethra. Patients feel the need to urinate frequently, often urgently and at night. Urination may be painful. It is difficult to start the flow of urine and to totally empty the bladder. The urine stream may be weak or split. Dribbling or incontinence may occur after attempts to urinate. Urination may also be infrequent. With severe prostatitis blood or small stones of calcium may be passed in the urine.
- Pain. In addition to pain when urinating caused by prostate swelling, stimulation of nerves in the prostate gland may cause pain in the penis, one or both

KEY TERMS

Catheterization—The placing of a flexible tube into the urethra or other body part.

Culture—A test in which a sample of body fluid, such as prostatic fluid, is grown to identify an organism causing infection.

Cystoscopy—The passing of a viewing instrument called a cystoscope up the urethra into the region of the prostate.

Ejaculation—The process by which semen is ejected by the erect penis.

Granuloma—A mass of chronically inflamed tissue.

Perineum—An area close to the prostate, between the scrotum and anus.

Prostate—The walnut-shaped gland that surrounds the urethra at the neck of the bladder in males and supplies fluid for semen.

Urethra—The tube in the penis that discharges urine from the bladder to the outside of the body.

testicles, the lower stomach, the lower back, and the perineum. Some patients experience pain during or after ejaculation, while sitting or walking, or during bowel movements.

- Sex and fertility. Prostatitis pain can make it impossible to enjoy sex. Men with prostatitis may be troubled by early release of sperm (premature ejaculation). Occasionally there is blood in the semen. Some of the drugs prescribed to ease the flow of the urine can dampen sexual desire. Because normal prostate secretions contribute to semen, prostatitis may severely reduce the number of sperm and make them less mobile, thereby lowering fertility.
- General health and psychological problems. These include fatigue, malaise, and depression.

Diagnosis

Examination

Acute prostatitis can usually be diagnosed from the symptoms and a **physical examination**. Diagnosis of chronic prostatitis can be much more difficult. The family physician or urologist will perform a digital rectal exam (DRE) using a lubricated gloved finger in the rectum to feel the prostate, which may be enlarged or tender. Physical examination may also reveal:

- urethral discharge
- enlarged or tender lymph nodes in the groin
- swelling and tenderness of the scrotum

Tests

- Gently squeezing the prostate produces a few drops of fluid that can be cultured to identify organisms that are causing infection and determine an appropriate treatment. Infected fluid also typically contains a large number of white blood cells for fighting infection. However, excessive pressure on the prostate can force bacteria into the blood and cause serious systemic or general infection. Other tests may include:
- examining and culturing a urine sample for concurrent infection
 - a prostate specific antigen (PSA) blood test, since prostatitis, as well as prostate cancer, can increase PSA levels
 - semen analysis

Procedures

- Procedures used to check for prostatitis include:
- cystoscopy—the insertion of a special instrument called a cystoscope into the penis to directly examine the prostate for inflammation
 - transrectal ultrasound
 - urine flow studies to measure the pressure of the flow and detect problems with the prostate, urethra, or pelvic muscles

Treatment

Traditional

- Nonbacterial prostatitis or prostatitis that does not respond to **antibiotics** may require other treatment:
- Balloon dilation—in which a collapsed balloon is inserted at the obstructed site and inflated—can temporarily widen a narrowed urethra.
 - Suprapubic catheterization can drain the bladder through the abdomen if the swollen prostate is severely restricting urine flow through the urethra.
 - Rarely—and usually only in older men—the prostate is surgically removed, sometimes by a minimally invasive laparoscopic prostatectomy.
 - Psychiatric treatment may be required for serious psychological problems resulting from prostatitis.

Drugs

Acute bacterial prostatitis is usually initially treated with broad-spectrum antibiotics—most often

trimethoprim-sulfamethoxazole (Bactrim or Septra), **fluoroquinolones** (Floxin or Cipro), or tetracycline or a tetracycline derivative such as doxycycline—for at least four weeks. Prostatitis caused by an STI is usually treated with a ceftriaxone injection, followed by a seven-day course of doxycycline. Severe acute prostatitis may require intravenous antibiotics and hospitalization. Chronic bacterial prostatitis is usually treated with antibiotics for one to three months or longer. Drugs are also used to treat fungal and parasitic infections of the prostate.

Other drugs used to treat prostatitis include:

- nonsteroidal anti-inflammatory medications (NSAIDs), such as aspirin or ibuprofen
- steroid anti-inflammatories
- alpha-adrenergic blockers or muscle relaxants to reduce muscle tension and ease urine flow, including doxazosin (Cardura), tamulosin (Flomax), or terazosin (Hytrin)
- pain medication
- allopurinol to reduce uric acid levels
- stool softeners to ease painful bowel movements
- diazepam (Valium) or another tranquilizer for stress

Alternative

There are various alternative treatments for prostatitis, which may be used in conjunction with antibiotics for bacterial infections:

- prostate drainage or massage, in which a finger is inserted into the rectum at regular intervals to exert pressure on the prostate and drain the ducts
- acupuncture and Chinese herbal medicine
- saw palmetto (*Serenoa repens*) to support the prostate
- quercetin and/or pollen extract (Cernitin)
- nutritional supplements thought to support the prostate and help reduce pain and promote healing, including zinc, omega-3 fatty acids, several amino acids, and anti-inflammatory nutrients and herbs
- hot and cold contrast sitz baths to help reduce inflammation
- biofeedback or relaxation techniques
- pelvic physical therapy including trigger point release
- pelvic floor muscle relaxation and strengthening techniques

Alternative treatments should be used with caution, as the benefits of many such treatments have not been confirmed by scientific research.

Home remedies

Home remedies for relieving symptoms of prostatitis are especially helpful for chronic prostatitis or until antibiotics have taken effect. Home remedies include:

- Heat. Exposing the perineum to very hot water with a sitz bath or hot water bottle or using a heating pad for 20 minutes or longer can relieve pain.
- Ice. When heat is ineffectual, ice packs or simply placing a small ice cube in the rectum may relieve pain for hours.
- Water. Although the discomfort of frequent urination may cause a patient to reduce his fluid intake, this can cause dehydration and increase the risk of bladder infection. Patients should continue to drink plenty of water (64–128 ounces daily) and urinate frequently and completely.
- Diet. Most doctors recommend reducing or eliminating caffeine (as in coffee or tea), alcohol, citrus juices, and all hot, spicy, or acidic foods, which may irritate the bladder. Constipation should be avoided because large, hard bowel movements may press on the swollen prostate and cause severe pain. Bran cereals and whole-grain breads are helpful for constipation.
- Exercise. It is especially important for patients with chronic prostatitis to maintain their activity levels. Walking is often helpful, although sometimes it can make the pain worse. Some exercises, such as bicycle riding, should be avoided.
- Frequent ejaculation. Ejaculating two or three times a week is often recommended, especially when taking antibiotics.

Prognosis

Acute bacterial prostatitis is usually relieved rapidly with antibiotics. Approximately 75% of chronic bacterial prostatitis cases are cured with a long course of antibiotics. Antibiotics often relieve symptoms of nonbacterial prostatitis as well, because many of them have direct anti-inflammatory actions. However, most antibiotics do not readily reach the prostate, and infection may recur or develop into chronic prostatitis, which has a much worse prognosis. Failure to control acute prostatitis can lead to complications including a prostatic **abscess**, kidney infection, or blood infection (septicemia). Even when chronic prostatitis cannot be cured, urinary symptoms can be controlled and quality of life can usually be maintained with low doses of antibiotics and other measures.

Prevention

The best prevention for bacterial prostatitis is to avoid potential sources of infection. This includes good perineal hygiene to prevent urinary tract infections and practicing safer sex to avoid STIs. Sex should be avoided whenever a female partner has an active bacterial infection of the vagina. Prompt treatment for infections of the kidneys, bladder, or other genitourinary organs can prevent prostatitis. The best way to prevent chronic prostatitis is to treat an initial acute episode promptly and effectively with a full course of antibiotics.

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ORGANIZATIONS

- American Urological Association (AUA), 1000 Corporate Blvd., Linthicum, MD, 21090, (410) 689-3700, (866)

RING-AUA (746-4282), (410) 689-3800, aua@AUAnet.org, <http://www.auanet.org>.

National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), Building 31, Room 9A06, 31 Center Dr., MSC 2560, Bethesda, MD, 20892-2560, (301) 496-3583, <http://www2.niddk.nih.gov>.

Prostatitis Foundation (PF), 1063 30th St., Smithshire, IL, 61478, (888) 891-4200, <http://www.prostatitis.org>.

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Prosthetic joint infection see **Infectious arthritis**

Protease inhibitors

Definition

A protease inhibitor is a type of drug that cripples the enzyme protease. An enzyme is a substance that triggers chemical reactions in the body. The human **immunodeficiency virus (HIV)** uses protease in the final stages of its reproduction (replication) process.

Purpose

The drug is used to treat selected patients with HIV infection. Blocking protease interferes with HIV reproduction, causing it to make copies of itself that cannot infect new cells. The drug may improve symptoms and suppress the infection but does not cure it.

Precautions

Patients should not discontinue this drug even if symptoms improve without consulting a doctor.

These drugs do not necessarily reduce the risk of transmitting HIV to others through sexual contact, so patients should avoid sexual activities or use **condoms**.

Description

Protease inhibitors are considered one of the most potent medications for HIV developed so far.

This class of drugs includes indinavir (Crixivan), ritonavir (Norvir), nelfinavir (Viracept), amprenavir (Agenerase), lopinavir plus ritonavir (Kaletra), saquinavir (Fortovase), and atazanavir (Reyataz). Reyataz received approval from the U.S. Food and Drug Administration (FDA) in mid-2003 and was the first

KEY TERMS

Human immunodeficiency virus (HIV)—The virus that causes AIDS.

protease inhibitor approved for once-daily dosing. Several weeks or months of drug therapy may be required before the full benefits are apparent.

The drug should be taken at the same time each day. Some types should be taken with a meal to help the body absorb them. Each of the types of protease inhibitor may have to be taken in a different way. In most cases, protease inhibitors are part of a combination therapy, used in conjunction with other classes of HIV drugs.

Risks

Common side effects include **diarrhea**, stomach discomfort, **nausea**, and mouth sores. Less often, patients may experience rash, muscle **pain**, **headache**, or weakness. Rarely, there may be confusion, severe skin reaction, or seizures. Some of these drugs can have interactions with other medication, and indinavir can be associated with **kidney stones**. Diabetes or high blood pressure may become worse when these drugs are taken. Reyataz has been shown to have fewer side effects than some protease inhibitors, though it can interact with other medications, including certain heart medications and antidepressants.

Experts do not know whether the drugs pass into breast milk, so **breastfeeding** mothers should avoid them or should stop nursing until the treatment is completed.

Resources

PERIODICALS

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LoBuono, Charlotte. "FDA Gives Nod to First Once-daily Protease Inhibitor." *Drug Topics* July 21, 2003: 16.

ORGANIZATIONS

National AIDS Treatment Advocacy Project, 580 Broadway, Ste. 1010, New York, NY, 10012, (212) 219-0106, (212) 219-8473, (866) 26-NATAP, info@natap.org, <http://www.natap.org>.

Carol A. Turkington
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Protein-calorie malnutrition see
Protein-energy malnutrition

Protein components test

Definition

Protein components tests measure the amounts and types of protein in the blood. Proteins are constituents of muscle, enzymes, hormones, transport proteins, hemoglobin, and other functional and structural elements of the body. Albumin and globulin make up most of the protein within the body and are measured in the total protein of the blood and other body fluids. Thus, the serum (blood) protein components test measures the total protein, as well as its albumin and globulin components in the blood.

Purpose

The protein components test is used to diagnose diseases that either affect proteins as a whole, or that involve a single type of protein. The test is also used to monitor the course of disease in certain cancers, intestinal and kidney protein-wasting states, immune disorders, liver dysfunction, and impaired **nutrition**.

Precautions

Drugs that may cause increased protein levels include the anabolic **steroids**, androgens (male hormones), growth hormone, insulin, and progesterone. Drugs that may decrease protein levels include estrogen, drugs poisonous to the liver, and **oral contraceptives**.

Description

Proteins are large molecules (complex organic compounds) that consist of amino acids, sugars, and lipids. There are two main types of proteins: those that are made of fiber and form the structural basis of body tissues, such as hair, skin, muscle, tendons, and cartilage; and globular proteins (generally water soluble), which interact with hormones, various other proteins in the blood (including hemoglobin and antibodies), and all the enzymes (substances that promote biochemical reactions in the body).

Proteins are needed in the diet to supply the body with amino acids. Ingested proteins are broken down in the digestive system to amino acids, which are then absorbed and rebuilt into new body proteins. One of the most important functions of proteins in the body is

to contribute to the osmotic pressure (the movement of water between the bloodstream and tissues). An example of this is seen in diseases that result in damage to the filtering units of the kidneys (**nephrotic syndrome**). A severe loss of protein from the bloodstream into the urine (proteinuria) results, lowering the protein content of the blood and resulting in fluid retention, or **edema**.

Albumin and globulin are two key components of protein. Albumin is made in the liver and constitutes approximately 60% of the total protein. The main function of albumin is to maintain osmotic pressure and to help transport certain blood constituents around the body via the bloodstream. Because albumin is made in the liver, it is one element that is used to monitor liver function.

Globulin is the basis for antibodies, glycoproteins (protein-carbohydrate compounds), lipoproteins (proteins involved in fat transport), and clotting factors. Globulins are divided into three main groups, the alpha, beta, and gamma globulins. Alpha globulins include enzymes produced by the lungs and liver, and haptoglobin, which binds hemoglobin together. The beta globulins consist mostly of low-density lipoproteins (LDLs), substances involved in fat transport. All of the gamma globulins are antibodies, proteins produced by the immune system in response to infection, during allergic reaction, and after organ transplants.

Both serum albumin and globulin are measures of nutrition. Malnourished patients, especially after surgery, demonstrate greatly decreased protein levels, while burn patients and those who have protein-losing syndromes show low levels despite normal synthesis. **Pregnancy** in the third trimester is also associated with reduced protein levels.

The relationship of albumin to globulin is determined by ratio, so when certain diseases cause the albumin levels to drop, the globulin level will be increased by the body in an effort to maintain a normal total protein level. For example, when the liver is unable to synthesize sufficient albumin in chronic **liver disease**, the albumin level will be low, but the globulin levels will be normal or higher than normal. In such cases, the protein components test is an especially valuable diagnostic aid because it determines the ratio of albumin to globulin, as well as the total protein level. It should be noted, however, that when globulin is provided as a calculation (total protein – albumin = globulin), the result is much less definitive than other methods of determining globulin.

Consequently, when the albumin/globulin ratio (A/G ratio) is less than 1.0, more precise tests should

KEY TERMS

Nephrotic syndromes—A collection of symptoms that result from damage to the filtering units of the kidney (glomeruli), causing severe loss of protein from the blood into the urine.

be ordered. These tests include **protein electrophoresis**, a method of separating the different blood proteins into groups. If the protein electrophoresis indicates a rise, or “spike” at the globulin level, an even more specific test for globulins, called **immunolectrophoresis**, should be ordered to separate out the various globulins according to type. Some diseases characterized by dysproteinemia (derangement of the protein content of the blood) have typical electrophoretic globulin peaks.

Preparation

Unless this is requested by the physician, there is no need that the patient restrict food or fluids before the test.

Risks

Risks posed by this test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or lightheadedness after venipuncture, or hematoma (blood accumulating under the puncture site).

Normal results

Reference values vary from laboratory to laboratory, but can generally be found within the following ranges: total protein: 6.4–8.3 g/dL; albumin: 3.5–5.0 g/dL; globulin: 2.3–3.4 g/dL.

Abnormal results

Increased total protein levels are seen in **dehydration**, in some cases of chronic liver disease (like **autoimmune hepatitis** and **cirrhosis**), and in certain tropical diseases (for example, **leprosy**). Very low total protein levels (less than 4.0 g/dL) and low albumin cause the edema (water retention) usually seen in nephrotic syndromes. Decreased protein levels may be seen in pregnancy, chronic **alcoholism**, prolonged **immobilization**, **heart failure**, **starvation**, and malabsorption or **malnutrition**.

Increased albumin levels are found in dehydration. Decreased albumin levels are indicative of liver disease, protein-losing syndromes, malnutrition,

inflammatory disease, and familial idiopathic (of unknown cause) dysproteinemia, a genetic disease in which the albumin is significantly reduced and globulins increased.

Increased globulin levels are found in **multiple myeloma** and **Waldenström's macroglobulinemia**, two cancers characterized by overproduction of gammaglobulin from proliferating plasma cells. Increased globulin levels are also found in chronic inflammatory diseases such as **rheumatoid arthritis**, acute and chronic infection, and cirrhosis. Decreased globulin levels are seen in genetic immune disorders and secondary immune deficiency.

Resources

BOOKS

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Janis O. Flores

Protein electrophoresis

Definition

Electrophoresis is a technique used to separate different elements (fractions) of a blood sample into individual components. Serum protein electrophoresis (SPEP) is a screening test that measures the major blood proteins by separating them into five distinct fractions: albumin, alpha₁, alpha₂, beta, and gamma proteins. Protein electrophoresis can also be performed on urine.

Purpose

Protein electrophoresis is used to evaluate, diagnose, and monitor a variety of diseases and conditions. It can be used for these purposes because the levels of different blood proteins rise or fall in response to such disorders as **cancer**, intestinal or kidney protein-wasting syndromes, disorders of the immune system, liver dysfunction, impaired **nutrition**, and chronic fluid-retaining conditions.

Precautions

Certain other diagnostic tests or prescription medications can affect the results of SPEP tests. The administration of a contrast dye used in some other tests may falsely elevate protein levels. Drugs that can

alter results include **aspirin**, bicarbonates, chlorpromazine (Thorazine), **corticosteroids**, isoniazid (INH), and neomycin (Mycifradin).

Description

Proteins are major components of muscle, enzymes, hormones, hemoglobin, and other body tissues. Proteins are composed of elements that can be separated from one another by several different techniques: chemical methods, ultracentrifuge, or electrophoresis. There are two major types of electrophoresis: protein electrophoresis and **immunolectrophoresis**. Immunoelectrophoresis is used to assess the blood levels of specific types of proteins called immunoglobulins. An immunolectrophoresis test is usually ordered if a SPEP test has a “spike,” or rise, at the immunoglobulin level. Protein electrophoresis is used to determine the total amount of protein in the blood, and to establish the levels of other types of proteins called albumin, alpha₁ globulin, alpha₂ globulin, and beta globulin.

Blood proteins

ALBUMIN. Albumin is a protein that is made in the liver. It helps to retain elements like **calcium**, some hormones, and certain drugs in the circulation by binding to them to prevent their being filtered out by the kidneys. Albumin also acts to regulate the movement of water between the tissues and the bloodstream by attracting water to areas with higher concentrations of salts or proteins.

GLOBULINS. Globulins are another type of protein, larger in size than albumin. They are divided into three main groups: alpha, beta, and gamma.

- Alpha globulins. These proteins include alpha₁ and alpha₂ globulins. Alpha₁ globulin is predominantly alpha₁ antitrypsin, an enzyme produced by the lungs and liver. Alpha₂ globulin, which includes serum haptoglobin, is a protein that binds hemoglobin to prevent its excretion by the kidneys. Various other alpha globulins are produced as a result of inflammation, tissue damage, autoimmune disorders, or certain cancers.
- Beta globulins. These include low-density substances involved in fat transport (lipoproteins), iron transport (transferrin), and blood clotting (plasminogen and complement).
- Gamma globulins. All of the gamma globulins are antibodies—proteins produced by the immune system in response to infection, allergic reactions, and organ transplants. If serum protein electrophoresis

has demonstrated a significant rise at the gamma globulin level, immunoelectrophoresis is done to identify the specific globulin that is involved.

Electrophoretic measurement of proteins

All proteins have an electrical charge. The SPEP test is designed to make use of this characteristic. There is some difference in method, but basically the sample is placed in or on a special medium (e.g., a gel), and an electric current is applied to the gel. The protein particles move through the gel according to the strength of their electrical charges, forming bands or zones. An instrument called a densitometer measures these bands, which can be identified and associated with specific diseases. For example, a decrease in albumin with a rise in the alpha₂ globulin usually indicates an acute reaction of the type that occurs in infections, **burns**, **stress**, or **heart attack**. On the other hand, a slight decrease in albumin with a slight increase in gamma globulin and a normal alpha₂ globulin is more indicative of a chronic inflammatory condition, as might be seen in **cirrhosis** of the liver.

Protein electrophoresis is performed on urine samples to classify kidney disorders that cause protein loss. Certain band patterns are specific to different diseases. For example, the identification of a specific protein called the Bence Jones protein (by performing the **Bence Jones protein test**) during the procedure suggests **multiple myeloma**.

Preparation

The serum protein electrophoresis test requires a blood sample. It is not necessary for the patient to restrict food or fluids before the test. The urine protein electrophoresis test requires either an early morning urine sample or a 24-hour urine sample according to the physician's request. The doctor should check to see if the patient is taking any medications that may affect test results.

Risks

Risks posed by the blood test are minimal but may include slight bleeding from the puncture site, **fainting** or lightheadedness after the blood is drawn, or the development of a small bruise at the puncture site.

Normal results

The following values are representative, although there is some variation among laboratories and specific

KEY TERMS

Albumin—A blood protein that is made in the liver and helps to regulate water movement in the body.

Electrophoresis—A technique for separating various blood fractions by running an electric current through a gel containing a blood sample.

Globulins—A group of proteins in blood plasma whose levels can be measured by electrophoresis in order to diagnose or monitor a variety of serious illnesses.

Haptoglobin—A protein in blood plasma that binds hemoglobin.

Immunoglobulins—Any of several types of globulin proteins that function as antibodies.

methods. These values are based on the agarose system:

- total protein: 5.9–8.0 g/dL
- albumin: 4.0–5.5 g/dL
- alpha₁ globulin: 0.15–0.25 g/dL
- alpha₂ globulin: 0.43–0.75 g/dL
- beta globulin: 0.5–1.0 g/dL
- gamma globulin: 0.6–1.3 g/dL

Abnormal results

Albumin levels are increased in **dehydration**. They are decreased in **malnutrition**, **pregnancy**, **liver disease**, inflammatory diseases, and such protein-losing syndromes as **malabsorption syndrome** and certain kidney disorders.

Alpha₁ globulins are increased in inflammatory diseases. They are decreased or absent in juvenile pulmonary **emphysema**, which is a genetic disease.

Alpha₂ globulins are increased in a kidney disorder called **nephrotic syndrome**. They are decreased in patients with an overactive thyroid gland (**hyperthyroidism**) or severe liver dysfunction.

Beta globulin levels are increased in conditions of high cholesterol levels (**hypercholesterolemia**) and **iron deficiency anemia**. They are decreased in malnutrition.

Gamma globulin levels are increased in chronic inflammatory disease (for example, **rheumatoid arthritis**, **systemic lupus erythematosus**), cirrhosis, acute and chronic infection, and a cancerous disease characterized by uncontrolled multiplication of plasma cells in

the bone marrow (multiple myeloma). Gammaglobulins are decreased in a variety of genetic immune disorders, and in secondary immune deficiency related to steroid use, leukemia, or severe infection.

Resources

BOOKS

Pagana, Kathleen Deska, and Timothy J. Pagana. *Mosby's Manual of Diagnostic and Laboratory Tests*. 4th ed. St. Louis: Mosby, 2009.

Janis O. Flores

Protein-energy malnutrition

Definition

Protein-energy **malnutrition** (PEM) is a potentially fatal body-depletion disorder. It is the leading cause of **death** in children in developing countries.

Demographics

Although PEM is not prevalent among the general population of the United States, it is often seen in elderly people who live in nursing homes and in children whose parents are poor. PEM occurs in one of every two surgical patients and in 48% of all other hospital patients.

Description

PEM is also referred to as protein–calorie malnutrition. It develops in children and adults whose consumption of protein and energy (measured by calories) is insufficient to satisfy the body's nutritional needs. While pure protein deficiency can occur when a person's diet provides enough energy but lacks the protein minimum, in most cases the deficiency will be dual. PEM may also occur in persons who are unable to absorb vital nutrients or convert them to energy essential for healthy tissue formation and organ function.

Types of PEM

Primary PEM results from a diet that lacks sufficient sources of protein and/or energy. Secondary PEM is more common in the United States, where it usually occurs as a complication of **AIDS**, **cancer**, **chronic kidney failure**, inflammatory bowel disease, or other illnesses that impair the body's ability to absorb or use nutrients or to compensate for nutrient

losses. PEM can develop gradually in a patient who has a chronic illness or experiences chronic semi-starvation. It may appear suddenly in a patient who has an acute illness.

Kwashiorkor

Kwashiorkor, also called wet protein-energy malnutrition, is a form of PEM characterized primarily by protein deficiency. This condition usually appears at the age of about 12 months when **breastfeeding** is discontinued, but it can develop at any time during a child's formative years. It causes fluid retention (**edema**); dry, peeling skin; and hair discoloration.

Marasmus

Primarily caused by energy deficiency, marasmus is characterized by stunted growth and wasting of muscle and tissue. Marasmus usually develops between the ages of six months and one year in children who have been weaned from breast milk or who suffer from weakening conditions like chronic **diarrhea**.

Causes and symptoms

Secondary PEM symptoms range from mild to severe, and can alter the form or function of almost every organ in the body. The type and intensity of symptoms depend on the patient's prior nutritional status and on the nature of the underlying disease and the speed at which it is progressing.

Mild, moderate, and severe classifications have not been precisely defined, but patients who lose 10%–20% of their body weight without trying are usually said to have moderate PEM. This condition is also characterized by a weakened grip and inability to perform high-energy tasks.

Losing 20% of body weight or more is generally classified as severe PEM. People with this condition can't eat normal-sized meals. They have slow heart rates and low blood pressure and body temperatures. Other symptoms of severe secondary PEM include baggy, wrinkled skin; **constipation**; dry, thin, brittle hair; lethargy; and pressure sores or other **skin lesions**.

Kwashiorkor

People who have kwashiorkor often have extremely thin arms and legs, but liver enlargement and **ascites** (abnormal accumulation of fluid) can distend the abdomen and disguise weight loss. Hair may turn red or yellow. Anemia, diarrhea, and fluid and **electrolyte disorders** are common. The body's immune

system is often weakened, behavioral development is slow, and **mental retardation** may occur. Children may grow to normal height but are abnormally thin.

Kwashiorkor-like secondary PEM usually develops in patients who have been severely burned, suffered trauma, or had **sepsis** (tissue-destroying infection) or another life-threatening illness. The condition's onset is so sudden that body fat and muscle mass of normal-weight people may not change. Some obese patients even gain weight.

Marasmus

Profound weakness accompanies severe marasmus. Since the body breaks down its own tissue to use as calories, people with this condition lose all their body fat and muscle strength, and acquire a skeletal appearance most noticeable in the hands and in the temporal muscle in front of and above each ear. Children with marasmus are small for their age. Since their immune systems are weakened, they suffer from frequent infections. Other symptoms include loss of appetite, diarrhea, skin that is dry and baggy, sparse hair that is dull brown or reddish yellow, mental retardation, behavioral retardation, low body temperature (**hypothermia**), and slow pulse and breathing rates.

The absence of edema distinguishes marasmus-like secondary PEM, a gradual wasting process that begins with weight loss and progresses to mild, moderate, or severe malnutrition (cachexia). It is usually associated with cancer, **chronic obstructive pulmonary disease** (COPD), or another chronic disease that is inactive or progressing very slowly.

Some individuals have both kwashiorkor and marasmus at the same time. This most often occurs when a person who has a chronic, inactive condition develops symptoms of an acute illness.

Hospitalized patients

Difficulty chewing, swallowing, and digesting food, as well as **pain**, **nausea**, and lack of appetite are among the most common reasons that many hospital patients don't consume enough nutrients. Nutrient loss can be accelerated by bleeding, diarrhea, abnormally high sugar levels (glycosuria), **kidney disease**, malabsorption disorders, and other factors. **Fever**, infection, surgery, and benign or malignant tumors increase the amount of nutrients hospitalized patients need. So do trauma, **burns**, and some medications.

KEY TERMS

Ascites—Abnormal accumulation of fluid in the abdomen, making the abdomen appear distended.

Cachexia—Severe malnutrition involving muscle wasting and organ damage.

Edema—Fluid retention, generally seen in the limbs.

Hypothermia—Low body temperature.

Diagnosis

A thorough **physical examination** and a health history that probes eating habits and weight changes, checks body-fat composition and muscle strength, and assesses gastrointestinal symptoms, underlying illness, and nutritional status is often as accurate as blood tests and urinalyses used to detect and document abnormalities.

Some doctors further quantify a patient's nutritional status by:

- comparing height and weight to standardized norms
- calculating body mass index (BMI)
- measuring skinfold thickness or the circumference of the upper arm

Treatment

Treatment is designed to provide adequate **nutrition**, restore normal body composition, and cure the condition that caused the deficiency. Tube feeding or intravenous feeding is used to supply nutrients to patients who can't or won't eat protein-rich foods.

In patients with severe PEM, the first stage of treatment consists of correcting fluid and electrolyte imbalances, treating infection with **antibiotics** that don't affect protein synthesis, and addressing related medical problems. The second phase involves replenishing essential nutrients slowly to prevent taxing the patient's weakened system with more food than it can handle. **Physical therapy** may be beneficial to patients whose muscles have deteriorated significantly.

Prognosis

Most people can lose up to 10% of their body weight without side effects, but losing more than 40% is almost always fatal. Death usually results from **heart failure**, an electrolyte imbalance, or low

body temperature. Patients with certain symptoms, including semi-consciousness, persistent diarrhea, **jaundice**, and low blood **sodium** levels, have a poorer prognosis than other patients. Recovery from marasmus usually takes longer than recovery from kwashiorkor. The long-term effects of childhood malnutrition are uncertain. Some children recover completely, while others may have a variety of life-long impairments, including an inability to properly absorb nutrients in the intestines and mental retardation. The outcome appears to be related to the length and severity of the malnutrition, as well as to the age of the child when the malnutrition occurred.

Prevention

Breastfeeding a baby for at least six months is considered the best way to prevent early childhood malnutrition. Preventing malnutrition in developing countries is a complicated and challenging problem. Providing food directly during famine can help in the short term, but more long-term solutions are needed, including agricultural development, public health programs (especially programs that monitor growth and development, as well as programs that provide nutritional information and supplements), and improved food distribution systems. Programs that distribute infant formula and discourage breastfeeding should be discontinued, except in areas where many mothers are infected with HIV.

Every patient being admitted to a hospital should be screened for the presence of illnesses and conditions that could lead to PEM. The nutritional status of patients at higher-than-average risk should be more thoroughly assessed and periodically reevaluated during extended hospital stays or nursing home residence.

Resources

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- Robertson, Cathie. *Safety, Nutrition and Health in Early Education*, 4th ed. Florence, KY: Wadsworth Publishing, 2009.
- Shalim, Judith., and Sari Edelstein. *Essentials of Life Cycle Nutrition*. New York, NY: Jones & Bartlett Publishers, 2010.

ORGANIZATIONS

American Academy of Family Physicians, P. O. Box 11210, Shawnee Mission, KS, 66207, (913) 906-6000, (913) 906-6075, (800) 274-2237, <http://www.aafp.org>.

American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007-1098, (847) 434-4000, <http://www.aap.org>.

American College of Nutrition, 722 Robert E. Lee Drive, Wilmington, NC, 20412-0927, (919) 152-1222.

American Institute of Nutrition, 9650 Rockville Pike, Bethesda, MD, 20814-3990, (301) 530-7050.

Food and Nutrition Information Center, National Agricultural Library, 10301 Baltimore Avenue, Room 105, Beltsville, MD, 20705, <http://www.nal.usda.gov/fnic>.

National Institute of Child Health and Human Development (NICHD), P.O. Box 3006, Rockville, MD, 30847, (866) 760-5947, (800) 370-2943, (800) 320-6942 (TTY), NICHDInformationResourceCenter@mail.nih.gov, <http://www.nichd.nih.gov>.

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Protein-modified diet see **Diets**

Prothrombin time

Definition

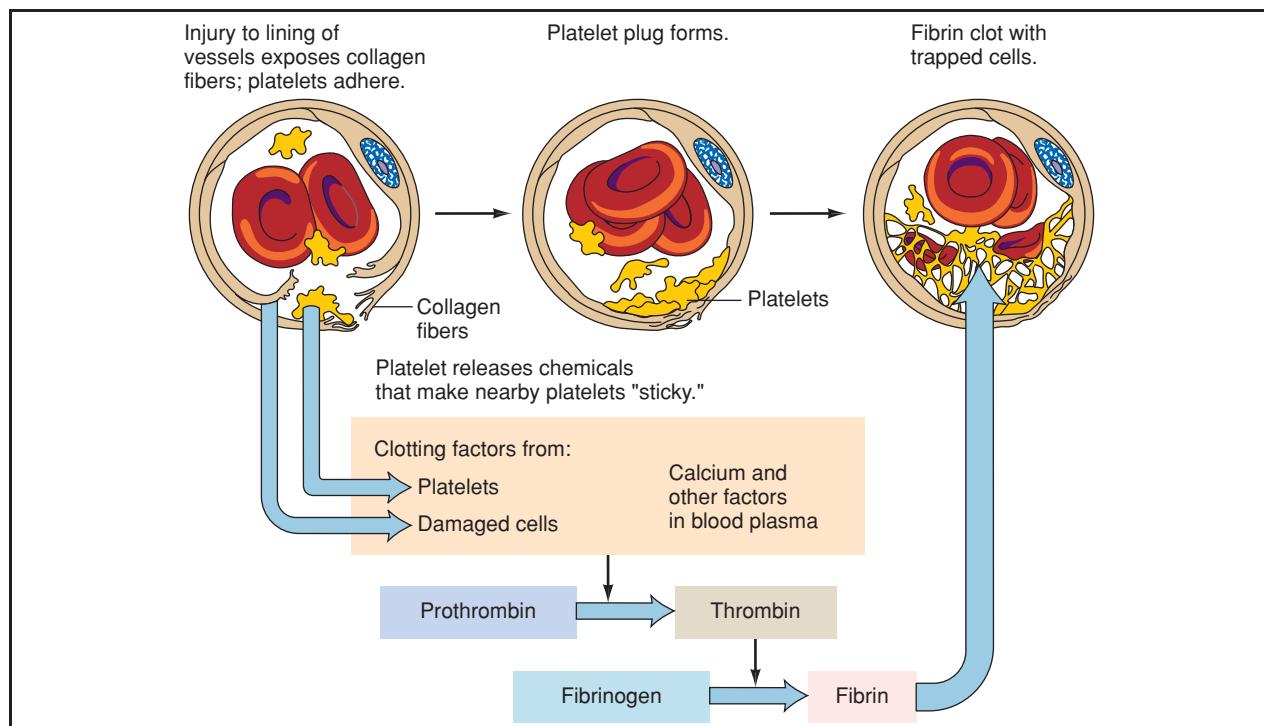
The prothrombin time test belongs to a group of blood tests that assess the clotting ability of blood. The test is also known as the pro time or PT test.

Purpose

The PT test is used to monitor patients taking certain medications as well as to help diagnose clotting disorders.

Diagnosis

Patients who have problems with delayed blood clotting are given a number of tests to determine the cause of the problem. The prothrombin test specifically evaluates the presence of factors VII, V, and X; prothrombin; and fibrinogen. Prothrombin is a protein in the liquid part of blood (plasma) that is converted to thrombin as part of the clotting process. Fibrinogen is a type of blood protein called a globulin; it is converted to fibrin during the clotting process. A drop in the concentration of any of these factors will



The blood clotting process. ((Illustration by Hans & Cassady, Inc. Reproduced by permission of Gale, a part of Cengage Learning.)

KEY TERMS

Disseminated intravascular coagulation (DIC)—A condition in which spontaneous bleeding and clot formation occur throughout the circulatory system. DIC can be caused by transfusion reactions and a number of serious illnesses.

Fibrin—The protein formed as the end product of the blood clotting process when fibrinogen interacts with thrombin.

Fibrinogen—A type of blood protein called a globulin that interacts with thrombin to form fibrin.

Plasma—The liquid part of blood, as distinct from blood cells.

Prothrombin—A protein in blood plasma that is converted to thrombin during the clotting process.

Thrombin—An enzyme in blood plasma that helps to convert fibrinogen to fibrin during the last stage of the clotting process.

Thromboplastin—A protein in blood that converts prothrombin to thrombin.

Warfarin—A drug given to control the formation of blood clots. The PT test can be used to monitor patients being treated with warfarin.

cause the blood to take longer to clot. The PT test is used in combination with the **partial thromboplastin time (PTT)** test to screen for **hemophilia** and other hereditary clotting disorders.

Monitoring

The PT test is also used to monitor the condition of patients who are taking warfarin (Coumadin). Warfarin is a drug that is given to prevent clots in the deep veins of the legs and to treat **pulmonary embolism**. It interferes with blood clotting by lowering the liver's production of certain clotting factors.

Description

A sample of the patient's blood is obtained by venipuncture. The blood is collected in a tube that contains **sodium citrate** to prevent the clotting process from starting before the test. The blood cells are separated from the liquid part of blood (plasma). The PT test is performed by adding the patient's plasma to a protein in the blood (thromboplastin) that converts prothrombin to thrombin. The mixture is then kept in a warm water bath at 37°C for one to two minutes. **Calcium** chloride is added to the mixture in order to counteract the sodium citrate and allow clotting to proceed. The test is timed from the addition of the calcium chloride until the plasma clots. This time is called the prothrombin time.

Preparation

The doctor should check to see if the patient is taking any medications that may affect test results. This precaution is particularly important if the patient is taking warfarin, because there are a number of

medications that can interact with warfarin to increase or decrease the PT time.

Aftercare

Aftercare consists of routine care of the area around the puncture mark. Pressure is applied for a few seconds and the wound is covered with a bandage.

Risks

The primary risk is mild **dizziness** and the possibility of a bruise or swelling in the area where the blood was drawn. The patient can apply moist warm compresses.

Normal results

The normal prothrombin time is 11–15 seconds, although there is some variation depending on the source of the thromboplastin used in the test. (For this reason, laboratories report a normal control value along with patient results.) A prothrombin time within this range indicates that the patient has normal amounts of clotting factors VII and X.

Abnormal results

A prolonged PT time is considered abnormal. The prothrombin time will be prolonged if the concentration of any of the tested factors is 10% or more below normal plasma values. A prolonged prothrombin time indicates a deficiency in any of factors VII, X, V, prothrombin, or fibrinogen. It may mean that the patient has a **vitamin K deficiency**, a **liver disease**, or disseminated intravascular coagulation (DIC). The prothrombin time of patients receiving warfarin therapy will also be prolonged—usually in the range

of one and one half to two times the normal PT time. A PT time that exceeds approximately two and a half times the control value (usually 30 seconds or longer) is grounds for concern, as abnormal bleeding may occur.

Resources

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John T. Lohr, PhD

Proton pump inhibitors

Definition

Proton pump inhibitors are drugs that reduce the secretion of acid in the stomach by temporarily disabling an enzyme, sometimes referred to as the proton pump, in parietal cells of the stomach wall that produces acid.

Purpose

These drugs are approved for short-term use (4–8 weeks) in treating the excessive production of stomach acid in peptic ulcer disease, gastroesophageal reflux (GERD), and Zollinger-Ellison Syndrome.

Proton pump inhibitors are also approved for longer-term use in conditions where there are abnormally high secretions of stomach acid, and as part of a multi-drug treatment program for reducing the recurrence of duodenal ulcers.

Proton pump inhibitors may be used to protect the stomach against ulcers in patients who regularly take **nonsteroidal anti-inflammatory drugs** or who take corticosteroid drugs long term.

Description

The class of proton pump inhibitors includes esomeprazole (Nexium), lansoprazole (Prevacid), omeprazole (Prilosec), pantoprazole (Protonix), and rabeprazole (Aciphex).

The drugs are usually formulated to be absorbed in the intestine after leaving the stomach. They do not provide immediate relief from symptoms of stomach distress or reflux (**heartburn**), and are not suitable alternatives to prompt-acting **antacids**.

Proton pump inhibitors disable the acid-producing enzyme and reduce acid production for 24 hours.

Recommended dosage

Doses of these drugs may vary, depending on conditions being treated.

Commonly prescribed doses include:

- Esomeprazole: 20 to 40 mg once a day
- Lansoprazole: 15 to 30 mg once a day
- Omeprazole: 20 to 40 mg once a day
- Pantoprazole: 40 mg once or twice a day
- Rabeprazole: 20 mg once a day

Lower doses are usually adequate for treating gastroesophageal reflux (GERD)

Higher doses for longer durations of time are sometimes required for treating chronic peptic ulcers or other conditions where there are abnormally high amounts of acid produced.

Precautions

Proton pump inhibitors should not be used in patients who have severe **liver disease**.

Though their effects in fetuses and nursing babies have not been thoroughly studied, these drugs cross the placenta and pass into breast milk. They should be used in **pregnancy** only when their value outweighs potential risks.

These drugs may increase the risk of intestinal bacterial infections.

Proton pump inhibitors may relieve the symptoms and mask the presence of **stomach cancer**.

People should not use the proton pump inhibitors that are available without prescription for more than two weeks without consulting a physician.

Treatment programs using these drugs should not be repeated more often than every four months.

Side effects

Adverse effects from these drugs include appetite changes, abdominal **pain**, **constipation**, **diarrhea**, **dizziness**, **headache**, and skin rash.

Interactions

Proton pump inhibitors interfere with and reduce the effects of clopidogrel (Plavix), taken to reduce the recurrence of heart attacks.

KEY TERMS

Antacids—Substances that counteract or neutralize acidity. They act promptly and have short durations of actions.

GERD—A condition where gastric acid refluxes or regurgitates into the esophagus, causing heartburn and acid indigestion. Over time, this can injure the esophageal lining and may lead to cancer.

Parietal cells—Cells of the gastric glands in the stomach lining that secrete hydrochloric acid.

Recurrent ulcer—Peptic ulcers that flare up after healing. They may be caused by a *helicobacter pylori* bacterial infection and are treated with a combination of antibiotics and gastric acid-reducing medications, like proton pump inhibitors.

Proton pump inhibitors may reduce the effectiveness of atazanavir (Ratataz) and indinavir (Crixivan), **antiretroviral drugs** used to treat **AIDS**.

By reducing the amount of acid in the stomach, this class of drugs may reduce the effectiveness of anti-fungal drugs like itraconazole (Sporonox) or ketoconazole (Nizoral).

Lansoprazole (Prevacid) may increase the likelihood of adverse effects from taking the antibiotic clarithromycin (Biaxin).

St John's Wort may reduce the effectiveness of omeprazole (Prilosec).

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International Foundation for Functional Gastrointestinal Disorders, P.O. Box 17864, Milwaukee, WI, 53217-8076, (414) 964-1799, (414) 964-7176, (888) 964-2001, iffd @iffgd.org, http://www.iffgd.org.

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Provence (sipuleucel-T)

Definition

Provence (sipuleucel-T) is the first drug to be developed and approved in a new class of drugs called autologous cellular immunotherapies.

Purpose

Sipuleucel-T was approved by the U.S. Food and Drug Administration (FDA) in 2010 for the treatment of advanced **prostate cancer** in men whose **cancer** is not yet causing symptoms and for men whose prostate cancer has metastasized and is causing minimal symptoms but has been resistant to treatment with hormonal therapies designed to treat prostate cancer.

Description

The exact mechanism of action of sipuleucel-T is not yet known. Sipuleucel-T works by harnessing the actions of the patient's own immune cells to target and treat that patient's biologically unique prostate cancer. Each dose of the drug is designed specifically for an individual patient using the process of leukapheresis, which is scheduled three days prior to a scheduled treatment with Provence. In leukapheresis, a cell collection process, some of the patient's own immune cells are collected. The collected cells are then packaged, labeled, and shipped to a drug manufacturing facility. Once at the facility, the patient's cells are placed in a culture with a human recombinant protein. This protein works to activate the patient's immune cells to function specifically as a prostate-associated antigen whose purpose is to trigger the patient's own immune system to recognize and kill prostate cancer cells. Another purpose of the end product is to stimulate the immune system. Once the process is completed at the drug manufacturing facility, the

activated cells are shipped back to a treatment center and are reinfused into the patient from whom they were originally collected. The patient's immune system is then activated to destroy prostate cancer cells.

Biologically active components of Provenge include autologous presenting cells (APCs) and remnants of a recombinant human protein designated as PAP–GM–CSF. PAP–GM–CSF consists of prostatic acid phosphatase (PAP), an antigen expressed by prostate cancer tissue, which is then combined with granulocyte-macrophage colony-stimulating factor (GM–CSF). GM–CSF stimulates immune cells to activate. In addition to the components derived from the recombinant human protein, the precise cellular composition of Provenge varies depending on the exact composition of the cells obtained from the patient during leukapheresis. The resulting product will likely contain immune cells such as T cells, B cells, natural killer cells and other cells, in addition to the autologous APCs.

Each dose of the drug is placed in suspension in 250 milliliters of Ringer's Lactate solution in a sealed infusion bag that is to be administered intravenously to one specific patient. No additives or preservatives are added to the solution during the manufacturing process.

The completed product can not be administered to any patient other than the original patient whose immune cells were collected during the leukapheresis process.

Recommended dosage

According to the manufacturer, each dose of Provenge contains a minimum of 50 million CD54+ cells that have been activated by human recombinant protein technology. The exact number of cells present in each dose varies. CD54 is a molecule on the surface of cells and is considered to be a marker of immune cell activity.

Administration of Provenge occurs in a three-dose schedule at intervals spaced about every two weeks. The drug is administered intravenously over a period of about an hour. No further treatment with Provenge is required after the initial three doses.

Precautions

This drug is not to be administered to patients other than the patient from whom immune cells were collected during leukapheresis. The identity of the patient receiving Provenge must match the patient identifying information on the infusion bag.

KEY TERMS

Antigen—A substance such as bacteria, enzymes, or other toxins that causes the body's immune system to react by stimulating production of antibodies.

Autologous—Derived from the same individual's body.

Immunotherapy—Therapy that stimulates, enhances, or suppresses the body's immune response; includes products such as monoclonal antibodies, vaccines, and growth factors.

Recombinant protein—A manipulated or modified form of a protein that results in the ability to produce the modified protein on a large scale.

A cell filter must not be used during infusion of Provenge.

Prior to infusion, the patient should be premedicated with oral **acetaminophen** and an antihistamine such as diphenhydramine. The infusion time may have to be slowed or stopped if severe infusion reactions occur during administration of this product.

The product has not been tested for infectious diseases that can be transmitted to others. Therefore, any infectious diseases that are present may be transmitted to health care workers during product handling or administration. Universal precautions should be adhered to by health care workers when handling this product.

If a patient is unable to receive a scheduled dose of Provenge, the patient will be required to undergo an additional leukapheresis procedure if treatment is to be continued. Patients should be informed of this possibility prior to the beginning of the treatment process.

Side effects

In clinical trials, the most common adverse reactions associated with Provenge administration included chills, **fever**, **fatigue**, back **pain**, **nausea**, pain in the joints, and **headache**. Some patients may experience acute and severe infusion reactions during administration of Provenge. Should these occur, the infusion may be slowed or stopped depending on the severity of the reaction. Severe reactions may occur after the administration is complete and typically occur within one day of drug administration.

Interactions

The concurrent use of **chemotherapy** and other medications that have the potential to suppress the immune system with Provence has not been studied. The use of immunosuppressive drugs concurrently with the use of Provence may result in decreased effectiveness of Provence. Patients on immunosuppressive drugs such as **corticosteroids** may be required to reduce or discontinue the use of these drugs during therapy with Provence.

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Pruritus see **Itching**

PSA test see **Prostate-specific antigen test**

Pseudoephedrine see **Decongestants**

Pseudogout

Definition

Pseudogout is a form of arthritis that causes **pain**, redness, and inflammation in one or more joints.

Description

Pseudogout is also known by another name: **calcium** pyrophosphate dihydrate deposition disease (CPPD), the basis of which is derived from the calcium deposits that collect in the joint. The deposits or crystals, as they are sometimes called, cause pain and inflammation in the joint. According to the Arthritis Foundation, this can eventually weaken the cartilage, which serves as padding between the bones, “allowing bone to rub against bone.” Pseudogout typically affects the large joints, such as the knees, wrists, and

ankles. In general, it occurs with equal frequency in men and women.

Most often seen in older adults, pseudogout can also affect younger patients, especially those with diseases that put them at a greater risk of developing it, such as **hemochromatosis**, **hypercalcemia**, **hypothyroidism**, ochronosis, or Wilson’s disease. Some people, according to an article for the American College of Rheumatology, experience attacks of pseudogout “following joint surgery or other surgery. Because many older people have calcium crystal deposits in their joints, any kind of insult to the joint can trigger the release of the calcium crystals, which then induce a painful inflammatory response.” Pseudogout affects about 3% of elderly people. Not all will experience severe attacks. By their 90s, 50% of people will have joint deposits. Although researchers have noticed that some people with pseudogout also have a family history of the disease, it is not clear what role genetics might play in its development.

Causes and symptoms

As the Arthritis Foundation points out, it is unclear what causes the crystals to form, but some speculation exists that “an abnormality in the cartilage cells or connective tissue could be responsible” for their development. Acute pain and fluid accumulation that leads to joint swelling are typical symptoms of pseudogout. When the crystals move into a joint, the Arthritis Foundation categorizes the pain as “sudden and severe.” Many patients report that joint motion is limited. In 50% of the cases, the patient will run a **fever**. Half of all the acute pseudogout attacks will involve a knee. The experts at MedlinePlus identify “chronic (long term) arthritis” as a symptom that can be present at the time of an acute pseudogout attack. The word “acute” implies short term; therefore, acute attacks of pseudogout will come and go, but chronic arthritis may remain. In addition, progressive degenerative arthritis is sometimes seen in numerous joints.

Diagnosis

Pseudogout and **gout** have similar symptoms, which can be confusing. However, uric acid is associated with gout, whereas calcium pyrophosphate crystals are associated with pseudogout. After a patient’s detailed medical history is obtained, a diagnosis can be made based on the symptoms and medical tests.

Using a needle, the physician can take a sample of the synovial fluid from the swollen or painful joint to ascertain the presence of calcium pyrophosphate crystals. The fluid will also contain white blood cells, which can be counted to assist in the diagnosis. Synovial fluid is the lubricating fluid that's secreted by the membranes that line the joints.

X rays may also be taken to confirm the presence of crystals. The x rays may show joint damage or that crystals have led to a condition called chondrocalcinosis, which is calcification of the cartilage. Other possible causes such as gout, **rheumatoid arthritis**, or infection must be ruled out. Blood tests can also help to confirm the diagnosis.

Treatment

There are a variety of treatment options. If patients have an adequate support system, such as family and friends willing to help, it makes it easier for patients to recover faster. Patients are often advised to avoid putting pressure on the affected joint. In some cases, it is appropriate for the patient to engage in special isometric exercises designed to help their specific condition heal faster. Once the inflammation and pain subsides, exercises are sometimes suggested to regain range of motion.

Medications can be prescribed to ease the pain, typically **nonsteroidal anti-inflammatory drugs** (NSAIDs). Ibuprofen (Motrin) and naproxen (Aleve) are two NSAIDs that are used often, as they are generally well tolerated and highly effective. Patients with kidney problems, stomach ulcers, or those on blood thinners may not be able to take NSAIDs. Indomethacin (Indocin) may be prescribed for those patients.

When no infection is present, **steroids**, such as prednisone, may be prescribed. Much of the literature discussing treatment options also suggests a medication called colchicine, which is only available as a generic. It is generally prescribed in low doses and should not be used by anyone with significant bone marrow dysfunction or renal insufficiency. Patients should talk with their physicians regarding any other reasons why colchicine may not be suitable for them.

In order to relieve some of the pressure, the excess fluid around the joint can be removed (aspirated) with a needle.

Anti-inflammatory treatments help slow joint degeneration, a consequence of untreated pseudogout. If joint degeneration does occur, surgery is available to

KEY TERMS

Hemochromatosis—A condition where the body absorbs too much iron.

Hypercalcemia—A condition where the bones absorb too much calcium.

Hypothyroidism—A disease of the thyroid caused by an underactive thyroid gland.

Ochronosis—A rare hereditary condition that usually leads to arthritis in adulthood.

Wilson's disease—Wilson's disease causes the body to retain copper, which ultimately can lead to liver damage.

replace or repair damaged joints; however, it is better for patients to engage in preventative measures that will help them avoid the need for surgery.

Prognosis

With regard to an acute attack of pseudogout, the prognosis is usually very good. The symptoms usually go away within two weeks. However, over time, joint degeneration may occur.

Prevention

There are no specific techniques applicable to every patient to prevent the formulation of the crystals; however, some patients with certain diseases are at greater risk of developing them. Diagnoses and treatment of underlying disorders is one of the most important aspects of managing crystal-induced arthropathies. Once a causative crystal is identified and a diagnosis has been established, a long-term management plan can be devised.

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Lee Ann Paradise

- Pseudohermaphroditism see **Intersex states**
 Pseudomembranous enterocolitis see **Antibiotic-associated colitis**
Pseudomonas aeruginosa infection see **Pseudomonas infections**

Pseudomonas infections

Definition

A pseudomonas infection is caused by a bacterium, *Pseudomonas aeruginosa*, and may affect any part of the body. In most cases, however, pseudomonas infections strike only persons who are very ill, usually those who are hospitalized.

Description

P. aeruginosa is a rod-shaped organism that can be found in soil, water, plants, and animals. Because it rarely causes disease in healthy persons, but infects those who are already sick or who have weakened immune systems, it is called an opportunistic pathogen. Opportunistic pathogens are organisms that do not ordinarily cause disease, but multiply freely in persons whose immune systems are weakened by illness or medication. Such persons are said to be immunocompromised. Patients with **AIDS** have an increased risk of developing serious pseudomonas infections. Hospitalized patients are another high-risk group, because *P. aeruginosa* is often found in hospitals. Infections that can be acquired in the hospital are sometimes called nosocomial diseases.

Of the two million nosocomial infections each year, 10% are caused by *P. aeruginosa*. The bacterium is the second most common cause of nosocomial **pneumonia** and the most common cause of intensive care unit (ICU) pneumonia. Pseudomonas infections can be spread within hospitals by health care workers, medical equipment, sinks, disinfectant solutions, and food. These infections are a very serious problem in hospitals for two reasons. First, patients who are critically ill can die from a pseudomonas infection. Second, many *Pseudomonas* bacteria are resistant to certain **antibiotics**, which makes them difficult to treat.

P. aeruginosa is able to infect many different parts of the body. Several factors make it a strong opponent. These factors include:

- the ability to stick to cells
- minimal food requirements
- resistance to many antibiotics
- production of proteins that damage tissue
- a protective outer coat

Infections that can occur in specific body sites include:

- Heart and blood. *P. aeruginosa* is the fourth most common cause of bacterial infections of the blood (bacteremia). Bacteremia is common in patients with blood cancer and patients who have pseudomonas infections elsewhere in the body. *P. aeruginosa* infects the heart valves of intravenous drug abusers and persons with artificial heart valves.
- Bones and joints. Pseudomonas infections in these parts of the body can result from injury, the spread of infection from other body tissues, or bacteremia. Persons at risk for pseudomonas infections of the bones and joints include diabetics, intravenous drug abusers, and bone surgery patients.
- Central nervous system. *P. aeruginosa* can cause inflammation of the tissues covering the brain and spinal cord (meningitis) and brain abscesses. These infections may result from brain injury or surgery, the spread of infection from other parts of the body, or bacteremia.
- Eye and ear. *P. aeruginosa* can cause infections in the external ear canal—so-called “swimmer’s ear”—that usually disappear without treatment. The bacterium can cause a more serious ear infection in elderly patients, possibly leading to hearing problems, facial paralysis, or even death. Pseudomonas infections of the eye usually follow an injury. They can cause ulcers of the cornea that may cause rapid tissue destruction and eventual blindness. Risk factors for pseudomonas eye infections include wearing soft extended-wear contact lenses, using topical corticosteroid eye medications, being in a coma, having extensive burns, undergoing treatment in an ICU, and having a tracheostomy or endotracheal tube.
- Urinary tract. Urinary tract infections can be caused by catheterization, medical instruments, and surgery.
- Lung. Risk factors for *P. aeruginosa* pneumonia include cystic fibrosis, chronic lung disease, immunocompromised condition, being on antibiotic

therapy or a respirator, and congestive heart failure. Patients with cystic fibrosis often develop pseudomonas infections as children and suffer recurrent attacks of pneumonia.

- Skin and soft tissue. Even healthy persons can develop a pseudomonas skin rash following exposure to the bacterium in contaminated hot tubs, water parks, whirlpools, or spas. This skin disorder is called pseudomonas or “hot tub” folliculitis, and is often confused with chickenpox. Severe skin infection may occur in patients with *P. aeruginosa* bacteremia. The bacterium is the second most common cause of burn wound infections in hospitalized patients.

Causes and symptoms

P. aeruginosa can be sudden and severe, or slow in onset and cause little pain. Risk factors for acquiring a pseudomonas infection include having a serious illness, being hospitalized, undergoing an invasive procedure such as surgery, having a weakened immune system, and being treated with antibiotics that kill many different kinds of bacteria (broad-spectrum antibiotics).

Each of the infections listed above has its own set of symptoms. **Pseudomonas bacteremia** resembles other bacteremias, producing fever, tiredness, muscle pains, joint pains, and chills. Bone infections are marked by swelling, redness, and pain at the infected site and possibly fever. **Pseudomonas meningitis** causes fever, headache, irritability, and clouded consciousness. Ear infection is associated with pain, ear drainage, facial paralysis, and reduced hearing. Pseudomonas infections of the eye cause ulcers that may spread to cover the entire eye, pain, reduced vision, swelling of the eyelids, and pus accumulation within the eye.

P. aeruginosa pneumonia is marked by chills, fever, productive cough, difficult breathing, and blue-tinted skin. Patients with **cystic fibrosis** with pseudomonas lung infections experience coughing, decreased appetite, weight loss, tiredness, wheezing, rapid breathing, fever, blue-tinted skin, and abdominal enlargement. Skin infections can cause a range of symptoms from a mild rash to large bleeding ulcers. Symptoms of pseudomonas **folliculitis** include a red itchy rash; headache; dizziness; earache; sore eyes, nose, and throat; breast tenderness; and stomach pain. Pseudomonas wound infections may secrete a blue-green colored fluid and have a fruity smell. Burn wound infections usually occur one to two weeks after the burn and cause discoloration of the burn

scab, destruction of the tissue below the scab, early scab loss, bleeding, swelling, and a blue-green drainage.

Diagnosis

Diagnosis and treatment of pseudomonas infections can be performed by specialists in **infectious disease**. Because *P. aeruginosa* is commonly found in hospitals, many patients carry the bacterium without having a full-blown infection. Consequently, the mere presence of *P. aeruginosa* in patients does not constitute a diagnostic finding. Cultures, however, can be easily done for test purposes. The organism grows readily in laboratory media; results are usually available in two to three days. Depending on the location of the infection, body fluids that can be tested for *P. aeruginosa* include blood, urine, cerebrospinal fluid, sputum, pus, and drainage from an infected ear or eye. X rays and other imaging techniques can be used to assess infections in deep organ tissues.

Treatment

Medications

Because *P. aeruginosa* is commonly resistant to antibiotics, infections are usually treated with two antibiotics at once. Pseudomonas infections may be treated with combinations of ceftazidime (Ceftaz, Fortraz, Tazicef), ciprofloxacin (Cipro), imipenem (Primaxin), gentamicin (Garamycin), tobramycin (Nebcin), ticarcillin-clavulanate (Timentin), or piperacillin-tazobactam (Zosyn). Most antibiotics are administered intravenously or orally for two to six weeks. Treatment of an eye infection requires local application of antibiotic drops.

Surgery

Surgical treatment of pseudomonas infections is sometimes necessary to remove infected and damaged tissue. Surgery may be required for brain abscesses, eye infections, bone and joint infections, ear infections, heart infections, and wound infections. Infected **wounds** and **burns** may cause permanent damage requiring arm or leg **amputation**.

Prognosis

Most pseudomonas infections can be successfully treated with antibiotics and surgery. In immunocompromised persons, however, *P. aeruginosa* infections have a high mortality rate, particularly following

KEY TERMS

Bacteremia—Bacterial infection of the blood.

“Hot tub” folliculitis—A skin infection caused by *P. aeruginosa* that often follows bathing in a hot tub or public swimming pool.

Immunocompromised—Having a weak immune system due to disease or the use of certain medications.

Nosocomial infection—An infection that is acquired in the hospital.

Opportunistic—Causing disease only under certain conditions, as when a person is already sick or has a weak immune system.

Pathogen—Any microorganism that produces disease.

bacteremia or infections of the lower lung. Mortality rates range from 15% to 20% of patients with severe ear infections to 89% of patients with infections of the left side of the heart.

Prevention

Most hospitals have programs for the prevention of nosocomial infections. Patients with cystic fibrosis may be given periodic doses of antibiotics to prevent episodes of pseudomonas pneumonia.

Minor skin infections can be prevented by avoiding hot tubs with cloudy water; avoiding public swimming pools at the end of the day; removing wet swimsuits as soon as possible; bathing after sharing a hot tub or using a public pool; cleaning hot tub filters every six weeks; and using appropriate amounts of chlorine in the water.

Resources

OTHER

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Pseudomonas pseudomallei infection see
Melioidosis

Pseudostrabismus see **Strabismus**

Pseudotuberculosis see **Sarcoidosis**

Pseudoxanthoma elasticum

Definition

Pseudoxanthoma elasticum (PXE) is an inherited connective tissue disorder in which the elastic fibers present in the skin, eyes, and cardiovascular system gradually become calcified and inelastic.

Description

PXE was first reported in 1881 by Rigal, but the defect in elastic fibers was described in 1986 by Darier, who gave the condition its name. PXE is also known as Grönblad-Strandberg-Touraine syndrome and systemic elastorrhexis.

The course of PXE varies greatly between individuals. Typically it is first noticed during adolescence as yellow-orange bumps on the side of the neck. Similar bumps may appear at other places where the skin bends a lot, like the backs of the knees and the insides of the elbows. The skin in these areas tends to get thick, leathery, inelastic, and acquire extra folds. These skin problems have no serious consequences, and for some people, the disease progresses no further.

Bruch’s membrane, a layer of elastic fibers in front of the retina, becomes calcified in some people with PXE. Calcification causes cracks in Bruch’s membrane, which can be seen through an ophthalmoscope as red, brown, or gray streaks called angiod streaks. The cracks can eventually (in 10–20 years) cause bleeding, and the usual resultant scarring leads to central vision deterioration. However, peripheral vision is unaffected.

Arterial walls and heart valves contain elastic fibers that can become calcified. This leads to a greater susceptibility to the conditions that are associated with hardening of the arteries in the normal **aging** population—high blood pressure, **heart attack**, **stroke**, and arterial obstruction—and, similarly, **mitral valve prolapse**. Heart disease and **hypertension** associated with PXE have been reported in children as young as 4 to 13 years of age. Although often appearing at a younger age, the overall incidence of these conditions is only slightly higher for people with PXE than it is in the general population.

Arterial inelasticity can lead to bleeding from the gastrointestinal tract and, rarely, acute **vomiting** of blood.

PXE is rare and occurs in about 1 in every 160,000 people in the general population. It is likely, though, that PXE is underdiagnosed, because of the presence

of mild symptoms in some affected persons and the lack of awareness of the condition among primary care physicians.

Causes and symptoms

PXE is caused by changes in the genetic material, called mutations, that are inherited in either a dominant or recessive mode. A person with the recessive form of the disease (which is most common) must possess two copies of the PXE gene to be affected, and, therefore, must have received one from each parent. In the dominant form, one copy of the defective gene is sufficient to cause the disease. In some cases, a person with the dominant form inherits the abnormal gene from a parent with PXE. More commonly, the mutation arises as a spontaneous change in the genetic material of the affected person. These cases are called "sporadic" and do not affect parents or siblings, although each child of a person with sporadic PXE has a 50% risk to inherit the condition.

Both males and females develop PXE, although the skin findings seem to be somewhat more common in females.

The actual genetic causes of this condition were not discovered until 2000. The recessive, dominant, and sporadic forms of PXE all appear to be caused by different mutations or deletions in a single gene called ABCC6 (also known as MRP6), located on chromosome 16. Although the responsible gene has been identified, how it causes PXE is still unknown.

Genetic researchers have since identified mutations in a number of persons with PXE, most of whom have been found to have the recessive type. Affected individuals in these families had mutations in both copies of the gene and parents, who are obligate carriers, had a mutation in only one copy. Contrary to the usual lack of symptoms in carriers of recessive genes, some carriers of recessive PXE have been found to have cardiovascular symptoms typical of PXE.

Although the recessive type is the most common, there are also familial and sporadic cases that have been found to be caused by dominant mutations in the ABCC6 gene.

A wide range in the type and severity of symptoms exists between people with PXE. The age of onset also varies, although most people notice initial symptoms during adolescence or early adulthood. Often, the first symptoms to appear are thickened skin with yellow bumps in localized areas such as the folds of the groin, arms, knees, and armpits. These changes can also occur in the mucous membranes, most often in the

inner portion of the lower lip. The appearance of the skin in PXE has been likened to a plucked chicken or Moroccan leather.

Angiod streaks in front of the retina are present in most people with PXE and an ophthalmologic examination can be used as an initial screen for the condition. Persons with PXE often complain of sensitivity to light. Because of the progressive breakdown of Bruch's membrane, affected persons are at increased risk for bleeding and scarring of the retina, which can lead to decreased central vision but does not usually cause complete blindness.

Calcium deposits in the artery walls contribute to early-onset **atherosclerosis**, and another condition called claudication, inadequate blood flow that results in **pain** in the legs after exertion. Abnormal bleeding, caused by calcification of the inner layer of the arteries, can occur in the brain, retina, uterus, bladder, and joints, but is most common in the gastrointestinal tract.

Diagnosis

The presence of calcium in elastic fibers, as revealed by microscopic examination of biopsied skin, unequivocally establishes the diagnosis of PXE.

Treatment

PXE cannot be cured, but **plastic surgery** can treat **PXE skin lesions**, and **laser surgery** is used to prevent or slow the progression of vision loss. Excessive blood loss due to bleeding into the gastrointestinal tract or other organ systems may be treated by **transfusion**. Mitral valve prolapse (protrusion of one or both cusps of the mitral heart valve back into the atrium during heart beating) can be corrected by surgery, if necessary.

Measures should be taken to prevent or lessen cardiovascular complications. People with PXE should control their cholesterol and blood pressure and maintain normal weight. They should **exercise** for cardiovascular health and to prevent or reduce claudication later in life. They should also avoid the use of tobacco, thiazide **antihypertensive drugs**, blood thinners like Coumadin, and **nonsteroidal anti-inflammatory drugs** like **aspirin** and ibuprofen. In addition, they should avoid strain, heavy lifting, and contact sports, since these activities could trigger retinal and gastrointestinal bleeding.

People with PXE should have regular eye examinations by an ophthalmologist and report any eye problems immediately. Regular checkups with a

KEY TERMS

Angiod streaks—Gray, orange, or red wavy branching lines in Bruch's membrane.

Bruch's membrane—A membrane in the eye between the choroid membrane and the retina.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring.

Claudication—Pain in the lower legs after exercise caused by insufficient blood supply.

Connective tissue—A group of tissues responsible for support throughout the body; includes cartilage, bone, fat, tissue underlying skin, and tissues that support organs, blood vessels, and nerves throughout the body.

Deletion—The absence of genetic material that is normally found in a chromosome. Often, the genetic material is missing due to an error in replication of an egg or sperm cell.

Dominant trait—A genetic trait in which one copy of the gene is sufficient to yield an outward display of

the trait. Dominant genes mask the presence of recessive genes and can be inherited from a single parent.

Elastic fiber—Fibrous, stretchable connective tissue made primarily from proteins, elastin, collagen, and fibrillin.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Mitral valve—The heart valve that prevents blood from flowing backwards from the left ventricle into the left atrium. Also known as bicuspid valve.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease, and can be transmitted to offspring.

Recessive trait—An inherited trait or characteristic that is outwardly obvious only when two copies of the gene for that trait are present.

physician are also recommended, including periodic blood pressure readings.

Some people have advocated a calcium-restricted diet, but it is not yet known whether this aids the problems brought about by PXE. It is known, however, that calcium-restriction can lead to bone disorders.

Prognosis

The prognosis for PXE is a normal life span with an increased chance of cardiovascular and circulatory problems, hypertension, gastrointestinal bleeding, and impaired vision. However, now that the gene for PXE has been identified, the groundwork for research to provide effective treatment has been laid. Studying the role of the ABCC6 protein in elastic fibers may lead to drugs that will ameliorate or arrest the problems caused by PXE.

Genetic tests are now available that can provide knowledge needed to both diagnose PXE in symptomatic persons and predict it prior to the onset of symptoms in persons at risk. Prenatal diagnosis of PXE, by testing fetal cells for mutations in the ABCC6 gene, can be done in early **pregnancy** by

procedures such as **amniocentesis** or **chorionic villus sampling**. For most people, PXE is compatible with a reasonably normal life, and prenatal diagnosis is not likely to be highly desired.

Genetic testing to predict whether an at-risk child will develop PXE may be helpful for medical management. A child who is found to carry a mutation can be monitored more closely for eye problems and bleeding, and can begin the appropriate lifestyle changes to prevent cardiovascular problems.

ORGANIZATIONS

National Association for Pseudoxanthoma Elasticum, 8760 Manchester Road, St. Louis, MO, 63144-2724, (314) 962-0100, napestlouis@sbcglobal.net, <http://www.pxeape.org>.

PXE International, 4301 Connecticut Ave, NW, Suite 404, Washington, DC, 20008-2369, (202) 362-9599, (202) 966-8553, info@pxe.org, <http://www.pxe.org>.

Barbara J. Pettersen

Psittacosis see **Parrot fever**

Psoas abscess see **Abscess**

Psoriasis

Definition

Named for the Greek word *psōra* meaning “itch,” psoriasis is a chronic, non-contagious disease characterized by inflamed lesions covered with silvery-white scabs of dead skin.

Demographics

Psoriasis, which affects at least four million Americans, is slightly more common in women than in men. Although the disease can develop at any time, 10%–15% of all cases are diagnosed in children under 10, and the average age at the onset of symptoms is 28. Psoriasis is most common in fair-skinned people and extremely rare in dark-skinned individuals.

Description

Normal skin cells mature and replace dead skin every 28–30 days. Psoriasis causes skin cells to mature in less than a week. Because the body can't shed old skin as rapidly as new cells are rising to the surface, raised patches of dead skin develop on the arms, back, chest, elbows, legs, nails, folds between the buttocks, and scalp.

Psoriasis is considered mild if it affects less than 5% of the surface of the body, moderate if 5%–30% of the skin is involved, and severe if the disease affects more than 30% of the body surface.

Types of psoriasis

Dermatologists distinguish different forms of psoriasis according to what part of the body is



Psoriasis, a chronic skin disorder, may appear on any area of the body, including the elbow, as shown above. (© Scott Camazine/Photo Researchers, Inc.)

affected, how severe symptoms are, how long they last, and the pattern formed by the scales.

PLAQUE PSORIASIS. Plaque psoriasis (*psoriasis vulgaris*), the most common form of the disease, is characterized by small, red bumps that enlarge, become inflamed, and form scales. The top scales flake off easily and often, but those beneath the surface of the skin clump together. Removing these scales exposes tender skin, which bleeds and causes the plaques (inflamed patches) to grow.

Plaque psoriasis can develop on any part of the body, but most often occurs on the elbows, knees, scalp, and trunk.

SCALP PSORIASIS. At least 50 of every 100 people who have any form of psoriasis have scalp psoriasis. This form of the disease is characterized by scale-capped plaques on the surface of the skull.

NAIL PSORIASIS. The first sign of nail psoriasis is usually pitting of the fingernails or toenails. Size, shape, and depth of the marks vary, and affected nails may thicken, yellow, or crumble. The skin around an affected nail is sometimes inflamed, and the nail may peel away from the nail bed.

GUTTATE PSORIASIS. Named for the Latin word *gutta*, which means “a drop,” guttate psoriasis is characterized by small, red, drop-like dots that enlarge rapidly and may be somewhat scaly. Often found on the arms, legs, and trunk and sometimes in the scalp, guttate psoriasis can clear up without treatment or disappear and resurface in the form of plaque psoriasis.

PUSTULAR PSORIASIS. Pustular psoriasis usually occurs in adults. It is characterized by blister-like lesions filled with non-infectious pus and surrounded by reddened skin. Pustular psoriasis, which can be limited to one part of the body (localized) or can be widespread, may be the first symptom of psoriasis or develop in a patient with chronic plaque psoriasis.

Generalized pustular psoriasis is also known as Von Zumbusch pustular psoriasis. Widespread, acutely painful patches of inflamed skin develop suddenly. Pustules appear within a few hours, then dry and peel within two days.

Generalized pustular psoriasis can make life-threatening demands on the heart and kidneys.

Palmar-plantar pustulosis (PPP) generally appears between the ages of 20 and 60. PPP causes large pustules to form at the base of the thumb or on the sides of the heel. In time, the pustules turn brown and peel. The disease usually becomes much less active for a while after peeling.

Acrodermatitis continua of Hallopeau is a form of PPP characterized by painful, often disabling, lesions on the fingertips or the tips of the toes. The nails may become deformed, and the disease can damage bone in the affected area.

INVERSE PSORIASIS. Inverse psoriasis occurs in the armpits and groin, under the breasts, and in other areas where skin flexes or folds. This disease is characterized by smooth, inflamed lesions and can be debilitating.

ERYthrodermic Psoriasis. Characterized by severe scaling, **itching**, and **pain** that affects most of the body, erythrodermic psoriasis disrupts the body's chemical balance and can cause severe illness. This particularly inflammatory form of psoriasis can be the first sign of the disease, but often develops in patients with a history of plaque psoriasis.

PSORIATIC ARTHRITIS. About 10% of patients with psoriasis develop a complication called **psoriatic arthritis**. This type of arthritis can be slow to develop and mild, or it can develop rapidly. Symptoms of psoriatic arthritis include:

- joint discomfort, swelling, stiffness, or throbbing
- swelling in the toes and ankles
- pain in the digits, lower back, wrists, knees, and ankles
- eye inflammation or pink eye (conjunctivitis)

Causes and symptoms

The cause of psoriasis is unknown, but research suggests that an immune-system malfunction triggers the disease. Factors that increase the risk of developing psoriasis include:

- family history
- stress
- exposure to cold temperatures
- injury, illness, or infection
- steroids and other medications
- race

Trauma and certain bacteria may trigger psoriatic arthritis in patients with psoriasis.

Diagnosis

A complete medical history and examination of the skin, nails, and scalp are the basis for a diagnosis of psoriasis. In some cases, a microscopic examination of skin cells is also performed.

Blood tests can distinguish psoriatic arthritis from other types of arthritis. **Rheumatoid arthritis**, in

KEY TERMS

Arthritis—An inflammation of joints.

particular, is diagnosed by the presence of a particular antibody present in the blood. That antibody is not present in the blood of patients with psoriatic arthritis.

Treatment

Age, general health, lifestyle, and the severity and location of symptoms influence the type of treatment used to reduce inflammation and decrease the rate at which new skin cells are produced. Because the course of this disease varies with each individual, doctors must experiment with or combine different treatments to find the most effective therapy for a particular patient.

Mild-moderate psoriasis

Steroid creams and ointments are commonly used to treat mild or moderate psoriasis, and **steroids** are sometimes injected into the skin of patients with a limited number of lesions. In mid-1997, the United States Food and Drug Administration (FDA) approved the use of tazarotene (Tazorac) to treat mild-to-moderate plaque psoriasis. This water-based gel has chemical properties similar to vitamin A.

Brief daily doses of natural sunlight can significantly relieve symptoms. **Sunburn** has the opposite effect.

Moisturizers and bath oils can loosen scales, soften skin, and may eliminate the itch. So can adding a cup of oatmeal to a tub of bath water. Salicylic acid (an ingredient in **aspirin**) can be used to remove dead skin or increase the effectiveness of other therapies.

Moderate psoriasis

Administered under medical supervision, ultraviolet light B (UVB) is used to control psoriasis that covers many areas of the body or that has not responded to topical preparations. Doctors combine UVB treatments with topical medications to treat some patients and sometimes prescribe home **phototherapy**, in which the patient administers his or her own UVB treatments.

Photochemotherapy (PUVA) is a medically supervised procedure that combines medication with exposure to ultraviolet light (UVA) to treat localized or widespread psoriasis. An individual with wide-

spread psoriasis that has not responded to treatment may enroll in one of the day treatment programs conducted at special facilities throughout the United States. Psoriasis patients who participate in these intensive sessions are exposed to UVB and given other treatments for six to eight hours a day for two to four weeks.

Severe psoriasis

Methotrexate (MTX) can be given as a pill or as an injection to alleviate symptoms of severe psoriasis or psoriatic arthritis. Patients who take MTX must be carefully monitored to prevent liver damage.

Psoriatic arthritis can also be treated with **non-steroidal anti-inflammatory drugs** (NSAIDs), like **acetaminophen** (Tylenol) or aspirin. Hot compresses and warm water soaks may also provide some relief for painful joints.

Other medications used to treat severe psoriasis include etrentinate (Tegison) and isotretinoin (Accutane), whose chemical properties are similar to those of vitamin A. Most effective in treating pustular or erythrodermic psoriasis, Tegison also relieves some symptoms of plaque psoriasis. Tegison can enhance the effectiveness of UVB or PUVA treatments and reduce the amount of exposure necessary.

Accutane is a less effective psoriasis treatment than Tegison, but both have similar side effects, including nosebleeds, inflammation of the eyes and lips, bone spurs, hair loss, and **birth defects**. Tegison is stored in the body for an unknown length of time and should not be taken by a woman who is pregnant or planning to become pregnant. A woman should use reliable birth control while taking Accutane and for at least one month before and after her course of treatment.

Cyclosporin emulsion (Neoral) is used to treat stubborn cases of severe psoriasis. Cyclosporin is also used to prevent rejection of transplanted organs, and Neoral, approved by the FDA in 1997, should be particularly beneficial to psoriasis patients who are young children or African Americans, or those who have diabetes.

Other conventional treatments for psoriasis include:

- Capsaicin (*Capsicum frutescens*), an ointment that can stop production of the chemical that causes the skin to become inflamed and halts the runaway production of new skin cells. Capsaicin is available without a prescription, but should be used under a doctor's supervision to prevent burns and skin damage.

- Hydrocortisone creams, topical ointments containing a form of vitamin D called calcitriol, and coal-tar shampoos and ointments can relieve symptoms. Hydrocortisone creams have been associated with such side effects as folliculitis (inflammation of the hair follicles), while coal-tar preparations have been associated with a heightened risk of skin cancer.

Alternative treatment

Nontraditional psoriasis treatments include:

- Soaking in warm water and German chamomile (*Matricaria recutita*) or bathing in warm salt water.
- Drinking as many as three cups a day of hot tea made with one or a combination of the following herbs: burdock (*Arctium lappa*) root, dandelion (*Taraxacum mongolicum*) root, Oregon grape (*Mahonia aquifolium*), sarsaparilla (*Smilax officinalis*), and balsam pear (*Momordica charantia*).
- Taking two 500-mg capsules of evening primrose oil (*Oenothera biennis*) a day. Pregnant women should not use evening primrose oil, and patients with liver disease or high cholesterol should use it only under a doctor's supervision.
- Eating a diet that includes plenty of fish, turkey, celery (for cleansing the kidneys), parsley, lettuce, lemons (for cleansing the liver), limes, fiber, and fruit and vegetable juices.
- Eating a diet that eliminates animal products high in saturated fats, since they promote inflammation.
- Drinking plenty of water (at least eight glasses) each day.
- Taking nutritional supplements including folic acid, lecithin, vitamin A (specific for the skin), vitamin E, selenium, and zinc.

Other helpful alternative approaches include identifying and eliminating food allergens from the diet, enhancing the function of the liver, augmenting the hydrochloric acid in the stomach, and completing a **detoxification** program. Constitutional homeopathic treatment, if properly prescribed, may also help resolve psoriasis.

Prognosis

Most cases of psoriasis can be controlled, and most people who have psoriasis can live normal lives.

Some people who have psoriasis are so self-conscious and embarrassed about their appearance that they become depressed and withdrawn. The Social Security Administration grants disability benefits to about 400 psoriasis patients each year, and a

comparable number die from complications of the disease.

Prevention

A doctor should be notified if:

- psoriasis symptoms appear or reappear after treatment
- pustules erupt on the skin and the patient experiences fatigue, muscle aches, and fever
- unfamiliar, unexplained symptoms appear

Resources

BOOKS

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- Goldman, L. and Ausiello D., eds. *Cecil Textbook of Internal Medicine*. 23rd ed. Philadelphia: Saunders, 2008.
- Habif, T.P. *Clinical Dermatology*. 5th ed. St. Louis: Mosby, 2009.
- Rakel, R. *Textbook of Family Medicine* 2007. 7th ed. Philadelphia: Saunders Elsevier, 2009.
- Rakel, R.E., and Bope, E.T. *Conn's Current Therapy*. 60th ed. Philadelphia: Saunders Elsevier, 2009.

ORGANIZATIONS

- American Academy of Dermatology, 930 N. Meacham Road, P.O. Box 4014, Schaumburg, IL, 60168-4014, (847) 330-0230, (847) 330-0050, <http://www.aad.org>.
- American Skin Association, Inc., 150 E. 58th St., 3rd floor, New York, NY, 10155-0002, (212) 688-6547, <http://www.americanskin.org>.
- National Psoriasis Foundation, 6600 S.W. 92nd Ave., Suite 300, Portland, OR, 97223, (800) 723-9166, <http://www.psoriasis.org>.

Maureen Haggerty

Psoriatic arthritis

Definition

Psoriatic arthritis is a form of arthritic joint disease associated with the chronic skin scaling and fingernail changes seen in **psoriasis**.

Description

Physicians recognize a number of different forms of psoriatic arthritis. In some patients, the arthritic symptoms will affect the small joints at the ends of the fingers and toes. In others, symptoms will affect joints on one side of the body but not on the other. In addition, there are patients whose larger joints on both sides of the body simultaneously become

affected, as in **rheumatoid arthritis**. Some people with psoriatic arthritis experience arthritis symptoms in the back and spine; in rare cases, called psoriatic arthritis mutilans, the disease destroys the joints and bones, leaving patients with gnarled and club-like hands and feet. In many patients, symptoms of psoriasis precede the arthritis symptoms; a clue to possible joint disease is pitting and other changes in the fingernails.

Most people develop psoriatic arthritis at ages 35–45, but it has been observed earlier in adults and children. Both the skin and joint symptoms will come and go; there is no clear relationship between the severity of the psoriasis symptoms and arthritis **pain** at any given time. It is unclear how common psoriatic arthritis is. Recent surveys suggest that between one in five people and one in two people with psoriasis may also have some arthritis symptoms.

Causes and symptoms

The cause of psoriatic arthritis is unknown. As in psoriasis, genetic factors appear to be involved. People with psoriatic arthritis are more likely than others to have close relatives with the disease, but they are just as likely to have relatives with psoriasis but no joint disease. Researchers believe genes increasing the susceptibility to developing psoriasis may be located on chromosome 6p and chromosome 17, but the specific genetic abnormality has not been identified. Like psoriasis and other forms of arthritis, psoriatic arthritis also appears to be an autoimmune disorder, triggered by an attack of the body's own immune system on itself.

Symptoms of psoriatic arthritis include dry, scaly, silver patches of skin combined with joint pain and destructive changes in the feet, hands, knees, and spine. Tendon pain and nail deformities are other hallmarks of psoriatic arthritis.

Diagnosis

Skin and nail changes characteristic of psoriasis with accompanying arthritic symptoms are the hallmarks of psoriatic arthritis. A blood test for rheumatoid factor, antibodies that suggest the presence of rheumatoid arthritis, is negative in nearly all patients with psoriatic arthritis. X rays may show characteristic damage to the larger joints on either side of the body, as well as fusion of the joints at the ends of the fingers and toes.

Treatment

Treatment for psoriatic arthritis is meant to control the **skin lesions** of psoriasis and the joint

KEY TERMS

Psoriasis—A common recurring skin disease that is marked by dry, scaly, and silvery patches of skin that appear in a variety of sizes and locations on the body.

Psoriatic arthritis mutilans—A severe form of psoriatic arthritis that destroys the joints of the fingers and toes and causes the bones to fuse, leaving patients with gnarled and club-like hands and feet.

Rheumatoid arthritis—A systemic disease that primarily affects the joints, causing inflammation, changes in structure, and loss of function.

Rheumatoid factor—A series of antibodies that signal the presence of rheumatoid arthritis. May also be present in Sjögren's syndrome and systemic lupus erythematosus, among others.

inflammation of arthritis. **Nonsteroidal anti-inflammatory drugs**, gold salts, and sulfasalazine are standard arthritis treatments, but have no effect on psoriasis. Antimalaria drugs and **systemic corticosteroids** should be avoided because they can cause **dermatitis** or exacerbate psoriasis when they are discontinued.

Several treatments are useful for both the skin lesions and the joint inflammation of psoriatic arthritis. Etretinate, a vitamin A derivative; methotrexate, a potent suppressor of the immune system; and ultraviolet **light therapy** have all been successfully used to treat psoriatic arthritis.

Alternative treatment

Food allergies/intolerances are believed to play a role in most **autoimmune disorders**, including psoriatic arthritis. Identification and elimination of food allergens from the diet can be helpful. Constitutional homeopathy can work deeply and effectively with this condition, if the proper prescription is given. **Acupuncture**, Chinese herbal medicine, and Western herbal medicine can all be useful in managing the symptoms of psoriatic arthritis. **Nutritional supplements** can contribute added support to the healing process. Alternative treatments recommended for psoriasis and rheumatoid arthritis may also be helpful in treating psoriatic arthritis.

Prognosis

The prognosis for most patients with psoriatic arthritis is good. For many, the joint and other

arthritis symptoms are much milder than those experienced in rheumatoid arthritis. One in five people with psoriatic arthritis, however, face potentially crippling joint disease. In some cases, the course of the arthritis can be far more mutilating than in rheumatoid arthritis.

Prevention

There are no preventive measures for psoriatic arthritis.

ORGANIZATIONS

American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168-4014, (847) 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

American College of Rheumatology, 2200 Lake Boulevard NE, Atlanta, GA, 30319, (404) 633-3777, (404) 633-1870, acr@rheumatology.org, <http://www.rheumatology.org>.

Richard H. Camer

PSP see **Progressive supranuclear palsy**

Psychiatric confinement

Definition

Psychiatric confinement is the use of restraints to detain a person in need of care and further evaluation.

Purpose

The primary purpose for psychiatric confinement is typically an urgent or emergent condition that could cause danger to the affected person or others, or cause severe disability to the extent whereby the affected person is unable to care for his/herself.

Precautions

Because psychiatric restraint has medicolegal implications, clinicians utilizing this form of patient and public safety should perform a comprehensive **mental status examination** and document the findings. This approach helps provide clear records establishing the specific presenting problems and symptoms, avoiding future ambiguities.

Description

Confinement with restraints can be categorized as urgent or emergent. Emergent causes can include patients exhibiting abnormal vital signs (breathing, pulse rate, temperature, blood pressure), threatening or violent behavior, and those who present with signs and symptoms of alcohol or illicit drug intoxication. Urgent use of confinement is indicated in patients showing suicidal thoughts, extreme **anxiety**, homicidal tendencies, violence, or a danger to self or the public at large.

Preparation

Those categorized as emergent should be prepared for further testing, which can include blood chemistry and psychological assessment and evaluation. Initially the patient is restrained with four-point leather restraints (both arms and both legs) and placed in a quiet room with a sitter. For those with urgent needs, restraint is initiated and initial management is directed to assess for an underlying medical cause and address psychological needs.

Aftercare

Further assessment, testing, and evaluation is necessary for a definitive diagnosis and to devise an appropriate treatment plan.

Risks

A deficiency in record-keeping can lead to legal problems. The criteria and specifications for confinement should be clearly indicated. Meticulous clinical examination and documentation is essential for a definitive diagnosis. Persons who are confined due to **substance abuse** problems may have legal issues. There is proposed legislation concerning the misuse of restraints for psychiatric inpatients, which in the past has been responsible for numerous wrongful deaths. There are currently no federal laws that regulate the use of inpatient restraints nor any requirements for reporting injuries or **death**.

Resources

PERIODICALS

Reeves, R., et al. "Medicolegal Errors in the ED Related to the Involuntary Confinement of Psychiatric Patients." *American Journal of Emergency Medicine* (November 1998).

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Psychoanalysis

Definition

Psychoanalysis is a method of talk therapy for mental, emotional, and behavioral dysfunctions developed by Sigmund Freud (1856-1939), a Viennese physician. Psychoanalysis is classified as an insight-oriented rather than a supportive form of therapy, because it is based on the notion that people are better able to make changes in their lives when they have improved their understanding of themselves through identifying their assumptions about life and the early life experiences that gave rise to them. In the words of the American Psychoanalytic Association, "Psychoanalytic treatment gives patients the opportunity to examine these assumptions, understand their origins in their lives, modify them if necessary, and make better choices for themselves."

Purpose

The basic purpose of psychoanalysis is to help people move forward in their lives by resolving inner conflicts leading to a range of mental and emotional problems. Psychoanalysts maintain that they can successfully treat **phobias**, **anxiety** attacks, obsessions and compulsions, **mood disorders**, repetitive patterns of failure in relationships and employment, unresolved grief, and a more general feeling of alienation or estrangement from others.

Demographics

As of 2010, there are 35 training institutes for psychoanalysts in the United States approved by the American Psychoanalytic Association and between 75 and 100 independent training institutes. It is difficult to obtain precise statistics on the number of patients who undergo psychoanalysis in an average year; however, because of the time and expense involved in this form of mental health treatment, considerably fewer people consult psychoanalysts than other mental health professionals.

Description

Origins

Developed by Sigmund Freud (1856-1939), a Viennese neurologist and professor of medicine, psychoanalysis is based on an approach in which the therapist helps the patient better understand him- or herself through examination of the deeply hidden feelings, relationships, and events that have shaped

the patient's motivations and behavior. Freud's interest began when he encountered patients who were clearly suffering physical symptoms for which he could find no organic (biological) cause. Freud's first attempt to uncover the psychological causes of his patients' **pain** was through hypnosis, which he studied in Paris in 1885 under Jean-Martin Charcot, a French neurologist who specialized in research in what were then called hysterical disorders. Freud found Charcot's approach, which depended heavily on hypnosis, to be less fruitful than he had hoped, however, and he soon borrowed from a Viennese contemporary the idea of getting a patient to simply talk about his or her problems. Freud expanded upon this practice by introducing a method that he called free association, in which a patient is encouraged to speak in a non-narrative or rambling manner under the expectation that he or she will eventually reveal or uncover the unconscious heart of the problem. This sort of undirected self-exploration became one of the signature tenets of psychoanalysis.

Continuing his research into the mind and the unconscious, Freud published *The Interpretation of Dreams* in 1900. In this work he outlined his ideas about the construction of the mind and human personality. This book was followed by the major publications of the Freudian canon: *The Psychopathology of Everyday Life* in 1904, and *A Case of Hysteria* as well as *Three Essays on the Theory of Sexuality* in 1905. By the second decade of the twentieth century, Freud had become an internationally renowned thinker, largely as a result of his accepting an invitation to give a series of lectures at Clark University in Massachusetts in September 1909. Although Freud delivered his talks in German, they were translated into English within weeks and made his name a household word among English-speaking physicians and psychologists. Psychoanalysis had emerged as a significant intellectual achievement on a par with the work of Albert Einstein in physics and in many ways comparable to the modernist movement in the visual arts. Psychoanalysis was in its prime by the 1920s; after the shock and disillusionment following World War I, Western intellectuals made undergoing psychoanalysis a fashionable form of treatment.

Major concepts

Freud believed that human personality is constructed of three parts: the id, the ego, and the superego. The id, according to this schema, is comprised largely of such instinctual drives as desires for food or sexual pleasure. These drives are essentially unconscious, yielding satisfaction when they are fulfilled and

frustration or anxiety when they are thwarted. The ego is linked to the id, but because the ego is the component that has undergone socialization and recognizes that instant gratification of the id's urges is not always possible, a person may experience inner conflicts. The superego acts in many ways like the ego, as a moderator of behavior—but whereas the ego moderates urges based on social constraints, the superego operates as an arbiter of right and wrong. It moderates the id's urges on the basis of a moral code. Having constructed this framework of human personality, Freud used it to demonstrate the ways in which instinctual drives inevitably run aground on strictly social codes (upheld by the ego) and notions of morality (upheld by the superego). The resultant conflicts, according to psychoanalytic theory, lie at the heart of most **anxiety disorders** and other neurotic problems.

In dealing with these conflicts, Freud's psychoanalytic theory suggests that the human mind constructs three forms of adaptive mechanisms: namely, defense mechanisms, neurotic symptoms, and dreams. Freud believed dreams were vivid representations of repressed urges; they emanate from the id's speaking out in wildly incongruous nighttime parables. He considered dreams to have two parts: the manifest content, which is the narrative that one is able to remember upon waking; and the latent content, which is the underlying and largely symbolic message of the dream. Because Freud believed dreams to represent unfulfilled longings of the id, psychoanalysis deals heavily with dream interpretation.

Psychoanalytic theory also regards various neurotic symptoms as symbolic acts representing the repressed longings of the id. For Freud, a neurotic symptom was what we now consider a psychosomatic disorder; that is, some physical symptom that has a psychological, or in Freud's terms, neurological, origin. Psychoanalytic theory suggests that conditions like blindness, **paralysis**, and severe headaches can result from unfulfilled longings that the patient is unable to confront on a conscious level. Because of this inability, the patient develops some acceptable symptom, such as headaches, for which he or she can then seek medical attention.

The final adaptive mechanisms that Freud described are defense mechanisms. Freud identified several defense mechanisms, such as repression, displacement, denial, rationalization, projection, and identification. Each has its own peculiar dynamic, but all work to distance a person from a conflict that is too difficult to confront realistically. These conflicts, according to psychoanalytic theory, originate during one of the four developmental stages Freud identified.

CARL GUSTAV JUNG (1875–1961)



(The Library of Congress.)

Carl Gustav Jung was born in Kesswil, Switzerland, on July 26, 1875, to a Protestant clergyman who moved his family to Basel when Jung was four. While growing up, Jung exhibited an interest in many diverse areas of study

but finally decided to pursue medicine at the University of Basel and the University of Zurich, earning his degree in 1902. He also studied psychology in Paris. In 1903, Jung married Emma Rauschenbach, his companion and collaborator. The couple had five children.

Jung's professional career began in 1900 at the University of Zurich where he worked as an assistant to Eugene Blueler in the psychiatric clinic. During his internship, he and some coworkers used an experiment that revealed groups of ideas in the unconscious psyche, which he named *complexes*. Jung sent his publication *Studies in Word Association* (1904) to Sigmund Freud after finding his own beliefs confirmed by Freud's work. Jung and Freud became friends and collaborators until 1913, when Jung's ideas began to conflict with Freud's. During the time following this split, Jung published *Two Essays on Analytical Psychology* (1916, 1917) and *Psychological Types* (1921). Jung's later work developed from the concepts in his *Two Essays* publication and he became known as a founder of modern depth psychology.

In 1944, Jung gave up his psychological practice and his explorations after he suffered a severe heart attack. Jung received honorary doctorates from numerous universities and in 1948 he founded the C. G. Jung Institute in Zurich. Jung died on June 6, 1961.

These stages, and the infantile sexuality he identified as occurring within them, are some of the most controversial aspects of psychoanalytic theory. Freud suggested that adult neuroses result from and can be traced back to frustrated sexual drives during these stages. Freud defined the stages as the oral stage, birth to one year; the anal stage, one to three years; the phallic stage, three to five years; and latency, five years to the beginning of **puberty**. Each of these stages is in turn divided into substages. In each of the major stages, the infant has sexual longings which, because of social mores, are left largely unfulfilled and lead to the formation of neuroses.

Freud hypothesized that children in the phallic stage of development form the Oedipus complex, easily the most renowned and controversial theoretical construction of the Freudian canon. The Oedipus complex refers to the notion that a child begins associating his genitals with sexual pleasure during the phallic stage and becomes erotically attracted to the parent of the opposite sex while at the same time developing an intense jealousy of the same-sex parent. Freud derived the name of the complex from Oedipus,

a figure in Greek mythology who ended up murdering his father and marrying his mother. While Freud's original theory excludes consideration of females, his contemporary, Carl Jung (1875–1961), expanded this particular dynamic and described an Electra complex for women in which the same psychodrama of erotic attraction and jealousy is played out from the young girl's point of view. Electra is the name of the Greek princess who, according to legend, killed her mother Clytemnaestra in order to avenge her mother's murder of her father, Agamemnon.

Criticisms of Freudian psychoanalysis

From nearly the beginning, Freud and his construction of psychoanalytic theory have faced intense criticism. His most famous dissenter is Carl Jung, his former disciple. Jung split with Freud in 1913 over a variety of issues including, but certainly not limited to, Freud's emphasis on infantile sexuality. Jung had a different view of the construction of human personality and had different ideas about how dreams should be interpreted and viewed as part of psychoanalysis.

Alfred Adler (1870–1937), another early disciple of Freud, broke with the master over the concept of infantile sexuality, positing a view that infants and children are driven primarily by a need for self-affirmation rather than sexual gratification.

More recently, Freud has been the target of criticism from many corners. Feminists especially criticize his understanding of **hysteria** and his notion of penis envy, which postulates that women feel inferior to men from the moment they discover that the male genitalia differ from their own, and spend the rest of their lives trying to compensate in various ways for the perceived lack of a penis.

In addition, many historians regard Freud's theories as shaped by his particular culture. Vienna was the capital of an empire that held together a loose assortment of nationalities and languages and underwent a number of economic and political crises from the 1870s up to the outbreak of World War I in 1914. Freud himself was an outsider, a Jew in an anti-Semitic society in which professional advancement in medicine was difficult for Jews. Many of his patients suffered from neurotic disorders that were byproducts of very strict methods of childrearing rather than outgrowths of universal characteristics of human nature. Historians of medicine in particular have observed that some of the emotional disorders that Freud regarded as common or widespread in the Europe of his day are rarely seen in contemporary patients.

Recent developments

Psychoanalysis has lost favor since the 1980s as an approach to therapy for a number of reasons. One is the expense; in traditional psychoanalysis, the patient sees the psychoanalyst several times a week, often for years on end. The cost of a classical analysis can easily run into thousands of dollars very quickly. With the coming of managed care, most health maintenance organizations (HMOs) will not reimburse patients for the cost of psychoanalytic therapy. Other approaches, such as **cognitive-behavioral therapy** (CBT), are preferred because they are short-term. It is significant that the National Institute of Mental Health (NIMH)'s 2010 pamphlet on different approaches to **psychotherapy** does not even mention classical psychoanalysis as a treatment option.

Another reason for growing opposition to psychoanalysis in the United States in the early 1990s was the publicity given to several malpractice scandals involving psychoanalysts, including one in which the analyst was forced to surrender her medical license following the **suicide** of a medical student with

whom she had a highly sexualized relationship. A male analyst was also forced to surrender his license in 1992 following lawsuits from five patients that he had sexually abused.

Next, a series of outcome studies in the 1980s and 1990s, following the lead of Hans J. Eysenck (1916–1997), a psychologist opposed to Freudian theory, found that psychoanalysis is not necessarily more effective than treatment with medications, short-term psychotherapy, or a placebo. Although many psychoanalysts still maintain that the Freudian approach is more comprehensive and hence more powerful in easing the patient's psychological suffering, proof of its superiority in these respects has not been established to the satisfaction of therapists from other schools of thought.

The other major change that has led to a decline in interest in traditional psychoanalysis is the rise of biological psychiatry as an alternative to talk therapies in general. Biological psychiatry is an approach to therapy that seeks to understand mental disorders in terms of biological dysfunction in the nervous system, whether alterations in the levels of various neurotransmitters, organic changes in brain tissue (as in Alzheimer's disease), or the effects of such contagious diseases as **syphilis** on the brain. Biological psychiatry has pioneered the use of drugs to treat a range of mental disorders from **schizophrenia** to depression. While most psychiatrists combine medications with some form of psychotherapy in ordinary practice, it is nonetheless true that increasing interest in biological explanations for mental disorders is another reason for the decline of classical psychoanalysis.

Psychoanalysis of children

Although traditional Freudian psychoanalysis was developed for adults and is not frequently used with children in its unmodified form, long-term therapy for problems of childhood and adolescence may be based on an approach that shares the Freudian emphasis on uncovering unconscious motivations and analyzing defenses. Freud's youngest daughter, Anna Freud (1895–1982), became a noteworthy child psychiatrist, beginning her practice with children in 1923.

With younger children, the psychoanalytic process takes place through play or storytelling. Young children, guided in play by a therapist, create situations or tell stories in which they reenact their problems. The therapist helps the child understand the feelings she expresses through her play scenarios, and assists the child in developing strategies for changing behavior. Older children and adolescents are

KEY TERMS

Analysand—A person undergoing psychoanalysis.

Biological psychiatry—An approach to psychiatry that aims to understand mental disorders in terms of the biological and biochemical functions of the central nervous system.

Cognitive-behavioral therapy (CBT)—An approach to psychotherapy that emphasizes the correction of distorted thinking patterns and changing one's behaviors accordingly.

Countertransference—The analyst's emotional reaction to or entanglement with the analysand.

Insight-oriented therapy—An approach to psychotherapy based on helping the client understand the existence of previously unconscious conflicts and the origin of maladaptive behavior in order to change it. Psychoanalysis is one form of insight-oriented therapy.

Psychodynamic psychotherapy—A less intensive form of insight-oriented therapy than psychoanalysis

that typically involves greater interaction between therapist and patient than classical psychoanalysis.

Psychosis—A severe mental disorder characterized by loss of contact with reality, as evidenced by delusions and hallucinations.

Supportive therapy—An approach to psychotherapy that seeks to encourage the patient or offer emotional support to him or her, as distinct from insight-oriented or educational approaches to treatment.

Talk therapy—A general term for any form of psychotherapy based on conversational interaction between a trained therapist and a client. It includes psychodynamic therapy, humanistic therapy, and the various behavioral therapies as well as psychoanalysis.

Transference—The redirection of feelings and thoughts from early childhood experiences toward a person in the present, usually the analyst.

encouraged to talk about their feelings and the situations that are causing them problems.

Benefits

Many therapists maintain that psychoanalysis is the most effective technique to identify and deal with internal conflicts and feelings that contribute to dysfunctional behavior. Through psychoanalysis, the patient increases his understanding of himself and his internal conflicts so that they will no longer exert as much influence on mental and emotional health. Psychoanalysis appears to be most beneficial, however, to people with moderate to severe problems who have not been helped by briefer or less intense forms of therapy.

Precautions

Some groups of people do not benefit from traditional psychoanalysis:

- People diagnosed as psychotic. While some analysts have adapted psychoanalysis to working with people diagnosed with schizophrenia and other psychoses, most analysts limit their practices to people less severely disturbed.
- People actively abusing alcohol or other drugs.

- People with limited intelligence or verbal ability. Insight-oriented therapies work best with people who feel comfortable with verbal discussion and are able to describe their problems in detail.

- Very young children. Some psychoanalysts do practice psychoanalysis of children and adolescents as noted above, consulting the parents (except in the case of older adolescents) in order to obtain a full picture of the child's problems. Children who are too young to talk with some fluency, however, generally do not benefit from a psychoanalytic approach.

- People who are emotionally fragile. Psychoanalysis requires a willingness to challenge oneself in depth; some people do not have the emotional sturdiness to cope with the results of questioning the beliefs or persons who have given structure to their lives.

- People who have severe difficulty trusting others. Psychoanalysis requires the analysand to form a stable long-term working relationship with the analyst.

Preparation

Preparation for psychoanalysis should include a consultation with either an analyst or another mental health professional to see whether a shorter-term form of therapy might be equally beneficial. Given the investment of time and money involved in psychoanalysis, most people should at least acquaint

themselves with other forms of talk therapy, particularly if their health insurance limits the number of sessions allowed for a mental health provider. The American Psychoanalytic Association also maintains a list of low-fee clinics on its website for those whose finances are limited.

The next step is an appointment with a credentialed psychoanalyst, who will make the decision as to whether the patient is a suitable candidate for this type of therapy and whether there is a good “fit” between the patient and the analyst. If the only issue is interpersonal compatibility, most psychoanalysts will refer the patient to a colleague. In some cases the patient will be advised to undertake a course of psychodynamic psychotherapy (a less intense type of insight-oriented therapy) prior to beginning a full psychoanalysis. Psychoanalysis itself typically involves scheduling regular sessions of 45–50 minutes in length with the analyst, three to five times each week for a number of years.

Aftercare

Aftercare may include occasional sessions after the analysis is complete to evaluate the patient's progress or explore the impact of major life changes on the patient's adjustment.

Risks

The chief risk of psychoanalysis is an unsatisfactory relationship between analyst and analysand. The analysand is expected to reenact feelings and sometimes behaviors from earlier relationships (usually with parents and other family members) by unconsciously redirecting early experiences to the analyst. This phenomenon is called transference. The analyst's response (including emotions) toward the analysand is called the countertransference. One focus in psychoanalysis is identifying the transference in the relationship and exploring its meaning in order to shed light on the analysand's unconscious. If the relationship between analyst and analysand is either fragile or distorted in some way, however, the analysis will not be productive or successful.

Research and general acceptance

There are relatively few research studies comparing psychoanalysis with other forms of treatment or other talk therapies. As of mid-2010, there are five clinical research studies of psychoanalysis under way, compared to 19 studies of psychodynamic psychotherapy and 834 studies of cognitive-behavioral therapy.

Training and certification

In the early years of psychoanalysis, credentialed analysts were all medical doctors (psychiatrists). Since 1978, however, the profession has been opened to clinical psychologists with doctorates (Psy.D.), social workers holding an M.S.W., or other mental health professionals holding a Ph.D. Training is long and rigorous; a candidate must undergo a personal analysis and complete 600 hours of classroom instruction over a four-year period. The trainee must then conduct between two and four cases, the exact number depending on the specific institute, under a supervising analyst. The cases must include both men and women. The supervision usually continues for several years and takes place in the supervisor's office.

Resources

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- Lombardi, R. “Mental Models and Language Registers in the Psychoanalysis of Psychosis: An Overview of a Thirteen-Year Analysis.” *International Journal of Psychoanalysis* 84 (August 2003): 843–863.
- Roland, A. “Psychoanalysis Across Civilizations: A Personal Journey.” *Journal of the American Academy of Psychoanalysis and Dynamic Psychiatry* 31 (Summer 2003): 275–295.

ORGANIZATIONS

- American Academy of Child and Adolescent Psychiatry (AACAP), 3615 Wisconsin Avenue, N.W., Washington, DC, 20016-3007, 202-966-7300, 202-966-2891, <http://www.aacap.org>.
- American Academy of Psychoanalysis and Psychodynamic Psychiatry, One Regency Drive, Bloomfield, CT, 06002, 888-691-8281, 860-286-0787, info@AAPDP.org, <http://www.aapdp.org>.
- American College of Psychoanalysts (ACOPSA), P.O. Box 570218, Dallas, TX, 75357, 972-613-0985, <http://www.acopsa.org/index.php>.
- American Psychoanalytic Association (APsaA), 309 East 49th Street, New York, NY, 10017, 212-752-0450, 212-593-0571, info@apsa.org, <http://www.apsa.org>.
- Association for Child Psychoanalysis (ACP), 7820 Enchanted Hills Blvd., #A-233, Rio Rancho, NM, 87144, 505-771-0372, <http://www.childanalysis.org>.

International Psychoanalytic Association (IPA), Broom-hills, Woodside Lane, London, United Kingdom, N12 8UD, +44 20 8446 8324, +44 20 8445 4729, ipa@ipa.org.uk, <http://www.ipa.org.uk/Public>.

National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD, 20892-9663, 301-443-4513, 866-615-6464, 301-443-4279, nimhinfo@nih.gov, <http://www.nimh.nih.gov/index.shtml>.

Rebecca J. Frey, PhD

Psychogenic disorder see **Somatoform disorders**

Psychological tests

Definition

Psychological tests are written, visual, or verbal evaluations administered to assess the cognitive and emotional functioning of children and adults.

Purpose

Psychological tests are used to assess a variety of mental abilities and attributes, including achievement and ability, personality, and neurological functioning.

Achievement and ability tests

For children, academic achievement, ability, and intelligence tests may be used as a tool in school placement, in determining the presence of a learning disability or a developmental delay, in identifying giftedness, or in tracking intellectual development. Intelligence testing may be used with adults to determine vocational ability (e.g., in career counseling) or to assess adult intellectual ability in the classroom.

Personality tests

Personality tests are administered for a wide variety of reasons, from diagnosing psychopathology (e.g., personality disorder, depressive disorder) to screening job candidates. They may be used in an educational or vocational setting to determine personality strengths and weaknesses, or in the legal system to evaluate parolees.

Neuropsychological tests

Patients who have experienced a traumatic brain injury, brain damage, or organic neurological problems (for example, **dementia**) are administered neuropsychological tests to assess their level of functioning

and identify areas of mental impairment. They may also be used to evaluate the progress of a patient who has undergone treatment or **rehabilitation** for a neurological injury or illness. In addition, certain neuropsychological measures may be used to screen children for developmental delays and/or learning disabilities.

Precautions

Psychological testing requires a clinically trained examiner. All psychological tests should be administered, scored, and interpreted by a trained professional, preferably a psychologist or psychiatrist with expertise in the appropriate area.

Psychological tests are only one element of a psychological assessment. They should never be used alone as the sole basis for a diagnosis. A detailed history of the test subject and a review of psychological, medical, educational, or other relevant records are required to lay the groundwork for interpreting the results of any psychological measurement.

Cultural and language differences in the test subject may affect test performance and may result in inaccurate test results. The test administrator should be informed before psychological testing begins if the test taker is not fluent in English and/or belongs to a minority culture. In addition, the subject's motivation and motives may also affect test results.

Description

Psychological tests are formalized measures of mental functioning. Most are objective and quantifiable; however, certain projective tests may involve some level of subjective interpretation. Also known as inventories, measurements, questionnaires, and scales, psychological tests are administered in a variety of settings, including preschools, primary and secondary schools, colleges and universities, hospitals, outpatient health care settings, social agencies, prisons, and employment or human resource offices. They come in a variety of formats, including written, verbal, and computer administered.

Achievement and ability tests

Achievement and ability tests are designed to measure the level of an individual's intellectual functioning and cognitive ability. Most achievement and ability tests are standardized, meaning that norms were established during the design phase of the test by administering the test to a large representative sample of the test population. Achievement and ability tests follow a uniform testing protocol or procedure (i.e., test instructions, test conditions, and scoring procedures) and their scores can be interpreted in relation to established norms. Common achievement

and ability tests include the **Wechsler intelligence test** (WISC-III and WAIS) and the **Stanford-Binet intelligence scales**.

Personality tests

Personality tests and inventories evaluate the thoughts, emotions, attitudes, and behavioral traits that comprise personality. The results of these tests determine an individual's personality strengths and weaknesses, and may identify certain disturbances in personality, or psychopathology. Tests such as the **Minnesota multiphasic personality inventory (MMPI-2)** and the Millon clinical multiaxial inventory III (MMPI-III), are used to screen individuals for specific psychopathologies or emotional problems.

Another type of personality test is the projective personality assessment. A projective test asks a subject to interpret some ambiguous stimuli, such as a series of inkblots. The subject's responses provide insight into his or her thought processes and personality traits. For example, the Rorschach inkblot test and the **Holtzman ink blot test (HIT)** use a series of inkblots that the test subject is asked to identify. Another projective assessment, the **Thematic apperception test (TAT)**, asks the subject to tell a story about a series of pictures. Some consider projective tests to be less reliable than objective personality tests. If the examiner is not well-trained in psychometric evaluation, subjective interpretations may affect the evaluation of these tests.

Neuropsychological tests

Many insurance plans cover all or a portion of diagnostic neuropsychological or psychological testing. Medicare reimburses for psychological and neuropsychological testing. Billing time typically includes test administration, scoring and interpretation, and reporting.

Preparation

Prior to the administration of any psychological test, the administrator should provide the test subject with information on the nature of the test and its intended use, complete standardized instructions for taking the test (including any time limits and penalties for incorrect responses), and information on the confidentiality of the results. After these disclosures are made, informed consent should be obtained from the test subject before testing begins (except in cases of legally mandated testing, where consent is not required of the subject).

KEY TERMS

Norms—A fixed or ideal standard; normative or mean score for a particular age group.

Psychopathology—A mental disorder or illness, such as schizophrenia, personality disorder, or major depressive disorder.

Quantifiable—Can be expressed as a number. The results of quantifiable psychological tests can be translated into numerical values, or scores.

Representative sample—A random sample of people that adequately represent the test taking population in age, gender, race, and socioeconomic standing.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

Normal results

All psychological and neuropsychological assessments should be administered, scored, and interpreted by a trained professional. When interpreting test results for test subjects, the test administrator will review with subjects what the test evaluates, its precision in evaluation, any margins of error involved in scoring, and what the individual scores mean in the context of overall test norms and the background of the test subject.

ORGANIZATIONS

American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (202) 336-5500, (800) 374-2721, <http://www.apa.org>.

Paula Anne Ford-Martin

Psychosis

Definition

Psychosis is a symptom or feature of mental illness typically characterized by radical changes in personality, impaired functioning, and a distorted or non-existent sense of objective reality.

Description

Patients suffering from psychosis have impaired reality testing; that is, they are unable to distinguish

personal subjective experience from the reality of the external world. They experience **hallucinations** and/or **delusions** that they believe are real, and may behave and communicate in an inappropriate and incoherent fashion. Psychosis may appear as a symptom of a number of mental disorders, including mood and **personality disorders**. It is also the defining feature of **schizophrenia**, schizopreniform disorder, **schizoaffective disorder**, delusional disorder, and the psychotic disorders (i.e., brief psychotic disorder, shared psychotic disorder, psychotic disorder due to a general medical condition, and substance-induced psychotic disorder).

Causes and symptoms

Psychosis may be caused by the interaction of biological and psychosocial factors, depending on the disorder in which it presents; psychosis can also be caused by purely social factors, with no biological component.

Biological factors that are regarded as contributing to the development of psychosis include genetic abnormalities and substance use. With regard to chromosomal abnormalities, studies indicate that 30% of patients diagnosed with a psychotic disorder have a microdeletion at chromosome 22q11. Another group of researchers has identified the gene G72/G30 at chromosome 13q33.2 as a susceptibility gene for childhood-onset schizophrenia and psychosis not otherwise specified.

With regard to **substance abuse**, several different research groups reported in 2004 that cannabis (**marijuana**) use is a risk factor for the onset of psychosis.

Migration is a social factor that influences people's susceptibility to psychotic disorders. Psychiatrists in Europe have noted the increasing rate of schizophrenia and other psychotic disorders among immigrants to almost all Western European countries. Black immigrants from Africa or the Caribbean appear to be especially vulnerable. The stresses involved in migration include family breakup, the need to adjust to living in large urban areas, and social inequalities in the new country.

Schizophrenia, schizopreniform disorder, and schizoaffective disorder

Psychosis in schizophrenia and perhaps schizopreniform disorder appears to be related to abnormalities in the structure and chemistry of the brain, and appears to have strong genetic links, but its course and severity can be altered by social factors such as **stress** or a lack of support within the family. The cause

of schizoaffective disorder is less clear cut, but biological factors are also suspected.

Delusional disorder

The exact cause of delusional disorder has not been conclusively determined, but potential causes include heredity, neurological abnormalities, and changes in brain chemistry. Some studies have indicated that delusions are generated by abnormalities in the limbic system, the portion of the brain on the inner edge of the cerebral cortex that is believed to regulate emotions. Delusional disorder is also more likely to develop in persons who are isolated from others in their society by language difficulties and/or cultural differences.

Brief psychotic disorder

Trauma and stress can cause a short-term psychosis (less than a month's duration) known as brief psychotic disorder. Major life-changing events such as the **death** of a family member or a natural disaster have been known to stimulate brief psychotic disorder in patients with no prior history of mental illness.

Psychotic disorder due to a general medical condition

Psychosis may also be triggered by an organic cause, termed a psychotic disorder due to a general medical condition. Organic sources of psychosis include neurological conditions (for example, **epilepsy** and cerebrovascular disease), metabolic conditions (for example, porphyria), endocrine conditions (for example, hyper- or **hypothyroidism**), renal failure, electrolyte imbalance, or **autoimmune disorders**.

Substance-induced psychotic disorder

Psychosis is also a known side effect of the use, **abuse**, and withdrawal from certain drugs. So-called recreational drugs, such as hallucinogenics, PCP, amphetamines, **cocaine**, marijuana, and alcohol, may cause a psychotic reaction during use or withdrawal. Certain prescription medications such as **steroids**, anticonvulsants, chemotherapeutic agents, and antiparkinsonian medications may also induce psychotic symptoms. Toxic substances such as carbon monoxide have also been reported to cause substance-induced psychotic disorder.

Shared psychotic disorder

Shared psychotic disorder, also known as *folie à deux* or psychosis by association, is a relatively rare delusional disorder involving two (or more) people with close emotional ties. In the West, shared

KEY TERMS

Brief psychotic disorder—An acute, short-term episode of psychosis lasting no longer than one month. This disorder may occur in response to a stressful event.

Delirium—An acute but temporary disturbance of consciousness marked by confusion, difficulty paying attention, delusions, hallucinations, or restlessness. Delirium may be caused by drug intoxication, high fever related to infection, head trauma, brain tumors, kidney or liver failure, or various metabolic disturbances.

Delusional disorder—Individuals with delusional disorder suffer from long-term, complex delusions that fall into one of six categories: persecutory, grandiose, jealous, erotomanic, somatic, or mixed.

Delusions—An unshakable belief in something untrue that cannot be explained by religious or cultural factors. These irrational beliefs defy normal reasoning and remain firm even when overwhelming proof is presented to refute them.

Hallucinations—False or distorted sensory experiences that appear to be real perceptions to the person experiencing them.

Paranoia—An unfounded or exaggerated distrust of others, sometimes reaching delusional proportions.

Porphyria—A disease of the metabolism characterized by skin lesions, urine problems, neurologic disorders, and/or abdominal pain.

Schizoaffective disorder—Schizophrenic symptoms occurring concurrently with a major depressive or manic episode.

Schizophrenia—A debilitating mental illness characterized by delusions, hallucinations, disorganized speech and behavior, and inappropriate or flattened affect (a lack of emotions) that seriously hampers the afflicted individual's social and occupational functioning. Approximately 2 million Americans suffer from schizophrenia.

Schizophreniform disorder—A short-term variation of schizophrenia that has a total duration of one to six months.

Shared psychotic disorder—Also known as *folie à deux*, shared psychotic disorder is an uncommon disorder in which the same delusion is shared by two or more individuals.

Tardive dyskinesia—Involuntary movements of the face and/or body that are a side effect of the long-term use of some older antipsychotic (neuroleptic) drugs. Tardive dyskinesia affects 15%-20% of patients on long-term neuroleptic treatment.

psychosis most commonly develops between two sisters or between husband and wife, while in Japan the most common form involves a parent and a son or daughter. Shared psychosis occasionally involves an entire nuclear family.

Psychosis is characterized by the following symptoms:

- Delusions. Those delusions that occur in schizophrenia and its related forms are typically bizarre (i.e., they could not occur in real life). Delusions occurring in delusional disorder are more plausible, but still patently untrue. In some cases, delusions may be accompanied by feelings of paranoia.
- Hallucinations. Psychotic patients see, hear, smell, taste, or feel things that aren't there. Schizophrenic hallucinations are typically auditory or, less commonly, visual; but psychotic hallucinations can involve any of the five senses.
- Disorganized speech. Psychotic patients, especially those with schizophrenia, often ramble on in incoherent, nonsensical speech patterns.

- Disorganized or catatonic behavior. The catatonic patient reacts inappropriately to his/her environment by either remaining rigid and immobile or by engaging in excessive motor activity. Disorganized behavior is behavior or activity that is inappropriate for the situation, or unpredictable.

Diagnosis

Patients with psychotic symptoms should undergo a thorough **physical examination** and history to rule out such possible organic causes as seizures, **delirium**, or alcohol withdrawal, and such other psychiatric conditions as dissociation or panic attacks. If a psychiatric cause such as schizophrenia is suspected, a mental health professional will typically conduct an interview with the patient and administer one of several clinical inventories, or tests, to evaluate mental status. This assessment takes place in either an outpatient or hospital setting.

Psychotic symptoms and behaviors are considered psychiatric emergencies, and persons showing signs of

psychosis are frequently taken by family, friends, or the police to a hospital emergency room. A person diagnosed as psychotic can be legally hospitalized against his or her will, particularly if he or she is violent, threatening to commit **suicide**, or threatening to harm another person. A psychotic person may also be hospitalized if he or she has become malnourished or ill as a result of failure to feed, dress appropriately for the climate, or otherwise take care of him- or herself.

Treatment

Psychosis that is symptomatic of schizophrenia or another psychiatric disorder should be treated by a psychologist and/or psychiatrist. An appropriate course of medication and/or psychosocial therapy is employed to treat the underlying primary disorder. If the patient is considered to be at risk for harming himself or others, inpatient treatment is usually recommended.

Treatment of shared psychotic disorder involves separating the affected persons from one another as well as using antipsychotic medications and **psychotherapy**.

Antipsychotic medication such as thioridazine (Mellaril), haloperidol (Haldol), chlorpromazine (Thorazine), clozapine (Clozaril), sertindole (Serlect), olanzapine (Zyprexa), or risperidone (Risperdal) is usually prescribed to bring psychotic symptoms under control and into remission. Possible side effects of antipsychotics include **dry mouth**, drowsiness, muscle stiffness, and **tardive dyskinesia** (involuntary movements of the body). Agranulocytosis, a potentially serious but reversible health condition in which the white blood cells that fight infection in the body are destroyed, is a possible side effect of clozapine. Patients treated with this drug should undergo weekly blood tests to monitor white blood cell counts for the first six months, then every two weeks thereafter.

After an acute psychotic episode has subsided, antipsychotic drug maintenance treatment is typically employed and psychosocial therapy and living and vocational skills training may be attempted.

Prognosis

Prognosis for brief psychotic disorder is quite good; for schizophrenia, less so. Generally, the longer and more severe a psychotic episode, the poorer the prognosis is for the patient. Early diagnosis and treatment are critical to improving outcomes for the patient across all psychotic disorders.

Approximately 10% of America's permanently disabled population is comprised of schizophrenic individuals. The mortality rate of schizophrenic individuals is also high—approximately 10% of schizophrenics commit suicide, and 20% attempt it. However, early diagnosis and long-term follow up care can improve the outlook for these patients considerably. Roughly 60% of patients with schizophrenia will show substantial improvement with appropriate treatment.

Resources

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ORGANIZATIONS

American Psychiatric Association (APA), 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209, (888) 357-7924, apa@psych.org, <http://www.psych.org>.

American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (202) 336-5500, (800) 374-2721, <http://www.apa.org>.

National Alliance for the Mentally Ill (NAMI), 3803 N. Fairfax Dr., Ste. 100, Arlington, VA, 22203, (703) 524-7600, (703) 524-9094, (800) 950-6264, <http://www.nami.org>.

National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Bethesda, MD, 20892, (301) 443-4513, (301) 443-4279, (866) 615-6464, nimhinfo@nih.gov, <http://www.nimh.nih.gov>.

Paula Anne Ford-Martin
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Psychosocial disorders

Definition

A psychosocial disorder is a mental illness caused or influenced by life experiences, as well as maladjusted cognitive and behavioral processes.

Description

The term psychosocial refers to the psychological and social factors that influence mental health. Social influences such as peer pressure, parental support, cultural and religious background, socioeconomic status, and interpersonal relationships all help to shape personality and influence psychological makeup. Individuals with psychosocial disorders frequently have difficulty functioning in social situations and may have problems effectively communicating with others.

The American Psychiatric Association distinguishes 16 different subtypes (or categories) of mental illness. Although psychosocial variables arguably have some degree of influence on all subtypes of mental illness, the major categories of mental disorders thought to involve significant psychosocial factors include:

- Substance-related disorders. Disorders related to alcohol and drug use, abuse, dependence, and withdrawal.
- Schizophrenia and other psychotic disorders. These include the schizoid disorders (schizophrenia, schizophreniform, and schizoaffective disorder), delusional disorder, and psychotic disorders.
- Mood disorders. Affective disorders such as depression (major, dysthymic) and bipolar disorders.
- Anxiety disorders. Disorders in which a certain situation or place triggers excessive fear and/or anxiety symptoms (e.g., dizziness, racing heart), such as panic disorder, agoraphobia, social phobia, obsessive-compulsive disorder, post-traumatic stress disorder, and generalized anxiety disorders.
- Somatoform disorders. Somatoform disorders involve clinically significant physical symptoms that

cannot be explained by a medical condition (e.g., somatization disorder, conversion disorder, pain disorder, hypochondriasis, and body dysmorphic disorder).

- Factitious disorders. Disorders in which an individual creates and complains of symptoms of a non-existent illness in order to assume the role of a patient (or sick role).
- Sexual and gender identity disorders. Disorders of sexual desire, arousal, and performance. It should be noted that the categorization of gender identity disorder as a mental illness has been a point of some contention among mental health professionals.
- Eating disorders. Anorexia and bulimia nervosa.
- Adjustment disorders. Adjustment disorders involve an excessive emotional or behavioral reaction to a stressful event.
- Personality disorders. Maladjustments of personality, including paranoid, schizoid, schizotypal, anti-social, borderline, histrionic, narcissistic, avoidant, dependent, and obsessive-compulsive personality disorders (the latter not to be confused with the anxiety disorder OCD).
- Disorders usually first diagnosed in infancy, childhood, or adolescence. Some learning and developmental disorders (e.g., ADHD) may be partially psychosocial in nature.

Causes and symptoms

It is important to note that the causes of mental illness are diverse and not completely understood. The majority of psychological disorders are thought to be caused by a complex combination of biological, genetic (hereditary), familial, and social factors or biopsychosocial influences. In addition, the role that each of these play can differ from person to person, so that a disorder such as depression that is caused by genetic factors in one person may be caused by a traumatic life event in another.

The symptoms of psychosocial disorders vary depending on the diagnosis in question. In addition to disorder-specific symptoms, individuals with psychosocial dysfunction usually have difficulty functioning normally in social situations and may have trouble forming and maintaining close interpersonal relationships.

Diagnosis

Patients with symptoms of psychosocial disorders or other mental illness should undergo a thorough **physical examination** and patient history to rule out

an organic cause for the illness (such as a neurological disorder). If no organic cause is suspected, a psychologist or other mental health care professional will meet with the patient to conduct an interview and take a detailed social and medical history. If the patient is a minor, interviews with a parent or guardian may also be part of the diagnostic process. The physician may also administer one or more **psychological tests** (also called clinical inventories, scales, or assessments).

Treatment

Counseling is typically a front-line treatment for psychosocial disorders. A number of counseling or talk therapy approaches exist, including **psychotherapy**, cognitive therapy, behavioral therapy, and **group therapy**. Therapy or counseling may be administered by social workers, nurses, licensed counselors and therapists, psychologists, or psychiatrists.

Psychoactive medication may also be prescribed for symptom relief in patients with mental disorders considered psychosocial in nature. For disorders such as major depression or **bipolar disorder**, which may have psychosocial aspects but also have known organic causes, drug therapy is a primary treatment approach. In cases such as personality disorder that are thought to not have biological roots, psychoactive medications are usually considered a secondary, or companion, treatment to psychotherapy.

Many individuals are successful in treating psychosocial disorders through regular attendance in self-help groups or 12-step programs such as Alcoholics Anonymous. This approach, which allows individuals to seek advice and counsel from others in similar circumstances, can be extremely effective.

In some cases, treating mental illness requires hospitalization of the patient. This hospitalization, also known as inpatient treatment, is usually employed in situations where a controlled therapeutic environment is critical for the patient's recovery (e.g., **rehabilitation** treatment for **alcoholism** or other drug addictions), or when there is a risk that the patient may harm himself (**suicide**) or others. It may also be necessary when the patient's physical health has deteriorated to a point where life-sustaining treatment is necessary, such as with severe **malnutrition** associated with **anorexia nervosa**.

Alternative treatment

Therapeutic approaches such as **art therapy** that encourage self-discovery and empowerment may be useful in treating psychosocial disorders. **Art therapy**, the use of the creative process to express and

KEY TERMS

Affective disorder—An emotional disorder involving abnormal highs and/or lows in mood.

Bipolar disorder—An affective mental illness that causes radical emotional changes and mood swings, from manic highs to depressive lows. The majority of bipolar individuals experience alternating episodes of mania and depression.

Bulimia—An eating disorder characterized by binge eating and inappropriate compensatory behavior such as vomiting, misusing laxatives, or excessive exercise.

Cognitive processes—Thought processes (e.g., reasoning, perception, judgment, memory).

Learning disorders—Academic difficulties experienced by children and adults of average to above-average intelligence that involve reading, writing, and/or mathematics, and which significantly interfere with academic achievement or daily living.

Schizophrenia—A debilitating mental illness characterized by delusions, hallucinations, disorganized speech and behavior, and flattened affect (i.e., a lack of emotions) that seriously hampers normal functioning.

understand emotion, encompasses a broad range of humanistic disciplines, including visual arts, dance, drama, music, film, writing, literature, and other artistic genres. This use of the creative process is believed to provide the patient with a means to gain insight to emotions and thoughts they might otherwise have difficulty expressing. After the artwork is created, the patient continues the therapeutic journey by interpreting its meaning under the guidance of a trained therapist.

Prognosis

According to the National Institute of Mental Health, more than 90% of Americans who commit suicide have a diagnosable mental disorder, so swift and appropriate treatment is important. Because of the diversity of the types of mental disorders influenced by psychosocial factors, and the complexity of diagnosis and treatment, the prognosis for psychosocial disorders is highly variable. In some cases, they can be effectively managed with therapy and/or medication. In others, mental illness can cause long-term disability.

Prevention

Participating in therapy or self-help groups can encourage patients to take an active part in their treatment program and to recognize symptoms of a relapse of their condition. In addition, educating friends and family members on the nature of the psychosocial disorder can assist them in knowing how and when to provide support to the patient.

Resources

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ORGANIZATIONS

National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD 20892-9663, (866) 615-6464, nimhinfo@nih.gov, <http://www.nimh.nih.gov/site-info/contact-nimh.shtml>.

Paula Anne Ford-Martin

Psychosurgery

Definition

Psychosurgery involves severing or otherwise disabling areas of the brain to treat a personality disorder, behavior disorder, or other mental illness. Modern psychosurgical techniques target the pathways between the limbic system (the portion of the brain on the inner edge of the cerebral cortex), which is believed to regulate emotions, and the frontal cortex, where thought processes are seated.

Purpose

Lobotomy is a psychosurgical procedure involving selective destruction of connective nerve fibers or tissue. It is performed on the frontal lobe of the brain and its purpose is to alleviate mental illness and chronic **pain** symptoms. The bilateral cingulotomy, a modern psychosurgical technique that has replaced the lobotomy, is performed to alleviate mental disorders such as major depression, **bipolar disorder**, or **obsessive-compulsive disorder** (OCD), which have not responded to **psychotherapy**, behavioral therapy, electroshock, or pharmacologic treatment. Bilateral cingulotomies are also performed to treat chronic pain in **cancer** patients.

Precautions

Psychosurgery should be considered only after all other nonsurgical psychiatric therapies have been fully explored. Much is still unknown about the biology of the brain and how psychosurgery affects brain function.

Description

Psychosurgery, and lobotomy in particular, reached the height of use just after World War II. Between 1946 and 1949, the use of the lobotomy grew from 500 to 5,000 annual procedures in the United States. At that time, the procedure was viewed as a possible solution to the overcrowded and understaffed conditions in state-run mental hospitals and asylums. Known as prefrontal or transorbital lobotomy, depending on the surgical technique used and area of the brain targeted, these early operations were performed with surgical knives, electrodes, suction, or ice picks, to cut or sweep out portions of the frontal lobe.

Today's psychosurgical techniques are much more refined. Instead of going in "blind" to remove large sections on the frontal lobe, as in these early operations, neurosurgeons use a computer-based process called stereotactic **magnetic resonance imaging** to guide a small electrode to the limbic system (brain structures involved in autonomic or automatic body functions and some emotion and behavior). There, an electrical current **burns** in a small lesion (usually 0.5 in. [1.3 cm] in size). In a bilateral cingulotomy, the cingulate gyrus, a small section of brain that connects the limbic region of the brain with the frontal lobes, is targeted. Another surgical technique uses a noninvasive tool known as a gamma knife to focus beams of radiation at the brain. A lesion forms at the spot where the beams converge in the brain.

Preparation

Candidates for cingulotomies or other forms of psychosurgery undergo a rigorous screening process to ensure that all possible nonsurgical psychiatric treatment options have been explored. Psychosurgery is only performed with the patient's informed consent.

Aftercare

Ongoing behavioral and medication therapy is often required in OCD patients who undergo cingulotomy. All psychosurgery patients should remain under a psychiatrist's care for follow-up evaluations and treatment.

Risks

As with any type of brain surgery, psychosurgery carries the risk of permanent brain damage, though

KEY TERMS

Gamma knife—A surgical tool that focuses beams of radiation at the head, which converge in the brain to form a lesion.

Lesion—Any discontinuity of tissue. Often a cut or wound.

Limbic system—A portion of the brain on the inner edge of the cerebral cortex that is thought to regulate emotions.

Psychosurgery—Brain surgery performed to alleviate chronic psychological conditions such as obsessive-compulsive disorder (OCD), depression, and bipolar disorder.

Stereotactic technique—A technique used by neurosurgeons to pinpoint locations within the brain. It employs computer imaging to create an external frame of reference.

the advent of non-invasive neurosurgical techniques, such as the gamma knife, has reduced the risk of brain damage significantly.

Normal results

In a 1996 study at Massachusetts General Hospital, over one-third of patients undergoing cingulotomy demonstrated significant improvements after the surgery. In contrast to the bizarre behavior and personality changes reported with lobotomy patients in the 1940s and 1950s, modern psychosurgery patients have demonstrated little post-surgical losses of memory or other high-level thought processes.

ORGANIZATIONS

International OCD Foundation, PO Box 961029, Boston, MA, 60219, (617) 973-5801, (617) 973-5803, info@ocfoundation.org, <http://www.ocfoundation.org>.

Massachusetts General Hospital. Functional and Stereotactic Neurosurgery Cingulotomy Unit, 55 Fruit St., Gray 502, Boston, MA, 02114, (617) 724-6590, (617) 724-0339, <http://neurosurgery.mgh.harvard.edu/functional>.

National Alliance for the Mentally Ill (NAMI), 3803 N. Fairfax Dr., Ste. 100, Arlington, VA, 22203, (703) 524-7600, (703) 524-9094, (800) 950-6264, <http://www.nami.org>.

Paula Anne Ford-Martin

Psychotherapy

Definition

Psychotherapy is the treatment of mental or emotional disorders and adjustment problems through the use of psychological techniques rather than through physical or biological means.

Description

Psychoanalysis, the first modern form of psychotherapy, was called the “talking cure,” and the many varieties of therapy practiced today are still characterized by their common dependence on a verbal exchange between the counselor or therapist and the person seeking help. The therapeutic interaction is characterized by mutual trust, with the goal of helping individuals change destructive or unhealthy behaviors, thoughts, and emotions. It is common for experienced therapists to combine several different approaches or techniques. Therapy is terminated when the treatment goals have been met or if the client and/or therapist conclude that it is not working. It can be effective to phase out treatment by gradually reducing the frequency of therapy sessions. Even after regular therapy has ended, the client may return for periodic follow-up and reassessment sessions.

Psychodynamic approach

Freudian psychoanalysis places emphasis on uncovering unconscious motivations and breaking down defenses. Therapy sessions may be scheduled once or even twice a week for a year or more. This type of therapy is appropriate when internal conflicts contribute significantly to a person’s problems.

Behavioral techniques

In contrast to the psychodynamic approach, behavior-oriented therapy is geared toward helping people see their problems as learned behaviors that can be modified, without looking for unconscious motivations or hidden meanings. According to the theory behind this approach, once behavior is changed, feelings will change as well. Probably the best-known type of behavioral therapy is behavior modification, which focuses on eliminating undesirable habits by providing positive reinforcement for the more desirable behaviors.

Another behavioral technique is systematic desensitization, in which people are deliberately and gradually exposed to a feared object or experience to help them overcome their fears. A person who is afraid of

dogs may first be told to visualize a dog, then is given a stuffed toy dog, then exposed to a real dog seen at a distance, and eventually forced to interact with a dog at close range. Relaxation training is another popular form of behavior therapy. Through such techniques as deep breathing, visualization, and progressive muscle relaxation, clients learn to control fear and **anxiety**.

Cognitive methods

Some behavior-oriented therapy methods are used to alter not only overt behavior, but also the thought patterns that drive it. This type of treatment is known as **cognitive-behavioral therapy** (or just cognitive therapy). Its goal is to help people break out of distorted, harmful patterns of thinking and replace them with healthier ones. Common examples of negative thought patterns include: magnifying or minimizing the extent of a problem, “all or nothing” thinking (e.g., a person regards himself as either perfect or worthless), overgeneralization (arriving at broad conclusions based on one incident, for example), and personalization (continually seeing oneself as the cause or focus of events).

In cognitive-behavioral therapy, a therapist may talk to the client, pointing out illogical thought patterns, or use a variety of techniques, such as thought substitution, in which a frightening or otherwise negative thought is driven out by substituting a pleasant thought in its place. Clients may also be taught to use positive self-talk, a repetition of positive affirmations. Cognitive therapy usually takes a longer amount of time as it treats more serious problems.

Couples therapy

Couples therapy focuses on the relationship between two people, typically who have a romantic or sexual connection. The aim of the therapy is to concentrate on the problems of the relationship and make each partner feel that they have an equal role. The therapy can be administered by either a male or female therapist, but many couples feel that having both a male and female therapist in the session is beneficial.

Family and group therapy

Family therapy has proven effective in treating a number of emotional and adjustment problems. While the client's immediate complaint is the initial focus of attention, the ultimate goal of family therapy is to improve the interaction between all family members and enhance communication and coping

skills on a long-term basis (although therapy itself need not cover an extended time period). **Group therapy**, which is often combined with individual therapy, offers the support and companionship of other people experiencing the same or similar problems and issues.

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ORGANIZATIONS

- American Psychological Association, 750 First Street NE, Washington, DC, 20002-4242, 202-336-5500, 800-374-2721, <http://www.apa.org>.
- Association for Psychological Science, 1133 Fifteenth Street NW, Suite 1000, Washington, DC, 20005, (202) 293-9300, (202) 293-9350, <http://www.psychologicalscience.org>.

Ruth A. Wienclaw, PhD
Brenda W. Lerner

Psyllium preparations see **Laxatives**

PT see **Prothrombin time**

Pterygium see **Pinguecula and pterygium**

Ptomaine poisoning see **Food poisoning**

Ptosis

Definition

Ptosis is the term used for a drooping upper eyelid. Ptosis, also called blepharoptosis, can affect one or both eyes.

Description

The eyelids serve to protect and lubricate the outer eye. The upper eyelid is lifted by a muscle called the levator muscle. Inside the back part of the lid is a tarsal plate which adds rigidity to the lid. The levator muscle is attached to the tarsal plate by a flat tendon called the levator aponeurosis. When the muscle cannot lift the eyelid or lifts it only partially, the person is said to have a ptosis.

There are two types of ptosis, acquired and congenital. Acquired ptosis is more common. Congenital ptosis is present at birth. Both congenital and acquired ptosis can be, but are not necessarily, hereditary.

Causes and symptoms

Ptosis may occur because the levator muscle's attachment to the lid is weakening with age. Acquired ptosis can also be caused by a number of different things, such as disease that impairs the nerves, diabetes, injury, tumors, inflammation, or aneurysms. Congenital ptosis may be caused by a problem with nerve innervation or a weak muscle. Drooping eyelids may also be the result of diseases such as **myotonic dystrophy** or **myasthenia gravis**.



A close-up view of a drooping upper eyelid (ptosis) on an elderly woman's face. Ptosis is normally due to a weakness of the levator muscle of the upper eyelid or to interference with the nerve supply to the muscle. (Dr. P. Marazzi/Photo Researchers, Inc.)

KEY TERMS

Congenital—A condition existing at birth.

Hereditary—A condition passed from parent to child; a genetic condition.

The primary symptom of ptosis is a drooping eyelid. Adults will notice a loss of visual field because the upper portion of the eye is covered. Children who are born with a ptosis usually tilt their head back in an effort to see under the obstruction. Some people raise their eyebrows in order to lift the lid slightly and therefore may appear to be frowning.

Diagnosis

Diagnosis of ptosis is usually made by observing the drooping eyelid. Finding the cause of the condition will require testing for any of the illnesses or injuries known to have this effect. Some possible tests include x rays and blood tests.

Treatment

Ptosis is usually treated surgically. Surgery can generally be done on an outpatient basis under local anesthetic. For minor drooping, a small amount of the eyelid tissue can be removed. For more pronounced ptosis, the approach is to surgically shorten the levator muscle or connect the lid to the muscles of the eyebrow. If the aponeurosis has separated from the tarsal plate, it can be reattached. Correcting the ptosis is usually done only after determining the cause of the condition. For example, myasthenia gravis must be ruled out before performing any surgery. As with any surgery, there are risks, and they should be discussed with the surgeon.

Children with ptosis need not have surgery immediately, but their vision should be checked periodically to prevent lazy eye (**amblyopia**).

"Ptosis crutches" are also available. These can be attached to the frame of eyeglasses to hold up the eyelid. These devices are uncomfortable and usually not well tolerated.

Prognosis

After diagnosing the cause of a drooping eyelid, then correcting the condition, most people have no further problems related to the ptosis. The correction, however, may still not make the eyes symmetrical.

Patients should have reasonable expectations and discuss the outcome with their doctor prior to surgery.

Prevention

Ptosis cannot be prevented.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, (415) 561-8500, <http://www.aoa.org>.

American Medical Association, 515 N. State St., Chicago, IL, 60654, (800) 621-8335, <http://www.ama-assn.org>.

American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, (314) 991-4100, (314) 991-4101, (800) 365-2219, <http://www.aoa.org>.

Dorothy Elinor Stonely

PTSD see **Post-traumatic stress disorder**

PTT see **Partial thromboplastin time**

desultory fashion on past age 20. Unlike girls, African American boys begin puberty at roughly the same age as their counterparts in other racial groups.

The age at onset of puberty can be affected by such factors as nutritional status and geography. In general, malnourished youngsters begin puberty at later ages than those who are adequately nourished. Diseases that affect the digestive tract, such as inflammatory bowel disease (IBD) and intestinal parasites, are known to postpone the onset of puberty. **Tuberculosis** is another disease associated with later onset of puberty.

Puberty also begins later among people living at higher altitudes. As of 2010, the highest average age at onset of puberty is found among groups that practice subsistence farming in the high deserts of central Asia. In Europe, children living in the warmer climates of the countries bordering the Mediterranean begin puberty about six months earlier on average than those living in Scandinavia.

There has also been a general shift toward earlier puberty in the developed countries for the past century and a half. From 1840 to 1950 there was an average drop of four months per decade in the age of menarche (first menstrual period) in girls in Europe and North America. For example, Norwegian girls had their first period at the average age of 17 in 1840; the average age in Great Britain at that time was 16.5 years, and it was 15.3 years in southern France. In Japan, the shift toward earlier puberty took place later than in the West but was more rapid when it did occur; between 1945 and 1975, the average age of menarche in Japanese girls dropped by 11 months per decade. As of 2010, the average age of menarche in girls worldwide is 11.75 years; in the United States and Canada, it is 12.5 years.

It is thought that genetic factors account for about 46% of the variation in the timing of puberty in both boys and girls. Both early and delayed puberty are known to run in families; however, the genetic association for the timing of puberty is stronger between mothers and daughters than between fathers and sons. The remaining 54% of variation is thought to be accounted for by environmental factors.

Description

Beginning as early as age eight in girls—and two years later, on average, in boys—a group of endocrine glands known as the hypothalamic-pituitary-gonadal (HPG) axis signals the beginning of puberty. The hypothalamus (part of the brain) releases a hormone

Demographics

The age at onset of puberty is partly population-specific. In the United States, the first sign of puberty occurs on average at age 11 in girls, with menstruation and fertility following about a year and a half later. On the other hand, African American girls begin puberty about a full year earlier than Caucasian and Hispanic girls in the United States. The reason for this difference is not known as of 2010.

Boys lag behind girls by about two years. Puberty may not begin until age 16 in boys and continue in a

called gonadotropin-releasing hormone (Gn-RH) that stimulates the pituitary gland. In turn, the pituitary releases its own hormones called gonadotropins that stimulate the gonads and adrenals. From these glands come a flood of sex hormones—androgens and testosterone in the male, estrogens and progestins in the female—that regulate the growth and function of the sex organs. It is interesting to note that the gonadotropins are the same for males and females, but the sex hormones they induce are different.

Risk factors

Risk factors for differences in the timing of puberty include:

- Sex. Girls are more likely to develop precocious puberty than boys, but boys are more likely to have delayed puberty than girls.
- Race. African American girls are three times more likely to develop precocious puberty than Caucasian or Hispanic girls. This racial difference, however, does not hold true for boys.
- Obesity, particularly in girls, is a risk factor for early puberty. One study found that obese girls had an 80% chance of developing breasts before their ninth birthday and starting menstruation before age 12.
- History of injury to the central nervous system caused by trauma, surgery, or radiation therapy.
- History of brain tumors or structural abnormalities in the brain.
- Exposure to products containing sex hormones, including skin or hair products, vitamins, contraceptive pills, or dietary supplements.
- History of either precocious or delayed puberty in other family members.

Causes and symptoms

Puberty begins when the part of the brain called the hypothalamus secretes a hormone (gonadotropin-releasing hormone or Gn-RH) that triggers the pituitary gland to release gonadotropins. These protein hormones in turn stimulate the gonads (ovaries or testes) to produce sex hormones. These sex hormones (especially estrogen in girls and testosterone in boys) are what causes the onset of sexual maturity.

The signs of puberty in girls according to the Tanner stages (described below) are:

- Stage 1: Prepubertal; no development of sexual characteristics
- Stage 2: Thelarche, body odor, growth spurt, first pubic hair

- Stage 3: Breasts enlarge, pubic hair becomes curly, vaginal discharge appears
- Stage 4: Menarche, breasts assume mature female shape
- Stage 5: Adult sexual maturity; pubic hair extends to inner thighs, growth in height slows then stops

The signs of puberty in boys according to the Tanner stages are:

- Stage 1: Prepubertal; no development of sexual characteristics
- Stage 2: Testes enlarge, adult body odor develops
- Stage 3: Penis enlarges, nocturnal emissions (wet dreams) begin, pubic hair appears
- Stage 4: Height spurt, enlargement of penis and scrotum, pubic hair becomes coarser and curlier, male breasts develop
- Stage 5: Adult sexual maturity; pubic hair extends to inner thighs, growth in height slows then stops

Diagnosis

Puberty falling outside the age limits considered normal for any given population should prompt a search for the cause. Parents should consult their child's pediatrician if their child shows any of the signs or symptoms of either precocious or delayed puberty. The pediatrician may refer the child to an endocrinologist (doctor who specializes in disorders of the glands that regulate growth and sexual maturation as well as other body processes).

Some of the possible disturbances of normal puberty include:

- Excess hormone stimulation is the cause for precocious puberty. It can come from the brain in the form of gonadotrophins or from the gonads and adrenals. Overproduction may be caused by functioning tumors or simple overactivity. Brain overproduction can also be the result of brain infections or injury.
- Likewise, delayed puberty is due to insufficient hormone. If the pituitary output is inadequate, so will be the output from the gonads and adrenals. On the other hand, a normal pituitary will overproduce if it senses there are not enough hormones in the circulation.
- There are several congenital disorders (polyglandular deficiency syndromes) that include failure of hormone output. These children do not experience normal puberty, but it may be induced by giving them the proper hormones at the proper time.

KEY TERMS

Adrenals—Glands on top of the kidneys that produce four different types of hormones.

Computed tomography scan (CT)—A method of creating images of internal organs using x rays.

Embryo—The life in the womb during the first two months.

Endocrine system—A system of ductless glands that secrete hormones that regulate a variety of body processes, including growth and sexual maturation. A doctor who specializes in diseases and disorders of these glands is called an endocrinologist.

Gonadotropins—Protein hormones secreted by the pituitary gland.

Hormone—A chemical produced in one place that has an effect somewhere else in the body.

Hypothalamic-pituitary-gonadal (HPG) axis—A term used by doctors to refer to the combined effects of the hypothalamus, the pituitary gland, and the gonads. This group of glands controls sexual maturation in humans as well as other processes.

Hypothalamus—Part of the brain located deep in the center of the skull and just above the pituitary.

Gonads—Glands that make sex hormones and reproductive cells—testes in the male, ovaries in the female.

Magnetic resonance imaging (MRI)—A method of creating images of internal organs. Magnetic resonance imaging (MRI) uses magnet fields and radio-frequency signals.

Menarche—The first menstrual period in a human female, considered the central event of puberty in girls.

Pituitary—The “master gland” of the body, controlling many of the others by releasing stimulating hormones.

Precocious—Developing at an unusually early age.

Syndrome—A collection of abnormalities that occur often enough to suggest they have a common cause.

Tanner stages—A set of scales to measure sexual development during puberty, named for James Tanner (1920–2010), the British pediatrician who devised it.

Thelarche—The onset of breast development in girls. It is usually first noticed as a firm but tender lump directly under the center of the nipple.

- Finally, some females have abnormalities in hormone production that produce male characteristics—so called virilizing syndromes. Should one of these appear during adolescence, it will disturb the normal progress of puberty. Notice that virilizing requires abnormal hormones in the female, while feminizing results from absent hormones in the male. Each embryo starts out life as female. Male hormones transform it if they are present.

Examination

An office **physical examination** can reveal the development of sexual characteristics in a child. The doctor will weigh the child and compare his or her development to charts of normal development for the child’s sex and age. Puberty has been divided into five sexual maturity rating (SMR) stages by two British doctors, W. Marshall and J. M. Tanner. These ratings are often referred to as Tanner stages 1–5. Staging is based on pubic hair growth, on male genital development, and female breast development. In girls, breast development precedes menarche in most cases; the medical term for this change in girls is called

thelarche. Staging helps determine whether the child’s development is normal for a given age.

Both sexes also grow axillary (armpit) hair and pimples. Males develop muscle mass, a deeper voice, and facial hair. Females redistribute body fat. Along with the maturing of the sex organs, there is a pronounced growth spurt averaging 3–4 in. (8–10 cm) and culminating in full adult stature. Puberty can be precocious (early) or delayed. It all depends upon the timing of the release of sex hormones. **Precocious puberty** is usually defined in North American populations as puberty beginning before age 8 in girls and age 9 in boys; delayed puberty is defined as no menarche in girls by age 16 or no testicular enlargement in boys by age 14.

Tests

Delayed or precocious puberty requires measurement of the several hormones involved to determine which are lacking or which are in excess. There are blood tests for each one. If a tumor is suspected, imaging of the suspect organ needs to be done with x rays, **computed tomography scans** (CT scans), or **magnetic resonance imaging** (MRI).

Procedures

In the case of girls with precocious puberty, the doctor may perform a **pelvic ultrasound** to check for the presence of an ovarian cyst or tumor.

Treatment

Children who begin puberty within the normal age range and at roughly the same time as their peers do not ordinarily need treatment. Those who experience either precocious or delayed puberty may need hormonal therapy. Those whose signs of maturation are different from the usual order (for example, a girl who gets her first period long before any changes in her figure, or a boy whose voice changes abruptly before he begins to grow taller and develop facial hair) may benefit from psychological counseling.

Most teenagers experience some discomfort or embarrassment associated with the changes taking place in their bodies, ranging from **acne** and the development of adult body odor to sudden growth spurts and temporary loss of physical coordination as a result. Those who develop much earlier or later than their peers or show the signs of puberty in a different order from the usual pattern for their sex may become acutely self-conscious or anxious. Parents and other family members can help by focusing on the child's personal qualities rather than on physical appearance, and by not teasing the child about the changes taking place in his or her body. In many cases the child's doctor can provide additional reassurance and advice.

Traditional

Drugs

In precocious puberty, the offending gland or tumor may require surgical attention, although there are several drugs now that counteract the effects of hormones released too early. Drugs that have been developed to treat precocious puberty include:

- histrelin (Supprelin LA)
- nafarelin (Synarel)
- synthetic gonadotropin-releasing hormone agonist
- progestin (Depo-Provera)
- leuprolide (Lupron)

If delayed, puberty can be stimulated with the correct hormones. Treatment should not be delayed because necessary bone growth is also affected.

Some doctors, however, prefer to monitor the child's growth for a few months rather than prescribing hormones right away, particularly if there is a family history of either precocious or delayed puberty.

Prognosis

More than 99% of children begin puberty during the normal range of timing for their sex, and the small percentage of those who have precocious puberty (about 1 in every 160 children) or delayed puberty (about the same number) can usually be treated successfully.

Prevention

Some of the factors that influence the timing of puberty, like sex, race, and family history, cannot be changed. Parents can, however, keep children away from dietary supplements or adult medications containing sex hormones, and help their children maintain healthy weight levels.

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American Academy of Child and Adolescent Psychiatry (AACAP), 3615 Wisconsin Avenue, N.W., Washington, DC, 20016-3007, (202) 966-7300, (202) 966-2891, <http://www.aacap.org>.

American Academy of Pediatrics (AAP), 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007, (847) 434-4000, (847) 434-8000, <http://www.aap.org>.

National Institute of Child Health and Human Development (NICHD), Bldg 31, Room 2A32, MSC 2425, 31 Center Drive, Bethesda, MD, 20892, (800) 370-2943, (866) 760-5947, NICHDInformationResourceCenter@mail.nih.gov, <http://www.nichd.nih.gov>.

J. Ricker Polsdorfer, MD
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Pubic lice see **Lice infestation**

Puerperal infection

Definition

The term puerperal infection refers to a bacterial infection following **childbirth**. The infection may also be referred to as puerperal or postpartum fever. The genital tract, particularly the uterus, is the most commonly infected site. In some cases infection can spread to other points in the body. Widespread infection, or **sepsis**, is a rare, but potentially fatal complication.

Description

Puerperal infection affects an estimated 1–8% of new mothers in the United States. Given modern medical treatment and **antibiotics**, it very rarely advances to the point of threatening a woman's life. An estimated 2–4% of new mothers who deliver vaginally suffer some form of puerperal infection, but for cesarean sections, the figure is five to ten times that high.

Deaths related to puerperal infection are very rare in the industrialized world. It is estimated three in 100,000 births result in maternal **death** due to infection. However, the death rate in developing nations may be 100 times higher.

Postpartum fever may arise from several causes, not necessarily infection. If the **fever** is related to infection, it often results from endometritis, an inflammation of the uterus. Urinary tract, breast, and wound infections are also possible, as well as septic **thrombophlebitis**, a blood clot-associated inflammation of veins. A woman's susceptibility to developing an infection is related to such factors as **cesarean section**, extended labor, **obesity**, anemia, and poor prenatal **nutrition**.

Causes and symptoms

The primary symptom of puerperal infection is a fever at any point between birth and 10 days postpartum. A temperature of 100.4°F (38°C) on any two days during this period, or a fever of 101.6°F (38.6 °C) in the first 24 hours postpartum, is cause for suspicion. An assortment of bacterial species may cause puerperal infection. Many of these bacteria are normally found in the mother's genital tract, but other bacteria may be introduced from the woman's intestine and skin or from a health care provider.

The associated symptoms depend on the site and nature of the infection. The most typical site of infection is the genital tract. Endometritis, which affects the uterus, is the most prominent of these infections. Endometritis is much more common if a small part of the placenta has been retained in the uterus. Typically, several species of bacteria are involved and may act synergistically—that is, the bacteria's negative effects are multiplied rather than simply added together. Synergistic action by the bacteria can result in a stubborn infection such as an **abscess**. The major symptoms of a genital tract infection include fever, malaise, abdominal **pain**, uterine tenderness, and abnormal vaginal discharge. If these symptoms do not respond to antibiotic therapy, an abscess or blood clot may be suspected.

Other causes of postpartum fever include urinary tract infections, wound infections, septic thrombophlebitis, and **mastitis**. Mastitis, or breast infection, is indicated by fever, malaise, achy muscles, and reddened skin on the affected breast. It is usually caused by a clogged milk duct that becomes infected. Infections of the urinary tract are indicated by fever, frequent and painful urination, and back pain. An **episiotomy** and a cesarean section carry the risk of a

KEY TERMS

Abscess—A pus-filled area with definite borders.

Blood clot—A dense mat formed by certain components of the blood stream to prevent blood loss.

Cesarean section—Incision through the abdomen and uterus to facilitate delivery.

Computed tomography scan (CT scan)—Cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures.

Episiotomy—Incision of the vulva (external female genitalia) during vaginal delivery to prevent tissue tearing.

Heparin—A blood component that controls the amount of clotting. It can be used as a drug to reduce blood clot formation.

Heparin challenge test—A medical test to evaluate how readily the blood clots.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and

radio waves to generate signals from atoms in the body. These signals are used to construct images of internal structures.

Postpartum—Referring to the time period following childbirth.

Prophylactic—Measures taken to prevent disease.

Sepsis—The presence of viable bacteria in the blood or body tissues.

Septic—Referring to the presence of infection.

Thrombophlebitis—An inflammation of veins accompanied by the formation of blood clots.

Ultrasound examination—A medical test in which high frequency sound waves are directed at a particular internal area of the body. As the sound waves are reflected by internal structures, a computer uses the data to construct an image of the structures.

Warfarin—A drug that reduces the ability of the blood to clot.

wound infection. Such infections are suggested by a fever and pus-like discharge, inflammation, and swelling at wound sites.

Diagnosis

Fever is not an automatic indicator of puerperal infection. A new mother may have a fever owing to prior illness or an illness unconnected to childbirth. However, any fever within 10 days postpartum is aggressively investigated. Physical symptoms such as pain, malaise, loss of appetite, and others point to infection.

Many doctors initiate antibiotic therapy early in the fever period to stop an infection before it advances. A pelvic examination is done and samples are taken from the genital tract to identify the bacteria involved in the infection. The pelvic examination can reveal the extent of infection and possibly the cause. Blood samples may also be taken for blood counts and to test for the presence of infectious bacteria. A **urinalysis** may also be ordered, especially if the symptoms are indicative of a **urinary tract infection**.

If the fever and other symptoms resist antibiotic therapy, an ultrasound examination or computed tomography scan (CT scan) is done to locate potential abscesses or **blood clots** in the pelvic region. **Magnetic**

resonance imaging (MRI) may be useful as well, in addition to a heparin challenge test if blood clots are suspected. If a lung infection is suspected, a **chest x ray** may also be ordered.

Treatment

Antibiotic therapy is the backbone of puerperal infection treatment. Initial antibiotic therapy may consist of clindamycin and gentamicin, which fight a broad array of bacteria types. If the fever and other symptoms do not respond to these antibiotics, a third, such as ampicillin, is added. Other antibiotics may be used depending on the identity of the infective bacteria and the possibility of an allergic reaction to certain antibiotics.

Antibiotics taken together are effective against a wide range of bacteria, but may not be capable of clearing up the infection alone, especially if an abscess or blood clot is present. Heparin is combined with the antibiotic therapy in order to break apart blood clots. Heparin is used for five–seven days, and may be followed by warfarin for the following month. If the infection is complicated, it may be necessary to surgically drain the infected site. Infected episiotomies can be opened and allowed to drain, but abscesses and blood clots may require surgery.

Prognosis

Antibiotic therapy and other treatment measures are virtually always successful in curing puerperal infections.

Prevention

Careful attention to antiseptic procedures during childbirth is the basic underpinning of preventing infection. With some procedures, such as cesarean section, a doctor may administer prophylactic antibiotics as a preemptive strike against infectious bacteria.

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Julia Barrett

Pulmonary alveolar proteinosis

Definition

Pulmonary alveolar proteinosis (PAP) is a rare disease of the lungs.

Description

In this disease, also called alveolar proteinosis or phospholipidosis, gas exchange in the lungs is progressively impaired by the accumulation of phospholipids, compounds widely found in other living cells of the body. The alveoli are filled with this substance that renders them less effective in protecting the lung. This may explain why infections are often associated with the disease.

Pulmonary alveolar proteinosis most commonly affects people ages 20–50, although it has been reported in children and the elderly. The incidence is 5 out of every 1 million people. The disease is more common among males.

KEY TERMS

Alveoli—The small cavities, or air sacs, in the lungs.

Bronchoscopy—A bronchoscopy is the examination of the bronchi, the primary divisions of the trachea that penetrate the lung, through a tube called a bronchoscope.

Clubbing—Clubbing is the rounding of the ends and swelling of fingers found in people with lung disease.

Remission—Lessening of severity, or abatement of symptoms.

Transtracheal biopsy—A transtracheal biopsy is the removal of a small piece of tissue from across the trachea or windpipe for examination under a microscope.

Causes and symptoms

The cause of this disease is unknown. In some people, however, it appears to result from infection, immune deficiency, or from exposure to silica, aluminum oxide, and a variety of dusts and fumes.

Symptoms include mild **shortness of breath** associated with a nonproductive or minimally productive **cough**, weight loss, and **fatigue**. Acute symptoms such as **fever** or progressive shortness of breath suggest a complicating infection.

Diagnosis

Physical examination may reveal clubbing of the fingers or a bluish coloration of the skin as a result of decreased oxygen.

A **chest x ray** may show alveolar disease. An arterial blood gas reveals low oxygen levels in the blood. **Bronchoscopy** with transtracheal biopsy shows alveolar proteinosis. Specific diagnosis requires a **lung biopsy**.

Treatment

Treatment consists of periodic whole-lung lavage, a washing out of the phospholipids from the lung with a special tube placed in the trachea. This is performed under **general anesthesia**.

Prognosis

In some, spontaneous remission occurs, while in others progressive **respiratory failure** develops.

Disability from respiratory insufficiency is common, but **death** rarely occurs. Repeated lavage may be necessary. Lung transplant is a last resort option.

Prevention

There is no known prevention for this very rare disorder.

ORGANIZATIONS

American Association for Respiratory Care, 9425 N. Mac-Arthur Blvd, Suite 100, Irving, TX, 75063-4706, (972) 243-2272, (972) 484-2720, info@aarc.org, http://www.aarc.org.

American Lung Association, 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20001, (202) 758-3355, (202) 452-1805, (800) 548-8252, info@lungusa.org, http://www.lungusa.org/.

Lorraine Steefel, RN

Pulmonary artery catheterization

Definition

Pulmonary artery catheterization is a diagnostic procedure in which a small catheter is inserted through a neck, arm, chest, or thigh vein and maneuvered into the right side of the heart, in order to measure pressures at different spots in the heart.

Purpose

Pulmonary artery catheterization is performed to:

- evaluate heart failure
- monitor therapy after a heart attack
- check the fluid balance of a patient with serious burns, kidney disease, or after heart surgery
- check the effect of medications on the heart

Precautions

Pulmonary artery catheterization is a potentially complicated and invasive procedure. The doctor must decide if the value of the information obtained will outweigh the risk of catheterization.

Description

Pulmonary artery catheterization, sometimes called Swan-Ganz catheterization, is usually performed at the bedside of a patient in the intensive care unit. A catheter is threaded through a vein in the arm, thigh, chest, or neck until it passes through the right side of the

heart. This procedure takes about 30 minutes. **Local anesthesia** is given to reduce discomfort.

Once the catheter is in place, the doctor briefly inflates a tiny balloon at its end. This temporarily blocks the blood flow and allows the doctor to make a pressure measurement in the pulmonary artery system. Pressure measurements are usually recorded for the next 48–72 hours in different parts of the heart. During this time, the patient must stay in bed so the catheter stays in place. Once the pressure measurements are no longer needed, the catheter is removed.

Preparation

Before and during the test, the patient will be connected to an electrocardiograph, which makes a recording of the electrical stimuli that cause the heart to contract. The insertion site is sterilized and prepared. The catheter is often sutured to the skin to prevent dislodgment.

Aftercare

The patient is observed for any signs of infection or complications from the procedure.

Risks

Pulmonary artery catheterization is not without risks. Possible complications from the procedure include:

- infection at the site where the catheter was inserted
- pulmonary artery perforation
- blood clots in the lungs
- irregular heartbeat

Normal results

Normal pressures reflect a normally functioning heart with no fluid accumulation. These normal pressure readings are:

- right atrium: 1–6 mm of mercury (mm Hg)
- right ventricle during contraction (systolic): 20–30 mm Hg
- right ventricle at the end of relaxation (end diastolic): less than 5 mm Hg
- pulmonary artery during contraction (systolic): 20–30 mm Hg
- pulmonary artery during relaxation (diastolic): about 10 mm Hg
- mean pulmonary artery: less than 20 mm Hg
- pulmonary artery wedge pressure: 6–12 mm Hg
- left atrium: about 10 mm Hg

KEY TERMS

Cardiac shunt—A defect in the wall of the heart that allows blood from different chambers to mix.

Abnormal results

Abnormally high right atrium pressure can indicate:

- pulmonary disease
- right side heart failure
- fluid accumulation
- compression of the heart after hemorrhage (cardiac tamponade)
- right heart valve abnormalities
- pulmonary hypertension (high blood pressure)

Abnormally high right ventricle pressure may indicate:

- pulmonary hypertension (high blood pressure)
- pulmonary valve abnormalities
- right ventricle failure
- defects in the wall between the right and left ventricle
- congestive heart failure
- serious heart inflammation

Abnormally high pulmonary artery pressure may indicate:

- diversion of blood from a left-to-right cardiac shunt
- pulmonary artery hypertension
- chronic obstructive pulmonary disease or emphysema
- blood clots in the lungs
- fluid accumulation in the lungs
- left ventricle failure

Abnormally high pulmonary artery wedge pressure may indicate:

- left ventricle failure
- mitral valve abnormalities
- cardiac insufficiency
- compression of the heart after hemorrhage

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Tish Davidson, A.M.

Pulmonary edema

Definition

Pulmonary **edema** is a condition in which fluid accumulates in the lungs, usually because the heart's left ventricle does not pump adequately.

Description

The build-up of fluid in the spaces outside the blood vessels of the lungs is called pulmonary edema. Pulmonary edema is a common complication of heart disorders, and most cases of the condition are associated with **heart failure**. Pulmonary edema can be a chronic condition, or it can develop suddenly and quickly become life threatening. The life-threatening type of pulmonary edema occurs when a large amount of fluid suddenly shifts from the pulmonary blood vessels into the lung, due to lung problems, **heart attack**, trauma, or toxic chemicals. It can also be the first sign of coronary heart disease.

In heart-related pulmonary edema, the heart's main chamber, the left ventricle, is weakened and does not function properly. The ventricle does not completely eject its contents, causing blood to back up and cardiac output to drop. The body responds by increasing blood pressure and fluid volume to compensate for the reduced cardiac output. This, in turn, increases the force against which the ventricle must expel blood. Blood backs up, forming a pool in the pulmonary blood vessels. Fluid leaks into the spaces between the tissues of the lungs and begins to accumulate. This process makes it more difficult for the lungs to expand. It also impedes the exchange of air and gases between the lungs and blood moving through lung blood vessels.

Causes and symptoms

Most cases of pulmonary edema are caused by failure of the heart's main chamber, the left ventricle. It can be brought on by an acute heart attack, severe **ischemia**, volume overload of the heart's left ventricle, and mitral stenosis. Non-heart-related pulmonary edema is caused by lung problems like **pneumonia**, an excess of intravenous fluids, some types of **kidney disease**, bad **burns**, **liver disease**, nutritional problems, and **Hodgkin's disease**. Non-heart-related pulmonary edema can also be caused by conditions where the lungs do not drain properly, and other conditions where the respiratory veins are blocked.

Early symptoms of pulmonary edema include:

- shortness of breath upon exertion
- sudden respiratory distress after sleep

- difficulty breathing, except when sitting upright
- coughing

In cases of severe pulmonary edema, these symptoms will worsen to:

- labored and rapid breathing
- frothy, bloody fluid containing pus coughed from the lungs (sputum)
- a fast pulse and possibly serious disturbances in the heart's rhythm (atrial fibrillation, for example)
- cold, clammy, sweaty, and bluish skin
- a drop in blood pressure resulting in a thready pulse

Diagnosis

A doctor can usually diagnose pulmonary edema based on the patient's symptoms and a physical exam. Patients with pulmonary edema will have a rapid pulse, rapid breathing, abnormal breath and heart sounds, and enlarged neck veins. A **chest x ray** is often used to confirm the diagnosis. Arterial blood gas testing may be done. Sometimes **pulmonary artery catheterization** is performed to confirm that the patient has pulmonary edema and not a disease with similar symptoms (called **adult respiratory distress syndrome** or "noncardiogenic pulmonary edema").

Treatment

Pulmonary edema requires immediate emergency treatment. Treatment includes: placing the patient in a sitting position, oxygen, assisted or mechanical ventilation (in some cases), and drug therapy. The goal of treatment is to reduce the amount of fluid in the lungs, improve gas exchange and heart function, and, where possible, to correct the underlying disease.

To help the patient breathe better, he/she is placed in a sitting position. High concentrations of oxygen are administered. In cases where respiratory distress is severe, a mechanical ventilator and a tube down the throat (tracheal intubation) will be used to improve the delivery of oxygen. Non-invasive pressure support ventilation is a new treatment for pulmonary edema in which the patient breathes against a continuous flow of positive airway pressure, delivered through a face or nasal mask. Non-invasive pressure support ventilation decreases the effort required to breath, enhances oxygen and carbon dioxide exchange, and increases cardiac output.

Drug therapy could include morphine, nitroglycerin, **diuretics**, angiotensin-converting enzyme (ACE) inhibitors, and **vasodilators**. Vasopressors are used for cardiogenic shock. Morphine is very effective in reducing the patient's **anxiety**, easing breathing, and improving blood flow. Nitroglycerin reduces pulmonary

KEY TERMS

Edema—Swelling caused by accumulation of fluid in body tissues.

Ischemia—A condition in which the heart muscle receives an insufficient supply of blood and slowly starves.

Left ventricle—The large chamber on the lower left side of the heart. The left ventricle sends blood to the aorta and the rest of the body.

Mitral stenosis—Narrowing or constricting of the mitral valve, which separates the left atrium from the left ventricle.

Pulmonary—Referring to the lungs and respiratory system.

blood flow and decreases the volume of fluid entering the overloaded blood vessels. Diuretics, like furosemide (Lasix), promote the elimination of fluids through urination, helping to reduce pressure and fluids in the blood vessels. ACE inhibitors reduce the pressure against which the left ventricle must expel blood. In patients who have severe **hypertension**, a vasodilator such as nitroprusside sodium (Nipride) may be used. For cardiogenic shock, an adrenergic agent (like dopamine hydrochloride [Intropin], dobutamine hydrochloride [Dobutrex], or epinephrine) or a bipyridine (like amrinone lactate [Inocor] or milrinone lactate [Primacor]) are given.

Prognosis

Most patients with pulmonary edema who seek immediate treatment can be treated quickly and effectively.

Prevention

Cardiogenic pulmonary edema can sometimes be prevented by treating the underlying heart disease. These treatments can include maintaining a healthy diet, taking appropriate medications correctly, and avoiding excess alcohol and salt.

Resources

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Lori De Milto

Pulmonary embolism

Definition

Pulmonary **embolism** is an obstruction of a blood vessel in the lungs, usually due to a blood clot, which blocks a coronary artery.

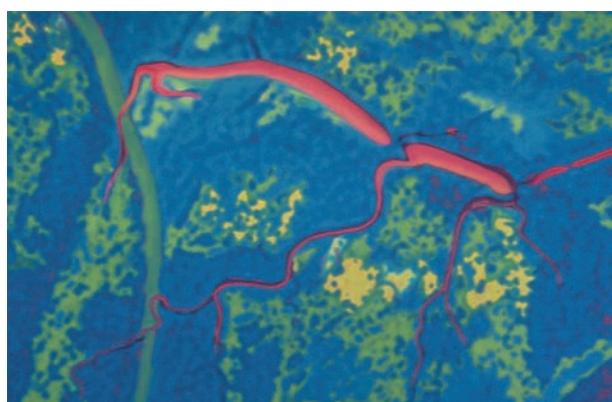
Description

Pulmonary embolism is a fairly common condition that can be fatal. According to the American Heart Association, an estimated 600,000 Americans develop pulmonary embolism annually; 60,000 die from it. As many as 25,000 Americans are hospitalized each year for pulmonary embolism, which is a relatively common complication in hospitalized patients. Even without warning symptoms, pulmonary embolism can cause sudden **death**. Treatment is not always successful.

Pulmonary embolism is difficult to diagnose. Less than 10% of patients who die from pulmonary embolism were diagnosed with the condition. It occurs when emboli block a pulmonary artery, usually due to a blood clot that breaks off from a large vein and travels to the lungs. More than 90% of cases of pulmonary embolism are complications of **deep vein thrombosis**, **blood clots** from the leg or pelvic veins. Emboli can also be comprised of fat, air, or tumor tissue. When emboli block the main pulmonary artery, pulmonary embolism can quickly become fatal.

Causes and symptoms

Pulmonary embolism is caused by emboli that travel through the blood stream to the lungs and block a pulmonary artery. When this occurs, circulation and



An angiography of a pulmonary embolism. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

oxygenation of blood is compromised. The emboli are usually formed from blood clots but are occasionally comprised of air, fat, or tumor tissue. Risk factors include: prolonged bed rest, surgery, **childbirth**, **heart attack**, **stroke**, congestive **heart failure**, **cancer**, **obesity**, a broken hip or leg, **oral contraceptives**, sickle cell anemia, congenital **coagulation disorders**, chest trauma, certain congenital heart defects, and old age.

Common symptoms of pulmonary embolism include:

- labored breathing, sometimes accompanied by chest pain
- a rapid pulse
- a cough that produces bloody sputum
- a low fever
- fluid build-up in the lungs

Less common symptoms include:

- coughing up a lot of blood
- pain caused by movement
- leg swelling
- bluish skin
- fainting
- swollen neck veins

In some cases there are no symptoms.

Diagnosis

Pulmonary embolism can be diagnosed through the patient's history, a physical exam, and diagnostic tests including **chest x ray**, lung scan, pulmonary **angiography**, **electrocardiography**, arterial blood gas measurements, and leg vein ultrasonography or **venography**.

A chest x ray can be normal or show fluid or other signs and rule out other diseases. The lung scan shows poor flow of blood in areas beyond blocked arteries. The patient inhales a small amount of a radiopharmaceutical and pictures of airflow into the lungs are taken with a gamma camera. Then a different radiopharmaceutical is injected into an arm vein and lung blood flow is scanned. A normal result essentially rules out pulmonary embolism. A lung scan can be performed in a hospital or an outpatient facility and takes about 45 minutes.

Pulmonary angiography is the most reliable test for diagnosing pulmonary embolism but it is not used often, because it carries some risk and is expensive, invasive, and not readily available in many hospitals. Pulmonary angiography is a radiographic test which involves injection of a pharmaceutical "contrast agent" to show up the pulmonary arteries. A cinematic camera records the blood flow through the lungs of the

patient, who lies on a table. Pulmonary angiography is usually performed in a hospital's radiology department and takes 30 minutes to one hour.

An electrocardiograph shows the heart's electrical activity and helps distinguish pulmonary embolism from a heart attack. Electrodes covered with conducting jelly are placed on the patient's chest, arms, and legs. Impulses of the heart's activity are traced on paper. The test takes about 10 minutes and can be performed in a physician's office or hospital lab.

Arterial blood gas measurements can be helpful, but they are rarely diagnostic for pulmonary embolism. Blood is taken from an artery instead of a vein, usually in the wrist and it is analyzed for oxygen, carbon dioxide and acid levels.

Venography is used to look for deep vein thrombosis, the most likely source of pulmonary embolism. It is very accurate, but it is not used often, because it is painful, expensive, exposes the patient to a fairly high dose of radiation, and can cause complications. Venography identifies the location, extent, and degree of attachment of the blood clots and enables the condition of the deep leg veins to be assessed. A contrast solution is injected into a foot vein through a catheter. The physician observes the movement of the solution through the vein with a fluoroscope while a series of x rays are taken. Venography takes between 30–45 minutes and can be done in a physician's office, a laboratory, or a hospital. Radionuclide venography, in which a radioactive isotope is injected, is occasionally used, especially if a patient has had reactions to contrast solutions. Most commonly performed tests are ultrasound and Doppler studies of leg veins.

Treatment

Patients with pulmonary embolism are hospitalized and generally treated with clot-dissolving and clot-preventing drugs. **Oxygen therapy** is often needed to maintain normal oxygen concentrations. For people who can't take anticoagulants and in some other cases, surgery may be needed to insert a device that filters blood returning to the heart and lungs. The goal of treatment is to maintain the patient's cardiovascular and respiratory functions while the blockage resolves, which takes 10–14 days, and to prevent the formation of other emboli.

Thrombolytic therapy to dissolve blood clots is the aggressive treatment for very severe pulmonary embolism. Streptokinase, urokinase, and recombinant tissue plasminogen activator (TPA) are thrombolytic agents. Heparin is the injectable anticoagulant (clot-

KEY TERMS

Deep vein thrombosis—A blood clot in the calf's deep vein. This frequently leads to pulmonary embolism if untreated.

Embolii—Clots or other substances that travel through the blood stream and get stuck in an artery, blocking circulation.

Thrombosis—The development of a blood clot inside a blood vessel.

preventing) drug of choice for preventing formation of blood clots. Warfarin, an oral anticoagulant, is usually continued when the patient leaves the hospital and doesn't need heparin any longer.

Prognosis

About 10% of patients with pulmonary embolism die suddenly within the first hour of onset of the condition. The outcome for all other patients is generally good; only 3% of patients who are properly diagnosed and treated die. In cases of undiagnosed pulmonary embolism, about 30% of patients die.

Prevention

Pulmonary embolism risk can be reduced in certain patients through judicious use of antithrombotic drugs such as heparin, venous interruption, gradient elastic stockings and/or intermittent pneumatic compression of the legs.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

Lori De Milto

Pulmonary fibrosis

Definition

Pulmonary fibrosis is scarring in the lungs.

Description

Pulmonary fibrosis develops when the alveoli, tiny air sacs that transfer oxygen to the blood, become damaged and inflamed. The body tries to heal the

KEY TERMS

Alveoli—Tiny air sacs in the lungs where oxygen and carbon dioxide are exchanged with the blood.

Autoimmune disease—A disease that develops when the immune system attacks normal cells or organs.

Bronchoscopy scan—The examination of the air passages through a flexible or rigid tube inserted into the nostril (or mouth). Sometimes cells are collected by washing the lungs with a small amount of fluid.

Computed tomography (CT)—A special x-ray technique that produces a cross sectional image of the organs inside the body.

Corticosteroids—A class of drugs, related to hormones naturally found in the body, that suppress the immune system. One example is prednisone, sold under many brand names including Deltasone.

End-stage lung disease—The final stages of lung disease, when the lung can no longer keep the blood supplied with oxygen. End-stage lungs in pulmonary fibrosis have large air spaces separated by bands of inflammation and scarring.

Farmer's lung—An allergic reaction to moldy hay, most often seen in farmers, that results in lung disease.

Immune suppressant drug—Any drug that dampens immune responses and decreases inflammation.

Inflammation—The body's reaction to an irritant, characterized by the accumulation of immune cells, redness, and swelling.

Lung function tests—Tests of how much air the lungs can move in and out, and how quickly and efficiently this can be done. Lung function tests are usually done by breathing into a device that measures air flow.

Mucous membranes—The moist coverings that line the mouth, nose, intestines, and other internal organs.

Pulmonary artery—The blood vessel that delivers blood from the heart to the lungs.

Sarcoidosis—A disease of unknown origin that results in clumps of immune cells and inflammation in organs throughout the body.

damage with **scars**, but these scars collapse the alveoli and make the lungs less elastic. If the cycle of inflammation and scarring continues, the lungs become increasingly unable to deliver oxygen to the blood. Changes in the lungs can also increase the blood pressure in the pulmonary artery. This condition, called **pulmonary hypertension**, makes the heart work harder and it may cause **heart failure**.

Pulmonary fibrosis can result from many different lung diseases including **sarcoidosis**, drug reactions, autoimmune diseases, environmental **allergies** such as Farmer's lung, and exposure to toxic dusts and gases.

Pulmonary fibrosis that develops without a known cause is called idiopathic pulmonary fibrosis. This disease is equally common in men and women. It is usually diagnosed between the ages of 40 and 60.

Causes and symptoms

The causes and risk factors vary with the underlying disease. They may include genetics, environmental factors, and infections.

The first symptom of pulmonary fibrosis is usually shortness of breath—at first, during **exercise**, but later also while resting. Patients may also have a dry **cough**, a rapid heartbeat, or enlargement of the fingertips and ends of the toes. Some people feel tired or have a **fever**, weight loss, muscle or joint pains. In late stages of the disease, the lack of oxygen in the blood can give the skin and mucus membranes a blue tinge known as **cyanosis**.

Diagnosis

Pulmonary fibrosis is often referred to a lung specialist. Several tests are usually needed to diagnose this disease and determine its cause. They include a **physical examination**, detailed history of the symptoms, chest x rays, lung function tests, and blood tests, including a measurement of the amount of oxygen in the blood. Computed tomography (CT scan) may give a more detailed picture of the lungs. **Bronchoscopy** may be done to examine the air passages and analyze the cells found deep in the lungs.

Lung biopsies are necessary to diagnose some diseases. Lung biopsies can be done through a needle

inserted into the chest through the skin, during bronchoscopy, or as a surgical procedure under **general anesthesia**.

Treatment

The treatment of pulmonary fibrosis depends on the underlying cause. Many diseases are treated by suppressing inflammation with **corticosteroids**. Stronger immune suppressants such as cyclophosphamide (Cytoxan) or azathioprine (Imuran) may also be tried. Some patients need supplemental oxygen. A lung transplant may be an option for incurable diseases. Approximately 60–80% of patients live for at least two years after the transplant.

There is no good treatment for idiopathic pulmonary fibrosis. Only 10–20% of patients with this disease respond to corticosteroids.

Alternative treatment

Anxiety and fear can make breathing difficulties worse. Some patients find that activities such as **yoga**, **prayer** or **meditation**, **music therapy**, or **biofeedback** help to relax them.

Prognosis

The prognosis depends on the specific disease. Some cases may stop progressing or improve, particularly if the cause can be identified and treated. Others may develop quickly or slowly into end-stage lung disease. The course of idiopathic pulmonary fibrosis is very difficult to predict; however, average survival is approximately five to seven years.

Prevention

There is no known prevention for idiopathic pulmonary fibrosis.

Some ways to prevent other causes of pulmonary fibrosis are:

- avoid exposure to particle dust such as asbestos, coal dust, and silica
- avoid exposure to chemical fumes
- do not smoke

Resources

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ORGANIZATIONS

American Lung Association, 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20001, (202) 758-3355, (202) 452-1805, (800) 548-8252, info@lungusa.org, <http://www.lungusa.org/>.

Pulmonary Fibrosis Association, 811 W Evergreen Avenue, Suite 303, Chicago, IL, 60642-2642, (888) 733-6741, info@pulmonaryfibrosis.org, <http://www.pulmonaryfibrosis.org>.

Anna Rovid Spickler, D.V.M., Ph.D.

Pulmonary function tests

Definition

Pulmonary function tests are a group of procedures that measure the function of the lungs, revealing problems in the way a patient breathes. The tests can determine the cause of **shortness of breath** and may help confirm the diagnosis of such lung diseases as **asthma**, **bronchitis**, or **emphysema**. The tests also are performed before any major **lung surgery** to make sure the person won't be disabled by having a reduced lung capacity.

Purpose

Pulmonary function tests can help a doctor diagnose a range of respiratory diseases that might not otherwise be obvious to the doctor or the patient. The tests are important since many kinds of lung problems can be successfully treated if detected early. The tests are also used to measure how a lung disease is progressing, and how serious the lung disease has become. Pulmonary function tests can also be used to assess how a patient is responding to different treatments.

The basic pulmonary function test is **spirometry** (from the Greco-Latin term meaning "to measure breathing"). This test, which can be given in a hospital or doctor's office, measures how much and how fast the air is moving in and out of the lungs. Specific measurements taken during the test include the volume of air from start to finish (the forced vital capacity or FVC); the fastest flow that is achieved; and the volume of air exhaled in the first second of the test.



Adult woman using a peak flow meter. (Edwige/Age Fotostock.)

This third measurement is known as the forced expiratory volume or FEV₁.

Precautions

Pulmonary function tests shouldn't be given to patients who have had a recent **heart attack**, or who have certain other types of heart disease. It is crucial that the patient cooperate with the health care team if accurate results are to be obtained.

Description

For a basic pulmonary function test (PFT), the patient places a clip over the nose and breathes through the mouth into a tube connected to a machine known as a spirometer. First the patient breathes in deeply and then exhales as quickly and forcefully as possible into the tube. The exhalation must last at least 6 seconds for the machine to work properly. Usually the patient repeats this test three times, and

the best of the three results is considered to be the measure of the patient's lung function. The results will help a doctor figure out which type of treatment to pursue.

A peak flow meter can determine how much a patient's airways have narrowed. The peak flow meter is a small handheld device that measures how rapidly a person suspected of asthma can exhale air. This measurement is called the peak expiratory flow rate or PEFR. The peak flow test is not considered the best way to evaluate a patient for asthma, however, as accurate measurement of PEFR requires training to use the meter correctly. Moreover, the normal expected value depends on the patient's sex, age and height and can be quite variable even under ordinary circumstances. Peak flow meters appear to be most useful to a small subset of asthma patients who need to monitor their use of medications.

Another test that can be given to evaluate patients with asthma is the inhalation challenge test. The patient is asked to inhale either cold air or a drug (usually histamine or methacholine) known to irritate the upper airway and produce bronchoconstriction, or narrowing of the airway. Patients with asthma will react to lower doses of the irritant than those with normal lungs.

A test of blood gases is a measurement of the concentration of oxygen and carbon dioxide in the blood, which shows how efficient the gas exchange is in the lungs.

Another type of lung function test reveals how efficient the lungs are in absorbing gas from the blood. This is measured by testing the volume of carbon monoxide a person breathes out after a known volume of the gas has been inhaled. Called the carbon monoxide diffusing test or the transfer factor test, this test consists of asking the patient to first breathe out as much air as possible. The patient is then asked to take a deep breath of a mixture of carbon monoxide (usually about 0.3%) and helium or some other inert tracer gas. The patient holds the gas mixture in the lungs for 10 seconds and then exhales it. The first part of the exhaled gas is discarded; the second portion, which represents the part of the gas that reached the alveoli in the lungs, is analyzed for its carbon monoxide content. This measurement allows the doctor to calculate how much carbon monoxide was taken up by the alveoli while the patient was holding his or her breath.

KEY TERMS

Alveoli (singular, alveolus)—Small spherical sacs at the ends of the bronchioles in the lungs in which blood gases are exchanged.

Body plethysmography—A very sensitive test given to measure damage to the lungs that might be missed by routine pulmonary function tests. The patient sits within a so-called airtight body box while various devices measure both the air pressure in the patient's alveoli and the airflow through the respiratory system.

Carbon monoxide diffusing test—Also called the transfer factor test, this test measures the ability of the patient's lungs to transfer blood gases.

Emphysema—A disease in which the small air sacs in the lungs become damaged, causing shortness of breath. In severe cases it can lead to respiratory or heart failure.

Forced expiratory volume (FEV1)—The maximum volume of air that the patient can forcibly blow out in the first second during the forced vital capacity (FVC) test.

Forced vital capacity (FVC)—A measurement of the volume of air that the patient can exhale from the lungs after taking a deep breath. To measure the FVC, the patient is asked to take the deepest breath

they can and then exhale into a sensor as hard as possible for as long as possible.

Functional residual capacity (FRC)—The volume of air left in the lungs at the end of passive expiration (breathing out). It can be measured by body plethysmography.

Inhalation challenge test—A test given to diagnose asthma by asking the patient to breathe cold air, methacholine, histamine, or another airway irritant and measuring the decline (if any) in the forced expiratory volume (FEV1).

Peak flow—A measurement of the maximum speed of the patient's expiration (breathing out). It is also known as the peak expiratory flow rate or PEFR. Peak flow is measured by a small handheld device called a peak flow meter.

Spirometer—A device used to measure the volume of air inhaled and exhaled by the patient's lungs. It can also be used to measure the rate at which the air is breathed in and out over a specified period of time.

Total lung capacity—The volume of air in the lungs at the end of a deep breath. The normal value in adults is between 4 and 6 quarts.

Body plethysmography is a sophisticated and highly sensitive test used to measure the volume of air in the lungs or the amount of airflow in patients who are too weak to perform multi-breath pulmonary function tests or whose loss of lung function might not be detected by conventional PFTs. The patient sits inside an airtight "body box" and breathes or pants into a mouthpiece connected to a transducer mounted in the wall of the box. Body plethysmography can be used to measure the total volume of air in the patient's lungs and the lungs' resistance to airflow. It can also be used to measure the patient's functional residual capacity (FRC), which is the amount of air remaining in the lungs at the end of a passive (unforced) exhalation. This information can help the doctor to distinguish between obstructive and restrictive lung disease, or to evaluate the patient's response to an inhalation challenge test.

Preparation

The patient should not eat a heavy meal before the test, nor smoke for four to six hours beforehand. The patient's doctor will issue specific instructions about

whether or not to use specific medications, including **bronchodilators** or inhalers, before the test. Sometimes medication may be administered as part of the test.

Patients preparing for an inhalation challenge test should tell their doctor if they have recently had a cold or other viral infection, or shots or immunizations, as these can affect the results of the test.

Body plethysmography requires a period of coaching and somewhat complex instructions for the subject prior to the test. It must also be conducted by a specially trained and certified pulmonary function technologist.

Risks

The risk is minimal for most people, although the test carries a slight risk of a collapsed lung in some patients with lung disease.

Normal results

Normal results are based on a person's age, height, and sex. Normal results are expressed as a percentage of

the predicted lung capacity. The prediction takes into account the patient's age, height, and sex.

Abnormal results

Abnormal results mean that the person's lung capacity is less than 80% of the predicted value. Such findings usually mean that there is some degree of chest or lung disease.

Resources

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Johns Hopkins School of Medicine. *Body Plethysmography Video*. This is a 2-minute video of a pulmonary function technologist coaching a patient in a "body box." http://oac.med.jhmi.edu/res_phys/Encyclopedia/BodyPleth/BodyPleth.HTML

ORGANIZATIONS

American Lung Association, 1301 Pennsylvania Ave. NW, Washington, DC, 20004, (202) 785-3355, <http://www.lungusa.org>.

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Pulmonary heart disease see **Cor pulmonale**

Pulmonary hypertension

Definition

Pulmonary **hypertension** is a rare lung disorder characterized by increased pressure in the pulmonary artery. The pulmonary artery carries oxygen-poor blood from the lower chamber on the right side of the heart (right ventricle) to the lungs where it picks up oxygen.

Description

Pulmonary hypertension is present when the blood pressure in the circulation of the lungs is measured at greater than 25 mm of mercury (Hg) at rest or 30 mm Hg during **exercise**. Pulmonary hypertension can be either primary or secondary:

- Primary pulmonary hypertension. The cause of pulmonary hypertension is unknown. It is rare, affecting one person per million. The illness most often occurs in young adults, especially women.
- Secondary pulmonary hypertension. Secondary pulmonary hypertension is increased pressure of the blood vessels of the lungs as a result of other medical conditions.

Regardless of whether pulmonary hypertension is primary or secondary, the disorder results in thickening of the pulmonary arteries and narrowing of these blood vessels. In response, the right side of the heart works harder to move the blood through these arteries and it becomes enlarged. Eventually overworking the right side of the heart may lead to right-sided **heart failure**, resulting in **death**.

Causes and symptoms

While the cause of primary pulmonary hypertension is uncertain, researchers think that in most people who develop the disease, the blood vessels are sensitive to certain factors that cause them to narrow. Diet suppressants, **cocaine**, and **pregnancy** are some of the factors that are thought to trigger constriction or narrowing of the pulmonary artery. In about 6–10% of cases, primary pulmonary hypertension is inherited.

Secondary pulmonary hypertension can be associated with breathing disorders such as **emphysema** and **bronchitis**, or diseases such as **scleroderma**, **systemic lupus erythematosus** (SLE) or **congenital heart disease** involving heart valves, and pulmonary thromboembolism.

Symptoms of pulmonary hypertension include **shortness of breath** with minimal exertion, general **fatigue**, **dizziness**, and **fainting**. Swelling of the ankles, bluish lips and skin, and chest **pain** are among other symptoms of the disease.

Diagnosis

Pulmonary hypertension is rarely detected during routine physical examinations and, therefore, often progresses to later stages before being diagnosed. In addition to listening to heart sounds with a stethoscope, physicians also use electrocardiogram, **pulmonary function tests**, perfusion lung scan, and/or right-heart **cardiac catheterization** to diagnose pulmonary hypertension.

Treatment

The aim of treatment for pulmonary hypertension is to treat the underlying cause, if it is known. For example, thromboendarterectomy is a surgical procedure performed to remove a blood clot on the lung that is causing the pulmonary hypertension. Lung transplants are another surgical treatment.

Some patients are helped by taking medicines that make the work of the heart easier. Anticoagulants, drugs that thin the blood, decrease the tendency of the blood to clot and allow blood to flow more freely. **Diuretics** decrease the amount of fluid in the body and reduce the amount of work the heart has to do. **Calcium channel blockers** relax the smooth muscle in the walls of the heart and blood vessels and improve the ability of the heart to pump blood.

One effective medical treatment that dilates blood vessels and seems to help prevent **blood clots** from forming is epoprostenol (prostacyclin). Prostacyclin is given intravenously to improve survival, exercise duration, and well-being. It is sometimes used as a bridge to help people who are waiting for a lung transplant. In other cases it is used for long-term treatment.

Some people require supplemental oxygen through nasal prongs or a mask if breathing becomes difficult.

Prognosis

Pulmonary hypertension is chronic and incurable with an unpredictable survival rate. Length of survival has been improving, with some patients able to live 15–20 years or longer with the disorder.

KEY TERMS

Hypertension—The medical term for abnormally high blood pressure.

Perfusion lung scan—A scan that shows the pattern of blood flow in the lungs.

Pulmonary—Having to do with the lungs.

Pulmonary function test—A test that measures how much air the lungs hold and the air flow in and out of the lungs.

Right-heart cardiac catheterization—A medical procedure during which a physician threads a catheter into the right side of the heart to measure the blood pressure in the right side of the heart and the pulmonary artery. The right heart's pumping ability can also be evaluated.

Prevention

Since the cause of primary pulmonary hypertension is still unknown, there is no way to prevent or cure this disease. A change in lifestyle may assist patients with daily activities. For example, relaxation exercises help to reduce **stress**. Good health habits such as a healthy diet, not **smoking**, and getting plenty of rest should be maintained.

Resources

OTHER

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ORGANIZATIONS

American Association for Respiratory Care, 9425 N. MacArthur Blvd, Suite 100, Irving, TX, 75063-4706, (972) 243-2272, (972) 484-2720, info@aarc.org, <http://ww.aarc.org>.

Pulmonary Hypertension Association, 801 Roeder Road, Ste. 1000, Silver Spring, MD, 20910, (301) 565-3004, (301) 565-3994, <http://www.phassoc.org.org>.

Lorraine Steefel, RN

Pulmonary incompetence see **Pulmonary valve insufficiency**

Pulmonary regurgitation see **Pulmonary valve insufficiency**

Pulmonary stenosis see **Pulmonary valve stenosis**

Pulmonary valve insufficiency

Definition

Pulmonary valve insufficiency is a disorder involving a defect of the valve located in the pulmonary artery.

Description

This disorder is also known as pulmonary valve regurgitation or pulmonary incompetence. The pulmonary valve is the structure in the pulmonary artery consisting of three flaps, which open and close during each heartbeat. The flaps keep blood from flowing back into the heart from the pulmonary artery—the artery that supplies blood to the lungs. With pulmonary valve insufficiency, the flaps may allow the blood to flow backward, resulting in a distinct murmur. The disorder may be congenital, but also often occurs in patients with severe **pulmonary hypertension**.

Causes and symptoms

There are generally few to no symptoms with pulmonary valve insufficiency. It may be initially noticed as a murmur in a routine exam of the heart and chest with a stethoscope. The most common causes of the disorder are severe pulmonary **hypertension**, or the presence of high pressure in the arteries and veins of the lungs. Pulmonary hypertension is usually caused by chronic lung disease, lung **blood clots**, and sometimes other diseases, such as **endocarditis**, an inflammation of the lining of the heart and valves. Previous surgery for **congenital heart disease** may also cause pulmonary valve insufficiency.

Diagnosis

The pitch and location of the murmur will help a physician determine if the cause is pulmonary valve insufficiency. An electrocardiogram (EKG) can detect flow changes. **Echocardiography** with color Doppler can usually detect regurgitation of blood in the area. This exam is done with ultrasound imaging. A **chest x ray** may show prominence of the pulmonary artery. In some cases, angiography, or x ray of the arteries and vessels with injection of a dye, may be ordered.

Treatment

On its own, pulmonary valve insufficiency is seldom severe enough to require treatment. **Antibiotics** are usually recommended before dental work to

KEY TERMS

Congenital—Used to describe a condition or defect present at birth.

Endocarditis—Inflammation of the lining of the heart and valves.

Prophylaxis—Preventive. Antibiotic prophylaxis is the use of antibiotics to prevent a possible infection.

Pulmonary—Refers to the lungs and the breathing system and function.

Pulmonary hypertension—High blood pressure in the veins and arteries of the lungs.

reduce the possibility of bacterial endocarditis. Management of the primary condition, such as medications to manage pulmonary hypertension, may help control pulmonary valve insufficiency.

Alternative treatment

Since there are few or no symptoms and the disorder is a structural defect, alternative treatment may have only limited usefulness. Proper diet, **exercise**, and **stress reduction** may help control hypertension. Coenzyme Q10 and hawthorn (*Crataegus laevigata*) are two important nutrients to nourish the heart. Antioxidant supplements (including **vitamins A, C, and E**, selenium, and zinc) can help keep the tissues of the whole body, including the heart, in optimal condition.

Prognosis

Patients with this disorder may never experience limitations from pulmonary valve insufficiency. The disorder may only show up if complicated by pulmonary hypertension. There is an increased incidence of bacterial endocarditis in patients with pulmonary valve insufficiency. Endocarditis can progress rapidly and be fatal.

Prevention

Pulmonary valve insufficiency resulting from chronic lung diseases can be prevented by behaviors and interventions to prevent those primary diseases. Bacterial endocarditis resulting from pulmonary valve insufficiency can usually be prevented with the use of antibiotic **prophylaxis** in preparation for dental procedures or other procedures which may introduce bacteria into the bloodstream.

Resources

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ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

Teresa Odle

KEY TERMS

Congenital—Present at birth.

Pulmonary—Relating to the opening leading from the right large chamber of the heart into the lung artery.

Stenosis—A narrowing or constriction, in this case of various heart valves. Stenosis reduces or cuts off the flow of blood.

Valve—Tissue between the heart's upper and lower chambers that controls blood flow.

Pulmonary valve stenosis

Definition

Pulmonary valve stenosis is a congenital heart defect in which blood flow from the heart to the pulmonary artery is blocked.

Description

Pulmonary valve stenosis is an obstruction in the pulmonary valve, located between the right ventricle and the pulmonary artery. Normally, the pulmonary valve opens to let blood flow from the right ventricle to the lungs. When the pulmonary valve is malformed, it forces the right ventricle to pump harder to overcome the obstruction. In its most severe form, pulmonary valve stenosis can be life-threatening.

Patients with pulmonary valve stenosis are at increased risk for getting valve infections and must take antibiotics to help prevent this before certain dental and surgical procedures. Pulmonary valve stenosis is also called pulmonary stenosis.

Causes and symptoms

Pulmonary valve stenosis is caused by a congenital malformation in which the pulmonary valve does not open properly. In most cases, scientists don't know why it occurs. In cases of mild or moderate stenosis, there are often no symptoms. With more severe obstruction, symptoms include a bluish skin tint and signs of **heart failure**.

Diagnosis

Diagnosis of pulmonary valve stenosis begins with the patient's medical history and a **physical examination**.

Tests to confirm the diagnosis include **chest x ray**, echocardiogram, electrocardiogram, and catheterization. An electrocardiograph shows the heart's activity. Electrodes covered with conducting jelly are placed on the patient. The electrodes send impulses that are traced on a recorder. **Echocardiography** uses sound waves to create an image of the heart's chambers and valves. The technician applies gel to a wand (transducer) and presses it against the patient's chest. The returning sound waves are converted into an image displayed on a monitor. Catheterization is an invasive procedure used to diagnose, and in some cases treat, heart problems. A thin tube, called a catheter, is inserted into a blood vessel and threaded up into the heart, enabling physicians to see and sometimes correct the problems.

Treatment

Patients with mild to moderate pulmonary valve stenosis, and few or no symptoms, do not require treatment. In more severe cases, the blocked valve will be opened surgically, either through **balloon valvuloplasty** or surgical valvulotomy. For initial treatment, balloon valvuloplasty is the procedure of choice. This is a catheterization procedure in which a special catheter containing a deflated balloon is inserted in a blood vessel and threaded up into the heart. The catheter is positioned in the narrowed heart valve and the balloon is inflated to stretch the valve open.

In some cases, surgical valvulotomy may be necessary. This is open heart surgery performed with a heart-lung machine. The valve is opened with an incision and in some cases, hypertrophied muscle in the right ventricle is removed. Rarely does the pulmonary valve need to be replaced.

Alternative treatment

Pulmonary valve stenosis can be life threatening and always requires a physician's care. In mild to

moderate cases of pulmonary valve stenosis, general lifestyle changes, including dietary modifications, **exercise**, and **stress reduction**, can contribute to maintaining optimal wellness.

Prognosis

Patients with the most severe form of pulmonary valve stenosis may die in infancy. The prognosis for children with more severe stenosis who undergo balloon valvuloplasty or surgical valvulotomy is favorable. Patients with mild to moderate pulmonary stenosis can lead a normal life, but they require regular medical care.

Prevention

Pulmonary valve stenosis cannot be prevented.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

CHASER (Congenital Heart Anomalies Support, Education, and Resources), 2112 North Wilkins Road, Swanton, OH, 43558, (419) 825-5575, (419) 825-2880, CHASER@compuserve.com, http://www.csun.edu.

Congenital Heart Information Network (C.H.I.N.), 101 N. Washington Ave., Suite 1A, Margate City, NJ, 08402-1195, (609) 882-1572, (609) 822-1574, mb@tchin.org, http://tchin.org/.

Texas Heart Institute. Heart Information Service, MC 3-116, PO Box 20345, Houston, TX, 77225, (832) 355-4011, (800) 292-2221, http://www.texasheart.org.

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Punctures see **Wounds**

Purple coneflower see **Echinacea**

Purpura hemorrhagica see **Idiopathic thrombocytopenic purpura**

Pustule see **Skin lesions**

Pyelography see **Intravenous urography**

Pyelonephritis

Definition

Pyelonephritis is an inflammation of the kidney and upper urinary tract that usually results from non-contagious bacterial infection of the bladder (**cystitis**).

Description

Acute pyelonephritis is most common in adult females but can affect people of either sex and any age. Its onset is usually sudden, with symptoms that often are mistaken as the results of straining the lower back. Pyelonephritis often is complicated by systemic infection. Left untreated or unresolved, it can progress to a chronic condition that lasts for months or years, leading to scarring and possible loss of kidney function.

Causes and symptoms

The most common cause of pyelonephritis is the backward flow (reflux) of infected urine from the bladder to the upper urinary tract. Bacterial infections also may be carried to one or both kidneys through the bloodstream or lymph glands from infection that began in the bladder. Kidney infection sometimes results from urine that becomes stagnant due to obstruction of free urinary flow. A blockage or abnormality of the urinary system, such as those caused by stones, tumors, congenital deformities, or loss of bladder function from nerve disease, increases a person's risk of pyelonephritis. Other risk factors include **diabetes mellitus**, **pregnancy**, chronic bladder infections, a history of analgesic **abuse**, **paralysis** from **spinal cord injury**, or tumors. Catheters, tubes, or surgical procedures may also trigger a kidney infection.

The bacteria most likely to cause pyelonephritis are those that normally occur in the feces. *Escherichia coli* causes about 85% of acute bladder and kidney infections in patients with no obstruction or history of surgical procedures. *Klebsiella*, *Enterobacter*, *Proteus*, or *Pseudomonas* are other common causes of infection. Once these organisms enter the urinary tract, they cling to the tissues that line the tract and multiply in them.

Symptoms of acute pyelonephritis typically include **fever** and chills, burning or frequent urination, aching **pain** on one or both sides of the lower back or abdomen, cloudy or bloody urine, and **fatigue**. The patient also may have **nausea**, **vomiting**, and **diarrhea**. The flank pain may be extreme. The symptoms of chronic pyelonephritis include weakness, loss of appetite, **hypertension**, anemia, and protein and blood in the urine.

Diagnosis

The diagnosis of pyelonephritis is based on the patient's history, a **physical examination**, and the

results of laboratory and imaging tests. During the physical examination, the doctor will touch (palpate) the patient's abdomen carefully in order to rule out **appendicitis** or other causes of severe abdominal pain.

Laboratory tests

In addition to collecting urine samples for **urinalysis** and **urine culture** and sensitivity tests, the doctor will take a sample of the patient's blood for a blood cell count. If the patient has pyelonephritis, the urine tests will show the presence of white blood cells, and bacteria in the urine. Bacterial counts of 100,000 organisms or higher per milliliter of urine point to a **urinary tract infection**. The presence of antibody-coated bacteria (ACB) in the urine sample distinguishes kidney infection from bladder infection, because bacteria in the kidney trigger an antibody response that coats the bacteria. The blood cell count usually indicates a sharp increase in the number of white blood cells.

Imaging studies

The doctor may order ultrasound imaging of the kidney area if he or she suspects that there is an obstruction blocking the flow of urine. X rays may demonstrate scarring of the kidneys and ureters resulting from long-standing infection.

Treatment

Treatment of acute pyelonephritis may require hospitalization if the patient is severely ill or has complications. Therapy most often involves a 2- to 3-week course of **antibiotics**, with the first few days of treatment given intravenously. The choice of antibiotic is based on laboratory sensitivity studies. The antibiotics used most often include ciprofloxacin (Cipro), ampicillin (Omnipen), or trimethoprim-sulfamethoxazole (Bactrim, Septra). Several advances in antibiotic therapy have been made in recent years. In 2003, the U.S. Food and Drug Administration (FDA) approved Cipro extended release tablets (Cipro XR) that could be taken once daily for acute uncomplicated pyelonephritis. A study in Europe also showed that a shorter course than that normally used in the United States could eradicate the bacteria that cause the disease. The primary objective of antimicrobial therapy is the permanent eradication of bacteria from the urinary tract. The early symptoms of pyelonephritis usually disappear within 48 to 72 hours of the start of antibacterial treatment. Repeat urine cultures are done in order to evaluate the effectiveness of the medication.

KEY TERMS

Bacteremia—The presence of bacteria in the bloodstream.

Cystitis—Inflammation of the bladder, usually caused by bacterial infection.

Reflux—The backward flow of a fluid in the body. Pyelonephritis is often associated with the reflux of urine from the bladder to the upper urinary tract.

Chronic pyelonephritis may require high doses of antibiotics for as long as 6 months to clear the infection. Other medications may be given to control fever, nausea, and pain. Patients are encouraged to drink extra fluid to prevent **dehydration** and increase urine output. Surgery sometimes is necessary if the patient has complications caused by **kidney stones** or other obstructions, or to eradicate infection. Urine cultures are repeated as part of the follow-up of patients with chronic pyelonephritis. These repeat tests are necessary to evaluate the possibility that the patient's urinary tract is infected with a second organism as well as to assess the patient's response to the antibiotic. Some persons are highly susceptible to reinfection, and a second antibiotic may be necessary to treat the organism.

Prognosis

The prognosis for most patients with acute pyelonephritis is quite good if the infection is caught early and treated promptly. The patient is considered cured if the urine remains sterile for a year. Untreated or recurrent kidney infection can lead to bacterial invasion of the bloodstream (**bacteremia**), hypertension, chronic pyelonephritis with scarring of the kidneys, and permanent kidney damage.

Prevention

Persons with a history of urinary tract infections should urinate frequently, and drink plenty of fluids at the first sign of infection. Women should void after intercourse which may help flush bacteria from the bladder. Girls should be taught to wipe their genital area from front to back after urinating to avoid getting fecal matter into the opening of the urinary tract.

Resources

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American Urological Association Foundation, 1000 Corporate Blvd., Linthicum, MD, 21090, (410) 689-3700, (410) 689-3800, (866) 746-4282, auafoundation@auafoundation.org, <http://www.urologyhealth.org/>.

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Pyloric stenosis

Definition

Pyloric stenosis refers to a narrowing of the passage between the stomach and the small intestine. The condition, which affects infants during the first several weeks of life, can be corrected effectively with surgery.

Demographics

The condition affects one in 4,000 infants. Most are diagnosed between three and five weeks old, though some babies may show symptoms during the first or second week of life. Infants with a family history of pyloric stenosis are more at risk for the condition, which tends to occur less often in females, blacks, and Asians. Pyloric stenosis is also referred to as hypertrophic pyloric stenosis.

Description

Frequent **vomiting** may be an indication of pyloric stenosis. The pylorus is the passage between the stomach and the small intestine. During the digestive process food passes through the pylorus, which is located near the bottom of the stomach, on its way to the intestines. In pyloric stenosis, the muscular wall of the passage becomes abnormally thickened. This causes the pylorus to become too narrow, which prevents food from emptying out of the stomach in a normal fashion. The

partially digested contents of the stomach are forced upwards into the mouth. As a result, a baby with pyloric stenosis often vomits after feedings.

Causes and symptoms

The cause of pyloric stenosis is not known. The main symptom is **vomiting** after feedings. These episodes of vomiting usually get worse over time, happening more often and becoming more forceful (forceful vomiting is often called "projectile" vomiting). Other symptoms include increased appetite, weight loss, infrequent bowel movements, belching, and **diarrhea**. Due to **dehydration**, the infant may also have fewer wet diapers.

Diagnosis

The clinician will examine the baby and talk with the parents about their infant's symptoms. If a child has the condition, the doctor should be able to feel a hard mass (about an inch [two centimeters] wide and olive shaped) in the area above the belly button. If the doctor cannot detect the mass, ultrasonography will be done to confirm the diagnosis. A blood test may also be performed to see if the infant is dehydrated, in which case intravenous fluids can be used to correct the problem.

Treatment

Pyloric stenosis can be cured with a surgical procedure called a pyloromyotomy. In this operation, the surgeon makes an incision in the baby's abdomen. Then a small cut is made in the thickened muscle of the pylorus and it is spread apart. In this manner, the passage can be widened without removing any tissue. (The procedure may be performed with the aid of a laparoscope.) After surgery, the pylorus will heal itself. The thickening gradually goes away and the passage resumes a normal shape. The whole procedure (including anesthesia) takes about an hour.

Most babies go home one or two days after surgery. Any mild discomfort can be controlled with **acetaminophen** (Tylenol). The infant may still vomit occasionally after surgery, but this is not usually a cause for alarm. However, if vomiting occurs three or more times a day, or for several consecutive days, the baby's pediatrician should be notified.

Alternative treatment

None known.

Prognosis

Surgery often provides a complete cure. Most infants do not experience complications or long-term effects.

KEY TERMS

Laparoscope—A thin, camera-fitted tube that can be inserted into the abdomen in order to view internal organs.

Stenosis—The narrowing of a passage (such as the pylorus).

Ultrasonography—A non-invasive imaging procedure that uses high-frequency sound waves.

Prevention

It is not known how to prevent pyloric stenosis.

Resources

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ORGANIZATIONS

- American Academy of Family Physicians (AAFP), 11400 Tomahawk Creek Pkwy., Leawood, KS, 66211–2672, (913) 906–6000, <http://www.aafp.org>.
 American Academy of Pediatrics (AAP), 141 Northwest Point Blvd., Elk Grove Village, IL, 60007–1098, (847) 434–4000, <http://www.aap.org>.

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Pyloroplasty

Definition

Pyloroplasty is an elective surgical procedure in which the lower portion of the stomach, the pylorus, is cut and resutured, to relax the muscle and widen the opening into the intestine. Pyloroplasty is a treatment for high-risk patients for gastric or peptic ulcer

disease. A peptic ulcer is a well-defined sore on the stomach where the lining of the stomach or duodenum has been eaten away by stomach acid and digestive juices.

Purpose

The end of the pylorus is surrounded by a strong band of muscle (pyloric sphincter), through which stomach contents are emptied into the duodenum (the first part of the small intestine). Pyloroplasty widens this opening into the duodenum.

A pyloroplasty is performed to treat complications of gastric ulcer disease, or when conservative treatment is unsatisfactory. The longitudinal cut made in the pylorus is closed transversely, permitting the muscle to relax. By establishing an enlarged outlet from the stomach into the intestine, the stomach empties more quickly. A pyloroplasty is often done in conjunction with a **vagotomy**, a procedure in which the nerves that stimulate stomach acid production and gastric motility (movement) are cut. As these nerves are cut, gastric emptying may be delayed and the pyloroplasty compensates for that effect.

Preparation

As with any surgical procedure, the patient will be required to sign a consent form after the procedure is explained thoroughly. Blood and urine studies, along with various x rays, may be ordered as the doctor deems necessary. Food and fluids will be prohibited after midnight before the procedure. Cleansing enemas may be ordered to empty the intestine. If **nausea** or **vomiting** are present, a suction tube to empty the stomach may be used.

Aftercare

Post-operative care for the patient who has had a pyloroplasty, as for those who have had any major surgery, involves monitoring of blood pressure, pulse, respiration, and temperature. Breathing tends to be shallow because of the effect of anesthesia and the patient's reluctance to breathe deeply and experience **pain** that is caused by the abdominal incision. The patient is shown how to support the operative site while breathing deeply and coughing, and given pain medication as necessary. Fluid intake and output is measured, and the operative site is observed for color and wound drainage. Fluids are given intravenously for 24–48 hours, until the patient's diet is gradually advanced as bowel activity resumes. The patient is generally allowed to walk approximately eight hours after surgery and the average hospital stay, dependent

KEY TERMS

Gastric (or peptic) ulcer—An ulcer (sore) of the stomach, duodenum or other part of the gastrointestinal system. Though the causes are not fully understood, they include excessive secretion of gastric acid, stress, heredity, and the use of certain drugs, especially acetylsalicylic acid and nonsteroidal anti-inflammatory drugs.

Pylorus—The valve which releases food from the stomach into the intestines.

Vagotomy—Cutting of the vagus nerve. If the vagus nerves are cut as they enter the stomach (truncal vagotomy), gastric secretions are decreased, as is intestinal motility (movement) and stomach emptying. In a selective vagotomy, only those branches of the vagus nerve are cut that stimulate the secretory cells.

upon overall recovery status, ranges from six to eight days.

Risks

Potential complications of this abdominal surgery include:

- excessive bleeding
- surgical wound infection
- incisional hernia
- recurrence of gastric ulcer
- chronic diarrhea
- malnutrition

Normal results

Complete healing is expected without complications. Four to six weeks should be allowed for recovery from the surgery.

Abnormal results

The doctor should be made aware of any of the following problems after surgery:

- increased pain, swelling, redness, drainage, or bleeding in the surgical area
- headache, muscle aches, dizziness, or fever
- increased abdominal pain or swelling, constipation, nausea or vomiting, rectal bleeding, or black, tarry stools

Resources

OTHER

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ORGANIZATIONS

American Gastroenterological Association (AGA), 4930 Del Ray Avenue, Bethesda, MD 20814, (301) 654-2055, <http://www.gastro.org/>.

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Pylorus repair see **Pyloroplasty**

Pyorrhea see **Periodontal disease**

Pyrazinamide see **Antituberculosis drugs**

Pyridoxine deficiency see **Vitamin B₆ deficiency**

Pyrimethamine see **Antimalarial drugs**

Pyruvate kinase deficiency

Definition

Pyruvate kinase deficiency (PKD) is part of a group of disorders called hereditary nonspherocytic hemolytic **anemias**. Hereditary nonspherocytic anemias are rare genetic conditions that affect red blood cells. PKD is caused by a deficiency in the enzyme, pyruvate kinase.

Demographics

Although PKD is the second most common of the hereditary nonspherocytic anemias, it is still rare with the incidence estimated to be 51 cases per million in the Caucasian population.

In general, PKD does not appear to affect one gender more than another or be more common in certain regions. However, there are studies of an Amish group in Pennsylvania where a severe form of PKD is more common. Three mutations found in the PKLR gene have been linked to individuals of specific descents. Caucasians of northern and central European descent are more likely to have the 1529A mutation, individuals of southern European descent usually have the 1456T mutation, and individuals of Asian descent are more likely to have the 1468T mutation.

Description

In PKD, there is a functional abnormality with the enzyme pyruvate kinase. Pyruvate kinase acts as a catalyst in the glycolysis pathway and is considered an essential component in this pathway. Glycolysis is the method by which cells produce energy. A problem with any of the key components in glycolysis can alter the amount of energy produced. In red blood cells, glycolysis is the only method available to produce energy. Without the proper amount of energy, red blood cells do not function normally. Since pyruvate kinase is one of the key components in glycolysis, when there is a problem with this enzyme in red blood cells, there is a problem with the production of energy, causing red blood cells not to function properly.

There are four different forms of the pyruvate kinase enzyme in the human body. These forms, called isozymes, all perform the same function but each isozyme of pyruvate kinase is structurally different and works in different tissues and organs. The four isozymes of pyruvate kinase are labeled M1, M2, L, and R. The isozyme M1 is found in the skeletal muscle and the brain, isozyme M2 can be found in most fetal and adult tissues, isozyme L works in the liver, and isozyme R works in red blood cells. In PKD, only the pyruvate kinase isozyme found in red blood cells, called PKR, is abnormal. Therefore, PKD affects red blood cells only and does not directly affect energy production in other organs and tissues of the body.

Causes and symptoms

There are two PK genes and each gene produces two of the four isozymes of pyruvate kinase. The M1 and M2 isozymes are produced by the pyruvate kinase gene called PKM2 and pyruvate kinase isozymes, L and R, are products of the pyruvate kinase gene, PKLR. The PKLR gene is located on chromosome 1, on the q arm (the top half of the chromosome), in region 21 (written as 1q21). There have been over 125 different mutations described in the PKLR gene that have been detected in individuals with PKD.

PKD is inherited mainly in an autosomal recessive manner. There have been a few families where it appeared that PKD was inherited in either an autosomal dominant manner or where the carriers of PKD exhibited mild problems with their red blood cells. As with all autosomal recessive conditions, affected individuals have a mutation in both pair of genes. Most individuals with PKD are compound heterozygotes, meaning that each PKLR gene in a pair contains a different mutation. There are individuals who have

the same mutation on each PKLR gene, but these individuals tend to be children of parents who are related to each other.

There are three mutations in the PKLR gene called, 1529A, 1456T, and 1468T, that are seen more frequently in individuals with PKD than the other mutations. The mutation 1529A is most frequently seen in Caucasians of northern and central European descent and is the most common mutation seen in PKD. The mutation 1456T is more common in individuals of southern European descent and the mutation 1468T is more common in individuals of Asian descent.

In general, the more severe the PKD, the earlier in life symptoms tend to be detected. Individuals with the more severe form of PKD often show symptoms soon after birth, but most individuals with PKD begin to exhibit symptoms during infancy or childhood. In individuals with the milder form of PKD, the condition is sometimes not diagnosed until late adulthood, after an acute illness, or during a **pregnancy** evaluation.

For most of the mutations seen in the PKLR gene, no correlation between the specific mutation and the severity of the disorder has been observed. However, for two of the mutations, there has been speculation on their effect on the severity of PKD. When the mutation 1456T has been seen in the homozygous state (when both PKLR genes contain the same mutation), those rare individuals experienced very mild symptoms of PKD. Also, there have been individuals who were homozygous for the 1529A mutation. These individuals had a very severe form of PKD. Therefore, it is thought that the 1456T mutation is associated with a milder form of the disease and the 1529A mutation is associated with a more severe form of the disease. It is not known how these mutations affect the severity of PKD when paired with different mutations.

Symptoms of PKD are similar to those symptoms seen in individuals who have long-term **hemolytic anemia**. The more common symptoms include variable degrees of **jaundice** (a yellowish pigment of the skin and eyes), slightly to moderately enlarged spleen (splenomegaly), and increased incidence of **gallstones**. Other physical effects of PKD can include smaller head size and a prominent and rounded forehead (called frontal bossing). If children with PKD have their spleen removed, growth tends to improve. Even within the same family, individuals can have different symptoms and severity of PKD.

KEY TERMS

Anemia—A condition in which the amount of red blood cells is less than normal.

Catalyst—A substance that changes the rate of a reaction, but is not changed by the process.

Compound heterozygotes—Individuals who have one gene in a pair with one mutation and the other gene in the pair has a different mutation.

Enzyme—A protein produced by cells that acts as a catalyst in a biological reaction.

Glycolysis—The pathway in which a cell breaks down glucose into energy.

Hemolytic anemia—Anemia that results from red blood cells being destroyed sooner than normal.

Heterozygote—An individual who has one gene in a pair that has a mutation while the other gene in the pair is unaffected.

Homozygote—An individual who has both genes in a pair with the same mutation.

Homozygous—A condition in which both genes in a pair have the same mutation.

Isozyme—One of a group of enzymes that perform the same function, but are different from one another in their structure or the way in which they move.

Mutation—A change in a gene that causes it to alter its function.

Nonspherocytic—Not sphere shaped. Often in inherited hemolytic anemias, red blood cells are sphere shaped. In nonspherocytic hemolytic anemias, red blood cells are not sphere shaped.

In individuals with PKD, red blood cells are taken out of circulation earlier than normal (shorten lifespan). Because of this change, individuals with PKD will have hemolytic anemia. Additionally, the anemia or other symptoms of PKD may worsen during a sudden illness or pregnancy.

Diagnosis

A diagnosis of PKD can be made by measuring the amount of pyruvate kinase in red blood cells. Individuals with PKD tend to have 5–25% of the normal amount of pyruvate kinase. Carriers of PKD also can have less pyruvate kinase in their red blood cells, approximately 40–60% of the normal value. However, there is an overlap between the normal range of pyruvate kinase and the ranges seen with carriers of PKD.

Therefore, measuring the amount of pyruvate kinase in red blood cells is not a good method of detecting carriers of PKD. If the mutations causing PKD in a family are known, it may be possible to perform mutation analysis to determine carrier status of an individual and to help diagnose individuals with PKD.

Treatment

In the severest cases, individuals with PKD will require multiple blood transfusions. In some of those cases, the spleen may be removed (**splenectomy**). Red blood cells are normally removed from circulation by the spleen. By removing the spleen of an individual (usually a child), red blood cells are allowed to stay in circulation longer than normal, thereby reducing the severity of the anemia. After a splenectomy, or once an individual with PKD is older, the number of transfusions tends to decrease.

Prognosis

The prognosis for PKD is extremely variable. Early intervention and treatment of symptoms frequently improves the individual's health. Without treatment, individuals may experience severe complications that may become lethal. Individuals with a mild form of PKD may appear to have no symptoms at all.

Resources

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ORGANIZATIONS

National Heart, Lung, and Blood Institute (NHLBI), PO Box 30105, Bethesda, MD, 20824–0105, (301) 592–8573. TTY: (240) 629–3255, <http://www.nhlbi.nih.gov>.

National Institutes of Health (NIH), 9000 Rockville Pike, Bethesda, MD, 20892, (301) 496-4000, <http://www.nih.gov/index.html>.

National Library of Medicine (NLM), 8600 Rockville Pike, Bethesda, MD, 20894, <http://www.nlm.nih.gov/>.

National Organization for Rare Diseases (NORD), PO Box 8923, Fairfield, CT, 06812, (213) 745–6518, <http://www.rarediseases.org>.

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Q

Q fever

Definition

Q fever is an illness caused by a type of bacteria, *Coxiella burnetii*, resulting in a fever and rash.

Description

C. burnetii lives in many different kinds of animals, including cattle, sheep, goats, ticks, cats, rabbits, birds, and dogs. In sheep and cattle the bacteria tends to accumulate in large numbers in the female's uterus (the organ where lambs and calves develop) and udder. Other animals have similar patterns of bacterial accumulation within the females. As a result, *C. burnetii* can cause infection through contaminated milk, or when humans come into contact with the fluids or tissues produced when a cow or sheep gives birth. The bacteria can also survive in dry dust for months; therefore, if the female's fluids contaminate the ground, humans may become infected when they come in contact with the contaminated dust.

Persons most at risk for Q fever include anybody who works with cattle or sheep, or products produced from them. These include farm workers, slaughterhouse workers, workers in meat-packing plants, veterinarians, and wool workers. Since September 2001, however, Q fever has become an additional concern because of its potential as an agent of bioterrorism.

Q fever has been found all over the world, except in some areas of Scandinavia, Antarctica, and New Zealand.

Causes and symptoms

C. burnetii causes infection when a human breathes in tiny droplets, or drinks milk, containing the bacteria. After 3 to 30 days, symptoms of the illness appear.

The usual symptoms of Q fever include fever, chills, heavy sweating, **headache, nausea and vomiting, diarrhea,**

fatigue, and **cough**. Also, a number of other problems may present themselves, including inflammation of the liver (hepatitis); inflammation of the sac containing the heart (**pericarditis**); inflammation of the heart muscle itself (**myocarditis**); inflammation of the coverings of the brain and spinal cord, or of the brain itself (meningoencephalitis); and **pneumonia**.

Chronic Q fever occurs most frequently in patients with other medical problems, including diseased heart valves, weakened immune systems, or **kidney disease**. Such patients usually have about a year's worth of vague symptoms, including a low fever, enlargement of the spleen and/or liver, and fatigue. Testing almost always reveals that these patients have inflammation of the lining of the heart (**endocarditis**).

Diagnosis

Q fever is diagnosed by demonstrating that the patient's immune system is making increasing numbers of antibodies (special immune cells) against markers (antigens) that are found on *C. burnetii*.

Treatment

Doxycycline and quinolone **antibiotics** are effective for treatment of Q fever. Treatment usually lasts for two weeks. Rifampin and doxycycline together are given for chronic Q fever. Chronic Q fever requires treatment for at least three years.

Minocycline has been found to be useful in treating post-Q fever fatigue. The dosage is 100 mg per day for three months.

Prognosis

Death is rare from Q fever. Most people recover completely, although some patients with endocarditis will require surgery to replace their damaged heart valves.

KEY TERMS

Antibodies—Specialized cells of the immune system that can recognize organisms that invade the body (such as bacteria, viruses, and fungi). The antibodies are then able to set off a complex chain of events designed to kill these foreign invaders.

Antigens—Markers on the outside of bacteria or viruses, which can be recognized by antibodies.

Bioterrorism—The use of disease microorganisms to intimidate or terrorize a civilian population.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body, which work together to prevent foreign invaders (bacteria, viruses, fungi, etc.) from taking hold and growing.

Inflammation—The body's response to tissue damage. Includes increased heat, swelling, redness, and pain in the affected part.

Prevention

Q fever can be prevented by the appropriate handling of potentially infective substances. For example, milk should always be pasteurized, and people who work with animals giving birth should carefully dispose of the tissues and fluids associated with birth. Industries which process animal materials (meat, wool) should take care to prevent the contamination of dust within the plant.

Vaccines are available for workers at risk for Q fever.

Resources

BOOKS

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ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

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Qigong

Definition

Qigong (pronounced “chee-gung,” also spelled *chi kung*) is translated from the Chinese to mean “energy cultivation” or “working with the life energy.” Qigong is an ancient Chinese system of postures, exercises, breathing techniques, and meditations. Its techniques are designed to improve and enhance the body’s *qi*. According to traditional Chinese philosophy, *qi* is the fundamental life energy responsible for health and vitality.

Purpose

Qigong may be used as a daily routine to increase overall health and well-being, as well as for disease prevention and longevity. It can be used to increase energy and reduce **stress**. In China, qigong is used in conjunction with other medical therapies for many chronic conditions, including **asthma**, **allergies**, **AIDS**, **cancer**, headaches, **hypertension**, depression, mental illness, strokes, heart disease, and **obesity**.

Qigong is presently being used in Hong Kong to relieve depression and improve the overall psychological and social well-being of elderly people with chronic physical illnesses.

Description

Origins

Qigong originated before recorded history. Scholars estimate qigong to be as old as 5,000–7,000 years. Tracing the exact historical development of qigong is difficult because it was passed down in secrecy among monks and teachers for many generations. Qigong survived through many years before paper was invented, and it also survived the Cultural Revolutions in China of the 1960s and 1970s, which banned many traditional practices.

Qigong has influenced and been influenced by many of the major strands of Chinese philosophy. The Taoist philosophy states that the universe operates within laws of balance and harmony, and that people must live within the rhythms of nature—ideas that pervade qigong. When Buddhism was brought from India to China around the seventh century A.D., **yoga** techniques and concepts of mental and spiritual awareness were introduced to qigong masters. The Confucian school was concerned with how people should live their daily lives, a concern of qigong as well. The martial arts were highly influenced by qigong and many of them, such as t'ai chi and kung fu, developed directly from it. **Traditional Chinese medicine** also shares many of the central concepts of qigong, such as the patterns of energy flow in the body. **Acupuncture** and **acupressure** use the same points on the body that qigong seeks to stimulate. In China, qigong masters have been renowned physicians and healers. Qigong is often prescribed by Chinese physicians as part of the treatment.

Due to the political isolation of China, many Chinese concepts have been shrouded from the Western world. Acupuncture was “discovered” by American doctors only in the 1970s, although it had been in use for thousands of years. With an increased exchange of information, more Americans have gained access to the once-secret teachings of qigong. In 1988, the First World Conference for Academic Exchange of Medical Qigong was held in Beijing, China, where many studies were presented to attendees from around the world. In 1990, Berkeley, California, hosted the First International Congress of Qigong. In the past decade, more Americans have begun to discover the beneficial effects of qigong, which motivate an estimated 60 million Chinese to practice it every day.

Basic concepts

In Chinese thought, qi, or chi, is the fundamental life energy of the universe. It is invisible but present in air, water, food and sunlight. In the body, qi is the unseen vital force that sustains life. We are all born with inherited amounts of qi, and we also get acquired qi from the food we eat and the air we breathe. In qigong, the breath is believed to account for the largest quantity of acquired qi, because the body uses air more than any other substance. The balance of our physical, mental, and emotional levels also affect qi levels in the body.

Qi travels through the body along channels called meridians. There are 12 main meridians, corresponding to the 12 principal organs as defined by the traditional Chinese system: the lungs, large intestines, stomach, spleen, heart, small intestines, urinary bladder, kidneys,

liver, gallbladder, pericardium, and the “triple warmer,” which represents the entire torso region. Each organ has qi associated with it, and each organ interacts with particular emotions on the mental level. Qigong techniques are designed to improve the balance and flow of energy throughout the meridians, and to increase the overall quantity and volume of qi. In qigong philosophy, mind and body are not separated as they often are in Western medicine. In qigong, the mind is present in all parts of the body, and the mind can be used to move qi throughout the body.

Yin and yang are also important concepts in qigong. The universe and the body can be described by these two separate but complementary principles, which are always interacting, opposing, and influencing each other. One goal of qigong is to balance yin and yang within the body. Strong movements or techniques are balanced by soft ones, leftward movements by rightward, internal techniques by external ones, and so on.

Practicing qigong

There are thousands of qigong exercises. The specific ones used may vary depending on the teacher, school, and objective of the practitioner. Qigong is used for physical fitness, as a martial art, and most frequently for health and healing. Internal qigong is performed by those wishing to increase their own energy and health. Some qigong masters are renowned for being able to perform external qigong, by which the energy from one person is passed on to another for healing. This transfer may sound suspect to Western logic, but in the world of qigong there are some amazing accounts of healing and extraordinary capabilities demonstrated by qigong masters. Qigong masters generally have deep knowledge of the concepts of Chinese medicine and healing. In China, there are hospitals that use medical qigong to heal patients, along with herbs, acupuncture, and other techniques. In these hospitals, qigong healers use external qigong and also design specific internal qigong exercises for patients’ problems.

There are basic components of internal qigong sessions. All sessions require warm-up and concluding exercises. Qigong consists of postures, movements, breathing techniques, and mental exercises. Postures may involve standing, sitting, or lying down. Movements include stretches, slow motions, quick thrusts, jumping, and bending. Postures and movements are designed to strengthen, stretch, and tone the body to improve the flow of energy. One sequence of postures and movements is known as the “Eight Figures for Every Day.” This sequence is designed to quickly and

effectively work the entire body, and is commonly performed daily by millions in China.

Breathing techniques include deep abdominal breathing, chest breathing, relaxed breathing, and holding breaths. One breathing technique is called the “Six Healing Sounds.” This technique uses particular breathing sounds for each of six major organs. These sounds are believed to stimulate and heal the organs.

Meditations and mind exercises are used to enhance the mind and move qi throughout the body. These exercises are often visualizations that focus on different body parts, words, ideas, objects, or energy flowing along the meridians. One mental exercise is called the “Inner Smile,” during which the practitioner visualizes joyful, healing energy being sent sequentially to each organ in the body. Another mental exercise is called the “Microscopic Orbit Meditation,” in which the practitioner intently meditates on increasing and connecting the flow of qi throughout major channels.

Discipline is an important dimension of qigong. Exercises are meant to be performed every morning and evening. Sessions can take from 15 minutes to hours. Beginners are recommended to practice between 15–30 minutes twice a day. Beginners may take classes once or twice per week, with practice outside of class. Classes generally cost between \$10–\$20 per session.

Preparations

Qigong should be practiced in a clean, pleasant environment, preferably outdoors in fresh air. Loose and comfortable clothing is recommended. Jewelry should be removed. Practitioners can prepare for success at qigong by practicing at regular hours each day to promote discipline. Qigong teachers also recommend that students prepare by adopting lifestyles that promote balance, moderation, proper rest, and healthy diets, all of which are facets of qigong practice.

Precautions

Beginners should learn from an experienced teacher, as performing qigong exercises in the wrong manner may cause harm. Practitioners should not perform qigong on either full or completely empty stomachs. Qigong should not be performed during extreme weather, which may have negative effects on the body’s energy systems. Menstruating and pregnant women should perform only certain exercises.

Side effects

Side effects may occur during or after qigong exercises for beginners, or for those performing exercises incorrectly. Side effects may include **dizziness**, **dry mouth**, **fatigue**, headaches, **insomnia**, rapid heartbeat, **shortness of breath**, heaviness or **numbness** in areas of the body, emotional instability, **anxiety**, or decreased concentration. Side effects generally clear up with rest and instruction from a knowledgeable teacher.

Research and general acceptance

Western medicine generally does not endorse any of the traditional Chinese healing systems that utilize the concept of energy flow in the body, largely because this energy has yet to be isolated and measured scientifically. New research is being conducted using sophisticated equipment that may verify the existence of energy channels as defined by the Chinese system. Despite the lack of scientific validation, the results of energy techniques including qigong and acupuncture have gained widespread interest and respect. One California group of qigong practitioners now conducts twice-yearly retreats to improve their skills and energy level. Furthermore, qigong masters have demonstrated to Western observers astounding control over many physical functions, and some have even shown the ability to increase electrical voltage measured on their skin’s surface. Most of the research and documentation of qigong’s effectiveness for medical conditions has been conducted in China, and is slowly becoming more available to English readers. Papers from the World Conferences for Academic Exchange of Medical Qigong are available in English, and address many medical studies and uses of qigong. A video is now available that presents the basic concepts of medical qigong as well as specific **exercise** prescriptions for the treatment of **breast cancer**. The exercise prescriptions consist of movements, postures, visualizations, and positive affirmations.

In terms of mainstream research in the United States, the first ongoing long-term study of qigong began in 1999 at the Center for Alternative and Complementary Medicine Research in Heart Disease at the University of Michigan; it focuses on the speed of healing of graft **wounds** in patients undergoing coronary bypass surgery. The National Center for Complementary and Alternative Medicine (NCCAM) has been funding studies of qigong since 2000. The first such study was conducted by a researcher in Arizona with patients using heart devices (**pacemakers**, etc.).

The breathing techniques of qigong are being studied intensively by Western physicians as a form of

KEY TERMS

- Martial arts**—Group of diverse activities originating from the ancient fighting techniques of the Orient.
- Meridians**—Channels or conduits through which Qi travels in the body.
- Qi**—Basic life energy, according to traditional Chinese medicine.
- Yin/Yang**—Universal characteristics used to describe aspects of the natural world.

therapy for anxiety-related problems and for disorders involving the vocal cords. Qigong is also being used in the **rehabilitation** of patients with severe asthma or **chronic obstructive pulmonary disease** (COPD).

Training and certification

In China, qigong has been subject to much government regulation, from banning to increased requirements for teachers. In the United States at this time, qigong has not been regulated. Different schools may provide teacher training, but there are no generally accepted training standards. Qigong teachings may vary, depending on the founder of the school, who is often an acknowledged Chinese master. Qigong organizations can provide further information.

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Qi: The Journal of Traditional Eastern Health and Fitness. PO Box 221343, Chantilly, VA 22022, (202) 378-3859.

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National Center for Complementary and Alternative Medicine (NCCAM), P.O. Box 7923, Gaithersburg, MD, 20898, (866) 464-3616, (888) 644-6226, info@nccam.nih.gov, <http://nccam.nih.gov/>.

National Qigong Association, P.O. Box 270065, St. Paul, MN, 55127, (888) 359-9526, (888) 815-1893, <http://nqa.org>.

The Qigong Institute, 617 Hawthorne Avenue, Los Altos, CA, 94024, <http://www.qigonginstitute.or>.

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Quadriplegia see **Paralysis**

Quarantine see **Isolation**

Quinidine see **Antiarrhythmic drugs**

Quinine see **Antimalarial drugs**

R

Rabbit fever see **Tularemia**

Rabies

Definition

Rabies is an acute viral disease of the central nervous system that affects humans and other mammals but is most common in carnivores (flesh-eaters). It is sometimes referred to as a **zoonosis**, or disease of animals that can be communicated to humans. Rabies is almost exclusively transmitted through saliva from the bite of an infected animal. Another name for the disease is *hydrophobia*, which literally means “fear of water,” a symptom shared by half of all people infected with rabies. Other symptoms include **fever**, depression, confusion, painful **muscle spasms**, sensitivity to touch, loud noise, and light, extreme thirst, painful swallowing, excessive salivation, and loss of muscle tone. If rabies is not prevented by immunization, it is almost always fatal.

Description

Cases of rabies in humans are very infrequent in the United States and Canada, averaging one or two a year (down from over 100 cases annually in 1900), but, according to the World Health Organization, about 55,000 people worldwide die of the infection each year; about one person every ten minutes. Rabies is most common in developing countries in Africa, Latin America, and Asia, particularly India. Dog **bites** are the major origin of infection for humans in developing countries, but other important host animals may include the wolf, mongoose, raccoon, jackal, and bat. A group of researchers in India found that monkeys as well as dogs were frequent vectors of rabies. The team also reported that the male:female ratio of rabies patients in India is 4:1.

Most deaths from rabies in the United States and Canada result from bat. The **death** of a nine-year-old girl in Quebec in the fall of 2000 was the first case of human rabies in Canada since 1985. Public health officials eventually determined that the girl had been bitten while she was sleeping by a silver-haired bat that had gotten into the family’s home.

On October 18, 2004, a Wisconsin teenager was diagnosed with full-blown rabies after suffering from a minor bat bite on September 12, 2004. Miraculously, she was cured of rabies after doctors induced **coma** and administered four **antiviral drugs** to her.

People whose work frequently brings them in contact with animals are considered to be at higher risk than the general population. This would include those in the fields of veterinary medicine, animal control, wildlife work, and laboratory work involving live rabies virus. People in these occupations and residents of or travelers to areas where rabies is a widespread problem should consider being immunized.

In late 2002, rabies re-emerged as an important public health issue. Dr. Charles E. Rupprecht, director of the World Health Organization (WHO) Collaborating Center for Rabies Reference and Research, has listed several factors responsible for the increase in the number of rabies cases worldwide:

- Rapid evolution of the rabies virus. Bats in the United States have developed a particularly infectious form of the virus
- Increased diversity of animal hosts for the disease
- Changes in the environment that are bringing people and domestic pets into closer contact with infected wildlife
- Increased movement of people and animals across international borders. In one recent case, a man who

KEY TERMS

Active immunization—Treatment that provides immunity by challenging an individual's own immune system to produce antibody against a particular organism, in this case the rabies virus.

Antibody—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

Biopsy—The removal of a small sample of tissue for diagnostic purposes.

Efferent nerves—Nerves that convey impulses away from the central nervous system to the periphery.

Fluorescent antibody test (FA test)—A test in which a fluorescent dye is linked to an antibody for diagnostic purposes.

Lumbar puncture—A procedure that involves withdrawing a small sample of cerebrospinal fluid from the back around the spinal cord.

Lyssavirus—A genus of viruses that includes the rabies virus and related viruses that infect insects as well as mammals.

Passive immunization—Treatment that provides immunity through the transfer of antibodies obtained from an immune individual.

Rhabdovirus—A type of virus named for its rod- or bullet-like shape. The rabies virus belongs to a family of viruses called Rhabdoviridae.

Vector—An animal or insect that carries a disease-producing organism.

Zoonosis—Any disease of animals that can be transmitted to humans. Rabies is an example of a zoonosis.

had contracted rabies in the Philippines was not diagnosed until he began to feel ill in the United Kingdom

- Lack of advocacy about rabies

Causes and symptoms

Rabies is caused by a rod- or bullet-shaped virus that belongs to the family Rhabdoviridae. The rabies virus is a member of a genus of viruses called lyssaviruses, which include several related viruses that infect insects as well as mammals. The rabies virus is usually transmitted via an animal bite, however, cases have also been reported in which the virus penetrated the body through infected saliva, moist tissues such as the eyes or lips, a scratch on the skin, or the transplantation of infected tissues. Inhalation of the virus in the air, as might occur in a highly populated bat cave, is also thought to occur.

From the bite or other area of penetration, the virus multiplies as it spreads along nerves that travel away from the spinal cord and brain (efferent nerves) and into the salivary glands. The rabies virus may lie dormant in the body for several weeks or months, but rarely much longer, before symptoms appear. Initially, the area around the bite may burn and be painful. Early symptoms may also include a **sore throat**, low-grade fever, **headache**, loss of appetite, **nausea and vomiting**, and **diarrhea**. Painful spasms develop in the muscles that control breathing and swallowing. The individual may begin to drool thick saliva and

may have dilated or irregular pupils, increased tears and perspiration, and low blood pressure.

Later, as the disease progresses, the patient becomes agitated and combative and may exhibit increased mental confusion. The affected person usually becomes sensitive to touch, loud noises, and bright lights. The victim also becomes extremely thirsty, but is unable to drink because swallowing is painful. Some patients begin to dread water because of the painful spasms that occur. Other severe symptoms during the later stage of the disease include excessive salivation, **dehydration**, and loss of muscle tone. Death usually occurs three to 20 days after symptoms have developed. Unfortunately, recovery is very rare.

Diagnosis

After the onset of symptoms, blood tests and **cerebrospinal fluid (CSF) analysis** tests will be conducted. CSF will be collected during a procedure called a **lumbar puncture** in which a needle is used to withdraw a sample of CSF from the area around the spinal cord. The CSF tests do not confirm diagnosis but are useful in ruling out other potential causes for the patient's altered mental state.

The two most common diagnostic tests are the fluorescent antibody test and isolation of the rabies virus from an individual's saliva or **throat culture**. The fluorescent antibody test involves taking a small sample of skin (biopsy) from the back of the neck of the patient. If specific proteins, called antibodies,

that are produced only in response to the rabies virus are present, they will bind with the fluorescent dye and become visible. Another diagnostic procedure involves taking a corneal impression in which a swab or slide is pressed lightly against the cornea of the eye to determine whether viral material is present.

Treatment

Until the most recent successful cure of a late-term rabies case can be validated with further success and validation from the medical community, the historic treatment options for rabies prevention immediately following a bite remains the most viable treatment. Because of the extremely serious nature of a rabies infection, the need for rabies immunizations will be carefully considered for anyone who has been bitten by an animal, based on a personal history and results of diagnostic tests.

If necessary, treatment includes the following:

- The wound is washed thoroughly with medicinal soap and water. Deep puncture wounds should be flushed with a catheter and soapy water. Unless absolutely necessary, a wound should not be sutured.
- Tetanus toxoid and antibiotics will usually be administered.
- Rabies vaccination may or not be given, based on the available information. If the individual was bitten by a domestic animal and the animal was captured, the animal will be placed under observation in quarantine for ten days. If the animal does not develop rabies within four to seven days, then no immunizations are required. If the animal is suspected of being rabid, it is killed, and the brain is examined for evidence of rabies infection. In cases involving bites from domestic animals where the animal is not available for examination, the decision for vaccination is made based on the prevalence of rabies within the region where the bite occurred. If the bite was from a wild animal and the animal was captured, it is generally killed because the incubation period of rabies is unknown in most wild animals.
- If necessary, the patient is vaccinated immediately, generally through the administration of human rabies immune globulin (HRIG) for passive immunization, followed by human diploid cell vaccine (HDCV) or rabies vaccine adsorbed (RVA) for active immunization. Passive immunization is designed to provide the individual with antibodies from an already immunized individual, while active

immunization involves stimulating the individual's own immune system to produce antibodies against the rabies virus. Both rabies vaccines are equally effective and carry a lower risk of side effects than some earlier treatments. Unfortunately, however, in underdeveloped countries, these newer vaccines are usually not available. Antibodies are administered to the patient in a process called passive immunization. To do this, the HRIG vaccine is administered once, at the beginning of treatment. Half of the dose is given around the bite area, and the rest is given in the muscle. Inactivated viral material (antigenic) is then given to stimulate the patient's own immune system to produce antibodies against rabies. For active immunization, either the HDCV or RVA vaccine is given in a series of five injections. Immunizations are typically given on days one, three, seven, 14, and 28.

In those rare instances in which rabies has progressed beyond the point where immunization would be effective, the groundbreaking treatment involving a drug-induced coma and the administration of four different antiviral drugs will most likely be a radical treatment option. The traditional approach prior to October 2004 was to provide as much relief from **pain** and suffering as possible through medical intervention while waiting to see if survival was possible. The patient would be given medication to prevent seizures, relieve some of the **anxiety**, and relieve painful muscle spasms. Pain relievers would also be given. In the later stages, aggressive supportive care would be provided to maintain breathing and heart function. Survival via the traditional treatment is rare but can occur.

Prognosis

If preventative treatment is sought promptly, rabies need not be fatal. Immunization is almost always effective if started within two days of the bite. Chance of effectiveness declines, however, the longer **vaccination** is put off. It is, however, important to start immunizations, even if it has been weeks or months following a suspected rabid animal bite, because the vaccine can be effective even in these cases. If immunizations do not prove effective or are not received, rabies is nearly always fatal with a few days of the onset of symptoms.

Prevention

One promising preventive strategy that has been used since the early 2000s is the distribution of wildlife baits containing an oral vaccine against rabies.

This strategy has been used in Germany to vaccinate wild foxes, which are frequent carriers of the disease in Europe. In the United States, veterinary researchers at Kansas State University have developed an oral vaccine for fruit bats; early trials of the vaccine have given promising results. The cost of rabies prevention in the United States (mostly attributable to the vaccination of domestic animals) was estimated at \$300 million.

The following precautions should be observed in environments where humans and animals may likely come into contact.

- Domesticated animals, including household pets, should be vaccinated against rabies. If a pet is bitten by an animal suspected to have rabies, its owner should contact a veterinarian immediately and notify the local animal control authorities. Domestic pets with current vaccinations should be revaccinated immediately; unvaccinated dogs, cats, or ferrets are usually euthanized (put to sleep). Further information about domestic pets and rabies is available on the American Veterinary Medical Association (AVMA) web site.
- Wild animals should not be touched or petted, no matter how friendly they may appear. It is also important not to touch an animal that appears ill or passive, or whose behavior seems odd, such as failing to show the normal fear of humans. These are all possible signs of rabies. Many animals, such as raccoons and skunks, are nocturnal and their activity during the day should be regarded as suspicious.
- People should not interfere in fights between animals.
- Because rabies is transmitted through saliva, a person should wear rubber gloves when handling a pet that has had an encounter with a wild animal.
- Garbage or pet food should not be left outside the house or camp site because it may attract wild or stray animals.
- Windows and doors should be screened. Some victims of rabies have been attacked by infected animals, particularly bats, that entered through unprotected openings.
- State or county health departments should be consulted for information about the prevalence of rabies in an area. Some areas, such as New York City, have been rabies-free, only to have the disease reintroduced at a later time.
- Preventative vaccination against rabies should be considered if one's occupation involves frequent contact with wild animals or non-immunized domestic animals.

- Bites from mice, rats, or squirrels rarely require rabies prevention because these rodents are typically killed by any encounter with a larger, rabid animal, and would, therefore, not be carriers.
- Travelers should ask about the prevalence of the disease in countries they plan to visit.

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American Veterinary Medical Association (AVMA), 1931 North Meacham Road, Suite 100, Schaumburg, IL, 60173-4360, <http://www.avma.org>.

Centers for Disease Control and Prevention, 1600 Clifton Rd., NE, Atlanta, GA, 30333, (404) 639-3311, (800) 311-3435, <http://www.cdc.gov>.

Institut Pasteur, 25-28, Rue du Dr. Roux, 75015, Paris, France, + 3301 45 68 80 00, http://www.pasteur.fr/haut_ext.html.

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Radial keratotomy

Definition

Radial keratotomy (RK) is a type of eye surgery used to correct **myopia** (nearsightedness). It works by changing the shape of the cornea—the transparent part of the eye that covers the iris and the pupil.

Purpose

About 25-30% of all people in the world are nearsighted and need eyeglasses or **contact lenses** for distance vision to be clear. For a number of reasons, some people don't like wearing corrective lenses. Some feel unattractive in eyeglasses while others worry about not being able to see without their glasses in an emergency, such as a house fire or a burglary. Both glasses and contact lenses can be scratched, broken, or lost. Contact lenses require special care and can irritate the eyes.

Radial keratotomy was introduced in North America in 1978. Since then doctors have improved the technique, and its results have become more predictable. Radial keratotomy is one of several surgical techniques to correct nearsightedness, reducing or eliminating the need for corrective lenses. It is most successful in patients with a low to moderate amount of

nearsightedness—people whose eyes require up to -5.00 diopters of correction. A diopter (D) is a unit of measure of focusing power. Minus lenses correct nearsightedness.

Precautions

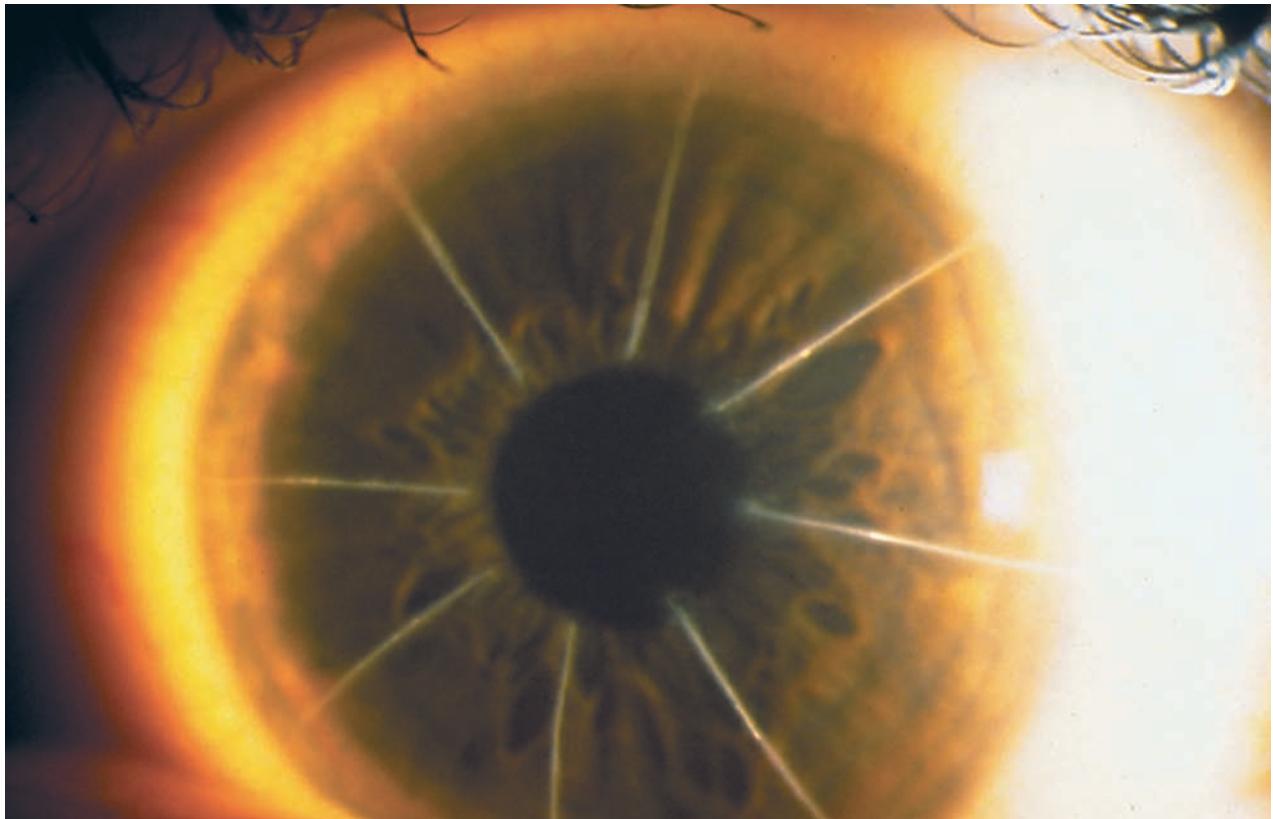
Not every nearsighted person is a good candidate for radial keratotomy. This type of surgery cannot help people whose nearsightedness is caused by keratoconus, a rare condition in which the cornea is cone shaped. The procedure usually is not done on patients under 18, because their eyes are still growing and changing shape. It is important that visual status is stable. Women who are pregnant, have just given birth, or are breast-feeding should not have the surgery because hormonal changes may cause temporary changes in the cornea. In addition, anyone with glaucoma or with any disease that interferes with healing (e.g., **rheumatoid arthritis**, lupus erythematosus, or uncontrolled diabetes) should not have RK.

Radial keratotomy weakens the cornea, making it vulnerable to injuries even long after the surgery. Getting hit in the head after having RK can cause the cornea to tear and can lead to blindness. For this reason, the procedure is not recommended for people who engage in sports that could result in a blow to the head (i.e., karate or racquetball).

It is important to keep in mind that RK is a permanent procedure and that success cannot be guaranteed. An experienced eye surgeon can estimate how likely it is that the surgery will help a particular patient, but that is just an estimate. There is no way to know for sure whether the surgery will improve eyesight enough to eliminate the need for corrective lenses. Vision usually improves after RK, but it is not always perfect. Anyone who decides to have RK should be prepared to accept less-than-perfect vision after surgery, which may necessitate the continued use of glasses or contact lenses. This surgery does not eliminate the need for reading glasses. Actually, someone who didn't need reading glasses before surgery because their myopia allowed near vision to be clear may find themselves needing reading glasses. Patients must ask about this prior to surgery.

Anyone considering RK should also be aware that certain professions, including branches of the military, are not open to people who have had the procedure.

A reputable ophthalmologist will discuss the risks of the procedure and should tell anyone considering it that perfect vision can't be guaranteed. Patients should be wary of any doctor who tries too hard to "sell" them on RK.



Radial keratotomy scars on the cornea of an eye. (© Bob Masini/Phototake. — All rights reserved.)

Description

In a person with clear vision, light passes through the cornea and the lens of the eye and focuses on a membrane lining the back of the eye called the retina. In a person with myopia, the eyeball is usually too long, so light focuses in front of the retina. Radial keratotomy reduces myopia by flattening the cornea. This reduces the focusing power of the cornea allowing light to focus further back onto the retina (or at least closer to it), forming a clearer image.

A surgeon performing RK uses a very small diamond-blade knife to makes four to eight radial incisions around the edge of the cornea. These slits are made in a pattern that resembles the spokes of wheel. As the cornea heals, its center flattens out.

Radial keratotomy is usually performed in an ophthalmologist's office. Before the surgery begins, the patient may be given medicine to help him or her relax. A local anesthetic—usually in the form of eye drops—is used to numb the eye, but the patient remains conscious during the procedure. The surgeon looks through a surgical microscope while making the slits. The treatment usually takes no more than 30 minutes.

Some ophthalmologists will perform RK on both eyes at once but others prefer to do one eye at a time. It once was thought that surgeons could use the results of the first eye to predict how well the procedure would work on the second eye. However, a study published in 1997 found that this was not the case. The authors of the study cautioned that there might be other reasons not to operate on both eyes at once, such as increased risk of infection and other complications.

The cost for RK depends on the surgeon, but usually ranges from \$1,000–\$1,500 per eye. Medical insurance usually does not cover RK, because it is considered an elective procedure—one that people choose to have done.

Preparation

Before beginning the procedure, the surgeon marks an area in the center of the cornea called the optical zone. This is the part of the cornea that one sees through (it is the area over the pupil). No cuts are made in this region. The surgeon also measures the cornea's thickness, to decide how deep the slits should be.

KEY TERMS

Cornea—The transparent part of the eye that covers the iris and the pupil.

Diopter (D)—Unit describing the amount of focusing power of a lens.

Iris—The colored part of the eye.

Laser-assisted in situ keratomileusis (LASIK)—A type of refractive eye surgery using a laser and another instrument to change the shape of the cornea.

Local anesthetic—Used to numb an area where surgery or another procedure is to be done, without causing the patient to lose consciousness.

Myopia—Nearsightedness. People with myopia cannot see distant objects clearly.

Ophthalmologist—A physician who specializes in treating eyes.

Photorefractive keratectomy (PRK)—A type of refractive eye surgery using a laser to change the shape of the cornea.

Pupil—The part of the eye that looks like a black circle in the center of the iris. It is actually an opening through which light passes.

Retina—A membrane lining the back of the eye onto which light is focused to form images.

Aftercare

After the surgery is over, the anesthetic wears off. Some patients feel slight **pain** and are given eye drops and medications to relieve their discomfort. For several days after the surgery, the eye that was treated may feel scratchy and look red. This is normal. The eye may also water, burn slightly, and be sensitive to light.

As with any type of surgery, it is important to guard against infection. Patients are given eye drops to protect against infection and may be told to use them for several weeks after the surgery. Because RK weakens the cornea it is important to protect the head and eyes.

The cornea heals slowly, and full recovery can take several months (another reason not to have the surgery done on both eyes at the same time). While the cornea is healing, patients may experience these problems:

- Variations in vision
- Temporary pain
- Increased glare
- Starburst or halo effects
- Hyperopic shift

As the cornea flattens, vision may become more farsighted (hyperopic). For this reason, the surgeon may initially undercorrect the patient. This gradual shift may occur over several years.

If RK does not completely correct a person's nearsightedness, glasses or contact lenses may be needed. In general, people who were able to wear contact lenses before the procedure can still wear them afterward. Even patients whose nearsightedness was corrected may still need glasses for reading. This is especially true for middle-aged and older patients. The lens of the eye

stiffens with age, making reading glasses necessary (**presbyopia**). Radial keratotomy does not correct this problem.

The surgeon who performs the RK procedure will tell the patient how often to return for follow-up visits. Often, two to four visits are needed, including one the day after surgery. It is also important to know what side effects should be reported immediately to the surgeon (e.g., pain or **nausea**).

Risks

Complications from RK are rare, but they can occur. These include:

- cataract a clouding of the lens of the eye, resulting in partial or total loss of vision
- serious infection
- lasting pain
- rips along an incision, especially after being hit in the head or eye
- loss of vision
- chance of overcorrection (hyperopic shift)

The chances of complications are reduced when the surgery is done by an ophthalmologist with a lot of experience in RK. Younger patients also tend to heal faster.

Normal results

The desired result of radial keratotomy is a reduction in myopia. A major study by the National Eye Institute, reported in 1994, tracked the success of RK in 374 patients who had had the procedure done 10 years earlier. The study found that:

- 85% had at least 20/40 vision (the acuity considered good enough to drive without glasses)
- 70% did not need glasses or contact lenses for distance vision
- 53% had 20/20 vision without glasses
- 30% still needed glasses or contact lenses to see clearly
- 1–3% had worse vision than before they had RK
- 40% had a hyperopic shift.

As with all surgeries, RK has risks. These risks include having worse vision than before the surgery; halos; glare; and although rare, blindness. Some after-effects, such as halos or glare may last for years. Other refractive surgeries, such as photorefractive keratectomy (PRK) and laser-assisted in situ keratomileusis (LASIK) use lasers to change the shape of the cornea and they may produce fewer side effects. It is important to speak with an experienced eye surgeon who has done many refractive surgeries to fully understand the options and risks involved before making a decision.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, 415 561-8500, <http://www.aoa.org>.

American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, 314 991-4100, (314) 991-4101, 800 365-2219, <http://www.aoa.org/>.

American Society of Cataract and Refractive Surgery, 4000 Legato Road, Suite 700, Fairfax, VA, 22033, (703) 591-2220, (703) 591-0614, <http://www.ascrs.org>.

Nancy Ross-Flanigan



This person's nose is inflamed and scaly due to radiation exposure. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

atmosphere, creating radiation. Radiation can either be electromagnetic or particulate.

The energy of electromagnetic radiation is a direct function of its frequency. The high-energy, high-frequency waves that can penetrate solids to various depths cause damage by separating molecules into electrically charged pieces, a process known as ionization. X rays are a type of electromagnetic radiation. Atomic particles come from radioactive isotopes as they decay to stable elements. Electrons are called beta particles when they radiate. Alpha particles are the nuclei of helium atoms—two protons and two neutrons—without the surrounding electrons. Alpha particles are too large to penetrate a piece of paper unless they are greatly accelerated in electric and magnetic fields. Both beta and alpha particles are types of particulate radiation. When over-exposure to ionizing radiation occurs, there is chromosomal damage in deoxyribonucleic acid (DNA). DNA is very good at repairing itself; both strands of the double helix must be broken to produce genetic damage.

Because radiation is energy, it can be measured. There are a number of units used to quantify radiation energy. Some refer to effects on air, others to effects on living tissue. The roentgen, named after Wilhelm Conrad Roentgen, who discovered x rays in 1895, measures ionizing energy in air. A rad expresses the energy transferred to tissue. The rem measures tissue response. A roentgen generates about a rad of effect and produces about a rem of response. The gray and the sievert are international units equivalent to 100 rads and rems, respectively. A curie, named after French physicists who experimented with radiation, is a measure of actual radioactivity given off by a radioactive element, not a measure of its effect. The average annual human

Radiation injuries

Definition

Radiation injuries are caused by ionizing radiation emitted by sources such as the sun, x-ray and other diagnostic machines, **tanning** beds, and radioactive elements released in nuclear power plant accidents and detonation of nuclear weapons during war and as terrorist acts.

Description

Ionizing radiation is made up of unstable atoms that contain an excess amount of energy. In an attempt to stabilize, the atoms emit the excess energy into the

exposure to natural background radiation is roughly 3 milliSieverts (mSv).

Any amount of ionizing radiation will produce some damage, however, there is radiation everywhere, from the sun (cosmic rays) and from traces of radioactive elements in the air (radon) and the ground (uranium, radium, carbon-14, potassium-40 and many others). Earth's atmosphere protects us from most of the sun's radiation. Living at 5,000 feet altitude in Denver, Colorado, doubles exposure to radiation, and flight in a commercial airliner increases it 150-fold by lifting us above 80% of that atmosphere. Because no amount of radiation is perfectly safe and because radiation is ever present, arbitrary limits have been established to provide some measure of safety for those exposed to unusual amounts. Less than 1% of them reach the current annual permissible maximum of 20 mSv.

A 2001 ruling by the Federal Court of Australia indicated that two soldiers died from **cancer** caused by minimal exposure to radiation while occupying Hiroshima in 1945. The soldiers were exposed to less than 5 mSv of radiation. The international recommendation for workers is safety level of up to 20 mSv. The ruling and its support by many international agencies suggests that even extremely low doses of radiation can be potentially harmful.

Ultraviolet (UV) radiation exposure from the sun and tanning beds

UV radiation from the sun and tanning beds and lamps can cause skin damage, premature **aging**, and skin cancers. **Malignant melanoma** is the most dangerous of skin cancers and there is a definite link between type UVA exposure used in tanning beds and its occurrence. UVB type UV radiation is associated with **sunburn**, and while not as penetrating as UVA, it still damages the skin with over exposure. Skin damage accumulates over time, and effects do not often manifest until individuals reach middle age. Light-skinned people who most often burn rather than tan are at a greater risk of skin damage than darker-skinned individuals that almost never burn. The U.S. Food and Drug Administration (FDA) and the Centers for Disease Control (CDC) discourage the use of tanning beds and sun lamps and encourage the use of sunscreen with at least an SPF of 15 or greater.

Over exposure during medical procedures

Ionizing radiation has many uses in medicine, both in diagnosis and in treatment. X rays, CT scanners, and fluoroscopes use it to form images of the body's insides. Nuclear medicine uses radioactive isotopes to diagnose

and to treat medical conditions. In the body, radioactive elements localize to specific tissues and give off tiny amounts of radiation. Detecting that radiation provides information on both anatomy and function. During the past 10 years, skin injuries caused by too much exposure during a medical procedure have been documented. In 1995, the FDA issued a recommendation to physicians and medical institutions to record and monitor the dosage of radiation used during medical procedures on patients in order to minimize the amount of skin injuries. The FDA suggested doses of radiation not exceed 1 Grey (Gy), which is roughly equivalent to a sievert. The FDA prepared further guidelines for fluoroscopy, the procedure most often associated with medical-related radiation skin injuries such as **rashes** and more serious **burns** and tissue **death**. Injuries occurred most often during **angioplasty** procedures using fluoroscopy.

CT scans of children have also been problematic. Oftentimes the dosage of radiation used for an adult isn't decreased for a child, leading to radiation over exposure. Children are more sensitive to radiation and a February 2001 study indicates 1,500 out of 1.6 million children under 15 years of age receiving CT scans annually will develop cancer. Studies show that decreasing the radiation by half for CT scans of children will effectively decrease the possibility of over exposure while still providing an effective diagnostic image. The benefits to receiving the medical treatment utilizing radiation is still greater than the risks involved, however, more stringent control over the amount of radiation used during the procedures will go far to minimize the risk of radiation injury to the patient.

Radiation exposure from nuclear accidents, weaponry, and terrorist acts

Between 1945 and 1987, there were 285 nuclear reactor accidents, injuring more than 1,550 people and killing 64. The most striking example was the meltdown of the graphite core nuclear reactor at Chernobyl in 1986, which spread a cloud of radioactive particles across the entire continent of Europe. Information about radiation effects is still being gathered from that disaster, however 31 people were killed in the immediate accident and 1,800 children have thus far been diagnosed with **thyroid cancer**. In a study published in May 2001 by the British Royal Society, children born to individuals involved in the cleanup of Chernobyl and born after the accident are 600% more likely to have genetic mutations than children born before the accident. These findings indicate that exposure to low doses of radiation can cause inheritable effects.

Since the terrorist attack on the World Trade Center and the Pentagon on September 11, 2001, the possibility of terrorist-caused nuclear accidents has been a growing concern. All 103 active nuclear power plants in the United States are on full alert, but they are still vulnerable to sabotage such as bombing or attack from the air. A no-fly zone of 12 miles below 18,000 feet has been established around nuclear power plants by the Federal Aviation Administration (FAA). There is also growing concern over the security of spent nuclear fuel—more than 40,000 tons of spent fuel is housed in buildings at closed plants around the country. Unlike the active nuclear reactors that are enclosed in concrete-reinforced buildings, the spent fuel is stored in non-reinforced buildings. Housed in cooling pools, the spent fuel could emit dangerous levels of radioactive material if exploded or used in makeshift weaponry. Radioactive medical and industrial waste could also be used to make “dirty bombs.” Since 1993, the Nuclear Regulatory Commission (NRC) has reported approximately 400 cases of stolen radioactive materials.

Causes and symptoms

Radiation can damage every tissue in the body. The particular manifestation will depend upon the amount of radiation, the time over which it is absorbed, and the susceptibility of the tissue. The fastest growing tissues are the most vulnerable, because radiation as much as triples its effects during the growth phase. Bone marrow cells that make blood are the fastest growing cells in the body. A fetus in the womb is equally sensitive. The germinal cells in the testes and ovaries are only slightly less sensitive. Both can be rendered useless with very small doses of radiation. More resistant are the lining cells of the body—skin and intestines. Most resistant are the brain cells, because they grow the slowest.

The length of exposure makes a big difference in what happens. Over time the accumulating damage, if not enough to kill cells outright, distorts their growth and causes scarring and/or cancers. In addition to leukemias, cancers of the thyroid, brain, bone, breast, skin, stomach, and lung all arise after radiation. Damage depends, too, on the ability of the tissue to repair itself. Some tissues and some types of damage produce much greater consequences than others.

There are three types of radiation injuries.

- External irradiation: as with x-ray exposure, all or part of the body is exposed to radiation that either is absorbed or passes through the body

- Contamination: as with a nuclear accident, the environment and its inhabitants are exposed to radiation. People are affected internally, externally, or with both internal and external exposure

- Incorporation: dependent on contamination, the bodies of individuals affected incorporate the radiation chemicals within cells, organs, and tissues and the radiation is dispersed throughout the body

Immediately after sudden irradiation, the fate of those affected depends mostly on the total dose absorbed. This information comes mostly from survivors of the atomic bomb blasts over Japan in 1945.

- Massive doses incinerate immediately and are not distinguishable from the heat of the source
- A sudden whole body dose over 50 Sv produces such profound neurological, heart, and circulatory damage that patients die within the first two days
- Doses in the 10–20 Sv range affect the intestines, stripping their lining and leading to death within three months from vomiting, diarrhea, starvation, and infection
- Victims receiving 6–10 Sv all at once usually escape an intestinal death, facing instead bone marrow failure and death within two months from loss of blood coagulation factors and the protection against infection provided by white blood cells
- Between 2–6 Sv gives a fighting chance for survival if victims are supported with blood transfusions and antibiotics
- One or two Sv produces a brief, non-lethal sickness with vomiting, loss of appetite, and generalized discomfort

Treatment

It is clearly important to have some idea of the dose received as early as possible, so that attention can be directed to those victims in the 2–10 Sv range that might survive with treatment. Blood transfusions, protection from infection in damaged organs, and possibly the use of newer stimulants to blood formation can save many victims in this category.

Local radiation exposures usually damage the skin and require careful wound care, removal of dead tissue, and **skin grafting** if the area is large. Again **infection control** is imperative.

One of the best known, and perhaps even mainstream, treatments of radiation injury is the use of *Aloe vera* preparations on damaged areas of skin. It has demonstrated remarkable healing properties even for chronic ulcerations resulting from radiation exposure.

KEY TERMS

DNA—Deoxyribonucleic acid. The chemical of chromosomes and hence the vehicle of heredity.

Isotope—An unstable form of an element that gives off radiation to become stable. Elements are characterized by the number of electrons around each atom. One electron's negative charge balances the positive charge of each proton in the nucleus. To keep all those positive charges in the nucleus from repelling each other (like the same poles of magnets), neutrons are added. Only certain numbers of neutrons work. Other numbers cannot hold the nucleus together, so it splits apart, giving off ionizing radiation. Sometimes one of the split products is not stable either, so another split takes place. The process is called radioactivity.

Alternative treatment

There is considerable interest these days in benevolent chemicals called "free radical scavengers." How well they work is yet to be determined, but population studies strongly suggest that certain **diets** are better than others, and that those diets are full of free radical scavengers, otherwise known as **antioxidants**. The recommended ingredients are beta-carotene, **vitamins E** and **C**, and selenium, all available as commercial preparations. Beta-carotene is yellow-orange and is present in yellow and orange fruits and vegetables. Vitamin C can be found naturally in citrus fruits. **Traditional Chinese medicine** (TCM) and **acupuncture**, botanical medicine, and homeopathy all have contributions to make to recovery from the damage of radiation injuries. The level of recovery will depend on the exposure. Consulting practitioners trained in these modalities will result in the greatest benefit.

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Jacqueline L. Longe

Radiation sickness see **Radiation injuries**

Radiation therapy

Definition

Radiation therapy, sometimes called radiotherapy, x-ray therapy, radiation treatment, cobalt therapy, electron beam therapy, or irradiation uses high energy, penetrating waves or particles such as x rays, gamma rays, proton rays, or neutron rays to destroy **cancer** cells or keep them from reproducing.

Purpose

The purpose of radiation therapy is to kill or damage cancer cells. Radiation therapy is a common form of cancer therapy. It is used in more than half of all cancer cases. Radiation therapy can be used:

- alone to kill cancer
- to prevent cancer cells from spreading into the area of tissue receiving radiation (prophylactic treatment)
- before surgery to shrink a tumor and make it easier to remove
- during surgery to kill cancer cells that may remain in surrounding tissue after the surgery (called intraoperative radiation)
- after surgery to kill cancer cells remaining in the body
- to shrink an inoperable tumor in order to reduce pain and improve quality of life (palliative treatment)
- in combination with chemotherapy or hormone therapy

For some kinds of cancers such as early-stage Hodgkin's disease, non-Hodgkin's lymphomas, and certain types of prostate or brain cancer, radiation therapy alone may cure the disease. In other cases, radiation therapy used in conjunction with surgery, **chemotherapy**, hormone therapy, or all three, increases survival rates over any of these therapies used alone.

Precautions

External radiation therapy does not make the body of the person having the treatments radioactive. In almost all cases, the benefits of this therapy outweigh the risks. However, radiation therapy can have serious consequences, so anyone contemplating it should be sure to understand why the treatment team believes it is the best possible treatment option for their cancer. Radiation therapy is often not appropriate for pregnant women, because the radiation can damage the cells of the developing baby. Women who think they might be pregnant should discuss this with their doctor.

KEY TERMS

Anemia—Insufficient red blood cells in the body.

Antibody—Protein molecule that recognizes and binds specifically to a foreign substance in the body in order to eliminate it.

Chemotherapy—Injecting drugs into the body where they circulate and kill cancer cells.

Computed tomography (CT or CAT) scan—Using X rays taken from many angles and computer modeling, CT scans help locate and size tumors and provide information on whether they can be surgically removed.

Fractionation—A procedure for dividing a dose of radiation into smaller treatment doses.

Gamma rays—Short wavelength, high energy electromagnetic radiation emitted by radioactive substances.

Hodgkin's disease—Cancer of the lymphatic system, characterized by lymph node enlargement and the presence of a large polypliod cells called Reed-Sternberg cells.

Magnetic resonance imaging (MRI)—MRI uses magnets and radio waves to create detailed cross-sectional pictures of the interior of the body.

Palliative—Referring to treatment intended to relieve pain rather than effect a cure.

Prophylactic—A medication or treatment intended to protect against or ward off disease.

Radiopharmaceuticals—Radioactive drugs used as tracers in the diagnosis and treatment of cancers. The most common element used in radiopharmaceuticals is an isotope of technetium known as Tc-99m.

Stereotactic—Characterized by precise positioning in space. When applied to radiosurgery, stereotactic refers to a system of three-dimensional coordinates for locating the target site.

Description

Radiation therapy is a local treatment that is painless. The radiation acts only on the part of the body that is exposed to the radiation. This form of treatment is very different from chemotherapy in which drugs circulate throughout the whole body. There are two main types of radiation therapy. In external radiation therapy a beam of radiation is directed from outside the body at the cancer. In internal radiation therapy, called

brachytherapy or implant therapy, a source of radioactivity is surgically placed inside the body near the cancer.

How radiation therapy works

The protein that carries the code controlling most activities in the cell is called deoxyribonucleic acid or DNA. When a cell divides, its DNA must also double and divide. High-energy radiation kills cells by damaging their DNA. This blocks their ability to grow and increase in number.

One of the characteristics of cancer cells is that they grow and divide faster than normal cells. This makes them particularly vulnerable to radiation. Radiation also damages normal cells, but because normal cells are growing more slowly, they are better able to repair radiation damage than are cancer cells. In order to give normal cells time to heal and reduce side effects, radiation treatments are often given in small doses over a six- or seven-week period.

External radiation therapy

External radiation therapy is the most common kind of radiation therapy. It is usually done during outpatient visits to a hospital clinic and is usually covered by insurance.

Once a doctor called a radiation oncologist determines the proper dose of radiation for a particular cancer, the dose is divided into smaller doses called fractions. One fraction is usually given each day, five days a week for six to seven weeks. However, each radiation plan is individualized depending on the type and location of the cancer and what other treatments are also being used. The actual administration of the therapy usually takes about half an hour daily, although radiation is only administered for one to five minutes at each session. It is important to attend every scheduled treatment to get the most benefit from radiation therapy.

Recently, trials have begun to determine if there are ways to deliver radiation fractions so that they kill more cancer cells or have fewer side effects. Some trials use smaller doses given more often. Up-to-date information on voluntary participation in clinical trials and where they are being held is available by entering the search term “radiation therapy” at the following web sites:

- National Cancer Institute: <http://cancertrials.nci.nih.gov> or (800) 4-CANCER
- National Institutes of Health Clinical Trials: <http://clinicaltrials.gov>
- Center Watch: A Clinical Trials Listing: <http://www.centerwatch.com>

The type of machines used to administer external radiation therapy and the material that provides the radiation vary depending on the type and location of the cancer. Generally, the patient puts on a hospital gown and lies down or sits in a special chair. Parts of the body not receiving radiation are covered with special shields that block the rays. A technician then directs a beam of radiation to a predetermined spot on the body where the cancer is located. The patient must remain still during the administration of the radiation so that no other parts of the body are affected. As an extra precaution in some treatments, special molds are made to make sure the body is in the same position for each treatment. However, the treatment itself is painless, like having a bone x-rayed.

Internal radiation therapy

Internal radiation therapy is called brachytherapy, implant therapy, interstitial radiation, or intracavitary radiation. With internal radiation therapy, a small amount of radioactive material is sealed in an implant (sometimes called a seed or capsule). The implant is then placed very close to the cancer. The advantage of internal radiation therapy is that it concentrates the radiation near the cancer and lessens the chance of damage to normal cells. Many different types of radioactive materials can be used in the implant, including cesium, iridium, iodine, phosphorus, and palladium.

The way in which the implant is positioned near the cancer depends on the size and location of the cancer. Internal radiation therapy is used for some cancers of the head, neck, thyroid, breast, female reproductive system, and prostate. Most people will have the radioactive capsule implanted by a surgeon while under either general or **local anesthesia** at a hospital or surgical clinic.

Patients receiving internal radiation therapy do become temporarily radioactive. They must remain in the hospital during the time that the implant stays in place. The length of time is determined by the type of cancer and the dose of radioactivity to be delivered. During the time the implant is in place, the patient will have to stay in bed and remain reasonably still.

While the implant is in place, the patient's contact with other people will be limited. Health care workers will make their visits as brief as possible to avoid exposure to radiation, and visitors, especially children and pregnant women, will be limited.

The implant usually can be removed in a simple procedure without an anesthetic. As soon as the implant is out of the body, the patient is no longer radioactive, and restrictions on being with other people are lifted. Generally people can return to a level of activity that feels comfortable to them as soon as the implant is removed. Occasionally the site of the implant is sore for some time afterwards. This discomfort may limit specific activities.

In some cases, an implant is left permanently inside the body. People who have permanent implants need to stay in the hospital and away from other people for the first few days. Gradually the radioactivity of the implant decreases, and it is safe to be around other people.

Radioimmunotherapy

Radioimmunotherapy is a promising way to treat cancer that has spread (metastasized) to multiple locations throughout the body. Antibodies are immune system proteins that specifically recognize and bind to only one type of cell. They can be designed to bind only with a certain type of cancer cell. To carry out radioimmunotherapy, antibodies with the ability to bind specifically to a patient's cancer cells are attached to radioactive material and injected into the patient's bloodstream. When these human-made antibodies find a cancer cell, they bind to it. Then the radiation kills the cancer cell. This process is still experimental, but because it can be used to selectively attack only cancer cells, it holds promise for eliminating cancers that have spread beyond the primary tumor.

Types of radiation used to treat cancer

PHOTON RADIATION. Early radiation therapy used x rays like those used to take pictures of bones, or gamma rays. X rays and gamma rays are high-energy rays composed of massless particles of energy (like light) called photons. The distinction between the two is that gamma rays originate from the decay of radioactive substances (like radium and cobalt-60), while x rays are generated by devices that excite electrons (such as cathode ray tubes and linear accelerators). These high-energy rays act on cells by disrupting the electrons of atoms within the molecules inside cells, disrupting cell functions, and, most importantly, by stopping their ability to divide and make new cells.

PARTICLE RADIATION. Particle radiation is radiation delivered by particles that have mass. Proton therapy has been used since the early 1990s. Proton rays consist of protons, a type of positively charged

atomic particle, rather than photons, which have neither mass nor charge. Like x rays and gamma rays, proton rays disrupt cellular activity. The advantage of using proton rays is that they can be shaped to conform to the irregular shape of the tumor more precisely than x rays and gamma rays. They allow delivery of higher radiation doses to tumors without increasing damage to the surrounding tissue.

Neutron therapy is another type of particle radiation. Neutron rays are very high-energy rays. They are composed of neutrons, which are particles with mass but no charge. The type of damage they cause to cells is much less likely to be repaired than that caused by x rays, gamma rays, or proton rays.

Neutron therapy can treat larger tumors than conventional radiation therapy. Conventional radiation therapy depends on the presence of oxygen to work. The center of large tumors lack sufficient oxygen to be susceptible to damage from conventional radiation. Neutron radiation works in the absence of oxygen, making it especially effective for the treatment of inoperable **salivary gland tumors**, bone cancers, and some kinds of advanced cancers of the pancreas, bladder, lung, prostate, and uterus.

Recent advances in radiation therapy

A newer mode of treating brain cancers with radiation therapy is known as stereotactic radiosurgery. Stereotactic radiosurgery allows the doctor to deliver a single high-level dose of precisely directed radiation to the tumor without damaging nearby healthy brain tissue. The treatment is planned with the help of three-dimensional computer-aided analysis of CT and MRI scans. The patient's head and neck are held steady in a skeletal fixation device during the actual treatment. Stereotactic radiosurgery can be used in addition to standard surgery to treat a recurrent **brain tumor**, or in place of surgery if the tumor cannot be reached by standard surgical techniques.

Three major forms of stereotactic radiosurgery are in use. The gamma knife is a stationary machine that is most useful for small tumors, blood vessels, or similar targets. Because it does not move, it can deliver a small, highly localized and precise beam of radiation. Gamma knife treatment is done all at once in a single hospital stay. The second type of radiosurgery uses a movable linear accelerator-based machine or LINAC that is preferred for larger tumors. This treatment is delivered in several small doses given over several weeks. The third form, in limited use in North America, is proton-beam radiosurgery. This form of radiosurgery uses a cyclotron to

generate protons, which are then steered by magnets toward the targeted tumor. Proton-beam radiosurgery can be used to treat cancers of the lung and prostate as well as cancers of the head and neck. Radiosurgery that is performed with divided doses is known as fractionated radiosurgery. The total dose of radiation is higher with a linear accelerator-based machine than with gamma knife treatment.

Another advance in intraoperative radiotherapy (IORT) is the introduction of mobile devices that allow the surgeon to use radiotherapy in early-stage disease and to operate in locations where it would be difficult to transport the patient during surgery for radiation treatment. Mobile IORT units have been used successfully in treating early-stage **breast cancer** and **rectal cancer**.

Radiation sensitizers are another recent innovation in radiation therapy. Sensitizers are medications that are given to make cancer cells easier to kill by radiation than normal cells. Gemcitabine (Gemzar) is one of the drugs most commonly used for this purpose.

3-D conformal radiation therapy and intensity-modulated radiation therapy (IMRT) are two newer techniques used to improve the effectiveness of external radiation therapy. 3-D conformal radiation therapy uses computers to enable doctors to measure the depth as well as the height and width of a tumor. CT scans or **magnetic resonance imaging** (MRI) can be used to obtain a three-dimensional image of the tumor. Special computer programs then design radiation beams that conform closely to the actual shape of the tumor. 3-D conformal radiation therapy has been reported to improve treatment outcomes for nasopharyngeal, prostate, lung, liver, and brain cancers.

IMRT is the most recent type of 3-D conformal radiation therapy. It uses x rays of different intensities to deliver different doses of radiation to small areas of tissue simultaneously. This technique allows the doctor to deliver larger doses of radiation to cancerous tissue and lower doses to nearby healthy tissue at the same time, thus reducing the risk of side effects. IMRT is delivered by a linear accelerator that rotates around the patient. Because the equipment is the size of a small car and is highly specialized, IMRT is not available in all cancer centers. It is used most often to treat cancers of the head, neck, and central nervous system, but has also been used to treat cancers of the breast, lung, uterus, thyroid, and digestive tract.

Radiopharmaceuticals

Radiopharmaceuticals are drugs containing radioactive materials that can be used as tracers in the

diagnosis as well as the treatment of cancer. There are about 28 different elements that can be used in radiopharmaceuticals; however, the element in most common use in cancer diagnosis is technetium. Its radioactive form is an isotope known as Tc99-m. Tc99-m is used to image the thyroid gland, bone marrow, lymph nodes, kidneys, lungs, and blood flow to the brain. Other radiopharmaceuticals include samarium 153 and strontium 89, approved by the FDA to treat **pain** caused by cancer that has metastasized to the bone.

Other drugs used as part of radiation therapy are radiosensitizers, used to make cancerous tissue more sensitive to radiation, and radioprotectors, used to protect healthy tissue from damage caused by radiation therapy. Amifostine is the only drug approved by the FDA as a radioprotector. It is used during radiation therapy of the head and neck to protect the patient's salivary glands and reduce the risk of **dry mouth**.

Preparation

Before radiation therapy, the size and location of the patient's tumor are determined very precisely using magnetic resonance imaging (MRI) and/or **computed tomography scans** (CT scans). The correct radiation dose, the number of sessions, the interval between sessions, and the method of application are calculated by a radiation oncologist based on the tumor type, its size, and the sensitivity of the nearby tissues.

The patient's skin is marked with a semi-permanent ink to help the radiation technologist achieve correct positioning for each treatment. Molds may be built to hold tissues in exactly the right place each time.

Aftercare

Many patients experience skin burn, **fatigue**, **nausea**, and **vomiting** after radiation therapy regardless of where the radiation is applied. After treatment, the skin around the site of the treatment may also become sore. Affected skin should be kept clean and can be treated like **sunburn**, with skin lotion or vitamin A and D ointment. Patients should avoid perfume and scented skin products and protect affected areas from the sun.

Nausea and vomiting are most likely to occur when the radiation dose is high or if the abdomen or another part of the digestive tract is irradiated. Sometimes nausea and **vomiting** occur after radiation to other regions, but in these cases the symptoms usually disappear within a few hours after treatment. Nausea and vomiting can be treated with **antacids**, Compazine, Tigan, or Zofran.

Fatigue frequently starts after the second week of therapy and may continue until about two weeks after

the therapy is finished. Patients may need to limit their activities, take naps, and get extra sleep at night.

Patients should see their oncologist (cancer doctor) at least once within the first few weeks after their final radiation treatment. They should also see an oncologist every six to twelve months for the rest of their lives so they can be checked to see if the tumor has reappeared or spread.

Risks

Radiation therapy can cause anemia, nausea, vomiting, **diarrhea**, hair loss (**alopecia**), skin burn, sterility, and rarely **death**. However, the benefits of radiation therapy almost always exceed the risks. Patients should discuss the risks with their doctor and get a second opinion about their treatment plan.

Normal results

The outcome of radiation treatment varies depending on the type, location, and stage of the cancer. For some cancers such as Hodgkin's disease, about 75% of the patients are cured. **Prostate cancer** also responds well to radiation therapy. Radiation to painful bony metastases is usually a dramatically effective form of pain control. Other cancers may be less sensitive to the benefits of radiation.

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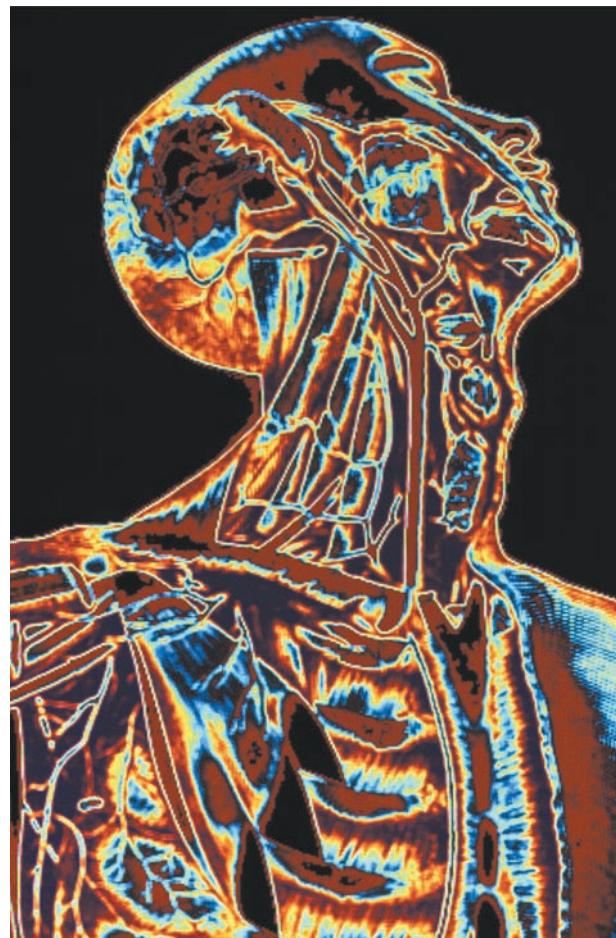
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ORGANIZATIONS

- American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329-4251, (800) ACS-2345, <http://www.cancer.org>.
- International Radiosurgery Support Association (IRSA), 3002 North Second Street, Harrisburg, PA, 17110, (717) 260-9808, <http://www.irsa.org>.
- National Association for Proton Therapy, 1301 Highland Drive, Silver Spring, MD, 20910, (301) 587-6100, <http://www.proton-therapy.org>.

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Radiation treatments see **Radiation therapy**



A digitized illustration of the human head and chest showing nasal passages, sinuses, trachea, vascular nerves, as well as ribs and parts of the lungs. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

dissection removes less tissue, and a selective neck dissection even less.

Radical neck dissection

Definition

Radical neck dissection is an operation used to remove cancerous tissue in the head and neck.

Purpose

The purpose of radical neck dissection is to remove lymph nodes and other structures in the head and neck that are likely or proven to be malignant. Variations on neck dissections exist depending on the extent of the **cancer**. A radical neck dissection removes the most tissue. It is done when the cancer has spread widely in the neck. A modified neck

Precautions

This operation should not be done if cancer has metastasized (spread) beyond the head and neck, or if the cancer has invaded the bones of the cervical vertebrae (the first seven vertebrae of the spinal column) or the skull. In these cases, the surgery will not effectively contain the cancer.

Description

Cancers of the head and neck (sometimes inaccurately called throat cancer) often spread to nearby tissues and into the lymph nodes. Removing these structures is one way of controlling the cancer.

KEY TERMS

Barium swallow—Barium is used to coat the throat in order to take x-ray pictures of the tissues lining the throat.

Computed tomography (CT or CAT) scan—Using x rays taken from many angles and computer modeling, CT scans help size and locate tumors and provide information on whether they can be surgically removed.

Lymph nodes—Small, bean-shaped collections of tissue found in lymph vessels. They produce cells and proteins that fight infection and filter lymph. Nodes are sometimes called lymph glands.

Lymphatic system—Primary defense against infection in the body. The tissues, organs, and channels (similar to veins) that produce, store, and transport lymph and white blood cells to fight infection.

Magnetic resonance imaging (MRI)—MRI uses magnets and radio waves to create detailed cross-sectional pictures of the interior of the body.

Malignant—Cancerous. Cells tend to reproduce without normal controls on growth and form tumors or invade other tissues.

Metastasize—Spread of cells from the original site of the cancer to other parts of the body where secondary tumors are formed.

Of the 600 hundred lymph nodes in the body, about 200 are in the neck. Only a small number of these are removed during a neck dissection. In addition, other structures such as muscles, veins, and nerves may be removed during a radical neck dissection. These include the sternocleidomastoid muscle (one of the muscles that functions to flex the head), internal jugular (neck) vein, submandibular gland (one of the salivary glands), and the spinal accessory nerve (a nerve that helps control speech, swallowing and certain movements of the head and neck). The goal is always to remove all the cancer but to save as many components surrounding the nodes as possible.

Radical neck dissections are done in a hospital under **general anesthesia** by a head and neck surgeon. An incision is made in the neck, and the skin is pulled back to reveal the muscles and lymph nodes. The surgeon is guided in what to remove by tests done prior to surgery and by examination of the size and texture of the lymph nodes.

Preparation

Radical neck dissection is a major operation. Extensive tests are done before the operation to try to determine where and how far the cancer has spread. These may include lymph node biopsies, CT (computed tomography) scans, MRI scans, and barium swallows. In addition, standard pre-operative blood and **liver function tests** are performed, and the patient will meet with an anesthesiologist before the operation. The patient should tell the anesthesiologist about all drug **allergies** and all medication (prescription, non-prescription, or herbal) that he or she is taking.

Aftercare

A person who has had a radical neck dissection will stay in the hospital several days after the operation, and sometimes longer if surgery to remove the primary tumor was done at the same time. Drains are inserted under the skin to remove the fluid that accumulates in the neck area. Once the drains are removed and the incision appears to be healing well, patients are usually discharged from the hospital, but will require follow-up doctor visits. Depending on how many structures are removed, a person who has had a radical neck dissection may require **physical therapy** to regain use of the arm and shoulder.

Risks

The greatest risk in a radical neck dissection is damage to the nerves, muscles, and veins in the neck. Nerve damage can result in **numbness** (either temporary or permanent) to different regions on the neck and loss of function (temporary or permanent) to parts of the neck, throat, and shoulder. The more extensive the neck dissection, the more function the patient is likely to lose. As a result, it is common following radical neck dissection for a person to have stooped shoulders, limited ability to lift the arm, and limited head and neck rotation and flexion due to the removal of nerves and muscles. Other risks are the same as for all major surgery: potential bleeding, infection, and allergic reaction to anesthesia.

Normal results

Normal lymph nodes are small and show no cancerous cells under the microscope.

Abnormal results

Abnormal lymph nodes may be enlarged and show malignant cells when examined under the microscope.

Resources

OTHER

The Voice Center at Eastern Virginia Medical School. <http://www.voice-center.com>.

ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329, (800) 227-2345, <http://www.cancer.org>.

NCI Office of Communications and Education, 6116 Executive Blvd. Suite 300, Bethesda, MD, 20892-8322, (800) 4-CANCER (422-6237), cancergovstaff@mail.nih.gov, <http://www.cancer.gov/aboutnci/cis>.

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Radioactive implants

Definition

Radioactive implants are devices that are placed directly within cancerous tissue or tumors in order to deliver radiation intended to kill cancerous cells. The practice of internal **radiation therapy** is also referred to as brachytherapy. The two main types of brachytherapy are intracavitary radiation, in which the radioactive source is placed in a body cavity, and interstitial radiation, which involves the placement of radioactive material into or near cancerous tissue but not in a body cavity.

Purpose

With the use of radioactive implants, a tumor is subjected to radioactive activity over a longer period of time and often at higher doses, as compared to radiation therapy that is delivered via external beam radiation therapy.

Description

Internal radiation therapy places the sources of radiation directly into the tumor and surrounding tissue. Brachytherapy can be used to treat many different types of **cancer** including tumors of the head, neck, prostate, cervix, skin, breast and other types of cancer.

Radioactive implants can be used alone or they can be used in combination with external radiation therapy. The implant may be permanent or removable. For example, a permanent implant of radioactive seeds,

KEY TERMS

External beam radiation therapy—Radiation therapy delivered from a source external to the body such as from a radiation therapy machine.

Sealed radioactive sources—A radioactive source contained or sealed within a ribbon, wire, needle, balloon, tube or catheter.

such as gold seeds, is placed directly into the organ. These seeds are usually very small in size, about the size of a grain of rice. Over several weeks or months, the seeds slowly deliver radiation to the tumor. This type of implant typically remains in the body permanently.

Depending on the type and location of the cancer being treated, brachytherapy implants can remain in place for a few minutes or for several days. Sealed radioactive sources, such as radioactive ribbons, wires, needles, balloons, and tubes, are placed into body cavities or tissues using devices called applicators. Applicators are usually put into the body in the operating room while the patient is under general or **local anesthesia**. Placement of applicators is done very precisely using x rays or **magnetic resonance imaging** (MRI) to guide the location of placement. The radioactive source is then placed into the applicators once the actual treatment is to begin. Some implants may require hospitalization and may necessitate as little movement as possible to minimize movement of the implant which is placed to target radiation precisely to the tumor. Other implants are left permanently in the body. In this situation, the applicator is removed once the implant is placed.

High-dose-rate brachytherapy is a technique which allows for treatment with a radioactive source which is placed in an applicator over a very short period of time, usually only a few minutes. Once the treatment time has elapsed, the radioactive materials are removed from the body. The applicator may or may not be removed. The treatment period ranges from several days to several weeks. This type of procedure is usually done in an outpatient radiation therapy center.

Preparation

The planning and procedures used for treatment with radioactive implants is becoming increasingly accurate and sophisticated as technology develops. Special imaging tools and computer software used by radiation physicists help radiation oncologists and radiation therapists visualize implant placements which very precisely target the cancerous tissue which is to be treated while minimizing radiation effects to non-cancerous tissue.

Aftercare

If an applicator is to remain in place, there may be some discomfort in the area of the applicator. Medications may be prescribed to minimize the discomfort.

In some cases, patients may be required to remain on bedrest with limited movement during brachytherapy treatments. The goal is to maintain precise placement of the applicator and of the radioactive source to ensure the radiation remains targeted to the site of the cancer while minimizing radiation to surrounding tissue.

Risks

To minimize radiation exposure to others, some patients who have received internal radiation may be required to remain in the hospital so that radiation levels can be monitored. Visitors may not be allowed during this time and staff is allowed in the room for only short periods of time. Pregnant women and children are not allowed to come into contact with the patient because of the exposure to radiation.

Patients who have received permanent implants may emit small doses of radiation for several weeks or months after the implants are placed. The risk to others is minimal. However, the patient may be asked to have little or no contact with children or pregnant women, especially when the implant is first placed.

Results

The goal of treatment with radioactive implants is to treat the cancer. Normal cells in or near the treatment site are subjected to the effects of radiation; any tissue near the radiation site may be damaged or destroyed. Some side effects are acute and temporary, while others develop over time and may be permanent.

In general, as compared to radiation that is delivered using external beam radiation therapy, radiation delivered using brachytherapy techniques usually results in less toxicity and has fewer side effects since the radiation is precisely targeted and delivered directly to the tumor.

Resources

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Radioactive iodine uptake test see **Thyroid nuclear medicine scan**

Radioallergosorbent test (RAST) see **Allergy tests**

Radiotherapy see **Radiation therapy**

Raloxifene see **Bone disorder drugs**

Range-of-motion exercises see **Exercise**

Rape and sexual assault

Definition

The various definitions of rape range from the broad (coercing a person to engage in any sexual act) to the specific (forcing a woman to submit to sexual intercourse). The United States Code includes the crime of rape under the more comprehensive term “sexual abuse.” Two types of sexual assault are defined in the code: **sexual abuse** and aggravated sexual **abuse**. Sexual abuse includes acts in which an individual is forced to engage in sexual activity by use of threats or other fear tactics (and when the individual does not, or is unable to provide, consent), or instances in which an individual is physically unable to decline. Aggravated sexual abuse occurs when an individual is forced to submit to sexual acts by use of physical force; threats of **death**, injury, or kidnapping; or substances that

Victim response to a rape or sexual assault

- Don't blame yourself
- Seek immediate medical attention
- Don't bathe or change your clothing
- Retain all evidence
- Avoid urinating before seeing a doctor, especially if you think you've been drugged
- Consider the “morning-after” pill

SOURCE: U.S. Department of Health and Human Safety, Office on Women's Health.

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

render that individual unconscious or impaired. In both cases, the act may be completed or may only be only attempted, but it still considered a sexual assault.

Demographics

The rape or sexual attack can happen to anyone—women, men, or children. The attacker can be anyone—a stranger, friend or acquaintance, family member, former or current partner, or person that is in a position of trust, confidence, authority, or power (such as clergies, teachers, or superiors). A man most commonly commits rape onto a woman. However, rape can also occur by a woman onto a man. Rape performed by a man onto another man, or by a woman onto another woman is also a type of rape. Such instances typically occur in closed environments, such as in prisons. According to *The New York Times*, the typical rape victim is a 16 to 24 years old woman and the average sexual attacker, or rapist, is a 25 to 44 year old man. From data provided to law enforcement across the country, the Federal Bureau of Investigation (FBI) estimates that a forcible rape occurs every 5.8 minutes in the United States.

Description

Many misconceptions exist about rape and sexual assault. It is often assumed that rape victims are all women who have been attacked by a total stranger and forced into having sexual intercourse. In reality, sexual assault can take many forms—it may be violent or nonviolent; the victim may be male or female, child or adult; the offender may be a stranger, relative, friend, authority figure, or spouse.

The number of sexual assaults reported depends on how those abuses are defined. The United States Code uses two terms to distinguish between different sexual activities:

- Sexual act: contact between penis and vagina or penis and anus that involves penetration; contact between the mouth and genitals or anus; penetration of the vagina or anus with an object; or direct touching (not through clothing) of the genitals of an individual under the age of 16 years.
- Sexual contact: intentional touching of the genitals, breasts, buttocks, anus, inner thigh, or groin without sexual penetration.

National statistics

According to the FBI's *Uniform Crime Reports*, there were an estimated 89,000 forcible rapes reported to U.S. law enforcement agencies in 2008. The FBI reports that the figure is the lowest number in the last twenty years. When compared to the 2007 estimate,

the 2008 estimate was 1.6% less. Just under 58 out of every 100,000 women were reported to be victims of rape in 2008, down 2.4% from 2007 (when it was just above 59 per 100,000). The actual number of rapes and sexual assaults, however, is in reality much larger; estimates of unreported rape range between 2 and 10 times the number reported to law enforcement. The National Violence Against Women Survey, jointly sponsored by the Centers for Disease Control and Prevention (CDC) and the National Institute of Justice (NIJ) and conducted in the latter part of the 2000s, found that one in six women (18%) and one in 34 men (about 3%) has experienced an attempted or completed rape. The survey estimated that nearly 18 million women and almost 3 million men in the United States have been raped or have had rape attempted as a child or adult, and that more than 300,000 women and about 93,000 men are estimated to have been raped in any given year.

The survey also stated that only one in five adult women report rapes to the police. There are numerous reasons why the majority of sexual assaults are never reported. Often the victim fears retaliation from the offender. He or she may be afraid of family, friends, the community, or the media learning about the offense. There may be a concern about being judged or blamed by others. The victim may think that no one will believe the assault occurred.

THE VICTIMS. The "Victim, Incident, and Offender Characteristics" published by the National Center for Juvenile Justice (NCJJ), analyzed sexual assault data collected by law enforcement agencies over a five-year span. The following characteristics were found to be significant among victims of sexual assault:

- Age: More than two-thirds of reported victims of sexual assault were juveniles under the age of 18 years. Twelve to 18 year olds represented the largest group of victims at 33%; 20% were between the ages of six and 11; children less than five years old and adults between 18 and 24 years of age each constituted 14% of victims; 12% were between the ages of 25 and 34; and 7% were over the age of 34. Persons over the age of 54 represented 1% of all victims. One out of every seven victims surveyed in the study were under the age of six.
- Gender: Females were more than six times more likely to be a victim of sexual assault than males; more than 86% of victims were females. The great majority (99%) of the victims of forcible rapes were women, while men constituted the majority (54%) of the victims of forcible sodomy (oral or anal intercourse). Females are most likely to be the victim of

sexual assault at age 14, while males are at most risk at age four.

- Location: The residence of the victim was the most commonly noted location of sexual assault (70%). Other common locations included schools, hotels/motels, fields, woods, parking lots, roadways, and commercial/office buildings.
- Weapons: A personal weapon (hands, feet, or fists) was used in 77% of cases. No weapon was noted in 14% of assaults; other weapons (knives, clubs, etc.) were used in 6% of cases. Firearms were involved in only 2% of assaults.

THE OFFENDERS. Similar statistics were gathered by the NCJJ regarding the perpetrators of rape and sexual assault. These characteristics included:

- Age: More than 23% of offenders were under the age of 18 years; juveniles were more likely to be perpetrators of forcible sodomy and fondling. The remaining 77% of offenders were adults and were responsible for 67% of juvenile victims. For younger juvenile victims (under the age of 12), juvenile offenders were responsible for approximately 40% of assaults.
- Gender: The great majority of all reported offenders were male (96%). The number of female offenders rose for victims under the age of six (12%), in contrast to 6% for victims aged six through 12, 3% for victims aged 12 through 17, and 1% for adult victims.
- Relationship with offender: Approximately 59% of offenders were acquaintances of their victims, compared to family members (27%) or strangers (14%). Family members were more likely to be perpetrators against juveniles (34%) than against adults (12%). In contrast, strangers accounted for 27% of adult victims and 7% of juveniles.
- Past offenses: In 19% of juvenile cases, the victim was not the only individual to be assaulted by the offender, compared to only 4% of adult cases.

Consequences

Victims of sexual assault may sustain a range of injuries; male victims are more likely than females to suffer severe physical trauma. The National Women's Study, funded by the National Institute of Drug Abuse, found that more than 70% of rape victims did not report any physical injuries as a result of their assault; only 4% sustain serious injuries that require hospitalization. At least 49% of victims, however, state that they feared severe injuries or death during their assault. Fatalities occur in approximately 0.1% of rape cases.

Sexually transmitted diseases (STDs) are a source of concern for many victims of sexual assault. The most

commonly transmitted diseases are **gonorrhea** (caused by *Neisseria gonorrhoeae*), chlamydia (caused by *Chlamydia trachomatis*), **trichomoniasis** (caused by *Trichomonas vaginalis*), and **genital warts** (caused by human papillomavirus). **Syphilis** (caused by *Treponema pallidum*) and human **immunodeficiency** virus (HIV) are also noted among some sexual assault victims. The transmission rate of STDs is estimated to be between 3.6% and 30% of rapes.

According to the National Women's Study, approximately 5% of adult female rape victims become pregnant as a result of their assault, leading to 32,100 pregnancies a year among women 18 years of age or older. Approximately 50% of pregnant rape victims had an abortion, 6% put the child up for adoption, and 33% kept the child (the remaining pregnancies resulted in **miscarriage**).

MENTAL HEALTH PROBLEMS. Also known as rape trauma syndrome, **post-traumatic stress disorder** (PTSD) is a mental health disorder that describes a range of symptoms often experienced by someone who has undergone a severely traumatic event. Approximately 31% of rape victims develop PTSD as a result of their assault; victims are more than six times more likely to develop PTSD than women who have not been victimized.

The symptoms of PTSD include:

- recurrent memories or flashbacks of the incident
- nightmares
- insomnia
- mood swings
- difficulty concentrating
- panic attacks
- emotional numbness
- depression
- anxiety

Persons who have been sexually assaulted have also been noted to have increased risk for developing other mental health problems. Over those who have not been victimized, rape victims are:

- three times more likely to have a major depressive episode
- four times more likely to have contemplated suicide
- thirteen times more likely to develop alcohol dependency problems
- twenty-six times more likely to develop drug abuse problems

Causes and symptoms

There is not a conclusive explanation as to why some people rape and sexually assault others. Some explanations include: desire for power and dominance, anger and hostility, need to inflict **pain**, and sexual gratification. Some sociologists point to the evolution of males in their role to propagate the species as one reason for sexual assault; that is, if they cannot convince a woman to copulate (have sexual intercourse), then they attempt violent means to accomplish the act.

Symptoms vary after being raped or sexually assaulted. Some of the more common symptoms include:

- confusion
- withdrawing from social events
- nervousness
- crying without apparent reason
- hostilities
- fearfulness
- inappropriate behaviors

Diagnosis

Sometimes a rape or sexual assault victim will not initially tell the medical profession of the attack. They will visit the doctor for presumably other reasons. In other cases, the victim will admit to the attack. In the former case, the assault may never be known by the doctor, or may be identified as the examination progresses. In the latter case, the medical professional should be supportive of the victim and help in any way possible. Many larger medical facilities possess special teams to deal with the emotional, physical, and legal issues involved with such assaults.

Law enforcement officials recommend that rape and sexual assault victims go to the hospital immediately after the attack. Ideally, the visit of the hospital or medical facility should occur without changing clothing, showering, or urinating so that evidence left by the perpetrator will not be removed. Psychologists recommend that a friend be present to help support the victim. If not possible, a nurse or other professional is often provided to assist.

Information about the attack should be provided, such as date and time of the rape, the location, and what happened. The presence of members of the local law enforcement agency may be recommended or required. In other cases, the police may be made aware of the situation after the diagnosis is complete.

The medical professional should ask about any existing or previous illnesses or injuries, along with any current medications. If the information is not previously available, the doctor should ask the female

victim for the date of her last menstrual period, along with her gynecological history. If the victim is a female, the possibility of **pregnancy** should be considered, both before and after the attack. A complete **physical examination** should be performed, including analysis of any suspected trauma or injury to the body. Samples of clothing, public hair, and fingernail scrapings may be taken. Evidence of sperm within the body's orifices may also be collected by the doctor. Tests for sexually transmitted diseases will also be taken.

Treatment

Once a victim of sexual assault reports the crime to local authorities, calls a rape crisis hotline, or arrives at the emergency room to be treated for injuries, a multidisciplinary team is often formed to address his or her physical, psychological, and judicial needs. This team usually includes law enforcement officers, physicians, nurses, mental health professionals, victim advocates, and/or prosecutors.

The victim of sexual assault may continue to feel fear and **anxiety** for some time after the incident, and in some instances this may significantly impact his or her personal or professional life. Follow-up counseling should therefore be provided for the victim, particularly if symptoms of PTSD become evident.

If the rapist has the possibility of being infected with HIV (human immunodeficiency virus), the doctor may recommend that an antiretroviral medication, generally called a post-exposure **prophylaxis** (PEP), be used to reduce the chance of infection.

After the examination is complete, the medical professional may also recommend the victim be referred to a local rape crisis center for further advice, counseling, and information. Medications to relieve symptoms, such as for depression and anxiety, are often also prescribed.

Forensic medical examination

Because rape is a crime, there are certain requirements for medical evaluation of the patient and for record keeping. The forensic medical examination is an invaluable tool for collecting evidence against a perpetrator that may be admissible in court. Since the great majority of victims know their assailant, the purpose of the medical examination is often not to establish identity but to establish nonconsensual sexual contact. The Sexual Assault Nurse Examiner program is an effective model that is used in many U.S. hospitals and clinics to collect and document evidence, evaluate and treat for STDs and pregnancy, and refer victims to follow-up medical care and counseling. The "Sexual Assault Nurse Examiner Development and Operation

Guide,” prepared by the Sexual Assault Resource Service, describes the ideal protocol for collecting evidence from a sexual assault victim. This includes:

- performing the medical examination within 72 hours of the assault
- taking a history of the assault
- documenting the general health of the victim, including menstrual cycle, potential allergies, and pregnancy status
- assessment for trauma and taking photographic evidence of injuries
- taking fingernail clippings or scrapings
- taking samples for sperm or seminal fluid
- combing head/pubic hair for foreign hairs, fibers, and other substances
- collection of bloody, torn, or stained clothing
- taking samples for blood typing and DNA screening

Prognosis

Emotional and health problems may arise after the attack has occurred. It is important to seek help after being assaulted. A medical care facility or hospital may be such a place to first seek assistance. A safe house (a place that is free of danger, safe from further abuse) may be contacted as a place to stay. Dialing 911 on the telephone will also bring assistance. Counseling is often helpful in dealing with the situation and its consequences. The prognosis for rape and sexual assault victims varies. Its outcome is more positive when the victim realizes that the fault lies with the attacker, not themselves.

Two phases after the assault are common. The acute phase occurs immediately after the assault. Here, the victim feels the physical pain and the mental emotions of the attack. The victim must cope with the situation over the first few days of the attack. The reorganization phase occurs about one week or so after the attack. It may last for several months or years. The victim continues to cope with the reality of the attack and how it affects one’s life. Psychological studies have shown that both phases can be handled better when **psychotherapy** and other counseling is provided to the victim.

Some victims never recover emotionally from the attack. Complications from PTSD often occur, such as nightmares, flashbacks, depression and anxiety, and inappropriate or deadened emotions. Alcohol or **substance abuse** may occur. Relationships with friends and family may also be adversely affected. Suicidal tendencies also happen. Therapies and medications usually help the victim recover.

Prevention

The prevention of rape and sexual assault can be achieved, according to law enforcement agencies, by education—making people aware of the possibility that rape and sexual assault can happen to anyone. The police suggest the following to minimize the risk of rape and sexual assault:

- Secure all home windows and doors with sturdy locks and other safety devices. Home security companies also provide security systems that can be installed
- Stay away from isolated or secluded areas when alone and outside (especially at night)
- Lock all car doors while driving and be aware of the immediate surroundings while driving and getting into and out of the vehicle
- Sit as near to the driver when taking public transportation
- Carry items that can help to alert others (such as whistles and personal alarms)
- Carry items that can provide defense if attacked (such as pepper spray)
- Do not hitchhike in any situation
- If having vehicle problems, call for assistance and wait inside the vehicle until help arrives
- Take a course in self-defense; know how to defend oneself

STD Prevention

While the concern of sexual assault victims of contracting an STD is often high, the actual risk of transmission is relatively low. The CDC estimates that the risk of contracting gonorrhea from an offender is between six and 12%, chlamydia between four and 17%, syphilis between 0.5 and 3%, and HIV less than 1%. Nonetheless, post-exposure prophylaxis (preventative treatment) against certain STDs is often provided for the victim. Treatment with zidovudine, for example, is recommended for individuals who are at a high risk of exposure to HIV. The CDC recommends the following prophylactic regimen be provided for victims of sexual assaults in which vaginal, oral, or anal penetration took place:

- a single dose of ceftriaxone, an antibiotic effective against *Neisseria gonorrhoeae*
- a single dose of metronidazole, an antibiotic effective against *Trichomonas vaginalis*
- a single dose of azithromycin or doxycycline, antibiotics effective against *Chlamydia trachomatis*
- inoculation with the post-exposure hepatitis B vaccine

In some instances, cultures may be taken during the medical examination and at time points afterward to test for gonorrhea or chlamydia. It is important that

KEY TERMS

Aggravated sexual abuse—When an individual is forced to submit to sexual acts by use of physical force; threats of death, injury, or kidnapping; or substances that render that individual unconscious or impaired.

Forcible sodomy—Forced oral or anal intercourse.

Forensic—Pertaining to or used during legal proceedings.

Post-traumatic stress disorder (PTSD)—Also known as rape trauma syndrome; a mental health disorder that describes a range of symptoms often experienced by someone who has undergone a severely traumatic event.

Sexual abuse—When an individual is forced to engage in sexual activity by use of threats or other fear tactics, or instances in which an individual is physically unable to refuse.

Sexual assault nurse examiner (SANE)—A registered nurse who is trained to collect and document evidence from a sexual assault victim, evaluate and treat for STDs and pregnancy, and refer victims to follow-up medical care and counseling.

Yupze regimen—A form of emergency contraception in which two oral contraceptive pills that contain both of the hormones estrogen and progestin are taken to prevent pregnancy.

the victim receive information regarding the symptoms of STDs and be counseled to return for further examination if any of these symptoms occur.

Pregnancy prevention

Female victims at risk for becoming pregnant after an assault should be counseled on the availability of **emergency contraception**. According to the Food and Drug Administration (FDA), emergency **contraception** is not effective if there is a pregnancy does not exist but works to prevent pregnancy from occurring by delaying or preventing ovulation, by affecting the transport of sperm, and/or by thinning the inner layer of the uterus (endometrium) so that implantation is prevented. It is therefore not a form of abortion.

A number of options are available for women if they choose to use emergency contraceptives to prevent pregnancy following a sexual assault. The Yupze regimen uses two oral contraceptive pills that contain the hormones estrogen and progestin. The risk of pregnancy is reduced by 75% after use of the Yupze regimen, reducing the average number of pregnancies after unprotected sex from eight in 100 to two in 100. Progestin-only **oral contraceptives** are also available and reduce the risk of pregnancy by 89 to 95%.

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ORGANIZATIONS

American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209-3901, (703) 907-7300, apa@psych.org, <http://www.psych.org>.
 Federal Bureau of Investigation, J. Edgar Hoover Building, 935 Pennsylvania Avenue NW, Washington, DC, 20535-0001, (202) 324-3000, <http://www.fbi.gov>.
 Office for Victims of Crime, U.S. Department of Justice, 810 7th Street NW, Washington, DC, 20531, (202) 544-1034, (800) 656-4673, <http://www.ojp.usdoj.gov/ovc/>.
 Rape, Abuse, and Incest National Network, 2000 L Street NW, Washington, DC, 20036, (202) 544-1034, (800) 656-4673, info@rainn.org, <http://www.rainn.org/>.

Stéphanie Dionne
 Rebecca J. Frey, PhD

Rashes

Definition

The popular term for a group of spots or red, inflamed skin that is usually a symptom of an underlying condition or disorder. Often temporary, a rash is only rarely a sign of a serious problem.

Description

A rash may occur on only one area of the skin, or it could cover almost all of the body. Also, a rash may or may not be itchy. Depending on how it looks, a rash may be described as:

- blistering (raised oval or round collections of fluid within or beneath the outer layer of skin)
- macular (flat spots)
- nodular (small, firm, knotty rounded mass)
- papular (small solid slightly raised areas)
- pustular (pus-containing skin blister)

Causes and symptoms

There are many theories as to the development of skin rashes, but experts are not completely clear what causes some of them. Generally a skin rash is an intermittent symptom, fading and reappearing. Rashes may accompany a range of disorders and conditions, such as:

- Infectious illness. A rash is symptom of many different kinds of childhood infectious illnesses, including chickenpox and scarlet fever. It may be triggered by other infections, such as Rocky Mountain spotted fever or ringworm.



An unidentified rash on a young boy's back. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

- Allergic reactions. One of the most common symptoms of an allergic reaction is an itchy rash. Contact dermatitis is a rash that appears after the skin is exposed to an allergen, such as metal, rubber, some cosmetics or lotions, or some types of plants (e.g. poison ivy). Drug reactions are another common allergic cause of rash; in this case, a rash is only one of a variety of possible symptoms, including fever, seizures, nausea and vomiting, diarrhea, heartbeat irregularities, and breathing problems. This rash usually appears soon after the first dose of the course of medicine is taken.
- Autoimmune disorders. Conditions in which the immune system turns on the body itself, such as systemic lupus erythematosus or purpura, often have a characteristic rash.
- nutritional disorders. For example, scurvy, a disease caused by a lack of Vitamin C, has a rash as one of its symptoms.

KEY TERMS

Purpura—A group of disorders characterized by purple or red brown areas of discoloration visible through the skin.

Scurvy—A nutritional disorder that causes skin bruising and hemorrhages.

- cancer. A few types of cancer, such as chronic lymphocytic leukemia, can be the underlying cause of a rash.

Rashes in infancy

Rashes are extremely common in infancy, and are usually not serious at all and can be treated at home.

Diaper rash is caused by prolonged skin contact with bacteria and the baby's waste products in a damp diaper. This rash has red, spotty sores and there may be an ammonia smell. In most cases the rash will respond within three days to drying efforts. A diaper rash that does not improve in this time may be a yeast infection requiring prescription medication. A doctor should be consulted if the rash is solid, bright red, causes **fever**, or the skin develops blisters, **boils**, or pus.

Infants also can get a rash on cheeks and chin caused by contact with food and stomach contents. This rash will come and go, but usually responds to a good cleaning after meals. About a third of all infants develop "acne" usually after the third week of life in response to their mothers' hormones before birth. This rash will disappear between weeks and a few months. Heat rash is a mass of tiny pink bumps on the back of the neck and upper back caused by blocked sweat glands. The rash usually appears during hot, humid weather, although a baby with a fever can also develop the rash.

A baby should see a doctor immediately if the rash:

- appears suddenly and looks purple or blood-colored
- looks like a burn
- appears while the infant seems to be sick

Diagnosis

A physician can make a diagnosis based on the medical history and the appearance of the rash, where it appears, and any other accompanying symptoms.

Treatment

Treatment of rashes focuses on resolving the underlying disorder and providing relief of the **itching** that often accompanies them. Soothing lotions or oral **antihistamines** can provide some relief, and **topical antibiotics** may be administered if the patient, particularly a child, has caused a secondary infection by scratching. The rash triggered by **allergies** should disappear as soon as the allergen is removed; drug rashes will fade when the patient stops taking the drug causing the allergy. For the treatment of diaper rash, the infant's skin should be exposed to the air as much as possible; ointments are not needed unless the skin is dry and cracked. Experts also recommend switching to cloth diapers and cleaning affected skin with plain water.

Prognosis

Most rashes that have an acute cause, such as an infection or an allergic reaction, will disappear as soon as the infection or irritant is removed from the body's system. Rashes that are caused by chronic conditions, such as **autoimmune disorders**, may remain indefinitely or fade and return periodically.

Prevention

Some rashes can be prevented, depending on the triggering factor. A person known to be allergic to certain drugs or substances should avoid those things in order to prevent a rash. Diaper rash can be prevented by using cloth diapers and keeping the diaper area very clean, breast feeding, and changing diapers often.

ORGANIZATIONS

American Academy of Dermatology, P.O. Box 4014, Schaumburg, IL, 60168-4014, 847 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

Carol A. Turkington

Rat-bite fever

Definition

Rat-bite fever refers to an infection which develops after having been bitten or scratched by an infected animal.

Description

Rat-bite fever occurs most often among laboratory workers who handle lab rats in their jobs, and among people who live in poor conditions, with rodent infestation. Children are particularly likely to be bitten by rodents infesting their home, and are therefore most likely to contract rat-bite fever. Other animals that can carry the types of bacteria responsible for this illness include mice, squirrels, weasels, dogs, and cats. One of the causative bacteria can cause the same illness if it is ingested, for example in unpasteurized milk.

Causes and symptoms

There are two variations of rat-bite fever, caused by two different organisms. In the United States, the bacteria *Streptobacillus moniliformis* is the most common cause (causing streptobacillary rat-bite fever). In other countries, especially Africa, *Spirillum minus* causes a different form of the infection (called spirillary rat-bite fever).

Streptobacillary rat-bite fever occurs up to 22 days after the initial bite or scratch. The patient becomes ill with fever, chills, **nausea and vomiting**, **headache**, and **pain** in the back and joints. A rash made up of tiny pink bumps develops, covering the palms of the hands and the soles of the feet. Without treatment, the patient is at risk of developing serious infections of the lining of the heart (**endocarditis**), the sac containing the heart (**pericarditis**), the coverings of the brain and spinal cord (**meningitis**), or lungs (**pneumonia**). Any tissue or organ throughout the body may develop a pocket of infection and pus, called an **abscess**.

Spirillary rat-bite fever occurs some time after the initial injury has already healed, up to about 28 days after the bite or scratch. Although the wound had appeared completely healed, it suddenly grows red and swollen again. The patient develops a fever. Lymph nodes in the area become swollen and tender, and the patient suffers from fever, chills, and headache. The skin in the area of the original wound sloughs off. Although rash is less common than with streptobacillary rat-bite fever, there may be a lightly rosy, itchy rash all over the body. Joint and muscle pain rarely occur. If left untreated, the fever usually subsides, only to return again in repeated two- to four-day cycles. This can go on for up to a year, although, even without treatment, the illness usually resolves within four to eight weeks.

Diagnosis

In streptobacillary rat-bite fever, found in the United States, diagnosis can be made by taking a sample of blood or fluid from a painful joint. In a

KEY TERMS

Abscess—A pocket of infection; a collection of pus.

Endocarditis—An inflammation of the lining of the heart.

Meningitis—An inflammation of the tissues covering the brain and spinal cord.

Pasteurization—A process during which milk is heated up and maintained at a particular temperature long enough to kill bacteria.

Pericarditis—An inflammation of the sac containing the heart.

laboratory, the sample can be cultured, to allow the growth of organisms. Examination under a microscope will then allow identification of the bacteria *Streptobacillus moniliformis*.

In spirillary rat-bite fever, diagnosis can be made by examining blood or a sample of tissue from the wound for evidence of *Spirillum minus*.

Treatment

Shots of procaine penicillin G or penicillin V by mouth are effective against both streptobacillary and spirillary rat-bite fever. When a patient is allergic to the **penicillins**, erythromycin may be given by mouth for streptobacillary infection, or tetracycline by mouth for spirillary infection.

Prognosis

With treatment, prognosis is excellent for both types of rat-bite fever. Without treatment, the spirillary form usually resolves on its own, although it may take up to a year to do so.

The streptobacillary form, found in the United States, however, can progress to cause extremely serious, potentially fatal complications. In fact, before **antibiotics** were available to treat the infection, streptobacillary rat-bite fever frequently resulted in **death**.

Prevention

Prevention involves avoiding contact with those animals capable of passing on the causative organisms. This can be an unfortunately difficult task for people whose economic situations do not allow them to move out of rat-infested buildings. Because streptobacillary rat-bite fever can occur after drinking contaminated

milk or water, only pasteurized milk, and water from safe sources, should be ingested.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Rosalyn Carson-DeWitt, MD

Rational-emotive therapy see
Cognitive-behavioral therapy

Raynaud's disease

Definition

Raynaud's disease refers to a disorder in which the fingers or toes (digits) suddenly experience decreased blood circulation. It is characterized by repeated episodes of color changes of the skin of digits on cold exposure or emotional stress.

Demographics

Women are five times more likely than men to develop primary Raynaud's disease. The average age of diagnosis is between 20 and 40 years. Approximately three out of ten people with primary Raynaud's disease eventually progress to secondary Raynaud's disease after diagnosis. About 15% of individuals improve.



A close-up view of a patient's fingers afflicted with Raynaud's disease. While this disorder may initially only affect the tips of the fingers and toes, eventually blood circulation of the entire finger or toe is affected. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

Raynaud's disease can be classified as one of two types: primary (or idiopathic) and secondary (also called Raynaud's phenomenon). Primary Raynaud's disease has no predisposing factor, is more mild, and causes fewer complications. About half of all cases of Raynaud's disease are of this type.

Secondary Raynaud's disease is the same as primary Raynaud's disease, but occurs in individuals with a predisposing factor, usually a form of collagen **vascular disease**. What is typically identified as primary Raynaud's is later identified as secondary once a predisposing disease is diagnosed. This occurs in approximately 30% of patients. As a result, the secondary type is often more complicated and severe, and is more likely to worsen.

Several related conditions that predispose persons to secondary Raynaud's disease include **scleroderma**, **systemic lupus erythematosus**, **rheumatoid arthritis**, and **polymyositis**. **Pulmonary hypertension** and some nervous system disorders such as herniated discs and tumors within the spinal column, strokes, and **polio** can progress to Raynaud's disease. Finally, injuries due to mechanical trauma caused by vibration (such as that associated with chain saws and jackhammers), repetitive motion (**carpal tunnel syndrome**), electrical shock, and exposure to extreme cold can lead to the development of Raynaud's disease. Some drugs used to control high blood pressure or migraine headaches have been known to cause Raynaud's disease.

The prevalence of Raynaud's phenomena in the general population varies 4-15%. Females are seven times more likely to develop Raynaud's diseases than are men. The problem has not been correlated with coffee consumption, dietary habits, occupational history (excepting exposure to vibration), or exposure to most drugs. An association between Raynaud's disease and migraine headaches has been reported. Secondary Raynaud's disease is common among individuals with systemic lupus erythematosus in tropical countries.

Causes and symptoms

There is significant familial aggregation of primary Raynaud's disease. However, no causative gene has been identified.

Risk factors

Risk factors for Raynaud's disease differ between males and females. Age and **smoking** seem to be associated with Raynaud's disease only in men, while the associations of marital status and alcohol use with Raynaud's disease are usually only observed in

KEY TERMS

Arteriole—The smallest type of artery.

Artery—A blood vessel that carries blood away from the heart to peripheral tissues.

Gangrene—Death of a tissue, usually caused by insufficient blood supply and followed by bacterial infection of the tissue.

Idiopathic—Of unknown origin.

Polymyositis—An inflammation of many muscles.

Pulmonary hypertension—A severe form of high blood pressure caused by diseased arteries in the lung.

Rheumatoid arthritis—Chronic, autoimmune disease marked by inflammation of the membranes surrounding joints.

Scleroderma—A relatively rare autoimmune disease affecting blood vessels and connective tissue that makes skin appear thickened.

Systemic lupus erythematosus—A chronic inflammatory disease that affects many tissues and parts of the body including the skin.

women. These findings suggest that different mechanisms influence the expression of Raynaud's disease in men and women.

Both primary and secondary Raynaud's disease signs and symptoms are thought to be due to arterioles over-reacting to stimuli. Cold normally causes the tiny muscles in the walls of arteries to contract, thus reducing the amount of blood that can flow through them. In people with Raynaud's disease, the extent of constriction is extreme, thus severely restricting blood flow. Attacks or their effects may be brought on or worsened by **anxiety** or emotional distress.

There are three distinct phases to an episode of Raynaud's disease. When first exposed to cold, small arteries respond with intense contractions (vasoconstriction). The affected fingers or toes (in rare instances, the tip of the nose or tongue) become pale and white because they are deprived of blood and, thus, oxygen. In response, capillaries and veins expand (dilate). Because these vessels are carrying deoxygenated blood, the affected area then becomes blue in color. The area often feels cold and tingly or numb. After the area begins to warm up, the arteries dilate. Blood flow is significantly increased. This changes the color of the area to a bright red. During this phase, persons often describe the affected area as feeling warm and throbbing painfully.

Raynaud's disease may initially affect only the tips of fingers or toes. As the disease progresses, it may eventually involve all of one or two digits. Ultimately, all the fingers or toes may be affected. About one person in ten, will experience a complication called sclerodactyly. In sclerodactyly, the skin over the involved digits becomes tight, white, thick, smooth and shiny. In approximately 1% of cases of Raynaud's disease, deep sores (ulcers) may develop in the skin. In rare cases of frequent, repetitive bouts of severe **ischemia** (decreased supply of oxygenated blood to tissues or organs), tissue loss, or **gangrene** may result and **amputation** may be required.

Diagnosis

Examination

Primary Raynaud's disease is diagnosed following the Allen Brown criteria. There are four components. The certainty of the diagnosis and severity of the disease increase as more criteria are met. The first is that at least two of the three color changes must occur during attacks provoked by cold and or stress. The second is that episodes must periodically occur for at least two years. The third is that attacks must occur in both the hands and the feet in the absence of vascular occlusive disease. The last is that there is no other identifiable cause for the Raynaud's episodes.

Tests

A cold stimulation test may also be performed to help confirm a diagnosis of Raynaud's disease. The temperature of affected fingers or toes is taken. The hand or foot is then placed completely into a container of ice water for 20 seconds. After removal from the water, the temperature of the affected digits is immediately recorded. The temperature is retaken every five minutes until it returns to the pre-immersion level. Most individuals recover normal temperature within 15 minutes. People with Raynaud's disease may require 20 minutes or more to reach their pre-immersion temperature. However, these results are often inconclusive for several reasons. Provocative testing such as the cold stimulation test, is difficult to interpret because there is considerable overlap between normal and abnormal results.

Laboratory testing is performed frequently. The **antinuclear antibody test** of blood is usually negative in Raynaud's disease. Capillary beds under finger nails usually appear normal. Erythrocyte sedimentation rates are often abnormal in people with connective tissue diseases. Unfortunately, this finding is not consistent in people with Raynaud's disease.

Treatment

There is no known cure for this condition. Avoidance of the trigger is the best supportive management available. Most cases of primary Raynaud's disease can be controlled with proper medical care and avoidance.

Drugs

People with severe cases of Raynaud's disease may need to be treated with medications to help keep the arterioles relaxed and dilated. Medications such as calcium-channel blockers, reserpine, or nitroglycerin may be prescribed to relax artery walls and improve blood flow.

Alternative

Because episodes of Raynaud's disease have been associated with stress and emotional upset, the condition may be improved by learning to manage stress. Regular **exercise** is known to decrease stress and lower anxiety. Hypnosis, relaxation techniques, and visualization are also useful methods to help control emotions.

Biofeedback training is a technique during which a patient is given continuous information on the temperature of his or her digits, and then taught to voluntarily control this temperature. Some alternative practitioners believe that certain dietary supplements and herbs may be helpful in decreasing the vessel spasm of Raynaud's disease. Suggested supplements include vitamin E (found in fruits, vegetables, seeds, and nuts), magnesium (found in seeds, nuts, fish, beans, and dark green vegetables), and fish oils. The circulatory herbs cayenne, ginger and prickly ash may help enhance circulation to affected areas.

Home remedies

Many people are able to find relief by simply adjusting their lifestyles. Affected individuals need to stay warm, and keep their hands and feet well covered in cold weather. Layered clothing, scarves, heavy coats, heavy socks, and mittens under gloves are suggested because gloves alone allow heat to escape. It is also recommended that patients cover or close the space between their sleeves and mittens. Indoors, they should wear socks and comfortable shoes. Smokers should quit as nicotine will worsen the problem. Avoid the use of vibrating tools as well.

Prognosis

The prognosis for most people with Raynaud's disease is very good. In general, primary Raynaud's disease has the best prognosis, with a relatively small chance

(1%) of serious complications. Approximately half of all affected individuals do well by taking simple precautions, and never require medication. The prognosis for people with secondary Raynaud's disease (or phenomenon) is less predictable. This prognosis depends greatly on the severity of other associated conditions such as scleroderma, lupus, or Sjögren syndrome.

Prevention

There is no way to prevent the development of Raynaud's disease. Once an individual realizes that he or she has the disorder, however, steps can be taken to reduce the frequency and severity of episodes.

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ORGANIZATIONS

- American College of Rheumatology, 2200 Lake Boulevard NE, Atlanta, GA, 30319, (404) 633-3777, (404) 633-1870, acr@rheumatology.org, <http://www.rheumatology.org>.

American Heart Association, 7272 Greenville Ave., Dallas, TX, 75231-4596, (214) 373-6300, (800) 242-8721, inquire@heart.org, <http://www.heart.org>.

Association of Applied Psychophysiology and Biofeedback, 10200 W. 44th Avenue, Suite 304, Wheat Ridge, CO, 80033, (303) 422-8436, (800) 477-8892, aapb@resourcecenter.com, <http://www.aapb.org>.

Biofeedback Certification International Alliance, 10200 W. 44th Avenue, Suite 310, Wheat Ridge, CO, 80033, (303) 420-2902, (866) 908-8713, info@bcia.org, <http://www.bcia.org>.

Irish Raynaud's and Scleroderma Society, P.O. Box 2958 Foxrock, Dublin, Ireland, 18, (01) 2020184, info@irishraynauds.com, <http://www.irishraynauds.com>.

National Heart, Lung, and Blood Institute, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov>.

National Institute of Arthritis and Musculoskeletal and Skin Disease Information Clearinghouse, National Institutes of Health, 1 AMS Circle, Bethesda, MD, 20892-3675, (877) 226-4267, NIAMSinfo@mail.nih.gov, <http://www.niams.nih.gov>.

National Organization for Rare Disorders (NORD), 55Kensosia Ave., P.O. Box 1968, Danbury, CT, 06813, (203) 744-0100, (800) 999-6673, (203) 798-2291, orphan@rarediseases.org, <http://www.rarediseases.org>.

Raynaud's & Scleroderma Association (UK), 112 Crewe Road, AlsagerCheshire, UK, ST7 2JA, 01270 872776, info@raynauds.org.uk, <http://www.raynauds.org.uk>.

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RDS see **Respiratory distress syndrome**

Reactive airway disease see **Asthma**

Reactive polycythemia see **Secondary polycythemia**

Reading disorder see **Learning disorders**

Recompression treatment

Definition

Recompression treatment is the use of elevated pressure to treat conditions within the body after it has been subjected to a rapid decrease in pressure. It also includes hyperbaric **oxygen** therapy.

Purpose

Recompression treatment is used to overcome the adverse effects of **gas embolism** and **decompression sickness** (sometimes called the bends) in underwater

divers who breathe compressed air. It is also approved for treatment of severe **smoke inhalation**, **carbon monoxide poisoning**, gas **gangrene**, radiation tissue damage, thermal **burns**, extreme blood loss, crush injuries, and **wounds** that won't heal.

Precautions

Hyperbaric oxygen therapy delivers greater amounts of oxygen more quickly to the body than breathing room air (which is only 21% oxygen) at regular pressure. Unmonitored, increased oxygen can produce toxic effects. Treatments must follow safe time-dose limits and may only be administered by a doctor.

Description

Recompression treatment is performed in a **hyperbaric chamber**, a sealed compartment in which the patient breathes normal air or "enhanced" air with up to 100% oxygen while exposed to controlled pressures up to three times normal atmospheric pressure. The patient may receive the oxygen through a face mask, a hood or tent around the head, or an endotracheal tube down the windpipe if the patient is already on a ventilator. When used to treat decompression sickness or gas **embolism**, the increased pressure reduces the size of gas bubbles in the patient's body. The increased oxygen concentration speeds the diffusion of the nitrogen within the bubbles out of the patient's body. As gas bubbles deflate, the trauma of gas embolism and decompression sickness begins to resolve. Treatment for diving emergencies typically involves one session, lasting four to six hours, at three atmospheres of pressure.

When used to treat other conditions, the increased pressure allows oxygen and other gases to dissolve more rapidly into the blood and thus be carried to oxygen-starved tissues to enhance healing. Elevated oxygen levels can also purge toxins such as carbon monoxide from the body. In addition, when body tissues are super-saturated with oxygen, the destruction of some bacteria is enhanced and the spread of certain toxins is halted. This makes hyperbaric oxygen therapy useful in treating gas gangrene and infections that cause tissue necrosis (death). Hyperbaric oxygen therapy also promotes the growth of new blood vessels.

Preparation

Oxygen is often administered to a patient as **first aid** while he or she is being transported to a hyperbaric chamber. The treatment begins with chamber compression; as the pressure of the chamber atmosphere increases, the temperature also rises and the patient's ears may fill as they would during an airplane landing. Swallowing and yawning are ways to relieve the inner

KEY TERMS

Atmosphere—A measurement of pressure. One atmosphere equals the pressure of air at sea level (14.7 pounds per square inch [psi]).

Compressed air—Air that is held under pressure in a tank to be breathed underwater by divers. A tank of compressed air is part of a diver's scuba (self-contained underwater breathing apparatus) gear.

Decompression—A decrease in pressure from the surrounding water that occurs with decreasing diving depth.

Decompression sickness—A condition found in divers in which gas bubbles of nitrogen form in tissues and blood vessels as a result of decreasing surrounding pressure, such as in ascent from a dive. It may be a painful condition, especially as nitrogen

bubbles invade the joints; persons stricken may walk stooped over in pain, in a bent stance that led to it being called "the bends."

Gas embolism—The presence of gas bubbles in the bloodstream that obstruct circulation. Also called air embolism.

Hyperbaric chamber—A sealed compartment in which patients are exposed to controlled pressures up to three times normal atmospheric pressure. Hyperbaric treatment may be used to regulate blood gases, reduce gas bubbles, and provide higher levels of oxygen more quickly.

Recompression—Restoring the elevated pressure of the diving environment to treat decompression sickness and gas embolism by decreasing bubble size.

ear pressure. Once the desired pressure is achieved, the patient is given pure oxygen to breathe. Because treatment is lengthy, patients are encouraged to sleep or listen to music. In larger chambers, patients may also read or watch videos.

Aftercare

Depending on the reason for treatment and the treatment outcome, the patient may be taken to a hospital for further care, or examined and released.

Risks

There is minimal risk when recompression treatment is administered by a competent physician. However, some common side effects are sinus **pain**, temporary changes in vision, and **fatigue**.

Normal results

With prompt and appropriate recompression treatment, most patients show marked improvement in their blood oxygen levels and tissue circulation, as well as other signs of healing. Divers treated for gas embolism or decompression sickness may recover with no lasting effects.

Abnormal results

When recompression treatment is not begun promptly or not conducted at adequate time-dose levels, patients with decompression sickness may develop bone necrosis. This significant destruction of bone, most commonly found in the hip and shoulder, produces chronic

pain and severe disability. Another result of delayed or inadequate treatment may be permanent neurological damage. When decompression sickness involves the spinal cord, partial **paralysis** may occur.

ORGANIZATIONS

American College of Hyperbaric Medicine, 9875 South Franklin Drive, Suite 300, Franklin, Wisconsin, 53132, (414) 385-2943, (414) 385-8721, <http://www.achm.org>.

Divers Alert Network, 6 West Colony Place, Durham, NC, 27705, 919 684-2948, 919 490-6630, (800) 446-2671, <http://www.diversalertnetwork.org>.

Undersea and Hyperbaric Medical Society, 21 West Colony Place, Suite 280, Durham, NC, 27705, (919) 490-5140, (919) 490-5149, (877) 533-UHMS (8467), uhms@uhms.org, <http://www.uhms.org>.

Bethany Thivierge

Reconstructive surgery see **Plastic, cosmetic, and reconstructive surgery**

Rectal cancer

Definition

The rectum is the portion of the large bowel that lies in the pelvis, terminating at the anus. **Cancer** of the rectum is the disease characterized by the development

of malignant cells in the lining or epithelium of the rectum. Malignant cells have changed such that they lose normal control mechanisms governing growth. These cells may invade surrounding local tissue or they may spread throughout the body and invade other organ systems.

Description

The rectum is the continuation of the colon (part of the large bowel) after it leaves the abdomen and descends into the pelvis. It is divided into equal thirds: the upper, mid, and lower rectum.

The pelvis and other organs in the pelvis form boundaries to the rectum. Behind, or posterior to the rectum is the sacrum (the lowest portion of the spine, closest to the pelvis). Laterally, on the sides, the rectum is bounded by soft tissue and bone. In front, the rectum is bounded by different organs in the male and female. In the male, the bladder and prostate are present. In the female, the vagina, uterus, and ovaries are present.

The upper rectum receives its blood supply from branches of the inferior mesenteric artery from the abdomen. The lower rectum has blood vessels entering from the sides of the pelvis. Lymph, a protein-rich fluid that bathes the cells of the body, is transported in small channels known as lymphatics. These channels run with the blood supply of the rectum. Lymph nodes are small filters through which the lymph flows on its way back to the blood stream. Cancer spreads elsewhere in the body by invading the lymph and vascular systems.

When a cell or cells lining the rectum become malignant, they first grow locally and may invade partially or totally through the wall of the rectum. The tumor here may invade surrounding tissue or the organs that bound it, a process known as local invasion. In this process, the tumor penetrates and may invade the lymphatics or the capillaries locally and gain access to the circulation in this way. As the malignant cells work their way to other areas of the body, they again become locally invasive in the new area to which they have spread. These tumor deposits, originating in the primary tumor in the rectum, are then known as metastasis. If metastases are found in the regional lymph nodes, they are known as regional metastases. If they are distant from the primary tumor, they are known as distant metastases. The patient with distant metastases may have widespread disease, also referred to as systemic disease. Thus the cancer originating in the rectum begins locally and, given time, may become systemic.

By the time the primary tumor is originally detected, it is usually larger than 1 cm (about 0.39 in) in size and has over one million cells. This amount of

growth is estimated to take about three to seven years. Each time the cells double in number, the size of the tumor quadruples. Thus like most cancers, the part that is identified clinically is later in the progression than would be desired. Screening becomes a very important endeavor to aid in earlier detection of this disease.

Passage of red blood with the stool, (noticeable bleeding with defecation), is much more common in rectal cancer than that originating in the colon because the tumor is much closer to the anus. Other symptoms (**constipation** and/ or **diarrhea**) are caused by obstruction and, less often, by local invasion of the tumor into pelvic organs or the sacrum. When the tumor has spread to distant sites, these metastases may cause dysfunction of the organ they have spread to. Distant metastasis usually occurs in the liver, less often to the lung(s), and rarely to the brain.

There are about 36,500 cases of rectal cancer diagnosed per year in the United States. Together, colon and rectal cancers account for 10% of cancers in men and 11% of cancers in women. It is the second most common site-specific cancer affecting both men and women. Nearly 57,000 people died from colon and rectal cancer in the United States in 2003. In recent years the incidence of this disease is decreasing very slightly, as has the mortality rate. It is difficult to tell if the decrease in mortality reflects earlier diagnosis, less **death** related to the actual treatment of the disease, or a combination of both factors.

Cancer of the rectum is felt to arise sporadically in about 80% of those who develop the disease. About 20% of cases probably arise from genetic predisposition; some people have a family history of rectal cancer occurring in a first-degree relative. Development of rectal cancer at an early age suggests a genetically transmitted form of the disease as opposed to the sporadic form.

Causes and symptoms

Causes of rectal cancer are probably environmental in sporadic cases (80%), and genetic in the heredity-predisposed (20%) cases. Since malignant cells have a changed genetic makeup, this means that in 80% of cases, the environment spontaneously induces change. Those born with a genetic predisposition are either destined to get the cancer, or it will take less environmental exposure to induce the cancer. Exposure to agents in the environment that may induce mutation is the process of carcinogenesis and is caused by agents known as **carcinogens**. Specific carcinogens have been difficult to identify; dietary factors, however, seem to be involved.

Rectal cancer is more common in industrialized nations. Dietary factors may be the reason. **Diets** high in fat, red meat, total calories, and alcohol seem to add to increased risk. Diets high in fiber are associated with a decreased risk. High-fiber diets may be related to less exposure of the rectal epithelium to carcinogens from the environment as the transit time through the bowel is faster with a high-fiber diet than with a low-fiber diet.

Age plays a definite role in rectal cancer risk. Rectal cancer is rare before age 40. This incidence increases substantially after age 50 and doubles with each succeeding decade.

There also is a slight increase of risk for rectal cancer in the individual who smokes.

Patients who suffer from an inflammatory disease of the colon known as ulcerative **colitis** are also at increased risk.

On chromosome 5 is the APC gene associated with familial adenomatous polyposis (FAP) syndrome. There are multiple mutations that occur at this site, yet they all cause a defect in tumor suppression that results in early and frequent development of **colon cancer**. This is transmitted to 50% of offspring and each of those affected will develop colon or rectal cancer, usually at an early age. Another syndrome, hereditary non-polyposis colon cancer (HNPCC), is related to mutations in any of four genes responsible for DNA mismatch repair. In patients with colon or rectal cancer, the p53 gene is mutated 70% of the time. When the p53 gene is mutated and ineffective, cells with damaged DNA escape repair or destruction, allowing the damaged cell to multiply. Continued replication of the damaged DNA may lead to tumor development. Though these syndromes (FAP and HNPCC) have a very high incidence of colon or rectal cancer, family history without the syndromes is also a substantial risk factor. When considering first-degree relatives, history of one with colon or rectal cancer raises the baseline risk from 2% to 6%; the presence of a second raises the risk to 17%.

The development of polyps of the colon or rectum commonly precedes the development of rectal cancer. Polyps are growths of the rectal lining. They can be unrelated to cancer, pre-cancerous, or malignant. Polyps, when identified, are removed for diagnosis. If the polyp, or polyps, are benign, the patient should undergo careful surveillance for the development of more polyps or the development of colon or rectal cancer.

Symptoms of rectal cancer most often result from the local presence of the tumor and its capacity to invade surrounding pelvic structure:

- bright red blood present with stool
- abdominal distention (stretching from internal pressure), bloating, inability to have a bowel movement
- narrowing of the stool, so-called ribbon stools
- pelvic pain
- unexplained weight loss
- persistent chronic fatigue
- rarely, urinary infection or passage of air in urine in males (late symptom)
- rarely, passage of feces through vagina in females (late symptom)

If the tumor is large and obstructing the rectum, the patient will not be evacuating stool normally and will get bloated and have abdominal discomfort. The tumor itself may bleed and, since it is near the anus, the patient may see bright red blood on the surface of the stool. Blood alone (without stool) may also be passed. Thus, **hemorrhoids** are often incorrectly blamed for bleeding, delaying the diagnosis. If anemia develops, which is rare, the patient will experience **chronic fatigue**. If the tumor invades the bladder in the male or the vagina in the female, stool will get where it does not belong and cause infection or discharge. (This condition is also rare.) Patients with widespread disease lose weight secondary to the chronic illness.

Diagnosis

Screening evaluation of the colon and rectum are accomplished together. Screening involves physical exam, simple laboratory tests, and the visualization of the lining of the rectum and colon. X rays (indirect visualization) and **endoscopy** (direct visualization) are used to visualize the organs' lining.

The **physical examination** involves the performance of a digital rectal exam (DRE). At the time of this exam, the physician checks the stool on the examining glove with a chemical to see if any occult (invisible), blood is present. At home, after having a bowel movement, the patient is asked to swipe a sample of stool obtained with a small stick on a card. After three such specimens are on the card, the card is then easily chemically tested for occult blood. These exams are accomplished as an easy part of a routine yearly physical exam.

Proteins are sometimes produced by cancers and these may be elevated in the patients blood. When this occurs the protein produced is known as a tumor marker. There is a tumor marker for cancer of the colon and rectum; it is known as carcinoembryonic antigen, (CEA). Unfortunately, this may be made by other adenocarcinomas as well, or it may not be

produced by a particular colon or rectal cancer. Therefore, screening by chemical analysis for CEA has not been helpful. CEA has been helpful in patients treated for colon or rectal cancer if their tumor makes the protein. It is used in a follow-up role, not a screening role.

Direct visualization of the lining of the rectum is accomplished using a scope or endoscope. The physician introduces the instrument into the rectum and is able to see the epithelium of the rectum directly. A simple rigid tubular scope may be used to see the rectal epithelium; however, screening of the colon is done at the same time. The lower colon may be visualized using a fiberoptic flexible scope in a procedure known as flexible **sigmoidoscopy**. When the entire colon is visualized, the procedure is known as total **colonoscopy**. Each type of endoscopy requires pre-procedure preparation (evacuation) of the rectum and colon.

The American Cancer Society has recommended the following screening protocol for colon and rectal cancers those over age 50:

- yearly fecal occult blood test
- flexible sigmoidoscopy at age 50
- flexible sigmoidoscopy repeated every 5 years
- double contrast barium enema every five years
- colonoscopy every 10 years

If there are predisposing factors such as positive family history, history of polyps, or a familial syndrome, screening evaluations should start sooner.

Evaluation of patients with symptoms

When patients visit their physician because they are experiencing symptoms that could possibly be related to colon or rectal cancer, the entire colon and rectum must be visualized. Even if a rectal lesion is identified, the entire colon must be screened to rule out a syndromous polyp or cancer of the colon. The combination of a flexible sigmoidoscopy and double contrast **barium enema** may be performed, but the much preferred evaluation of the entire colon and rectum is that of complete colonoscopy. Colonoscopy allows direct visualization, photography, as well as the opportunity to obtain a biopsy, (a sample of tissue), of any abnormality visualized. If, for technical reasons the entire colon is not visualized endoscopically, a double contrast barium enema should complement the colonoscopy. A patient who is identified to have a problem in one area of the colon or rectum is at greater risk to have a similar problem in area of the

colon or rectum. Therefore the entire colon and rectum need to be visualized during the evaluation.

The diagnosis of rectal cancer is actually made by the performance of a biopsy of any abnormal lesion in the rectum. Many rectal cancers are within reach of the examiner's finger. Identifying how close to the anus the cancer has developed is very important in planning the treatment. Another characteristic ascertained by exam is whether the tumor is mobile or fixed to surrounding structure. Again, this will have implications related to primary treatment. As a general rule, it is easier to identify and adequately obtain tissue for evaluation in the rectum as opposed to the colon. This is because the lesion is closer to the anus.

If the patient has advanced disease, areas where the tumor has spread, such as the liver, may require biopsy. Such biopsies are usually obtained using a special needle under **local anesthesia**.

Once a diagnosis of rectal cancer has been established by biopsy, in addition to the physical exam, an **endorectal ultrasound** will be performed to assess the extent of the disease. For rectal cancer, endorectal ultrasound is the most preferred method for staging both depth of tumor penetration and local lymph node status. Endorectal ultrasound:

- differentiates areas of invasion within large rectal adenomas that seem benign
- determines the depth of tumor penetration into the rectal wall
- determines the extent of regional lymph node invasion
- can be combined with other tests (chest x rays and computed tomography scans, or CT scans) to determine the extent of cancer spread to distant organs, such as the lungs or liver

The resulting rectal cancer staging allows physicians to determine the need for—and order of—radiation, surgery, and **chemotherapy**. In 2003, it was reported that **magnetic resonance imaging** (MRI) also may be useful in staging rectal cancer. MRI may help physicians determine if a tumor can be resected and risk of cancer recurrence.

Treatment

Once the diagnosis has been confirmed by biopsy and the endorectal ultrasound has been performed, the clinical stage of the cancer is assigned. The treating physicians use staging to plan the specific treatment protocol for the patient. In addition, the stage of the cancer at the time of presentation gives a statistical likelihood of the treatment outcome (prognosis).

Clinical staging

Rectal cancer first invades locally and then progresses to spread to regional lymph nodes or to other organs. Stage is derived using the characteristics of the primary tumor, its depth of penetration through the rectum, local invasion into pelvic structure, and the presence or absence of regional or distant metastases. A CT scan of the pelvis is helpful in staging because tumor invasion into the sacrum or pelvic sidewalls may mean surgical therapy is not initially possible. On this basis, clinical staging is used to begin treatment. The pathologic stage is defined when the results of analyzing the surgical specimen are available. (typically stage I and II).

Rectal cancer is assigned stages I through IV, based on the following general criteria:

- Stage I: the tumor is confined to the epithelium or has not penetrated through the first layer of muscle in the rectal wall
- Stage II: the tumor has penetrated through to the outer wall of the rectum or has gone through it, possibly invading other local tissue or organs
- Stage III: Any depth or size of tumor associated with regional lymph node involvement
- Stage IV: any of previous criteria associated with distant metastasis

Surgery

The first, or primary, treatment modality utilized in the treatment of rectal cancer is surgery. Stage I, II, and even suspected stage III disease are treated by surgical removal of the involved section of the rectum along with the complete vascular and lymphatic supply. Most Stage II and Stage III rectal cancers (based on endorectal ultrasound, CT scan, and **chest x ray**) are treated with radiation and possibly chemotherapy prior to surgery.

When determining primary treatment for rectal cancer, the surgeon's ability to reconnect the ends of the rectum. The pelvis is a confining space that makes the performance of the hook-up more difficult to do safely when the tumor is in the lower rectum. The upper rectum does not usually present a substantial problem to the surgeon restoring bowel continuity after the cancer has been removed. Mid-rectal tumors, (especially in males where the pelvis is usually smaller than a woman's), may present technical difficulties in hooking the proximal bowel to the remaining rectum. Technical advances in stapling instrumentation have largely overcome these difficulties. If the anastomosis (hook-up) leaks postoperatively, infection can occur. In the past,

this was a major cause of complications in resection of rectal cancers. Today, utilizing the stapling instrumentation, a hook-up at the time of original surgery is much safer. If the surgeon feels that the hook-up is compromised or may leak, a **colostomy** may be performed. A colostomy is performed by bringing the colon through the abdominal wall and sewing it to the skin. In these cases the stool is diverted away from the hook-up, allowing it to heal and preventing the infectious complications associated with leak. Later, when the hook-up has completely healed, the colostomy can be taken down and bowel continuity restored.

Stapling devices have allowed the surgeon to get closer to the anus and still allow the technical performance of a hook-up, but there are limits. It is generally felt that there should be at least three centimeters of normal rectum below the tumor or the risk of recurrence locally will be excessive. In addition, if there is no residual native rectum, the patient will not have normal sensation or control and will have problems with uncontrollable soiling, (incontinence). For these reasons, patients presenting with low rectal tumors may undergo total removal of the rectum and anus. This procedure is known as an abdominal-perineal resection. A permanent colostomy is performed in the lower left abdomen.

Radiation

As mentioned, for many late stage II or stage III tumors, **radiation therapy** can shrink the tumor prior to surgery. The other roles for radiation therapy are as an aid to surgical therapy in locally advanced disease that has been removed, and in the treatment of certain distant metastases. Especially when utilized in combination with chemotherapy, radiation used postoperatively has been shown to reduce the risk of local recurrence in the pelvis by 46% and death rates by 29%. Such combined therapy is recommended in patients with locally advanced primary tumors that have been removed surgically. Radiation has been helpful in treating effects of distant metastases, particularly in the brain. In very few cases, radiation alone may be the curative treatment for rectal cancer.

Chemotherapy

Adjuvant chemotherapy, (treating the patient who has no evidence of residual disease but who is at high risk for recurrence), is considered in patients whose tumors deeply penetrate or locally invade (late stage II and stage III). If the tumor was not locally advanced, this form of chemotherapeutic adjuvant therapy may be recommended without radiation. This therapy is identical to that of colon cancer and

KEY TERMS

Adenocarcinoma—Type of cancer beginning in glandular epithelium.

Adjuvant therapy—Treatment involving radiation, chemotherapy (drug treatment), or hormone therapy, or a combination of all three given after the primary treatment for the possibility of residual microscopic disease.

Anastomosis—Surgical re-connection of the ends of the bowel after removal of a portion of the bowel.

Anemia—The condition caused by too few circulating red blood cells, often manifest in part by fatigue.

Carcinogens—Substances in the environment that cause cancer, presumably by inducing mutations, with prolonged exposure.

Defecation—The act of having a bowel movement.

Epithelium—Cells composing the lining of an organ.

Lymphatics—Channels that are conduits for lymph.

Lymph nodes—Cellular filters through which lymphatics flow.

Malignant—Cells that have been altered such that they have lost normal control mechanisms and are capable of local invasion and spread to other areas of the body.

Metastasis—Site of invasive tumor growth that originated from a malignancy elsewhere in the body.

Mutation—A change in the genetic makeup of a cell that may occur spontaneously or be environmentally induced.

Occult blood—Presence of blood that cannot be appreciated visually.

Polyps—Localized growths of the epithelium that can be benign, pre-cancerous, or harbor malignancy.

Resect—To remove surgically.

Sacrum—Posterior bony wall of the pelvis.

Systemic—Referring to throughout the body.

leads to similar results. Standard therapy is treatment with 5-fluorouracil, (5-FU) combined with leucovorin for a period of six to 12 months. 5-FU is an antimetabolite and leucovorin improves the response rate. Another agent, levamisole, (which seems to stimulate the immune system), may be substituted for leucovorin. These protocols reduce rate of recurrence by about 15% and reduce mortality by about 10%. The regimens have some toxicity but usually are tolerated fairly well.

Similar chemotherapy is administered for stage IV disease or if a cancer progresses and metastasis develops. Results show response rates of about 20%. A response is a temporary regression of the cancer in response to the chemotherapy. Unfortunately, these patients eventually succumb to the disease. Clinical trials have now shown that the results can be improved with the addition of another agent to this regimen. Irinotecan does not seem to increase toxicity but has improved response rates to 39%, added two to three months to disease free survival, and prolonged overall survival by a little more than two months.

Alternative treatment

Most alternative therapies have not been studied in clinical trials. Large doses of **vitamins**, fiber, and

green tea are among therapies tried. A 2003 report on a large Harvard University study showed that people who took multivitamins for at least 15 years had a 34% reduction in risk of rectal cancer. Before initiating any alternative therapies, the patient should consult his or her physician to be sure that these therapies do not complicate or interfere with the recommended therapy.

Prognosis

Prognosis is the long-term outlook or survival after therapy. Overall, about 50% of patients treated for colon and rectal cancer survive the disease. As expected, the survival rates are dependent upon the stage of the cancer at the time of diagnosis, making early detection crucial.

About 15% of patients present with stage I disease, or are diagnosed with Stage I disease when they initially visit a doctor, and 85-90% survive. Stage II represents 20-30% of cases and 65-75% survive; 30-40% comprise the stage III presentation, of which 55% survive. The remaining 20-25% present with stage IV disease and are rarely cured.

Prevention

There is not an absolute method for preventing colon or rectal cancer. An individual can lessen risk or

identify the precursors of colon and rectal cancer. The patient with a familial history can enter screening and surveillance programs earlier than the general population. High-fiber diets and vitamins, avoiding **obesity**, and staying active lessen the risk. In fact, a 2003 report said that vigorous **exercise** (to the point of sweating or feeling out of breath) lowered risk of rectal cancer by nearly 40% compared to those who exercised less. Avoiding cigarettes and alcohol may be helpful. By controlling these environmental factors, an individual can lessen risk and to this degree prevent the disease.

By undergoing appropriate screening when uncontrollable genetic risk factors have been identified, an individual may be rewarded by the identification of benign polyps that can be treated as opposed to having these growths degenerate into a malignancy.

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Richard A. McCartney, MD
 Teresa G. Odle

Rectal examination

Definition

Rectal examination or **digital rectal examination** (DRE) is performed by means of inserting a gloved, lubricated finger into the rectum and palpating (feeling) for lumps.

Purpose

DRE is used as a screening tool to locate **rectal cancer** and **prostate cancer**. It is also used as a diagnostic test to find non-cancerous abnormalities within the rectum like **hemorrhoids**, anal fissures, or congenital deformities that can cause chronic **constipation**.

Precautions

There are no precautions when performing DRE, aside from routine sanitary procedures.

Description

DRE is performed in most instances as an annual routine procedure in colorectal **cancer** screening. Digital palpitation of the rectum can often find abnormal growths which may require further testing or commonplace hemorrhoids. It is a critical initial clinical test and is important in the assessment of the size and location of tumors.

This procedure is often not performed routinely on patients over 70, even though this population is at high risk for colorectal cancer. It also is not done as often in elderly women as in elderly men.

DRE has also been used as a screening tool for prostate cancer. It seems to be very effective for larger masses found in the prostate and correlated well with higher prostate-specific antigens.

Of less predictive value was DRE in routine rectovaginal examinations of women under the age of 50. These instances of DRE did not locate colorectal cancer or any other abnormality.

More gastroenterologists are recommending that pediatricians and family physicians perform DRE on pediatric patients exhibiting chronic constipation before those patients are referred to intestinal specialists. The pediatrician or family physician could identify fecal compaction and treat it themselves, and then only refer patients who have a specific abnormality to gastroenterologists.

KEY TERMS

DRE—Digital rectal examination.

Gastroenterologist—A physician who specializes in diseases of the digestive system.

Rectum—The last eight to ten inches of the colon, of which the anus is a part and the opening through which wastes are removed from the body.

Preparation

The physician must conduct DRE using a gloved hand. Some sort of lubricant should be used so that penetration of the rectum is easier and does not create the damage that the procedure is seeking.

Aftercare

There is no aftercare after a DRE is performed.

Risks

There are no risks to DRE and it is virtually painless.

Normal results

The physician finds a normal rectal canal with no abnormalities.

Abnormal results

Growths, tears, anal fissures, or congenital structural defects can be found inside the rectum with DRE.

Resources

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Janie F. Franz

Rectal polyps

Definition

Rectal polyps are tissue growths that arise from the wall of the rectum and protrude into it. They may be either benign or malignant (cancerous).

Description

The rectum is the last segment of the large intestine, ending in the anus, the opening to the exterior of the body. Rectal polyps are quite common. They occur in 7-50% of all people, and in two thirds of people over age 60.

Rectal polyps can be either benign or malignant, large or small. There are several different types of polyps. The type is determined by taking a sample of the polyp and examining it microscopically. Most polyps are benign. They are of concern, however, because 90% of colon and rectal cancers arise from polyps that are initially benign. For this reason, rectal polyps are usually removed when they are discovered.

Causes and symptoms

The cause of most rectal polyps is unknown, however a diet high in animal fat and red meat, and low in fiber, is thought to encourage polyp formation. Some types of polyps are hereditary. In an inherited disease called **familial polyposis**, hundreds of small, malignant and pre-malignant polyps are produced before the age of 40. Also, inflammatory bowel disease may cause growth of polyps and pseudo-polyps. Juvenile polyps (polyps in children) are usually benign and often outgrow their blood supply and disappear at **puberty**.

Most rectal polyps produce no symptoms and are discovered on routine digital or endoscopic examination of the rectum. Rectal bleeding is the most common complaint when symptoms do occur. Abdominal cramps, **pain**, or obstruction of the intestine occur with some large polyps. Certain types of polyps cause mucous-filled or watery **diarrhea**.

Diagnosis

Rectal polyps are commonly found by **sigmoidoscopy** (visual inspection with an instrument consisting of a tube and a light) or **colonoscopy**. If polyps are found in the rectum, a complete examination of the large intestine is done, as multiple polyps are common.

KEY TERMS

Colon—The part of the large intestine that extends from the cecum to the rectum. The sigmoid colon is the area of the intestine just above the rectum; linking the descending colon with the rectum. It is shaped like the letter S.

Rectum—The final part of the large intestine, ending in the anus.

Sigmoidoscopy—A procedure where a thin tube containing a camera and a light is inserted into the lower section of the large intestine so that the doctor can visually inspect the lower (sigmoid) colon and rectum. Colonoscopy examines the entire large intestine using the same techniques.

Polyps do not show up on regular x rays, but they do appear on **barium enema** x rays.

Treatment

Normally polyps are removed when they are found. Polypectomy is the name for the surgery that removes these growths. Polypectomy is performed at a hospital, outpatient surgical facility or in a doctor's office, depending on the number and type of polyps to be removed, and the age and health of the patient. The procedure can be done by a surgeon, gastroenterologist, or family practitioner.

Before the operation, a colonoscopy (examination of the intestine with an endoscope) is performed, and standard pre-operative blood and urine studies are done. The patient is also given medicated **enemas** to cleanse the bowel.

The patient is given a sedative and a narcotic pain killer. A colonoscope is inserted into the rectum. The polyps are located and removed with a wire snare, ultrasound, or laser beam. After they are removed, the polyps are examined to determine if they are malignant or benign. When polyps are malignant, it may be necessary to remove a portion of the rectum or colon to completely remove cancerous tissue.

Alternative treatment

In addition to a diet low in animal fat and high in fiber, nutritionists recommend antioxidant supplements (including **vitamins** A, C, and E, selenium, and zinc) to reduce rectal polyps.

Prognosis

For most people, the removal of polyps is an uncomplicated procedure. Benign polyps that are left in place can give rise to **rectal cancer**. People who have had rectal polyps once are more likely to have them again and should have regular screening examinations.

Prevention

Eating a diet low in red meat and animal fat and high in fiber is thought to help prevent rectal polyps.

ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329, (800) 227-2345, <http://www.cancer.org>.

National Cancer Institute (National Institutes of Health), NCI Office of Communications and Education, 6116 Executive Blvd. Suite 300, Bethesda, MD, 20892-8322, (800) 4-CANCER (422-6237), cancergovstaff@mail.nih.gov, <http://www.cancer.gov/>.

Tish Davidson, A.M.

Rectal prolapse

Definition

Rectal prolapse is protrusion of rectal tissue through the anus to the exterior of the body. The rectum is the final section of the large intestine.

Description

Rectal prolapse can be either partial or complete. In partial prolapse, only the mucosa layer (mucous membrane) of the rectum extends outside the body. The projection is generally 0.75–1.5 in (2–4 cm) long. In complete prolapse, called procidentia, the full thickness of the rectum protrudes for up to 4.5 in (12 cm).

Rectal prolapse is most common in people over age 60, and occurs much more frequently in women than in men. It is also more common in psychiatric patients. Prolapse can occur in normal infants, where it is usually transient. In children it is often an early sign of **cystic fibrosis** or is due to neurological or anatomical abnormalities.

Although rectal prolapse in adults may initially reduce spontaneously after bowel movements, it eventually becomes permanent. Adults who have had prior rectal or vaginal surgery, who have chronic **constipation**, regularly depend on **laxatives**, have **multiple**

sclerosis or other neurologic diseases, **stroke**, or **paralysis** are more likely to experience rectal prolapse.

Causes and symptoms

Rectal prolapse in adults is caused by a weakening of the sphincter muscle or ligaments that hold the rectum in place. Weakening can occur because of **aging**, disease, or in rare cases, surgical trauma. Prolapse is brought on by straining to have bowel movements, chronic laxative use, or severe **diarrhea**.

Symptoms of rectal prolapse include discharge of mucus or blood, **pain** during bowel movements, and inability to control bowel movements (**fecal incontinence**). Patients may also feel the mass of tissue protruding from the anus. With large prolapses, the patient may lose the normal urge to have a bowel movement.

Diagnosis

Prolapse is initially diagnosed by taking a patient history and giving a **rectal examination** while the patient is in a squatting position. It is confirmed by **sigmoidoscopy** (inspection of the colon with a viewing instrument called a endoscope) **Barium enema** x rays and other tests are done to rule out neurologic (nerve) disorders or disease as the primary cause of prolapse.

Treatment

In infants, conservative treatment, consisting of strapping the buttocks together between bowel movements and eliminating any causes of bowel straining, usually produces a spontaneous resolution of prolapse. For partial prolapse in adults, excess tissue is surgically tied off with special bands causing the tissue to wither in a few days.

Complete prolapse requires surgery. Different surgical techniques are used, but all involve anchoring the rectum to other parts of the body, and using plastic mesh to reinforce and support the rectum. In patients too old, or ill, to tolerate surgery, a wire or plastic loop can be inserted to hold the sphincter closed and prevent prolapse. Treatment should be undertaken as soon as prolapse is diagnosed, since the longer the condition exists, the more difficult it is to reverse.

Alternative treatment

Alternative therapies can act as support for conventional treatment, especially if surgery is required. **Acupuncture**, homeopathy, and botanical medicine can all be used to assist in resolution of the prolapse or in recovery from surgery.

KEY TERMS

Rectum—The part of the large intestine that ends at the anal canal.

Prognosis

Successful resolution of rectal prolapse involves prompt treatment and the elimination of any underlying causes of prolapse. Infants and children usually recover completely without complications. Recovery in adults depends on age, general health, and the extent of the prolapse.

Prevention

Reducing constipation by eating a diet high in fiber, drinking plenty of fluids, and avoiding straining during bowel movements help prevent the onset of prolapse. Exercises that strengthen the anal sphincter may also be helpful.

Resources

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Tish Davidson, A.M.

Recurrent fever see **Relapsing fever**

Recurrent miscarriage

Definition

Recurrent **miscarriage** is defined as three or more miscarriages of a fetus before 20 weeks of gestation (i.e., before the fetus can live outside the womb).

Description

Also referred to as spontaneous abortion, miscarriage occurs in 15-20% of all conceptions. The majority of miscarriages occur during the first trimester. The number of previous miscarriages does not affect subsequent full-term pregnancies.

Causes and symptoms

Recurrent miscarriage can be caused by several factors, including fetal, placental, or maternal abnormalities.

- In over half of all miscarriages, the fetus is abnormal. The abnormality can either be genetic or developmental. The fetus is very sensitive to ionizing radiation. Tobacco and even moderate alcohol consumption are known to cause fetal damage that may lead to miscarriage. There is some evidence that over four cups of coffee a day, because of the caffeine, adversely affect pregnancy, as well.
- Placental abnormalities, including abnormal implantation in the placental wall and premature separation of the placenta, can cause miscarriage.
- Maternal abnormalities include insufficient hormones (usually progesterone) to support the pregnancy, an incompetent cervix (mouth of the womb does not stay closed), or a deformed uterus (womb). A deformed uterus can be caused by diethylstilbestrol (DES) given to the mother's mother during her pregnancy. Some immunologic abnormalities may cause the mother to reject the fetus as if it were an infection or a transplant. Maternal blood clotting abnormalities may cut-off blood supply to the fetus, causing miscarriage.
- Maternal diabetes mellitus causes miscarriage if the diabetes is poorly controlled. Maternal infections may occasionally lead to miscarriage. There is some evidence that conceptions that take place between old eggs (several days after ovulation) or old sperm (that start out several days before ovulation) may be more likely to miscarry.

Symptoms of miscarriage include pink or brown colored discharge for several weeks, which develops into painful cramping and increased vaginal bleeding; dilation of the cervix; and expulsion of the fetus.

Diagnosis

A pelvic examination can detect a deformed uterus, and frequent examinations during **pregnancy** can detect an **incompetent cervix**. Blood tests can detect the presence of immunologic or blood-clotting problems in the mother. **Genetic testing** can also determine if chromosomal abnormalities may be causing the miscarriages.

Treatment

If a uterus is deformed, it may be surgically repaired. If a cervix is incompetent, it can be surgically fortified, until the fetus matures, by a procedure known as circlage

KEY TERMS

Fetus—A developing embryo in the womb after the first eight weeks of gestation.

Ionizing radiation—Radiation produced by x rays and radioactivity.

Ovulation—Release of an egg for fertilization from the ovary that happens about fourteen days before each menstrual period.

(tying the cervix closed). Supplemental progesterone may also help sustain a pregnancy. Experimental treatment of maternal immunologic abnormalities with white cell immunization (injecting the mother with white cells from the father) has been successful in some cases of recurrent miscarriage. Clotting abnormalities can be treated with **anticoagulant drugs**, such as heparin and **aspirin**, to keep blood flowing to the fetus.

Prognosis

If there is no underlying disease or abnormality present, the rate of successful pregnancy after several miscarriages approaches normal. Seventy to eighty-five percent of women with three or more miscarriages will go on to complete a healthy pregnancy.

Resources

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J. Ricker Polsdorfer, MD

Red blood cell indices

Definition

Red blood cell indices are measurements that describe the size and oxygen-carrying protein (hemoglobin) content of red blood cells. The indices are used to help in the differential diagnosis of anemia. They are also called red cell absolute values or erythrocyte indices.

Purpose

Anemia includes a variety of conditions with the same outcome: a person's blood cannot carry as much oxygen as it should. A healthy person has an adequate

number of correctly sized red blood cells that contain enough hemoglobin to carry sufficient oxygen to all the body's tissues. An anemic person has red blood cells that are either too small or too few in number. As a result, the heart and lungs must work harder to make up for the lack of oxygen delivered to the tissues by the blood.

Anemia is caused by many different diseases or disorders. The first step in finding the cause is to determine what type of anemia the person has. Red blood cell indices help to classify the **anemias**.

Precautions

Certain prescription medications may affect the test results. These drugs include zidovudine (Retrovir), phenytoin (Dilantin), and azathioprine (Imuran).

Description

Overview

Anemia has several general causes: blood loss; a drop in production of red blood cells; or a rise in the number of red blood cells destroyed. Blood loss can result from severe hemorrhage or a chronic slow bleed, such as the result of an accident or an ulcer. Lack of iron, vitamin B₁₂, or **folic acid** in the diet, as well as certain chronic diseases, lower the number of red blood cells produced by the bone marrow. Inherited disorders affecting hemoglobin, severe reactions to blood transfusions, prescription medications, or poisons can cause red blood cells to burst (hemolyze) well before the end of their usual 120-day lifespan.

Anemia of any type affects the results of one or more of the common blood tests. These tests are the **hematocrit**, hemoglobin, and red blood cell count. The hematocrit is a measure of red blood cell mass, or how much space in the blood is occupied by red blood cells. The **hemoglobin test** is a measure of how much hemoglobin protein is in the blood. The red blood cell count (RBC) measures the number of red blood cells present in the blood. Red blood cell indices are additional measurements of red blood cells based on the relationship of these three test results.

The relationships between the hematocrit, the hemoglobin level, and the RBC are converted to red blood cell indices through mathematical formulas. These formulas were worked out and first applied to the classification of anemias by Maxwell Wintrobe in 1934.

The indices include these measurements: mean corpuscular volume (MCV); mean corpuscular hemoglobin (MCH); mean corpuscular hemoglobin concentration (MCHC); and red cell distribution width

(RDW). They are usually calculated by an automated instrument as part of a **complete blood count** (CBC). Indices are covered by insurance when medically necessary. Results are available the same day that the blood is drawn or the following day.

Mean corpuscular volume (MCV)

MCV is the index most often used. It measures the average volume of a red blood cell by dividing the hematocrit by the RBC. The MCV categorizes red blood cells by size. Cells of normal size are called normocytic, smaller cells are microcytic, and larger cells are macrocytic. These size categories are used to classify anemias. Normocytic anemias have normal-sized cells and a normal MCV; microcytic anemias have small cells and a decreased MCV; and macrocytic anemias have large cells and an increased MCV. Under a microscope, stained red blood cells with a high MCV appear larger than cells with a normal or low MCV.

Mean corpuscular hemoglobin concentration (MCHC)

The MCHC measures the average concentration of hemoglobin in a red blood cell. This index is calculated by dividing the hemoglobin by the hematocrit. The MCHC categorizes red blood cells according to their concentration of hemoglobin. Cells with a normal concentration of hemoglobin are called normochromic; cells with a lower than normal concentration are called hypochromic. Because there is a physical limit to the amount of hemoglobin that can fit in a cell, there is no hyperchromic category.

Just as MCV relates to the size of the cells, MCHC relates to the color of the cells. Hemoglobin contains iron, which gives blood its characteristic red color. When examined under a microscope, normal red blood cells that contain a normal amount of hemoglobin stain pinkish red with a paler area in the center. These normochromic cells have a normal MCHC. Cells with too little hemoglobin are lighter in color with a larger pale area in the center. These hypochromic cells have a low MCHC. Anemias are categorized as hypochromic or normochromic according to the MCHC index.

Mean corpuscular hemoglobin (MCH)

The average weight of hemoglobin in a red blood cell is measured by the MCH. The formula for this index is the sum of the hemoglobin multiplied by 10 and divided by the RBC. MCH values usually rise or fall as the MCV is increased or decreased.

KEY TERMS

Anemia—A variety of conditions in which a person's blood can't carry as much oxygen as it should due to a decreased number or size of red blood cells.

Hypochromic—A descriptive term applied to a red blood cell with a decreased concentration of hemoglobin.

Macrocytic—A descriptive term applied to a larger than normal red blood cell.

Mean corpuscular hemoglobin (MCH)—A measurement of the average weight of hemoglobin in a red blood cell.

Mean corpuscular hemoglobin concentration (MCHC)—The measurement of the average concentration of hemoglobin in a red blood cell.

Mean corpuscular volume (MCV)—A measure of the average volume of a red blood cell.

Microcytic—A descriptive term applied to a smaller than normal red blood cell.

Normochromic—A descriptive term applied to a red blood cell with a normal concentration of hemoglobin.

Normocytic—A descriptive term applied to a red blood cell of normal size.

Red blood cell indices—Measurements that describe the size and hemoglobin content of red blood cells.

Red cell distribution width (RDW)—A measure of the variation in size of red blood cells.

Red cell distribution width (RDW)

The RDW measures the variation in size of the red blood cells. Usually red blood cells are a standard size. Certain disorders, however, cause a significant variation in cell size.

Obtaining the blood sample

The RBC indices test requires 0.17–24 oz (5–7 mL) of blood. A healthcare worker ties a tourniquet on the person's upper arm, locates a vein in the inner elbow region, and inserts a needle into that vein. Vacuum action draws the blood through the needle into an attached tube. Collection of the sample takes only a few minutes.

Preparation

The doctor should check to see if the patient is taking any medications that may affect test results. The patient does not need to fast before the test.

Aftercare

Aftercare consists of routine care of the area around the puncture mark. Pressure is applied for a few seconds and the wound is covered with a bandage.

Risks

The primary risk is mild **dizziness** and the possibility of a bruise or swelling in the area where the blood was drawn. The patient can apply moist warm compresses.

Normal results

Normal results for red blood cell indices are as follows:

- MCV 82–98 fl (femtoliters)
- MCHC 31–37 g/dL
- MCH 26–34 pg (picograms)
- RDW 11.5–14.5%

Abnormal results

The category into which a person's anemia is placed based on the indices provides a significant clue as to the cause of the anemia, but further testing is needed to confirm a specific diagnosis.

The most common causes of macrocytic anemia (high MCV) are vitamin B₁₂ and folic acid deficiencies. Lack of iron in the diet, **thalassemia** (a type of hereditary anemia), and chronic illness are the most common causes of microcytic anemia (low MCV). Normocytic anemia (normal MCV) can be caused by kidney and **liver disease**, bone marrow disorders, or excessive bleeding or hemolysis of the red blood cells.

Lack of iron in the diet and thalassemia are the most common causes of hypochromic anemia (low MCHC). Normocytic anemias are usually also normochromic and share the same causes (normal MCHC).

The RDW is increased in anemias caused by deficiencies of iron, vitamin B₁₂, or folic acid. Abnormal hemoglobins, such as in sickle cell anemia, can change the shape of red blood cells as well as cause them to hemolyze. The abnormal shape and the cell fragments resulting from hemolysis increase the RDW. Conditions that cause more immature cells to be released into the bloodstream, such as severe blood loss, will increase the RDW. The larger size of immature cells creates a distinct size variation.

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Nancy J. Nordenson

Red blood cell test see **Hemoglobin test**



Red reflex testing

Definition

Red reflex (RR) testing is an examination of the red reflex—light reflected back from the retina at the rear of the eye, which causes the pupil of the eye to appear red, as in flash photographs. RR testing is used to screen for abnormalities or obstructions of the retina—such as a cataract or tumor—that distort or eliminate the red reflex. The red reflex test is also called a light reaction test.

Purpose

Red reflex testing is considered to be an essential component of all healthcare visits for newborns, infants, and children through five years of age or until they are old enough to read an eye chart. Basic eye examinations in newborns, including RR testing, have become even more important as the number of premature and medically fragile infants born in the United States continues to increase. Although many more of these children are surviving, they often suffer **visual impairment**.

The primary purpose of RR testing is to screen for vision-threatening and life-threatening conditions, especially **retinoblastoma** and congenital, infantile, or juvenile **cataracts**. Retinoblastoma is an inherited malignant (cancerous) tumor of the retina that can grow to fill much of the eye and spread to other parts of the body. A cataract is a clouding of the lens of the eye or its surrounding transparent membrane that prevents the passage of light into the eye. Although these conditions are rare, they are much more likely to be successfully treated with early detection. However detection of and referral for these conditions are often delayed. It is estimated that 75% of American children under age five have never had a comprehensive eye exam and only about 22% of preschoolers have had any type of vision screening.

A toddler is evaluated by an orthoptist to detect any abnormalities in the eye. (Phanie/Photo Researchers, Inc.)

About 60% of children who develop retinoblastoma have no known family history that would cause them to be screened. As many as 80% of retinoblastomas are initially detected by the child's family or friends, often by noticing that the child has one red eye and one white eye in a flash photograph. However by the time the condition is this obvious, it is often too late to save the affected eye. The American Academy of Pediatrics (AAP) recommends RR testing with an ophthalmoscope for all infants within the first two months of life, in a darkened room to maximize pupil dilation. Pediatricians often fail to diagnose retinoblastoma early because the exams are conducted in well-lit rooms in which the pupils are very small. For this reason, some states have considered legislation that would mandate RR testing of eye-drop-dilated pupils for all newborns, and possibly at all six-week to eight-week and six-month to nine-month well-baby exams.

RR testing can detect other abnormalities in addition to retinoblastoma and cataracts, including:

- other opacities—areas of the eye that are not transparent to light—and abnormalities of the retina
- white spots in the eye
- strabismus—misalignment of the eyes caused by an imbalance in the muscles of the eyeball and resulting in an eye that turns in toward the nose (cross-eyed) or outward away from the nose (walleyed)
- high refractive error—the inability of an eye to focus light properly due to an irregularly shaped cornea, causing blurry vision and including nearsightedness, farsightedness, or astigmatism
- asymmetric refractive error—significant differences in refractive error between the two eyes

- congenital glaucoma—a disease characterized by increased pressure within the eyeball
- lenticonus, a rare, usually congenital condition, in which the lens of the eye has a conical surface
- certain systemic diseases that affect the eyes

Description

In red reflex testing, a bright light is transmitted from an ophthalmoscope through all of the parts of the eye that are normally transparent, including the tear film, cornea, aqueous humor (the transparent fluid in the space between the cornea and lens), crystalline lens, and vitreous body (the transparent jelly that fills the eyeball behind the lens). The light reflects off the ocular fundus—the part of the eye opposite the pupil—and is transmitted back through the eye and the aperture of the ophthalmoscope. Any obstruction of this optical pathway results in an abnormal red reflex.

RR testing is quick and is usually performed by a pediatrician or other primary care provider who has been trained in the technique. It should be performed in a dim or darkened room so that the baby's pupils dilate or widen to increase the retinal reflection and provide the examiner with a better view. A direct ophthalmoscope or retinoscope is used to detect the red reflex in each eye individually and then in both eyes simultaneously to compare the red light reflection from each pupil. The ophthalmoscope is held close to the examiner's eye, about 12–18 inches (30–46 cm) from the child's eye. Sometimes RR testing is performed at two different distances and lens settings. If the baby's eyelids do not open adequately, a lid speculum may be used to hold them open.

A Bruckner test is RR testing combined with a simultaneous corneal light reflex test. The latter involves observing the location of a shined light on each cornea with respect to the pupil to check for ocular misalignment or other abnormalities. With a Bruckner test, the direct ophthalmoscope is held 2–3 feet (60–90 cm) from the child and the red reflexes and corneal light reflexes of both eyes are viewed simultaneously.

Photoscreening is a newer type of RR testing in which a photograph of a child's eyes are analyzed for anomalies. It is especially useful for children who have trouble keeping still during an exam. RR testing for leukocoria—a white pupil reflex—is also sometimes performed by taking a flash photograph of the eyes after the child has been in a dark room for three to five minutes. Leukocoria may be more apparent in flash

photographs because the pupil is exposed to a large amount of light only briefly, so that it does not have time to contract.

Infants and children who are at high risk due to family history of retinoblastoma, congenital, infantile, or juvenile cataracts, congenital retinal dysplasia (abnormal growth in the retina), glaucoma, or other congenital disorders of the lens or retina should have initial RR testing performed with eye-drop-dilation of the pupils. These children should be examined by an experienced pediatric ophthalmologist, even when their RR test results are normal.

Preparation

Sometimes, particularly with high-risk infants and children or when an abnormality is suspected, the pupils are artificially dilated with ophthalmic eye drops or sprays called mydriatic agents prior to RR testing. This ensures that the pupils remain dilated when exposed to bright light. Mydriatic agents are administered about 15 minutes prior to RR testing.

- In infants younger than nine months, combination medication containing 0.2% or 0.25% cyclopentolate and 1% or 2.5% phenylephrine (Cyclomydril) are used.
- Lower concentrations are used for preterm infants.
- In babies older than nine months, one drop of 1% or less tropicamide and/or 2.5% phenylephrine are used.

Aftercare

There is no necessary aftercare for red reflex testing.

Risks

The use of mydriatic agents to dilate infants' pupils for RR testing is somewhat controversial. Some experts contend that infant pupils are so small that there is only a 30% chance of detecting retinoblastoma and other abnormal conditions inside the eye unless the pupil is dilated. Pediatric ophthalmologists routinely use mydriatic agents on infants older than two weeks and they are often used on premature infants in neonatal intensive care units. However, rare, but significant, medical complications have been reported in infants with all commercially available dilating agents and preterm infants may be particularly sensitive. Some practitioners believe that these side effects occur more often than the retinoblastomas detected by

KEY TERMS

Amblyopia—Lazy eye; poor vision in one eye with no apparent structural cause.

Aqueous humor—The clear, watery fluid between the cornea and the crystalline lens of the eye.

Astigmatism—A refractive error caused by an irregular-shaped cornea.

Cataract—Opacity or cloudiness of the eye lens, which can prevent a clear image from forming on the retina.

Cornea—The transparent covering of the iris and pupil that admits light to the interior of the eye.

Glaucoma—Damage to the optic nerve resulting in vision loss and usually accompanied by inflammation and increased pressure in the eye (intraocular pressure).

Iris—Pigmented tissue behind the cornea that gives color to the eye and varies the size of the pupil to control the amount of light entering the eye.

Lens—The transparent biconvex crystalline tissue that focuses light rays on the retina of the eye.

Lenticonus—A rare, usually congenital, condition in which the surface of the lens of the eye is conical.

Leukocoria—A pupil reflex that is white instead of red or that has white spots.

Mydriatic—Causing dilation or widening of the pupil of the eye.

Ocular fundus—The part of the eye opposite the pupil.

Opacity—An opaque spot in a normally transparent structure, such as the lens of the eye.

Ophthalmoscope—An instrument for examining the inner structure of the eye.

Pupil—The black circular opening at the center of the iris that regulates the amount of light that enters the eye.

Refractive error—The inability of the eye to properly focus light due to an irregularly shaped cornea, resulting in blurry vision, nearsightedness, farsightedness, or astigmatism.

Retina—The light-sensitive tissue at the back of the eye.

Retinoblastoma—A hereditary malignant tumor of the retina that develops during childhood.

Retinoscope—An instrument for determining the state of refraction of the eye by illuminating the retina with a mirror.

Strabismus—An imbalance of the eyeball muscles that prevents one eye from attaining binocular vision with the other.

Vitreous body—The transparent jelly that fills the eyeball behind the lens.

RR testing. The AAP recommends dilation with mydriatic agents only if there is evidence of an abnormality.

Reported complications of mydriatic agents include:

- increased blood pressure
- slowed or increased heart rate
- heart arrhythmias
- respiratory depression
- behavioral disturbances
- hives (urticaria)
- contact dermatitis
- discomfort or pain

Results

Results of red reflex testing are considered to be negative or normal if the reflections from the two eyes, viewed both individually and simultaneously, are

symmetrical—equivalent in color, brightness or intensity, size, and clarity—and without opacities or leukocoria (white spots). The reflected color is usually a bright reddish yellow. However the red reflex can vary significantly depending on the child's racial or ethnic background, because of differing levels of pigmentation of the ocular fundus. The reflex may be light gray in dark pigmented, brown-eyed children.

Normal red reflexes indicate that:

- the ocular media are free of opacities
- there are no large refractive errors
- the eyes are aligned

Positive or abnormal RR testing results that require referral to an ophthalmologist include:

- dark spots in the red reflex
- a diminished or blunted red reflex
- opacities or dark spots or white spots in one area of the red reflex
- a white reflex (leukocoria)

- absence of a red reflex
- any differences in red reflex between the two eyes

Abnormalities in red reflexes or asymmetries between the two eyes can be due to:

- mucus or other foreign bodies in the tear film, which move and disappear upon blinking
- unequal or high refractive errors that indicate the need for glasses
- strabismus (eye misalignment)
- opacities in the cornea or aqueous or vitreous media
- abnormalities of the iris that affect the pupil
- cataracts
- glaucoma
- retinoblastoma
- other retinal abnormalities

Specific abnormalities in red reflexes may suggest certain conditions:

- A brighter red reflex in one eye may suggest that the eye is misaligned or strabismic.
- A difference in red reflex color between the two eyes may indicate unequal refractive power in the eyes (anisometropia) and a risk for amblyopia.
- Leukocoria or a white pupil reflex is the most common symptom of retinoblastoma.
- Strabismus is the second most common sign of retinoblastoma.
- Leukocoria can also be due to coloboma—a congenital cleft, fissure, or slit in the eye.
- An absent, dull, or patchy red reflex may suggest a cataract. Cataracts can also cause the pupil to appear white or yellow.
- Although lenticonus is usually detected by a slit-lamp examination, it also appears as a dimple or oil droplet that moves with eye movement in the red reflex.

An abnormal RR testing result is usually followed by RR testing after pupil dilation of each eye and/or referral to a pediatric ophthalmologist. The ophthalmologist will conduct a complete eye exam including an ocular fundus examination by indirect ophthalmoscopy with pupil dilation.

All infants and children with a positive family history of retinoblastoma, congenital, infantile, or juvenile cataracts, glaucoma, or retinal abnormalities should be referred to a pediatric ophthalmologist, regardless of the results of RR testing. The age at which the child should be referred depends on the specific risk factor. However any infant with consistently white pupils requires examination for retinoblastoma. Any infant

with large eyes, excessive tearing, and cloudy corneas should be tested for congenital glaucoma, which can also lead to blindness.

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ORGANIZATIONS

- American Academy of Ophthalmology (AAO), PO Box 7424, San Francisco, CA, 94120–7424, (415) 561–8500, (415) 561–8533, <http://www.aoa.org>.
 American Academy of Pediatrics (AAP), 141 Northwest Point Blvd., Elk Grove Village, IL, 60007–1098, (874) 434–4000, (874) 434–8000, kidsdocs@aap.org, <http://www.aap.org>.
 National Eye Institute (NEI), 31 Center Drive MSC 2510, Bethesda, MD, 20992–3655, (301) 496–5248, 2020@nei.nih.gov, <http://www.nei.nih.gov>.

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Reflex sympathetic dystrophy

Definition

Reflex sympathetic dystrophy is the feeling of pain associated with evidence of minor nerve injury.

Description

Historically, reflex sympathetic dystrophy (RSD) was noticed during the civil war in patients who suffered pain following gunshot wounds that affected the median nerve (a major nerve in the arm). In 1867 the condition was called causalgia from the Greek term meaning "burning pain." Causalgia refers to pain

associated with major nerve injury. The exact causes of RSD are still unclear. Patients usually develop a triad of phases. In the first phase, pain and sympathetic activity is increased. Patients will typically present with swelling (**edema**), stiffness, pain, increased vascularity (increasing warmth), hyperhydrosis, and x-ray changes demonstrating loss of **minerals** in bone (demineralization). The second phase develops three to nine months later. It is characterized by increased stiffness and changes in the extremity that include a decrease in warmth and atrophy of the skin and muscles. The late phase commencing several months to years later presents with a pale, cold, painful, and atrophic extremity. Patients at this stage will also have **osteoporosis**.

It has been thought that each phase relates to a specific nerve defect that involves nerve tracts from the periphery spinal cord to the brain. Both sexes are affected, but the number of new cases is higher in women, adolescents, and young adults. RDS has been associated with other terms such as Sudeck's atrophy, post-traumatic osteoporosis, causalgia, shoulder-hand syndrome, and reflex neuromuscular dystrophy.

Causes and symptoms

The exact causes of RSD at present is not clearly understood. There are several theories such as sympathetic overflow (over activity), abnormal circuitry in nerve impulses through the sympathetic system, and as a post-operative complication for both elective and traumatic surgical procedures. Patients typically develop pain, swelling, temperature, color changes, and skin and muscle wasting.

Diagnosis

The diagnosis is simple and confirmed by a local anesthetic block along sympathetic nerve paths in the hand or foot, depending on whether an arm or leg is affected. A test called the **erythrocyte sedimentation rate** (ESR) can be performed to rule out diseases with similar presentation and arising from other causes.

Treatment

The preferred method to treat RSD includes sympathetic block and **physical therapy**. Pain is improved as motion of the affected limb improves. Patients may also require tranquilizers and mild **analgesics**. Patients who received repeated blocks should consider surgical sympathectomy (removal of the nerves causing pain).

KEY TERMS

Atrophy—Abnormal changes in a cell that lead to loss of cell structure and function.

Osteoporosis—Reduction in the quantity of bone.

Prognosis

The prognosis for treatment during phase one is favorable. As the disease progresses undetected into phase two or three the prognosis for recovery is poor.

Prevention

There is no known prevention since the cause is not clearly understood.

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Laith Farid Gulli, M.D.
Robert Ramirez, B.S.

Reflex tests

Definition

Reflex tests are simple physical tests of nervous system function.

Purpose

A reflex is a simple nerve circuit. A stimulus, such as a light tap with a rubber hammer, causes sensory neurons (nerve cells) to send signals to the spinal cord. Here, the signals are conveyed both to the brain and to nerves that control muscles affected by the stimulus. Without any brain intervention, these muscles may respond to an appropriate stimulus by contracting.

Reflex tests measure the presence and strength of a number of reflexes. In so doing, they help to assess the integrity of the nerve circuits involved. Reflex tests

are performed as part of a **neurological exam**, either a “mini-exam” done to quickly confirm integrity of the spinal cord, or a more complete exam performed to diagnose the presence and location of **spinal cord injury** or neuromuscular disease.

Deep tendon reflexes are responses to muscle stretch. The familiar “knee-jerk” reflex is an example; this reflex tests the integrity of the spinal cord in the lower back region. The usual set of deep tendon reflexes tested, involving increasingly higher regions of the spinal cord, are:

- ankle
- knee
- abdomen
- forearm
- biceps
- triceps

Another type of reflex test is called the Babinski test, which involves gently stroking the sole of the foot to assess proper development of the spine and cerebral cortex.

Precautions

Reflex tests are entirely safe, and no special precautions are needed.

Description

The examiner positions the patient in a comfortable position, usually seated on the examination table with legs hanging free. The examiner uses a rubber mallet to strike different points on the patient’s body, and observes the response. The examiner may position, or hold, one of the limbs during testing, and may require exposure of the ankles, knees, abdomen, and arms. Reflexes can be difficult to elicit if the patient is paying too much attention to the stimulus. To compensate for this, the patient may be asked to perform some muscle contraction, such as clenching teeth or grasping and pulling the two hands apart. When performing the Babinski reflex test, the doctor will gently **stroke** the outer soles of the patient’s feet with the mallet while checking to see whether or not the big toe extends out as a result.

Normal results

The strength of the response depends partly on the strength of the stimulus. For this reason, the examiner will attempt to elicit the response with the smallest stimulus possible. Learning the range of normal responses requires some clinical training. Responses should be the same for both sides of the body. A normal response to

the Babinski reflex test depends upon the age of the person being examined. In children under the age of one and a half years, the big toe will extend out with or without the other toes. This is due to the fact that the fibers in the spinal cord and cerebral cortex have not been completely covered in myelin, the protein and lipid sheath that aids in processing neural signals. In adults and children over the age of one and a half years, the myelin sheath should be completely formed, and, as a result, all the toes will curl under (planter flexion reflex).

Abnormal results

Weak or absent response may indicate damage to the nerves outside the spinal cord (**peripheral neuropathy**), damage to the motor neurons just before or just after they leave the spinal cord (motor neuron disease), or muscle disease. Excessive response may indicate spinal cord damage above the level controlling the hyperactive response. Different responses on the two sides of the body may indicate early onset of progressive disease, or localized nerve damage, as from trauma. An adult or older child who responds to the Babinski with an extended big toe may have a lesion in the spinal cord or cerebral cortex.

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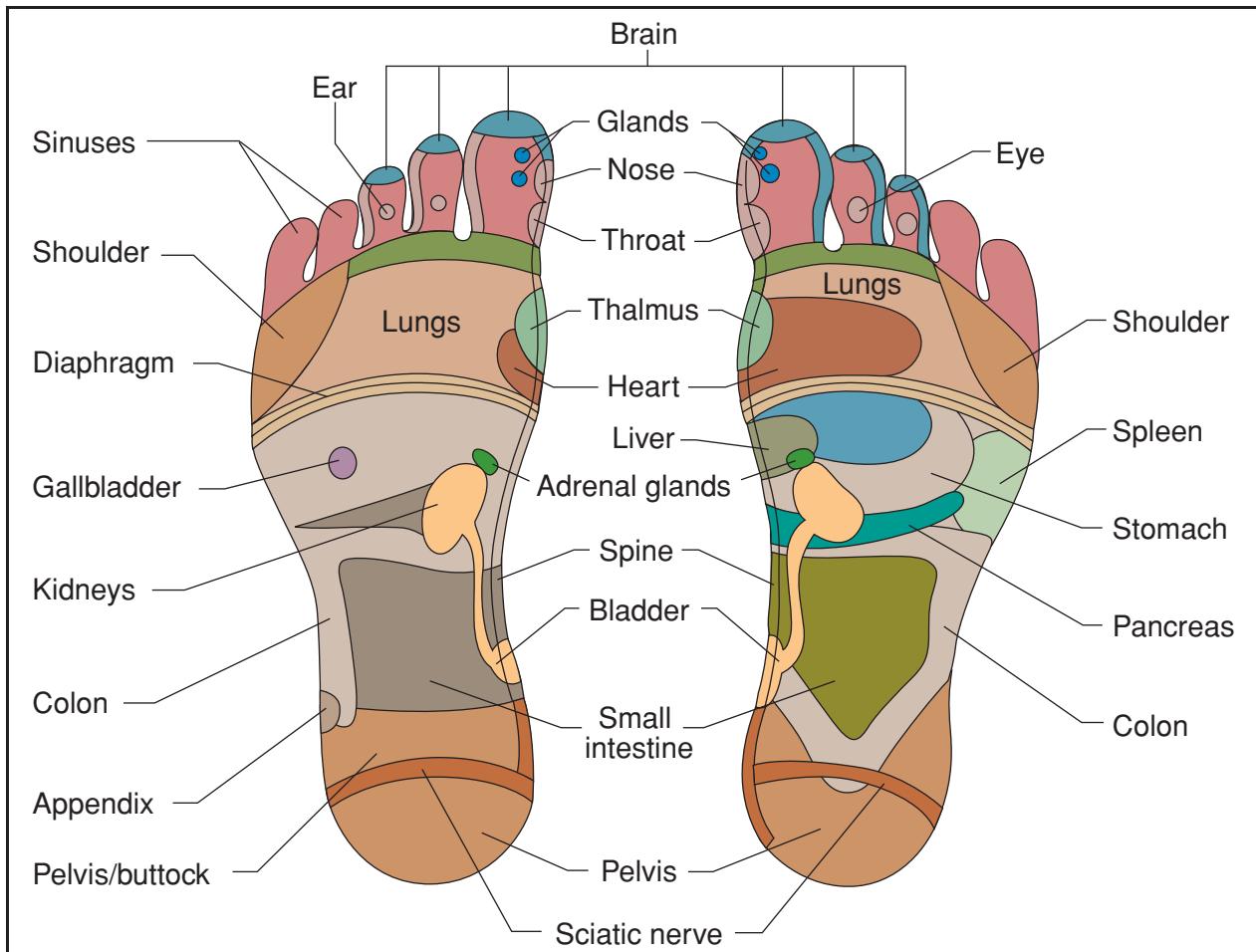
Richard Robinson

Reflexology

Definition

Reflexology is a therapeutic method of relieving **pain** by stimulating predefined pressure points on the feet and hands. This controlled pressure alleviates the source of the discomfort. In the absence of any particular malady or abnormality, reflexology may be as effective for promoting good health and for preventing illness as it may be for relieving symptoms of **stress**, injury, and illness.

Reflexologists work from maps of predefined pressure points that are located on the hands and feet. These pressure points are reputed to connect directly through the nervous system and affect the bodily organs and glands. The reflexologist manipulates the pressure points according to specific techniques of reflexology



Reflexology employs the principle that the reflex points on the feet, when hand pressure is applied, will reflexively stimulate energy to a related muscle or organ in the body and promote healing. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

therapy. By means of this touching therapy, any part of the body that is the source of pain, illness, or potential debility can be strengthened through the application of pressure at the respective foot or hand location.

Purpose

Reflexology promotes healing by stimulating the nerves in the body and encouraging the flow of blood. In the process, reflexology not only quells the sensation of pain, but relieves the source of the pain as well.

Anecdotally, reflexologists claim success in the treatment of a variety of conditions and injuries. One condition is fibromyalgia. People with this disease are encouraged to undergo reflexology therapy to alleviate any of a number of chronic bowel syndromes associated with the condition. Frequent brief sessions of reflexology therapy are also recommended as an

alternative to drug therapy for controlling the muscle pain associated with fibromyalgia and for relieving difficult breathing caused by tightness in the muscles of the patient's neck and throat.

Reflexology applied properly can alleviate allergy symptoms, as well as stress, back pain, and chronic **fatigue**. The techniques of reflexology can be performed conveniently on the hand in situations where a session on the feet is not practical, although the effectiveness of limited hand therapy is less pronounced than with the foot pressure therapy.

Description

Origins

Reflexology is a healing art of ancient origin. Although its origins are not well documented, there

are reliefs on the walls of a Sixth Dynasty Egyptian tomb (c. 2450 B.C.) that depict two seated men receiving massage on their hands and feet. From Egypt, the practice may have entered the Western world during the conquests of the Roman Empire. The concepts of reflexology have also been traced to pre-dynastic China (possibly as early as 3000 B.C.) and to ancient Indian medicine. The Inca civilization may have subscribed to the theories of reflexology and passed on the practice of this treatment to the Native Americans in the territories that eventually entered the United States.

In recent times, Sir Henry Head first investigated the concepts underlying reflexology in England in the 1890s. Therapists in Germany and Russia were researching similar notions at approximately the same time, although with a different focus. Less than two decades later, a physician named William H. Fitzgerald presented a similar concept that he called zone analgesia or zone therapy. Fitzgerald's zone analgesia was a method of relieving pain through the application of pressure to specific locations throughout the entire body. Fitzgerald divided the body into 10 vertical zones, five on each side, that extended from the head to the fingertips and toes, and from front to back. Every aspect of the human body appears in one of these 10 zones, and each zone has a reflex area on the hands and feet. Fitzgerald and his colleague, Dr. Edwin Bowers, demonstrated that by applying pressure on one area of the body, they could anesthetize or reduce pain in a corresponding part. In 1917, Fitzgerald and Bowers published *Relieving Pain at Home*, an explanation of zone therapy.

Later, in the 1930s, a physical therapist, Eunice D. Ingham, explored the direction of the therapy and made the startling discovery that pressure points on the human foot were situated in a mirror image of the corresponding organs of the body with which the respective pressure points were associated. Ingham documented her findings, which formed the basis of reflexology, in *Stories the Feet Can Tell*, published in 1938. Although Ingham's work in reflexology was inaccurately described as zone therapy by some, there are differences between the two therapies of pressure analgesia. Among the more marked differences, reflexology defines a precise correlation between pressure points and afflicted areas of the body. Furthermore, Ingham divided each foot and hand into 12 respective pressure zones, in contrast to the 10 vertical divisions that encompass the entire body in Fitzgerald's zone therapy.

In 1968 two siblings, Dwight Byers and Eusebia Messenger, established the National Institute of

EUNICE INGHAM (1889–1974)

Eunice D. Ingham was born on February 24, 1889. A physical therapist by occupation, she was a colleague of Dr. Shelby Riley, who along with Dr. W. H. Fitzgerald actively developed zone therapy, a similar but distinct therapy from reflexology. Unlike reflexology, zone therapy does not connect the zones with the body as a whole. In the 1930s, Ingham discovered an unmistakable pattern of reflexes on the human foot; she subsequently devoted the rest of her life to publicizing the message of reflexology until shortly before her death on December 10, 1974.

Ingham traveled and lectured widely about reflexology, initially to audiences of extremely desperate or aging patients who had lost hope in finding relief. Because of their sometimes astonishing improvement, reflexology became better known and respected among the medical community and gained credibility for its therapeutic value. Ingham described her theories of reflexology in her 1938 book, entitled *Stories the Feet Can Tell*, which included a map of the reflex points on the feet and the organs that they parallel. The book was translated into seven languages, although it was erroneously published as *Zone Therapy* in some countries, an error which led to misunderstanding about the true nature of reflexology and inaccurately linked it to zone therapy.

Reflexology. By the early 1970s the institute had grown and was renamed the International Institute of Reflexology.

In a typical reflexology treatment, the therapist and patient have a preliminary discussion prior to therapy, to enable the therapist to focus more accurately on the patient's specific complaints and to determine the appropriate pressure points for treatment.

A reflexology session involves pressure treatment that is most commonly administered in foot therapy sessions of approximately 40–45 minutes in duration. The foot therapy may be followed by a brief 15-minute hand therapy session. No artificial devices or special equipment are associated with this therapy. The human hand is the primary tool used in reflexology. The therapist applies controlled pressure with the thumb and forefinger, generally working toward the heel of the foot or the outer palm of the hand. Most reflexologists apply pressure with their thumbs bent; however, some also use simple implements, such as the eraser end of a pencil. Reflexology therapy is not massage, and it is not a substitute for medical treatment.

Reflexology is a complex system that identifies and addresses the mass of 7,000 nerve endings that are contained in the foot. Additional reflexology addresses the nerves that are located in the hand. This is a completely natural therapy that affords relief without the use of drugs. The Reflexology Association of America (RAA) formally discourages the use of oils or other preparations in performing this hands-on therapy.

Preparations

In order to realize maximum benefit from a reflexology session, the therapist as well as the patient should be situated so as to afford optimal comfort for both. Patients in general receive treatment in a reclining position, with the therapist positioned as necessary—to work on the bare feet, or alternately on the bare hands.

A reflexology patient removes both shoes and socks in order to receive treatment. No other preparation is involved. No prescription drugs, creams, oils, or lotions are used on the skin.

Precautions

Reflexology is extremely safe. It may even be self-administered in a limited form whenever desired. The qualified reflexologist offers a clear and open disclaimer that reflexology does not constitute medical treatment in any form, nor is reflexology given as a substitute for medical advice or treatment. The ultimate purpose of the therapy is to promote wellness; fundamentally it is a form of preventive therapy.

People with serious and long-term medical problems are urged to seek the advice of a physician. Diabetes patients in particular are urged to approach this therapy cautiously. Likewise pregnant women are cautioned emphatically to avoid reflexology during the early phases of **pregnancy** altogether, as accidentally induced labor and subsequent premature delivery can result from reflexology treatment.

A consultation with a reflexologist is recommended in order to determine the safety and appropriateness of reflexology therapy for a specific health problem or condition.

Side effects

Because reflexology is intended to normalize the body functions, the therapy does not cause a condition to worsen. Most patients find that pain diminishes over the course of the therapy. It has been noted, however, that some patients experience greater

discomfort in the second session than in the first session, because a significant easing of pain and tension is generally associated with the initial therapy session. As a result, when pressure is reapplied to the tender points of the foot during the second session, the sensitivity has been heightened. This increase in sensitivity may cause minor additional discomfort for the patient.

Research and general acceptance

Although only one controlled trial of reflexology therapy, done in 1993, has been documented in medical journals, this therapy is practiced worldwide at different levels of medical care. In Russia, for example, only licensed physicians may legally perform reflexology treatment. In contrast, the practice is a commonplace homestyle remedy in the Netherlands. The Internet “Home of Reflexology” lists at least 66 professional organizations worldwide, including New Zealand and Malaysia. Associations include the following:

- Academy of Reflexology Austria
- Association of Finnish Reflexologists
- Chinese Society of Reflexologists
- Hellenic Association of Reflexologists
- Indian Society for Promotion of Reflexology
- International Council of Reflexologists (HQ: San Diego, USA)
- Israeli Reflexology Association
- New Zealand Reflexology Association
- Polish Instytut of Reflexology (Polish Language)
- Reflexology Association of America
- Reflexology Association of Australia
- Rwo-Shr Health Institute International (Malaysia)
- The South African Reflexology Society

Regulatory status

Ongoing legislative debate ensued during the 1990s regarding the legal status of the reflexology trade. The reflexology community, along with legislators and other bodywork practitioners, engaged in reassessment of the reflexology business and its relationship to **massage therapy** and massage parlors. Organizations and individuals brought judicial appeals of certain court cases that threatened the legitimate licensing of reflexologists as practitioners of alternative medicine. Such professional reflexology interests as the RAA documented in detail the disparities between reflexology and massage, citing the purpose of reflexology, which is to stimulate internal body functions (glands and

organs) as opposed to the topical muscular and joint relief associated with massage. In a status update in 1998 the Association reported that 19 states had laws requiring the licensing of massage/reflexology therapists. Licensing laws established educational requirements and required candidates to pass written, oral, and/or practical examinations.

Also at issue was a trend among municipalities to license massage parlors (and reflexologists) under the business codes affecting the adult entertainment business. B. and K. Kunz reported that judicial decisions in two states—Tennessee and New Mexico—had excluded the practice of reflexology practice from the laws pertaining to massage parlors. Those courts held that reflexology is a business separate and distinct from massage parlors, and deserving of its own respective licensing standards. In Sacramento, California, reflexologists petitioned successfully to become licensed as practitioners of somatic therapy rather than as providers of adult entertainment. Likewise, in the Canadian province of Ontario, a nonprofit organization to register reflexology practitioners was established in order to define a distinct classification for therapists separate from erotic body rubbers, which was the original classification given to reflexologists. Other states where court proceedings or legislative attempts to legitimize reflexology have stalled include Pennsylvania, Florida, New Jersey, and New York.

Training and certification

Training programs

Reflexology is taught by means of a series of seminars, classes, and training films. Certification is earned after a six month program that includes 200 hours of training. The certification training breaks down as follows: 28 hours of preliminary seminar training; 14 hours of advanced seminar training; 58 hours of self-directed study; and 100 hours of practical experience, including administering reflexology to a minimum of 15 people.

Specific aspects of the training include instruction in the assessment of the pressure points on the feet and hands through a study of human anatomy. Students also learn to give reflexology sessions to patients along with specific techniques for working with the hands.

Certification and advanced certification

As part of its function, the independently organized American Reflexology Certification

KEY TERMS

Pressure points—Specific locations on the feet and hands that correspond to nerve endings that connect to the organs and glands of the human body via the spinal cord.

Zone therapy—Also called zone analgesia, a method of relieving pain by applying pressure to specific points on the body. It was developed in the early twentieth century by Dr. William Fitzgerald.

Board (ARCB) certifies the competency of reflexology practitioners on an individual basis. The ARCB does not evaluate schools and teachers. Prerequisites for individual certification include completion of educational requirements and passing a standard qualifying examination. Successful candidates receive the title of Board Certified Reflexologist.

Minimum qualifications to take the certification examination include attendance at an advanced seminar within two years prior to taking the examination. In addition, the applicant must have attended preliminary seminars for two full days—in addition to the required day of advanced seminar training—and the applicant is required to have a minimum of six months of practical experience in administering the therapy. Applicants are examined by means of both written tests and practical demonstrations.

Continuing education certification is available. Advanced training focuses on mastering the ability to perform hand reflexology. The therapist also receives instruction in new and advanced techniques of basic reflexology. Some reflexology training classes may be applied toward degree programs in other disciplines, depending on the specific course of study and the certification of the respective training institutions involved.

The RAA provides published standards of practice for reflexologists.

ORGANIZATIONS

International Institute of Reflexology, P.O. Box 12642, St. Petersburg, FL, 33733-2642, (727) 343-4811, (727) 381-2807, info@reflexology-usa.net, <http://www.reflexology-usa.net/>.

Reflexology Association of America, P.O. Box 714, Chepachet, RI, 02814, (980) 234-0159, (401) 568-6449, InfoRAA@ reflexology-usa.org, http://www.reflexology-usa.org.

Gloria Cooksey

Refsum's syndrome see **Lipidoses**

Regional anesthetic see **Anesthesia, local**

Regional enteritis see **Crohn's disease**



Rehabilitation

Definition

Rehabilitation is a treatment or treatments designed to facilitate the process of recovery from injury, illness, or disease to as normal a condition as possible.

Purpose

The purpose of rehabilitation is to restore some or all of the patient's physical, sensory, and mental capabilities that were lost due to injury, illness, or disease. Rehabilitation includes assisting the patient to compensate for deficits that cannot be reversed medically. It is prescribed after many types of injury, illness, or disease, including amputations, arthritis, **cancer**, cardiac disease, neurological problems, orthopedic injuries, spinal cord injuries, **stroke**, and traumatic brain injuries. The Institute of Medicine has estimated that as many as 14% of all Americans may be disabled at any given time.

Precautions

Rehabilitation should be carried out only by qualified therapists. Exercises and other physical interventions must take into account the patient's deficit. An example of a deficit is the loss of a limb.

Description

A proper and adequate rehabilitation program can reverse many disabling conditions or can help patients cope with deficits that cannot be reversed by medical care. Rehabilitation addresses the patient's physical, psychological, and environmental needs. It is achieved by restoring the patient's physical functions and/or modifying the patient's physical and

A man who suffered a stroke is helped with his rehabilitation by a physical therapist. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

social environment. The main types of rehabilitation are physical, occupational, and **speech therapy**.

Each rehabilitation program is tailored to the individual patient's needs and can include one or more types of therapy. The patient's physician usually coordinates the efforts of the rehabilitation team, which can include physical, occupational, speech, or other therapists; nurses; engineers; physiatrists (physical medicine); psychologists; orthotists (makes devices such as braces to straighten out curved or poorly shaped bones); prosthetists (a therapist who makes artificial limbs or prostheses); and vocational counselors. Family members are often actively involved in the patient's rehabilitation program.

Physical therapy

Physical therapy helps the patient restore the use of muscles, bones, and the nervous system through the use of heat, cold, massage, whirlpool baths, ultrasound, **exercise**, and other techniques. It seeks to relieve **pain**, improve strength and mobility, and train the patient to perform important everyday tasks. Physical therapy may be prescribed to rehabilitate a patient after amputations, arthritis, **burns**, cancer, cardiac disease, cervical

and lumbar dysfunction, neurological problems, orthopedic injuries, pulmonary disease, spinal cord injuries, stroke, traumatic brain injuries, and other injuries/illnesses. The duration of the physical therapy program varies depending on the injury/illness being treated and the patient's response to therapy.

Exercise is the most widely used and best known type of physical therapy. Depending on the patient's condition, exercises may be performed by the patient alone or with the therapist's help, or with the therapist moving the patient's limbs. Exercise equipment for physical therapy could include an exercise table or mat, a stationary bicycle, walking aids, a wheelchair, practice stairs, parallel bars, and pulleys and weights.

Heat treatment, applied with hot-water compresses, infrared lamps, short-wave radiation, high frequency electrical current, ultrasound, paraffin wax, or warm baths, is used to stimulate the patient's circulation, relax muscles, and relieve pain. Cold treatment is applied with ice packs or cold-water soaking. Soaking in a whirlpool can ease muscle spasm pain and help strengthen movements. Massage aids circulation, helps the patient relax, relieves pain and **muscle spasms**, and reduces swelling. Very low strength electrical currents applied through the skin stimulate muscles and make them contract, helping paralyzed or weakened muscles respond again.

Occupational therapy

Occupational therapy helps the patient regain the ability to do normal everyday tasks. This may be achieved by restoring old skills or teaching the patient new skills to adjust to disabilities through adaptive equipment, orthotics, and modification of the patient's home environment. Occupational therapy may be prescribed to rehabilitate a patient after **amputation**, arthritis, cancer, cardiac disease, head injuries, neurological injuries, orthopedic injuries, pulmonary disease, spinal cord disease, stroke, and other injuries/illnesses. The duration of the occupational therapy program varies depending on the injury/illness being treated and the patient's response to therapy.

Occupational therapy includes learning how to use devices to assist in walking (artificial limbs, canes, crutches, walkers), getting around without walking (wheelchairs or motorized scooters), or moving from one spot to another (boards, lifts, and bars). The therapist will visit the patient's home and analyze what the patient can and cannot do. Suggestions on modifications to the home, such as rearranging furniture or adding a wheelchair ramp, will be made. Health aids to bathing and grooming could also be recommended.

KEY TERMS

Orthotist—A health care professional who is skilled in making and fitting orthopedic appliances.

Physiatrist—A physician who specializes in physical medicine.

Prosthetist—A health care professional who is skilled in making and fitting artificial parts (prosthetics) for the human body.

Speech therapy

Speech therapy helps the patient correct **speech disorders** or restore speech. Speech therapy may be prescribed to rehabilitate a patient after a brain injury, cancer, neuromuscular diseases, stroke, and other injuries/illnesses. The duration of the speech therapy program varies depending on the injury/illness being treated and the patient's response to therapy.

Performed by a speech pathologist, speech therapy involves regular meetings with the therapist in an individual or group setting and home exercises. To strengthen muscles, the patient might be asked to say words, smile, close his mouth, or stick out his tongue. Picture cards may be used to help the patient remember everyday objects and increase his vocabulary. The patient might use picture boards of everyday activities or objects to communicate with others. Workbooks might be used to help the patient recall the names of objects and practice reading, writing, and listening. Computer programs are available to help sharpen speech, reading, recall, and listening skills.

Other types of therapists

Inhalation therapists, audiologists, and registered dietitians are other types of therapists. Inhalation therapists help the patient learn to use respirators and other breathing aids to restore or support breathing. Audiologists help diagnose the patient's **hearing loss** and recommend solutions. Dietitians provide dietary advice to help the patient recover from or avoid specific problems or diseases.

Rehabilitation centers

Rehabilitation services are provided in a variety of settings including clinical and office practices, hospitals, skilled-care nursing homes, sports medicine clinics, and some health maintenance organizations. Some therapists make home visits. Advice on choosing the

appropriate type of therapy and therapist is provided by the patient's medical team.

ORGANIZATIONS

National Rehabilitation Association, 633 S. Washington St., Alexandria, VA, 22314, (703) 836-0850.

National Rehabilitation Information Center, 8201 Corporate Drive, Suite 600, Landover, MD, 20785, (800) 346-2742, naricinfo@heitechservices.com, <http://www.naric.com>.

Rehabilitation International, 25 East 21st Street, 4th floor, New York, NY, 10010, (212) 420-1500, 212 505-0871, ri@riglobal.org, <http://www.riglobal.org>.

Lori De Milto

Rehydration see **Intravenous rehydration**

Reiki

Definition

Reiki is a form of therapy that uses simple hands-on, no-touch, and visualization techniques, with the goal of improving the flow of life energy in a person. Reiki (pronounced *ray-key*) means "universal life energy" in Japanese, and Reiki practitioners are trained to detect and alleviate problems of energy flow on the physical, emotional, and spiritual level. Reiki touch therapy is used in much the same way to achieve similar effects that traditional **massage therapy** is used—to relieve **stress** and **pain**, and to improve the symptoms of various health conditions.

Purpose

Reiki claims to provide many of the same benefits as traditional massage therapy, such as reducing stress, stimulating the immune system, increasing energy, and relieving the pain and symptoms of health conditions. Practitioners have reported success in helping patients with acute and chronic illnesses, from **asthma** and arthritis to trauma and recovery from surgery. Reiki is a gentle and safe technique, and has been used successfully in some hospitals. It has been found to be very calming and reassuring for those suffering from severe or fatal conditions. Reiki can be used by doctors, nurses, psychologists and other health professionals to bring touch and deeper caring into their healing practices.

Description

Origins

Reiki was developed in the mid-1800s by Dr. Mikao Usui, a Japanese scholar of religion. According to the story that has been passed down among reiki teachers, Usui was a Christian who was intrigued by the idea that Christ could heal sick people by touching them with his hands. Searching for clues that would explain the secrets of healing with hands, Usui made a long pilgrimage around the world, visiting many ancient religious sects and studying ancient books. Some reiki teachers claim that Usui found clues leading back nearly 10,000 years to healing arts that originated in ancient Tibet. During his intense studies, Usui claimed he had a spiritual experience, which enabled him to heal with his own hands by becoming aware of and tapping into the universal life force. After that, he dedicated his life to helping the sick and poor. His reputation grew as he healed sick people for many years in Kyoto, Japan. Before his **death**, Usui passed on his healing insights using universal life energy to Dr. Chujiru Hayashi, a close acquaintance. Hayashi, in turn, passed on the healing techniques in 1938 to Hawayo Takata, a Japanese woman from Hawaii, whom he had cured of life-threatening illness using reiki methods. Takata became a firm believer and proponent of reiki, and during the 1970s formed an initiation program for training reiki masters to preserve Usui's teachings. Before she died, she prepared her granddaughter, Phyllis Lei Furumoto, to continue the lineage. Takata had personally trained 21 practitioners before she died at the age of 80 in 1980. Along with other reiki masters authorized by Takata, Furumoto formed the reiki Alliance. A faction led by Barbara Ray, formed the American Reiki Association, which was known as Radiance Technique Association International. Today, there are over 1,000 reiki masters practicing around the world, whose methods can all be traced back directly to Dr. Usui.

The basic philosophy of reiki

The basic concept underlying reiki is that the body has an energy field that is central to its health and proper functioning, and this energy travels in certain pathways that can become blocked or weakened. This idea of energy flow in the body is also a central concept in **Ayurvedic medicine** and **traditional Chinese medicine**, including **acupuncture**.

Reiki practitioners believe that everyone has the potential to access the universal life energy, but that over time most people's systems become blocked and the energy becomes weakened in them. A reiki

MIKAO USUI (1865–1926)

Mikao Usui, born in the Gifu Prefecture (Japan), was an ethereal child who sought to unravel the mysteries of the universe. As an adult he developed an interest in the metaphysical healing talent of Buddha. Usui became determined to regenerate the healing secrets of Buddha in order to improve the lot of humanity. He traveled to many temples and spoke with holy people, but all said that the secret of Buddha's powers were lost to the world due to lack of use.

Eventually the abbot of a Zen monastery encouraged Usui to study the ancient writings containing the secrets on healing. Usui learned two new languages, Chinese and Sanskrit, in order to understand the writings better, and from his reading he obtained the formula for healing. The Sutras in particular provided the enlightenment that he sought.

Usui next set out to obtain the power to heal. It is widely believed that he developed that ability after spending 21 days in retreat and in fasting on the holy Mountain of Kori-yama, where he had a vision of light and received the knowledge of the symbols of reiki and their use in healing. He officially formulated Usui Reiki therapy in 1922 and touted as many as one million followers during his lifetime.

Prior to the transition (death) of Usui, he imparted the secrets of healing to 16 teachers in order that the secrets would not be lost again.

practitioner is trained to be able to detect these blockages, and practitioners will use their hands, thoughts, and own energy fields to improve the energy flow in a patient. Reiki is one of the more esoteric alternative medical practices, because no one is sure exactly how it works on the physiological level. Practitioners claim that it works on very subtle energy levels, or possibly works on the *chakra* system. The chakras are the system of seven energy centers along the middle of the body believed to be connected with the nervous and endocrine systems, as defined by **yoga** and Ayurvedic medicine. Reiki masters claim that healing energy can even be sent to a person from far away, noting that reiki works on the same principles that enables praying to work for some patients, although a practitioner needs advanced training to be able to send energy from afar.

According to the original principles of Usui, patients must also have a proper attitude for reiki to work most effectively. Patients must take responsibility for their own health, and must want to be healed.

Furthermore, when energy is received from a reiki healer, patients must be willing to give back energy to others, and to compensate the healer in some way, as well. Finally, Usui claimed that a healing attitude was free from worry and fear, was filled with gratitude for life and for others, and placed emphasis on each person finding honest and meaningful work in their lives—all this, in order to complete the picture of overall health.

A reiki session

Reiki sessions can take various forms, but most commonly resemble typical bodywork appointments, where the receiver lies clothed on his or her back on a flat surface or massage table. A session generally lasts from an hour to an hour and a half. Reiki is a simple procedure, consisting of calm and concentrated touching, with the practitioner focusing on healing and giving energy to specific areas on the receiver's body. Practitioners place their hands over positions on the body where the organs and endocrine glands reside, and the areas that correspond to the chakra centers. Practitioners also use mental visualization to send healing energy to areas of the receiver's body that need it. In special cases or with injuries, a no-touch technique is used, where the practitioner's hands are sometimes held just above the body without touching it. Advanced practitioners rely on intuition and experience to determine which areas of a body need the most energy healing.

The practitioner's hands are held flat against the receiver's body, with the fingertips touching. There can be more than 20 positions on both sides of the body where the hands are placed. The positions begin at the crown of the head and move towards the feet. The receiver usually turns over once during the session. The practitioner's hands are held in each position for a usually five minutes, to allow the transfer of energy and the healing process to take place. In each position, the hands are kept stationary, unlike typical massage where the hands move, and both the giver and receiver attempt to maintain an attitude of awareness, openness, and caring.

Reiki practitioners recommend that those receiving reiki for the first time go through a series of three to four initial treatments over the course of about a week, to allow for cleansing and the initial readjustment of energy. Reiki sessions can cost from \$30–100 per session. Insurance coverage is rare, and consumers should consult their individual policies as to whether or not such therapies are included.

Self-treatment with reiki

Although reiki practitioners believe that formal training is necessary to learn the proper methods of energy channeling and healing, individuals can still use some of the basic positions of reiki to relieve stress and to stimulate healing on themselves or another. The positions can be performed anywhere and for however long they are needed. Positions generally move from the top of the body down, but positions can be used wherever there is pain or stress. Mental attitude is important during reiki; the mind should be cleared of all stressful thoughts and concentrated on compassion, love, and peace as forms of energy that are surrounding, entering, and healing the body.

The following positions are illustrated in *Reiki: Energy Medicine*:

- Position one: Hands are placed on the top of the head, with the wrists near the ears and the fingertips touching on the crown of the head. Eyes should be closed. Hold for five minutes or more, until the mind feels clear and calm.
- Position two: Cup the hands slightly and place the palms over the closed eyes, with the fingers resting on the forehead.
- Position three: Place the hands on the sides of the head, with the thumbs behind the ear and the palms over the lower jaws, with the fingers covering the temples.
- Position four: Place one hand on the back of the neck, at the base of the skull, and put the other hand on the head just above it, parallel to it.
- Position five: Wrap the hands around the front of the throat, and rest them gently with the heels of the hands touching in front.
- Position six: Place each hand on top of a shoulder, close to the side of neck, on top of the trapezius muscle.
- Position seven: Form a T-shape with the hands over the chest, with the left hand covering the heart and the right hand above it, covering the upper part of the chest.
- Position eight: The hands are placed flat against the front of the body with fingertips touching. Hold for five minutes or so, and repeat four or five times, moving down a hand-width each time until the pelvic region is reached, which is covered with a v-shape of the hands. Then, for the final position, repeat this technique on the back, beginning as close to the shoulders as the hands can reach, and ending by forming a T-shape with the hands at the base of the spine.

KEY TERMS

Attunement—Life energy teaching given by Reiki master to a student.

Chakra—One of seven major energy centers in the body, as defined by Hindu and yoga philosophy.

Relaxation response—The human body's response to relaxation techniques, during which metabolism and stress levels decrease and immune response increases.

Side effects

Reiki generally has no side effects, as it is a very low impact and gentle procedure. Some receivers report **tingling** or sensations of heat or cold during treatment. Others have reported sadness or **anxiety** during treatment, which practitioners claim are buried or repressed emotions being released by the new energy flow.

Research and general acceptance

Reiki has been used in major clinics and hospitals as part of alternative healing practice, and doctors, dentists, nurses and other health professionals have been trained to use its gentle touch techniques as part of their practice. To date, the little scientific research that has been conducted with reiki implies that its techniques bring about the *relaxation response*, in which stress levels decrease, and immune response increases. Reiki practitioners claim that the most important measurement of their technique is whether the individual feels better after treatment. They also claim that science cannot measure the subtle energy changes that they are attempting to bring about.

There are differences of opinion within the mainstream medical community regarding the acceptability of reiki. On the one hand, medical professionals in Canada have proposed strategies to limit the popularity of reiki as well as several other alternative therapies by resisting the integration of these therapies with mainstream treatments and by opposing government research in complementary and alternative medicine. On the other hand, the U. S. National Center for Complementary and Alternative Medicine (NCCAM) is conducting a series of clinical trials to evaluate the efficacy of reiki. As of the summer of 2004, there were four NCCAM trials for reiki, measuring

its effectiveness in treating such disorders as fibromyalgia, neuropathy, **prostate cancer**, and advanced **AIDS**.

Training and certification

Reiki practitioners undergo a series of *attunements*, which are sessions with reiki masters that teach the basic methods of energy healing. Several organizations provide resources for reiki training. Reiki practitioners believe these attunements are necessary for correct technique. The masters teach each person how to activate the universal life energy in themselves before they can pass it on to others. These initiations often are held during weekend workshops. Trainees can achieve up to four levels of attunements, until they reach the level of master themselves. The certification process is not a formal one; masters approve students when they feel satisfied with their progress.

Resources

PERIODICALS

Hallett, A. "Narratives of Therapeutic Touch." *Nursing Standard* 19 (September 15, 2004): 33–37.

Kelner, M., B. Wellman, H. Boon, and S. Welch.

"Responses of Established Healthcare to the Professionalization of Complementary and Alternative Medicine in Ontario." *Social Science and Medicine* 59 (September 2004): 915–930.

OTHER

American Reiki Masters Association (ARMA). P.O. Box 130, Lake City, FL 32056-0130. (904) 755-9638.

Global Reiki Healing Network. <http://www.reiki.org>.

NCCAM Reiki Clinical Trials. <http://nccam.nih.gov/clinicaltrials/reiki.htm>.

Reiki Alliance. P.O. Box 41, Cataldo, ID 83810-1041, phone (208) 682-3535.

ORGANIZATIONS

International Association of Reiki Professionals, info@iarp.org, <http://www.iarpreeiki.org/>.

The International Center for Reiki Training, 21421 Hilltop Street, Unit #28, Southfield, MI, 48033, (248) 948-8112, (248) 948-9534, (800) 332-8112, center@reiki.org, <http://www.reiki.org/>.

National Center for Complementary and Alternative Medicine (NCCAM), P.O. Box 7923, Gaithersburg, MD, 20898, (866) 464-3616, (888) 644-6226, info@nccam.nih.gov, <http://nccam.nih.gov/>.

Douglas Dupler, MA
Rebecca J. Frey, PhD

Reiter's syndrome

Definition

Reiter's syndrome (RS), which is also known as arthritis urethritis, venereal arthritis, reactive arthritis, and polyarteritis enterica, is a form of arthritis that affects the eyes, urethra, and skin, as well as the joints. It was first described by Hans Reiter, a German physician, during World War I.

Description

Reiter's syndrome is marked by a cluster of symptoms in different organ systems of the body that may or may not appear simultaneously. The disease may be acute or chronic, with spontaneous remissions or recurrences.

RS primarily affects sexually active males between ages 20–40, particularly males who are HIV positive. Most women and children who develop RS acquire the disease in its intestinal form.

Causes and symptoms

The cause of Reiter's syndrome was unknown as of early 1998, but scientists think the disease results from a combination of genetic vulnerability and various disease agents. More than 80% of Caucasian patients and 50–60% of African Americans test positive for HLAB27, which suggests that the disease has a genetic component. In sexually active males, most cases of RS follow infection with *Chlamydia trachomatis* or *Ureaplasma urealyticum*. Other patients develop the



Keratoderma, a skin condition characterized by horny patches, is one symptom of Reiter's syndrome. (© Dr. Milton Reisch/Corbis.)

symptoms following gastrointestinal infection with *Shigella*, *Salmonella*, *Yersinia*, or *Campylobacter* bacteria.

The initial symptoms of RS are inflammation either of the urethra or the intestines, followed by acute arthritis four to 28 days later. The arthritis usually affects the fingers, toes, and weight-bearing joints in the legs. Other symptoms include:

- inflammation of the urethra, with painful urination and a discharge from the penis
- mouth ulcers
- inflammation of the eye
- keratoderma blennorrhagica, these are patches of scaly skin on the palms, soles, trunk, or scalp of RS patients

Diagnosis

Patient history

Diagnosis of Reiter's syndrome can be complicated by the fact that different symptoms often occur several weeks apart. The patient does not usually draw a connection between the arthritis and previous sexual activity. The doctor is likely to consider Reiter's syndrome when the patient's arthritis occurs together with or shortly following inflammation of the eye and the genitourinary tract lasting a month or longer.

Laboratory tests

There is no specific test for diagnosing RS, but the physician may have the urethral discharge cultured to rule out **gonorrhea**. Blood tests of RS patients are typically positive for the HLA-B27 genetic marker, with an elevated white blood cell (WBC) count and an increased sedimentation rate of red blood cells. The patient may also be mildly anemic.

Diagnostic imaging

X rays do not usually reveal any abnormalities unless the patient has had recurrent episodes of the disease. Joints that have been repeatedly inflamed may show eroded areas, signs of **osteoporosis**, or bony spurs when x rayed.

Treatment

There is no specific treatment for RS. Joint inflammation is usually treated with **nonsteroidal anti-inflammatory drugs** (NSAIDs.) Skin eruptions and eye inflammation can be treated with **corticosteroids**. Gold treatments may be given for eroded bone.

Patients with chronic arthritis are also given **physical therapy** and advised to **exercise** regularly.

KEY TERMS

Acute—Having a sudden onset and lasting a short time.

Chronic—Of long duration.

Keratoderma blennorrhagica—The medical name for the patches of scaly skin that occur on the arms, legs, and trunk of RS patients.

Reactive arthritis—Another name for Reiter's syndrome.

Prognosis

The prognosis varies. Most patients recover in three to four months, but about 50% have recurrences for several years. Some patients develop complications that include inflammation of the heart muscle, stiffening inflammation of the vertebrae, glaucoma, eventual blindness, deformities of the feet, or accumulation of fluid in the lungs.

Prevention

In males, Reiter's syndrome can be prevented by sexual abstinence or the use of **condoms**.

Resources

BOOKS

- Beers, Mark H., Robert S. Porter, and Thomas V. Jones, eds. *The Merck Manual of Diagnosis and Therapy*. 18th ed. Whitehouse Station, NJ: Merck Research Laboratories, 2006.
- McPhee, Stephen, and Maxine Papadakis. *Current Medical Diagnosis and Treatment*, 2010, 49th ed. New York: McGraw-Hill Medical, 2009.

Rebecca J. Frey, PhD

Relapsing fever

Definition

Relapsing **fever** refers to two similar illnesses, both of which cause high fevers. The fevers resolve, only to recur again within about a week.

Description

Relapsing fever is caused by spiral-shaped bacteria of the genus *Borrelia*. This bacterium lives in

rodents and in insects, specifically ticks and body lice. The form of relapsing fever acquired from ticks is slightly different from that acquired from body lice.

In tick-borne relapsing fever (TBRF), rodents (rats, mice, chipmunks, and squirrels) which carry *Borrelia* are fed upon by ticks. The ticks then acquire the bacteria, and are able to pass it on to humans. TBRF is most common in sub-Saharan Africa, parts of the Mediterranean, areas in the Middle East, India, China, and the south of Russia. Also, *Borrelia* causing TBRF exist in the western regions of the United States, particularly in mountainous areas. The disease is said to be endemic to these areas, meaning that the causative agents occur naturally and consistently within these locations.

In louse-borne relapsing fever (LBRF), lice acquire *Borrelia* from humans who are already infected. These lice can then go on to infect other humans. LBRF is said to be epidemic, as opposed to endemic, meaning that it can occur suddenly in large numbers in specific communities of people. LBRF occurs in places where poverty and overcrowding predispose to human infestation with lice. LBRF has flared during wars, when conditions are crowded and good hygiene is next to impossible. At this time, LBRF is found in areas of east and central Africa, China, and in the Andes Mountains of Peru.

Causes and symptoms

In TBRF, humans contract *Borrelia* when they are fed upon by ticks. Ticks often feed on humans at night, so many people who have been bitten are unaware that they have been. The bacteria is passed on to humans through the infected body fluids of the tick.

In LBRF, a louse must be crushed or smashed in order for *Borrelia* to be released. The bacteria then enter the human body through areas where the person may have scratched him or herself.

Both types of relapsing fever occur some days after having acquired the bacteria. About a week after becoming infected, symptoms begin. The patient spikes a very high fever, with chills, sweating, terrible **headache**, **nausea**, **vomiting**, severe **pain** in the muscles and joints, and extreme weakness. The patient may become dizzy and confused. The eyes may be bloodshot and very sensitive to light. A **cough** may develop. The heart rate is greatly increased, and the liver and spleen may be swollen. Because the substances responsible for blood clotting may be disturbed during the illness, tiny purple marks may appear on the skin, which are evidence of minor bleeding occurring under the skin. The patient may suffer from a **nosebleed**, or may cough up bloody

KEY TERMS

Endemic—Refers to a particular organism which consistently exists in a particular location under normal conditions.

Epidemic—Refers to a condition suddenly acquired by a large number of people within a specific community, and which spreads rapidly throughout that community.

Shock—A state in which the blood pressure is so low that organs and tissues are not receiving an appropriate flow of blood.

sputum. All of these symptoms last for about three days in TBRF, and about five days in LBRF.

With or without treatment, a crisis may occur as the bacteria are cleared from the blood. This crisis, called a Jarisch-Herxheimer reaction, results in a new spike in fever, chills, and an initial rise in blood pressure. The blood pressure then falls drastically, which may deprive tissues and organs of appropriate blood flow (shock). This reaction usually lasts for about a day.

Recurrent episodes of fever with less severe symptoms occur after about a week. In untreated infections, fevers recur about three times in TBRF, and only once or twice in LBRF.

Diagnosis

Diagnosis of relapsing fever is relatively easy, because the causative bacteria can be found by examining a sample of blood under the microscope. The characteristically spiral-shaped bacteria are easily identifiable. The blood is best drawn during the period of high fever, because the bacteria are present in the blood in great numbers at that time.

Treatment

Either tetracycline or erythromycin is effective against both forms of relapsing fever. The medications are given for about a week for cases of TBRF; LBRF requires only a single dose. Children and pregnant women should receive either erythromycin or penicillin. Because of the risk of the Jarish-Herxheimer reaction, patients must be very carefully monitored during the initial administration of antibiotic medications. Solutions containing salts must be given through a needle in the vein (intravenously) to keep the blood pressure from dropping too drastically. Patients with

extreme reactions may need medications to improve blood circulation until the reaction resolves.

Prognosis

In epidemics of LBRF, **death** rates among untreated victims have run as high as 30%. With treatment, and careful monitoring for the development of the Jarish-Herxheimer reaction, prognosis is good for both LBRF and TBRF.

Prevention

Prevention of TBRF requires rodent control, especially in and near homes. Careful use of insecticides on skin and clothing is important for people who may be enjoying outdoor recreation in areas known to harbor the disease-carrying ticks.

Prevention of LBRF is possible, but probably more difficult. Good hygiene and decent living conditions would prevent the spread of LBRF, but these may be difficult for those people most at risk for the disease.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Rosalyn Carson-DeWitt, MD

Relapsing polychondritis

Definition

Relapsing polychondritis is a disease characterized by autoimmune-like episodic or progressive inflammation of cartilage and other connective tissue, such as the nose, ears, throat, joints, kidneys, and heart.

Description

Cartilage is a tough, flexible tissue that turns into bone in many places in the body. Bones all start out as cartilage in the fetus. Consequently, children have more cartilage than adults. Cartilage persists in adults in the linings of joints, the ears, the nose, the airway and the ribs near the breast bone. All these sites are attacked by relapsing polychondritis, which usually occurs equally in middle-aged males and females. It is frequently diagnosed along with **rheumatoid arthritis**, **systemic lupus erythematosus**, and other connective tissue diseases.

KEY TERMS

Aorta—The biggest artery in the body, receiving blood directly from the heart.

Connective tissue—Several types of tissue that hold the body's parts together—tendons, ligaments, fascia, and cartilage.

Inflammation—The body's immune reaction to presumed foreign substances like germs. Inflammation is characterized by increased blood supply and activation of defense mechanisms. It produces redness, swelling, heat, and pain.

Causes and symptoms

The most common first symptom of relapsing polychondritis is **pain** and swelling of the external ear. Usually, both ears turn red or purple and are tender to the touch. The swelling can extend into the ear canal and beyond, causing ear infections, **hearing loss**, balance disturbances with vertigo and **vomiting**, and eventually a droopy ear. The nose is often afflicted as well and can deteriorate into a flattened nose bridge called saddle nose. Inflammation of the eye occurs less frequently, but can lead to blindness.

As relapsing polychondritis advances, it causes more dangerous symptoms such as deterioration of the cartilage that holds the windpipe open. Progressive disease can destroy the integrity of the airway and compromise breathing. Destruction of the rib cartilage can collapse the chest, again hindering breathing. Joints everywhere are involved in episodes of arthritis, with pain and swelling. Other tissues besides cartilage are also involved, leading to a variety of problems with the skin and other tissues. Occasionally, the aorta or heart valves are damaged.

The disease may occur in episodes with complete remission between, or it may smolder along for years, causing progressive destruction.

Diagnosis

A characteristic array of symptoms and physical findings will yield a diagnosis of relapsing polychondritis. Laboratory tests are sometimes helpful. Biopsies of the affected cartilage may confirm the diagnosis. Further diagnostic tests are done to confirm other associated conditions such as rheumatoid arthritis. It is important to evaluate the airway, although only 10% of patients will die from airway complications.

Treatment

Mild inflammations can be treated with **aspirin** or **nonsteroidal anti-inflammatory drugs** (NSAIDs) such as ibuprofen. **Corticosteroids** (most often prednisone) are usually prescribed for more advanced conditions and do improve the disease. They may have to be continued over long periods of time, in which case their usage must be closely watched to avoid complications. Immune suppression with cyclophosphamide, azathioprine, cyclosporine, or dapsone is reserved for more aggressive cases. A collapsed chest or airway may require surgical support, and a heart valve or aorta may need repair or replacing.

Prognosis

There is no known cure for relapsing polychondritis. It can only be combated with each onset of inflammation and deterioration of cartilaginous tissue. As the disease progresses over a period of years, the mortality rate increases. At five years duration, relapsing polychondritis has a 30% mortality rate.

Resources

BOOKS

Fauci, Anthony S., et al., eds. *Harrison's Principles of Internal Medicine*. 17th ed. New York: McGraw-Hill Professional, 2008.

J. Ricker Polsdorfer, MD

Renal artery occlusion

Definition

Renal artery occlusion is a blockage of the major arteries that supply blood to the kidneys caused by thrombosis or **embolism**.

Description

Renal artery occlusion occurs when the flow of blood from the arteries leading to the kidneys becomes blocked by a blood clot or cholesterol emboli. The lack of oxygenation can lead to necrosis (tissue **death**) and ultimately, **chronic kidney failure**.

Causes and symptoms

Renal arterial occlusion occurs when a thrombus or embolism (blood clot or cholesterol plaque) breaks

free and blocks the arteries leading to one or both kidneys.

Symptoms of an acute renal arterial occlusion may include:

- hypertension
- fever
- sudden pain in the lower back or flank
- nausea and vomiting
- protein and/or blood in the urine

An individual with renal arterial occlusion may have no overt symptoms, particularly if only one kidney is affected or if the blockage is only partial. Health problems from secondary complications such as chronic kidney failure may be the first indication that something is wrong.

Diagnosis

The high blood pressure that is sometimes associated with a renal artery blockage may be the first sign that it is present, particularly if the **hypertension** is not responding to standard treatment. Urine and blood tests may or may not be useful in diagnosing this condition. Blood tests may show an elevated plasma creatinine level. If kidney tissue infarction (cell death caused by a lack of blood supply) has occurred, lactic dehydrogenase (LDH) may also be present in the urine and blood.

An arteriogram, an x-ray study of the arteries that uses a radiopaque substance, or dye, to make the arteries visible under x ray, may also be performed. This test is used with caution in patients with impaired kidney function, as the contrast medium can cause further kidney damage. In patients with whom this is not an issue, a spiral computed tomography (CT) scan with contrast medium may also be used.

Treatment

Occlusions may be treated with anticoagulant (blood thinning) or thrombolytic (clot destroying) drugs. If the blockage is significant, surgical intervention or **angioplasty** may be required. Between 1996 and 2000, the number of these procedures performed on Medicare patients more than doubled, according to a 2004 report.

Alternative treatment

Renal arterial occlusion is a serious and potentially life-threatening condition, and should always be treated by a healthcare professional familiar with the disorder.

KEY TERMS

Angioplasty—A non-surgical procedure that uses a balloon-tipped catheter to open a blocked artery.

Artherosclerotic plaque—A deposit of fatty and calcium substances that accumulate in the lining of the artery wall, restricting blood flow.

Atrophy—Cell or tissue wasting or death.

Chronic kidney failure—End-stage renal disease (ESRD); chronic kidney failure is diagnosed as ESRD when kidney function falls to 5–10% of capacity.

Embolism—Blood vessel obstruction by a blood clot or other substance (i.e., air).

Thrombus—Formation of a blood clot within the vascular system. A thrombus becomes an embolism if it breaks away and blocks a blood vessel.

Prognosis

The outcome of renal arterial occlusion depends on the speed with which it is treated. Once the blood supply is minimized or cut off to the kidney, tissue death soon results, ultimately leading to chronic kidney failure (end-stage renal disease).

Prevention

Atherosclerosis may encourage the formation of cholesterol emboli, a potential cause of renal artery occlusion. Strategies for avoiding **vascular disease** include eating right, maintaining a desirable weight, quitting **smoking**, managing **stress**, and exercising regularly. People prone to emboli from **blood clots** can take blood thinning drugs to prevent potential emboli from lodging in the renal artery.

Resources

PERIODICALS

“Explosive Growth Seen in Renal Artery Interventional Procedures.” *Heart Disease Weekly* September 26, 2004: 20.

Truelove, Christiane. “First for Pulmonary Embolism.” *Med Ad News* August 2004: 82.

ORGANIZATIONS

American Kidney Fund (AKF), 6110 Executive Boulevard, Suite 1010, Rockville, MD, 20852, (800) 638-8299, <http://www.kidneyfund.org>.

National Institute of Diabetes and Digestive and Kidney Diseases, NIDDK, NIH Bldg 31, Rm 9A06 31 Center Drive, MSC 2560, Bethesda, MD, 20892-2560, (301) 496.3583, <http://www2.niddk.nih.gov/Footer>.

National Kidney Foundation, Inc., 30 East 33rd Street, New York, NY, 10016, (212) 889-2210, (212) 689-9261, (800) 622-9010, <http://www.kidney.org/>.

Paula Anne Ford-Martin
Teresa G. Odle

Renal artery stenosis

Definition

Renal artery stenosis is a blockage or narrowing of the major arteries that supply blood to the kidneys.

Description

Renal artery stenosis occurs when the flow of blood from the arteries leading to the kidneys is constricted by tissue or artherosclerotic plaque. This narrowing of the arteries diminishes the blood supply to the kidneys, which can cause them to atrophy and may ultimately lead to kidney failure. It may also cause **renovascular hypertension**, or high blood pressure related to renal artery blockage.

Causes and symptoms

The two main causes of renal artery stenosis are **atherosclerosis** and fibromuscular disease. Fibromuscular diseases such as fibromuscular dysplasia cause growth of fibrous tissues on the arterial wall. Stenosis may also occur when scar tissue forms in the renal artery after trauma to the kidney.

Renal arterial stenosis has no overt symptoms. Eventually, untreated renal arterial stenosis causes secondary complications such as **chronic kidney failure**, which may be characterized by frequent urination, anemia, **edema**, headaches, **hypertension**, lower back **pain**, and other signs and symptoms.

Diagnosis

The high blood pressure that is sometimes associated with renal artery stenosis may be the first sign that it is present, particularly if the hypertension is not responding to standard treatment. Presence of a *bruit*, a swooshing sound from the artery that indicates an obstruction, may be heard through a stethoscope.

An arteriogram, an x-ray study of the arteries that uses a radiopaque substance, or dye, to make the arteries visible under x ray, may also be performed. This test is used with caution in patients with impaired kidney function, as the contrast medium may cause further kidney damage.

KEY TERMS

Artherosclerotic plaque—A deposit of fatty and calcium substances that accumulate in the lining of the artery wall, restricting blood flow.

Atrophy—Cell or tissue wasting or death.

Chronic kidney failure—End-stage renal disease (ESRD); chronic kidney failure is diagnosed as ESRD when kidney function falls to 5–10% of capacity.

Edema—Swelling which occurs when body tissues retain fluid.

Stent—An expandable “scaffold-like” device, usually constructed of a stainless steel material, that is inserted into an artery to expand the inside passage and improve blood flow.

Treatment

Treatment for renal artery stenosis is either surgical, pharmaceutical, or with **angioplasty** or stenting. Angioplasty involves guiding a balloon catheter down into the renal artery and inflating the balloon to clear the blockage. A stent may be inserted into the artery to widen the opening. Some patients may be candidates for surgical revascularization, which involves restoring blood flow with an arterial bypass. Drugs known as angiotension-converting enzyme (ACE) inhibitors may be prescribed for some patients. The chosen treatment approach depends on the cause of the stenosis and factors such as the patient’s kidney function and blood pressure control.

Alternative treatment

Renal artery stenosis is a serious and potentially life-threatening condition, and should always be treated by a healthcare professional familiar with the disorder.

Prognosis

Untreated renal artery stenosis can cause hypertension (high blood pressure) and may ultimately lead to chronic kidney failure (end-stage renal disease).

Prevention

Maintaining a heart healthy lifestyle can help to prevent cases of renal arterial stenosis attributable to atherosclerosis. Strategies for avoiding **vascular disease** include eating right, maintaining a desirable

weight, quitting **smoking**, managing **stress**, and exercising regularly.

ORGANIZATIONS

American Kidney Fund (AKF), 6110 Executive Boulevard, Suite 1010, Rockville, MD, 20852, (800) 638-8299, <http://www.kidneyfund.org>.

National Institute of Diabetes and Digestive and Kidney Diseases, NIDDK, NIH Bldg 31, Rm 9A06 31 Center Drive, MSC 2560, Bethesda, MD, 20892-2560, (301) 496-3583, <http://www2.niddk.nih.gov/Footer>.

National Kidney Foundation, Inc., 30 East 33rd Street, New York, NY, 10016, 212 889-2210, 212 689-9261, (800) 622-9010, <http://www.kidney.org/>.

Paula Anne Ford-Martin

Renal calculi see **Kidney stones**

Renal failure see **Acute kidney failure; Chronic kidney failure**

Renal nuclear medicine scan see **Kidney nuclear medicine scan**

Renal tubular acidosis

Definition

Renal tubular acidosis (RTA) is a group of metabolic disorders in which acid accumulates in the body because the kidneys fail to keep the urine at a proper level of acidity. This condition is a form of metabolic acidosis—a condition in which there is too much acid in the body fluids. Although kidney failure may also be characterized by acidosis, the term RTA is applied only to patients with poor acidification of the urine whose kidneys are otherwise functioning normally. RTA was identified as a distinctive disorder only in the twentieth century, being first described in children in 1936 and in adults in 1945.

Demographics

Renal tubular acidosis is a relatively uncommon condition, although some of its types are rarer than others:

- Type I: Type I may be inherited as a result of genetic mutations or may be sporadic, associated with such autoimmune disorders as Sjögren syndrome or lupus. It appears to be more common in women than in men.

- Type II: This type of RTA is very rare, most often occurs in infancy, and is usually found in association with Fanconi syndrome or as a reaction to certain drugs.
- Type III: This type is extremely rare and is thought to occur primarily in the Maghreb region of West Africa as the result of a genetic mutation.
- Type IV: Type IV is thought to be the most common form of RTA but is still rare in the general population.

Description

Renal tubular acidosis is a disorder in which acid produced by the breakdown of food in the body accumulates in the blood instead of being removed from the blood during filtration by the kidneys. Under normal circumstances, as blood passes through the tubules of the kidneys, this acid is removed and excreted into the urine, and an alkaline substance called bicarbonate is returned to the blood. In RTA, the ability of the tubules to either remove the acid from the blood or to return the bicarbonate is partially impaired.

There are four basic types of RTA:

- Type I: Type I RTA, also called classical distal RTA or dRTA, is caused by a failure of the cells in the lower (distal) section of the kidney tubule to secrete enough hydrogen ions into the filtrate. The result is increased acidity of the urine, low potassium levels in the blood (hypokalemia), and leakage of calcium into the urine.
- Type II: Type II RTA is called proximal RTA or pRTA because the part of the kidney tubule that is defective lies closest to the point where fluid and wastes from the blood enter the tubule. In pRTA, this portion of the tubule fails to return bicarbonate to the blood.
- Type III: Type III RTA, a combination of dRTA and pRTA, is extremely rare and is not often used as a classification as of 2010 because it is thought to be a combination of Type I and Type II RTA.
- Type IV: Type IV RTA is sometimes called generalized RTA or hyperkalemic RTA. It is characterized by a general impairment of the distal portion of the renal tubule in transporting potassium, chloride, and sodium across cell membranes. This type of RTA is distinguished from dRTA by abnormally high levels of potassium in the blood (hyperkalemia) rather than low levels. It occurs when a person has low levels of a hormone called aldosterone or when the kidneys do not respond properly to the aldosterone that is produced in the body. Type IV RTA is not a genetic disorder; it may be secondary

to certain kidney disorders or to exposure to certain medications.

Risk factors

The risk factors for RTA include a wide range of inherited and acquired disorders, various prescription medications, and environmental factors.

Causes and symptoms

Causes

The causes of RTA vary according to its type. RTA may be either a primary or a secondary disorder; that is, it may arise by itself or as a complication of another disease.

- Type I: Type I RTA can be caused by certain genetic disorders or associated with such autoimmune disorders as lupus or Sjögren syndrome, or with sickle cell anemia. Other diseases and disorders associated with dRTA include cirrhosis, hyperparathyroidism, hyperthyroidism, rejection of a transplanted kidney, chronic urinary tract infections, and a hereditary form of deafness. Some of these disorders cause calcium to build up in the kidney tubule, which interferes with the functioning of the distal tubule. Drugs that may cause dRTA include lithium, amphotericin B, and ifosfamide.
- Type II: Type II, or pRTA, can be caused by such hereditary disorders as cystinosis, fructose intolerance, or Wilson's disease; by such acquired disorders as multiple myeloma; by exposure to heavy metals, particularly lead and cadmium; or by exposure to certain drugs, including ifosfamide, acetazolamide, or outdated tetracycline antibiotics.
- Type IV: Type IV RTA is not caused by genetic abnormalities but by a deficiency of the hormone aldosterone (primary aldosterone deficiency), or by certain diseases or disorders that affect kidney functioning and the body's ability to use the hormone. These include diabetes, HIV infection, and blockages of the urinary tract; lupus, amyloidosis, removal or destruction of both adrenal glands, and kidney transplant rejection. Type IV RTA can be worsened by such drugs as NSAIDs, cyclosporine (an immunosuppressant), ACE inhibitors (blood pressure drugs), potassium-sparing diuretics, trimethoprim or pentamidine (antibiotics), or heparin (a blood thinner).

Symptoms

Metabolic acidosis—of which RTA is one type—does not have any unique symptoms or signs. It does, however, affect the functioning of the body's muscles,

KEY TERMS

Acidosis—A condition in which the blood becomes increasingly acid.

Aldosterone—A hormone produced in the cortex of the adrenal gland that increases the reabsorption of sodium and water and the release of potassium in the kidneys.

Cystinosis—A genetic disorder characterized by a buildup of an amino acid called cystine in the body. It leads to abnormal amounts of carbohydrates and amino acids in the urine, excessive urination, and low blood levels of potassium and phosphates.

Distal tubule—The portion of the kidney tubule that lies furthest away from the point at which fluid from the blood enters the tubule.

Fanconi syndrome—A kidney disorder in which glucose, amino acids, uric acid, phosphate, and bicarbonate are passed into the urine instead of being reabsorbed into the blood.

Filtrate—The medical term for the water and small molecules filtered in the nephron of the kidney.

Hyperkalemia—An abnormally high level of potassium in the blood.

Hypokalemia—An abnormally low level of potassium in the blood.

Nephron—The basic structural unit of the kidney, responsible for regulating the concentration of water and soluble chemicals in the blood by filtering the blood, reabsorbing the compounds needed by the body, and excreting the rest in the urine. Each kidney in humans contains between 800,000 and one million nephrons.

Osteomalacia—Softening or weakening of the bones due to either a lack of vitamin D or the body's inability to make use of this vitamin.

Proximal tubule—The portion of the kidney tubule that lies closest to the point at which fluid from the blood enters the tubule.

Sjögren syndrome—An autoimmune disorder in which the body's immune system attacks the glands that produce saliva and tears.

Sporadic—Referring to a disease or disorder that occurs in isolated instances.

Tubule—A very small tube-shaped structure in the nephron of the kidney that removes certain ions and molecules from the blood and deposits them into the fluid within the tubule.

Wilson's disease—A genetic disorder in which copper accumulates in the body, leading to liver disease and a variety of neurological and psychiatric symptoms.

bones, heart, lungs, and nervous system; patients may have a variety of symptoms ranging from headaches, **fatigue**, and weakened bones or muscles to irregular heartbeat, **nausea and vomiting**, skeletal abnormalities, confusion, and rapid breathing.

Many patients with RTA have no symptoms for years; when symptoms do develop, the characteristic symptoms of the different types of RTA include:

- Type I: The hypokalemia characteristic of dRTA affects the muscles, including the heart, leading to weakness, irregular heartbeat, paralysis, and even death. Untreated Type I RTA causes growth retardation in children and progressive kidney and bone disease (osteomalacia) in adults because the acid levels in the blood lead to demineralization of bone. Calcium may build up in the kidneys, leading to the formation of kidney stones and eventual kidney failure.
- Type II: Symptoms of Type II RTA may include bone pain, muscle cramps, and bone demineralization due to the loss of phosphates in the urine.

Children can develop growth retardation with this type of RTA as well as with Type I.

- Type IV: This type of RTA is usually asymptomatic if the acidosis is mild; however, an irregular heartbeat or paralysis may develop if the hyperkalemia is severe.

Diagnosis

The diagnosis of RTA is based on the results of laboratory tests.

Examination

Since many patients are asymptomatic, an office **physical examination** is usually unrevealing unless the person has extreme muscle weakness and diminished reflexes.

Tests

The most common types of tests used to diagnose RTA are blood and urine tests:

- Acid-base tests of blood and urine samples. These measure the acidity or alkalinity of body fluids. If the blood is more acidic than normal or the urine is less acidic, RTA is a possible diagnosis but additional tests are needed to confirm the diagnosis and determine a patient's specific type of RTA.
- Urinalysis. This test measures the level of electrolytes in the blood and may show abnormal levels of phosphate, calcium, glucose, and amino acids.
- Acid loading test. This test is done to measure the ability of the kidney tubules to acidify the urine. The patient is given capsules of a chemical called ammonium chloride to take over a three-day period and urine and blood samples are then taken. Failure to acidify the urine indicates dRTA.
- Measure of blood potassium level. This test can help to distinguish between Type I RTA, in which blood potassium levels are low, and Type IV, in which they are higher than normal.
- Aldosterone test. This is a test done to measure the levels of aldosterone in the patient's blood. It can be used to evaluate patients suspected of having Type IV RTA.
- Bicarbonate infusion. To confirm the diagnosis of Type II RTA, the doctor may infuse a solution of sodium bicarbonate into the patient's vein and then measure the acidity of the patient's urine and the amount of bicarbonate that is excreted. If the urine turns alkaline and there is a large amount of bicarbonate excreted, the patient has Type II RTA.

Treatment

The primary goal of therapy in RTA is to neutralize the acid in the patient's blood, but the specific method of treatment depends on the underlying cause of the acidosis.

Traditional

The most common form of treatment for RTA is use of medications that can reduce the acidity of the patient's blood.

Drugs

RTA is treated primarily with medications:

- Type I: Type I RTA is treated by administering sodium bicarbonate and sodium citrate. These chemicals correct the low blood potassium levels, leakage of calcium into the urine, and salt depletion found in dRTA. They also reduce the risk of kidney stone formation and eventual kidney failure.

Infants with Type I RTA occasionally need potassium supplements.

- Type II: Children with pRTA are usually given large doses of sodium bicarbonate or potassium citrate to treat acidosis and prevent bone disorders, kidney stones, and growth failure. They may be given vitamin D supplements to reduce the risk of bone deformities.
- Type IV: Type IV RTA is treated with alkaline chemicals to lower the acidity of the blood and a diuretic like furosemide to lower the levels of potassium in the blood. The patient may also be asked to minimize their intake of foods that are high in potassium, such as meats and fish, apricots, bananas, cantaloupe, lima beans, citrus fruits, and tomatoes.

Prognosis

Most patients with RTA get better with treatment. Early recognition and prompt treatment is essential to prevent kidney failure. Patients require maintenance therapy and periodic monitoring of blood acidity throughout their lives.

Prevention

Most of the disorders that cause RTA are not preventable. Risk of Type II can be reduced by limiting exposure to heavy metals.

Resources

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ORGANIZATIONS

- American Kidney Fund (AKF), 6110 Executive Blvd., Suite 1010, Rockville, MD, 20852, (800) 638-8299, <http://www.kidneyfund.org>.
- American Society of Nephrology (ASN), 1725 I Street, NW, Suite 510, Washington, DC, 20006, (202) 659-0599, (202) 659-0709, email@asn-online.org, <http://www ASN-online.org>.
- American Urological Association (AUA), 1000 Corporate Boulevard, Linthicum, MD, 21090, (410) 689-3700, (866) RING AUA, (410) 689-3800, <http://www.auanet.org>.
- National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), Building 31, Rm 9A06, 31 Center Drive, MSC 2560, Bethesda, MD, 20892-2560, (301) 496-3583, <http://www2.niddk.nih.gov/Footer/ContactNIDDK.htm>, <http://www2.niddk.nih.gov>.
- National Kidney Foundation (NKF), 30 East 33rd Street, New York, NY, 10016, (212) 889-2210, (800) 622-9010, (212) 689-9261, <http://www.kidney.org>.

Rebecca J. Frey, PhD

Renal ultrasound see **Abdominal ultrasound**

Renal vein thrombosis

Definition

Renal vein thrombosis develops when a blood clot forms in the renal vein, which carries blood from the kidneys back to the heart. The disorder is not common.

Description

Renal vein thrombosis occurs in both infants and adults. Onset of the disorder can be rapid (acute) or gradual. The number of people who suffer from renal vein thrombosis is difficult to determine, as many people do not show symptoms, and the disorder is diagnosed only by specific tests. Ninety percent of childhood cases occur in children under one year old, and 75% occur in infants under one month of age. In adult women, oral contraceptive use increases the risk of renal vein thrombosis.

Causes and symptoms

In children, renal vein thrombosis almost always occurs rapidly after an episode of severe **dehydration**. Severe dehydration decreases blood volume and causes the blood to clot more readily.

In adults, renal vein thrombosis can be caused by injury to the abdomen or back, as a result of malignant kidney tumors growing into the renal vein, or as a result of kidney diseases that cause degenerative changes in the cells of the renal tubules (**nephrotic syndrome**).

Acute onset of renal vein thrombosis at any age causes **pain** in the lower back and side, **fever**, bloody urine, decreased urine output, and sometimes kidney failure. In adults, when the onset of the disorder is gradual, there is a slow decrease in kidney function, and protein appears in the urine. Many adults with renal vein thrombosis show few symptoms.

Diagnosis

Renal **venography**, where a contrast material (dye) is injected into the renal vein before x rays are taken, is one of the best ways to detect renal vein thrombosis. Other useful tests to detect a clot include **computed tomography scans** (CT scans), **magnetic resonance imaging** (MRI), and ultrasound.

Treatment

One of the major goals of treatment is to prevent the blood clot in the renal vein from detaching and moving into the lungs, where it can cause serious complications as a **pulmonary embolism**. The enzyme streptokinase may be given to help dissolve the renal clot. Anticoagulant medications are usually prescribed to prevent clots from recurring. Rarely, when there is a complete blockage of the renal vein in infants, the kidney must be surgically removed.

Prognosis

Most cases of renal vein thrombosis resolve without any permanent damage. **Death** from renal vein thrombosis is rare, and is often caused by the blood clot detaching and lodging in the heart or lungs.

Prevention

There is no specific prevention for renal vein thrombosis. Preventing dehydration reduces the risk that it will occur.

ORGANIZATIONS

National Kidney Foundation, Inc., 30 East 33rd Street, New York, NY, 10016, 212 889-2210, 212 689-9261, (800) 622-9010, <http://www.kidney.org/>.

Tish Davidson, A.M.

Rendu-Osler-Weber disease see **Hereditary hemorrhagic telangiectasia**

Renin assay see **Plasma renin activity**

Renovascular hypertension

Definition

Renovascular **hypertension** is a secondary form of high blood pressure caused by a narrowing of the renal artery.

Description

Primary hypertension, or high blood pressure, affects millions of Americans. It accounts for over 90% of all cases of hypertension and develops without apparent causes. It is helpful for the clinician to know if a secondary disease is present and may be contributing to the high pressure. If clinical tests indicate this is so, the term used for the rise in blood pressure is secondary hypertension.

Renal hypertension is the most common form of secondary hypertension and affects no more than one percent of all adults with primary hypertension. There are two forms of renovascular hypertension.

In atherosclerotic renovascular hypertension disease, plaque is deposited in the renal artery. The deposits narrow the artery, disrupting blood flow. Atherosclerotic renovascular hypertension is most often seen in men over age 45 and accounts for two-thirds of the cases of renovascular hypertension. In

most patients, it affects the renal arteries to both kidneys.

Renovascular hypertension caused by fibromuscular dysplasia occurs mainly in women under age 45. It is also the cause of hypertension in 10% of children with the disorder. In fibromuscular dysplasia, cells from the artery wall overgrow and cause a narrowing of the artery channel.

The risk of having hypertension is related to age, lifestyle, environment, and genetics. **Smoking, stress, obesity**, a diet high in salt, exposure to heavy metals, and an inherited predisposition toward hypertension all increase the chances that a person will develop both primary and renovascular hypertension.

Causes and symptoms

Narrowing of the renal artery reduces the flow of blood to the kidney. In response, the kidney produces the protein renin. Renin is released into the blood stream. Through a series of steps, renin is converted into an enzyme that causes **sodium** (salt) retention and constriction of the arterioles. In addition to atherosclerotic and fibromuscular dysplasia, narrowing of the renal artery can be caused by compression from an injury or tumor, or by **blood clots**.

Renovascular hypertension is suspected when hypertension develops suddenly in patients under 30 or over 55 years of age or abruptly worsens in any patient. Symptoms are often absent or subtle.

Diagnosis

No single test for renovascular hypertension is definitive. About half of patients with renovascular hypertension have a specific cardiovascular sound that is heard when a doctor listens to the upper abdomen with a stethoscope. Other diagnostic tests give occasional false positive and false negative results. Most tests are expensive, and some involve serious risks.

Imaging studies are used to diagnose renovascular hypertension. In **intravenous urography**, a dye is injected into the kidney, pictures are made, and the kidneys compared. In renal arteriography, contrast material is inserted into the renal artery and cinematic x rays (showing motion within the kidney) are taken. Studies of kidney function are performed. Tests are done to measure renin production. The results of these tests taken together are used to diagnose renovascular hypertension.

Treatment

Renovascular hypertension may not respond well to anti-hypertensive drugs. Percutaneous transluminal **angioplasty** (PTA), where a balloon catheter is used to dilate the renal artery and remove the blockage, is effective in improving the condition of about 90% of patients with fibromuscular dysplasia. One year later, 60% remain cured. It is less successful in patients with **atherosclerosis**, where renovascular hypertension recurs in half the patients. Where kidney damage occurs, surgery to repair or bypass the renal artery blockage is often effective. In some cases, the damaged kidney must be removed.

Alternative treatment

Alternative treatment stresses eliminating the root causes of hypertension. With renovascular hypertension, as with primary hypertension, the root causes generally cannot be totally reversed by any method. Lifestyle changes are recommended. These include stopping smoking, eating a diet low in animal fats and salt, avoiding exposure to heavy metals, stress control through **meditation**, and anger management. Herbal medicine practitioners recommend garlic (*Allium sativum*) to help lower blood pressure. Constitutional homeopathy and **acupuncture** also can be helpful in lowering blood pressure.

Prognosis

PTA is effective in many younger patients with fibromuscular dysplasia. Older patients are less responsive to this treatment. Surgery is also more risky and less successful in older patients.

Prevention

Renovascular hypertension is possibly preventable through lifestyles that prevent atherosclerosis and primary hypertension. It is unknown how to prevent fibromuscular hyperplasia.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review, personal.info@heart.org.

Tish Davidson, A.M.

Respiratory acidosis

Definition

Respiratory acidosis is a condition in which a build-up of carbon dioxide in the blood produces a shift in the body's pH balance and causes the body's system to become more acidic. This condition is brought about by a problem either involving the lungs and respiratory system or signals from the brain that control breathing.

Description

Respiratory acidosis is an acid imbalance in the body caused by a problem related to breathing. In the lungs, oxygen from inhaled air is exchanged for carbon dioxide from the blood. This process takes place between the alveoli (tiny air pockets in the lungs) and the blood vessels that connect to them. When this exchange of oxygen for carbon dioxide is impaired, the excess carbon dioxide forms an acid in the blood. The condition can be acute with a sudden onset, or it can develop gradually as lung function deteriorates.

Causes and symptoms

Respiratory acidosis can be caused by diseases or conditions that affect the lungs themselves, such as **emphysema**, chronic **bronchitis**, **asthma**, or severe **pneumonia**. Blockage of the airway due to swelling, a foreign object, or vomit can induce respiratory acidosis. Drugs like anesthetics, sedatives, and **narcotics** can interfere with breathing by depressing the respiratory center in the brain. Head injuries or brain tumors can also interfere with signals sent by the brain to the lungs. Such neuromuscular diseases as **Guillain-Barré syndrome** or **myasthenia gravis** can impair the muscles around the lungs making it more difficult to breathe. Conditions that cause chronic **metabolic alkalosis** can also trigger respiratory acidosis.

The most notable symptom will be slowed or difficult breathing. **Headache**, drowsiness, restlessness, tremor, and confusion may also occur. A rapid heart rate, changes in blood pressure, and swelling of blood vessels in the eyes may be noted upon examination. This condition can trigger the body to respond with symptoms of metabolic alkalosis, which may include **cyanosis**, a bluish or purplish discoloration of the skin due to inadequate oxygen intake. Severe cases of respiratory acidosis can lead to **coma** and **death**.

KEY TERMS

pH—A measurement of acid or alkali (base) of a solution based on the amount of hydrogen ions available. Based on a scale of 14, a pH of 7.0 is neutral. A pH below 7.0 is an acid; the lower the number, the stronger the acid. A pH above 7.0 is a base; the higher the number, the stronger the base. Blood pH is slightly alkali with a normal range of 7.36–7.44.

Diagnosis

Respiratory acidosis may be suspected based on symptoms. A blood sample to test for pH and arterial blood gases can be used to confirm the diagnosis. In this type of acidosis, the pH will be below 7.35. The pressure of carbon dioxide in the blood will be high, usually over 45 mmHg.

Treatment

Treatment focuses on correcting the underlying condition that caused the acidosis. In patients with chronic lung diseases, this may include use of a bronchodilator or steroid drugs. Supplemental oxygen supplied through a mask or small tubes inserted into the nostrils may be used in some conditions, however, an oversupply of oxygen in patients with lung disease can make the acidosis worse. **Antibiotics** may be used to treat infections. If the acidosis is related to an overdose of narcotics, or a **drug overdose** is suspected, the patient may be given a dose of naloxone, a drug that will block the respiratory-depressing effects of narcotics. Use of mechanical ventilation like a respirator may be necessary. If the respiratory acidosis has triggered the body to compensate by developing metabolic alkalosis, symptoms of that condition may need to be treated as well.

Prognosis

If the underlying condition that caused the respiratory acidosis is treated and corrected, there may be no long term effects. Respiratory acidosis may occur chronically along with the development of lung disease or **respiratory failure**. In these severe conditions, the patient may require the assistance of a respirator or ventilator. In extreme cases, the patient may experience coma and death.

Prevention

Patients with chronic lung diseases and those who receive sedatives and narcotics need to be monitored closely for development of respiratory acidosis.

Resources

BOOKS

West, John B. *Respiratory Physiology: The Essentials*.

7th ed. Philadelphia: Lippincott Williams & Wilkins, 2005.

Altha Roberts Edgren

Respiratory alkalosis

Definition

Respiratory alkalosis is a condition where the amount of carbon dioxide found in the blood drops to a level below normal range. This condition produces a shift in the body's pH balance and causes the body's system to become more alkaline (basic). This condition is brought on by rapid, deep breathing called hyperventilation.

Description

Respiratory alkalosis is an alkali imbalance in the body caused by a lower-than-normal level of carbon dioxide in the blood. In the lungs, oxygen from inhaled air is exchanged for carbon dioxide from the blood. This process takes place between the alveoli (tiny air pockets in the lungs) and the blood vessels that connect to them. When a person hyperventilates, this exchange of oxygen for carbon dioxide is speeded up, and the person exhales too much carbon dioxide. This lowered level of carbon dioxide causes the pH of the blood to increase, leading to alkalosis.

Causes and symptoms

The primary cause of respiratory alkalosis is hyperventilation. This rapid, deep breathing can be caused by conditions related to the lungs like **pneumonia**, lung disease, or **asthma**. More commonly, hyperventilation is associated with **anxiety**, **fever**, **drug overdose**, **carbon monoxide poisoning**, or serious infections. Tumors or swelling in the brain or nervous system can also cause this type of respiration. Other stresses to the body, including **pregnancy**, liver failure, high elevations, or

KEY TERMS

Hyperventilation—Rapid, deep breathing, possibly exceeding 40 breaths/minute. The most common cause is anxiety, although fever, aspirin overdose, serious infections, stroke, or other diseases of the brain or nervous system.

pH—A measurement of acid or alkali (base) of a solution based on the amount of hydrogen ions available. Based on a scale of 14, a pH of 7.0 is neutral. A pH below 7.0 is an acid; the lower the number, the stronger the acid. A pH above 7.0 is a base; the higher the number, the stronger the base. Blood pH is slightly alkali with a normal range of 7.36-7.44.

metabolic acidosis can also trigger hyperventilation leading to respiratory alkalosis.

Hyperventilation, the primary cause of respiratory alkalosis, is also the primary symptom. This symptom is accompanied by **dizziness**, light headedness, agitation, and **tingling** or numbing around the mouth and in the fingers and hands. Muscle twitching, spasms, and weakness may be noted. Seizures, irregular heart beats, and tetany (**muscle spasms** so severe that the muscle locks in a rigid position) can result from severe respiratory alkalosis.

Diagnosis

Respiratory alkalosis may be suspected based on symptoms. A blood sample to test for pH and arterial blood gases can be used to confirm the diagnosis. In this type of alkalosis, the pH will be elevated above 7.44. The pressure of carbon dioxide in the blood will be low, usually under 35 mmHg.

Treatment

Treatment focuses on correcting the underlying condition that caused the alkalosis. Hyperventilation due to anxiety may be relieved by having the patient breath into a paper bag. By rebreathing the air that was exhaled, the patient will inhale a higher amount of carbon dioxide than he or she would normally. **Antibiotics** may be used to treat pneumonia or other infections. Other medications may be required to treat fever, seizures, or irregular heart beats. If the alkalosis is related to a drug overdose, the patient may require treatment for **poisoning**. Use of mechanical ventilation like a respirator may be necessary. If the respiratory alkalosis has triggered the body to compensate by

developing metabolic acidosis, symptoms of that condition may need to be treated, as well.

Prognosis

If the underlying condition that caused the respiratory alkalosis is treated and corrected, there may be no long-term effects. In severe cases of respiratory alkalosis, the patient may experience seizures or heart beat irregularities that may be serious and life threatening.

Resources

BOOKS

West, John B. *Respiratory Physiology: The Essentials*. 7th ed. Philadelphia: Lippincott Williams & Wilkins, 2005.

Altha Roberts Edgren

Respiratory distress syndrome

Definition

Respiratory distress syndrome (RDS) of the newborn, also known as infant RDS, is an acute lung disease present at birth, which usually affects premature babies. Layers of tissue called hyaline membranes keep the oxygen that is breathed in from passing into the blood. The lungs are said to be “airless.” Without treatment, the infant will die within a few days after birth. If oxygen can be provided, and the infant receives modern treatment in a neonatal intensive care unit, complete recovery with no after-effects can be expected.

Demographics

RDS nearly always occurs in premature infants, and the more premature the birth, the greater is the chance that RDS will develop. RDS also is seen in some infants whose mothers are diabetic. Paradoxically, RDS is less likely in the presence of certain states or conditions which themselves are harmful: abnormally slow growth of the fetus; high blood pressure, a condition called **preeclampsia** in the mother; and early rupture of the birth membranes.

Description

If a newborn infant is to breathe properly, the small air sacs (alveoli) at the ends of the breathing tubes must remain open so that oxygen in the air can get into the tiny blood vessels that surround the

alveoli. Normally, in the last months of **pregnancy**, cells in the alveoli produce a substance called **surfactant**, which keep surface tension inside the alveoli low so that the sacs can expand at the moment of birth, and the infant can breathe normally. Surfactant is produced starting at about 34 weeks of pregnancy and, by the time the fetal lungs mature at 37 weeks, a normal amount is present.

If an infant is born prematurely, enough surfactant might not have formed in the alveoli, causing the lungs to collapse and making it very difficult for the baby to get enough air (and the oxygen it contains). Sometimes a layer of fibrous tissue called a hyaline membrane forms in the air sacs, making it even harder for oxygen to get through to the blood vessels. RDS in newborn infants used to be called hyaline membrane disease.

Causes and symptoms

Labored breathing (the “respiratory distress” of RDS) may begin as soon as the infant is born, or within a few hours. Breathing becomes very rapid, the nostrils flare, and the infant grunts with each breath. The ribs, which are very flexible in young infants, move inwards each time a breath is taken. Before long the muscles that move the ribs and diaphragm, so that air is drawn into the lungs, become fatigued. When the oxygen level in the blood drops severely the infant’s skin turns bluish in color. Tiny, very premature infants may not even have signs of trouble breathing. Their lungs may be so stiff that they cannot even start breathing when born.

There are two major complications of RDS. One is called **pneumothorax**, which means “air in the chest.” When the infant itself or a breathing machine applies pressure on the lungs in an attempt to expand them, a lung may rupture, causing air to leak into the chest cavity. This air causes the lung to collapse further, making breathing even harder and interfering with blood flow in the lung arteries. The blood pressure can drop suddenly, cutting the blood supply to the brain. The other complication is called intraventricular hemorrhage, bleeding into the cavities (ventricles) of the brain, which may be fatal.

Diagnosis

When a premature infant has obvious trouble breathing at birth or within a few hours of birth, RDS is an obvious possibility. If premature birth is expected, or there is some condition that calls for delivery as soon as possible, the amount of surfactant in the amniotic fluid will indicate how well the lungs have matured. If

little surfactant is found in an amniotic fluid sample taken by placing a needle in the uterus (**amniocentesis**), there is a definite risk of RDS. Often this test is done at regular intervals so that the infant can be delivered as soon as the lungs are mature. If the membranes have ruptured, surfactant can easily be measured in a sample of vaginal fluid.

The other major diagnostic test is a **chest x ray**. Collapsed lung tissue has a typical appearance, and the more lung tissue is collapsed, the more severe the RDS. An x ray also can demonstrate pneumothorax (air or gas in the area around the lung), if this complication has occurred. The level of oxygen in the blood can be measured by taking a blood sample from an artery, or, more easily, using a device called an oximeter, which is clipped to an earlobe. Pneumothorax may have occurred if the infant suddenly becomes worse while on ventilation; x rays can help make the diagnosis.

Treatment

If only a mild degree of RDS is present at birth, placing the infant in an oxygen hood may be the only treatment required. It is important to guard against too much oxygen, as this may damage the retina and cause loss of vision. Using an oximeter to keep track of the blood oxygen level, repeated artery punctures or heel sticks can be avoided. In more severe cases a drug very like natural surfactant (Exosurf Neonatal or Survanta), can be dripped into the lungs through a fine tube (endotracheal tube) placed in the infant’s windpipe (trachea). Typically the infant will be able to breathe more easily within a few days at the most, and complications such as lung rupture are less likely to occur. The drug is continued until the infant starts producing its own surfactant. There is a risk of bleeding into the lungs from surfactant treatment; about 10% of the smallest infants are affected.

Infants with severe RDS may require treatment with a ventilator, a machine that takes over the work of the lungs and delivers air under pressure. In tiny infants who do not breathe when born, ventilation through a tracheal tube is an emergency procedure. Assisted ventilation must be closely supervised, as too much pressure can cause further lung damage. A gentler way of assisting breathing, continuous positive airway pressure (CPAP), delivers an oxygen mixture through nasal prongs or a tube placed through the nose rather than an endotracheal tube. CPAP may be tried before resorting to a ventilator, or after an infant placed on a ventilator begins to improve. Drugs that stimulate breathing may speed the recovery process.

KEY TERMS

Alveoli—The small air sacs located at the ends of the breathing tubes of the lung, where oxygen normally passes from inhaled air to blood vessels.

Amniotic fluid—The fluid bathing the fetus, which may be sampled using a needle to determine whether the fetus is making enough surfactant.

Endotracheal tube—A metal or plastic tube inserted in the windpipe which may be attached to a ventilator. It also may be used to deliver medications such as surfactant.

Hyaline membranes—A fibrous layer that settles in the alveoli in RDS and prevents oxygen from escaping from inhaled air to the bloodstream.

Pneumothorax—Air in the chest, often a result of the lung's rupturing when oxygen is delivered under too high a pressure.

Preeclampsia—A disease of pregnancy in which the mother's blood pressure is elevated; associated with both maternal and fetal complications, and sometimes with fetal death.

Steroid—A natural body substance that often is given to women before delivering a very premature infant to stimulate the fetal lungs to produce surfactant, hopefully preventing RDS (or making it less severe).

Surfactant—A material normally produced in the fetal lungs in the last months of pregnancy, which helps the air sacs to open up at the time of birth so that the newborn infant can breathe freely.

Ventilator—A machine that can breathe for an infant having RDS until its lungs are producing enough surfactant and are able to function normally.

Pneumothorax is an emergency that must be treated right away. Air may be removed from the chest using a needle and syringe. A tube then is inserted into the lung cavity, and suction applied.

Prognosis

If an infant born with RDS is not promptly treated, lack of an adequate oxygen supply will damage the body's organs and eventually cause them to stop functioning altogether. **Death** is the result. The central nervous system (the brain and spinal cord) in particular is very dependent on a steady oxygen supply and is one of the first organ systems to feel the effects of RDS. On the other hand, if the infant's breathing is supported until the lungs mature and make their own surfactant, complete recovery within three to five days is the rule.

If an air leak causes pneumothorax, immediate removal of air from the chest will allow the lungs to re-expand. Bleeding into the brain is a very serious condition that worsens the outlook for an infant with RDS.

Prevention

The best way of preventing RDS is to delay delivery until the fetal lungs have matured and are producing enough surfactant—generally at about 37 weeks of pregnancy. If delivery cannot be delayed, the mother may be given a steroid hormone, similar to a natural substance produced in the body, which crosses the

barrier of the placenta and helps the fetal lungs to produce surfactant. The steroid should be given at least 24 hours before the expected time of delivery. If the infant does develop RDS, the risk of bleeding into the brain will be much less if the mother has been given a dose of steroid.

If a very premature infant is born without symptoms of RDS, it may be wise to deliver surfactant to its lungs. This may prevent RDS, or make it less severe if it does develop. An alternative is to wait until the first symptoms of RDS appear and then immediately give surfactant. Pneumothorax may be prevented by frequently checking the blood oxygen content, and limiting oxygen treatment under pressure to the minimum needed.

Resources

BOOKS

Cameron, Kristy M. *Mitchell's Gift—A Parent's Perspective on Surviving Life... With a Premature Baby in the NICU*. Santa Maria, CA: LP Publishing, 2009.

Gunter, Jennifer. *The Preemie Primer: A Complete Guide for Parents of Premature Babies—from Birth through the Toddler Years and Beyond*. Cambridge, MA: Da Capo Lifelong Books, 2010.

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ORGANIZATIONS

American Lung Association, 1301 Pennsylvania Ave. NW, Washington, DC, 20004, (202) 785-3355, (202) 452-1805, <http://www.lungusa.org>.

National Heart Lung and Blood Institute (NHLBI), P.O. Box 30105, Bethesda, MD, 20824–0105, (301) 592-8573. TTY: (240) 629-3255, <http://www.nhlbi.nih.gov>.

National Institutes of Health (NIH), 9000 Rockville Pike, Bethesda, MD, 20892, (301) 496-4000, <http://www.nih.gov/index.html>.

National Library of Medicine (NLM), 8600 Rockville Pike, Bethesda, MD, 20894, <http://www.nlm.nih.gov>.

National Organization for Rare Diseases (NORD), P.O. Box 8923, Fairfield, CT, 06812, (213) 745-6518, <http://www.rarediseases.org>.

Respiratory Distress Syndrome Foundation (RDSF), P.O. Box 723, Montgomeryville, PA, 18936, <http://membrane.com/phylanet/rds/index.html>.

David A. Cramer, MD.
Laura Jean Cataldo, RN, Ed.D.

Respiratory failure

Definition

Respiratory failure is nearly any condition that affects breathing function or the lungs themselves and can result in failure of the lungs to function properly. The main tasks of the lungs and chest are to get oxygen from the air that is inhaled into the bloodstream, and, at the same time, to eliminate carbon dioxide (CO_2) from the blood through air that is breathed out. In respiratory failure, the level of oxygen in the blood becomes dangerously low, and/or the level of CO_2 becomes dangerously high. There are two ways in which this can happen. Either the process by which oxygen and CO_2 are exchanged between the blood and the air spaces of the lungs (a process called “gas exchange”) breaks down, or the movement of air in and out of the lungs (ventilation) does not take place properly.

Description

Respiratory failure often is divided into two main types. One of them, called hypoxic respiratory failure, occurs when something interferes with normal gas exchange. Too little oxygen gets into the blood (hypoxemia), and all organs and tissues in the body suffer as a result. One common type of hypoxic failure, occurring in both adults and prematurely born infants, is **respiratory distress syndrome**, a condition in which fluid or tissue changes prevent oxygen from passing

out of the air sacs of the lungs into the circulating blood. Hypoxemia also may result from spending time at high altitudes (where there is less oxygen in the air); various forms of lung disease that separate oxygen from blood in the lungs; severe anemia (“low blood”); and blood vessel disorders that shunt blood away from the lungs, thus precluding the lungs from picking up oxygen.

The other main type of respiratory failure is ventilatory failure, occurring when, for any reason, breathing is not strong enough to rid the body of CO_2 . Then CO_2 builds up in the blood (hypercapnia). Ventilatory failure can result when the respiratory center in the brainstem fails to drive breathing; when muscle disease keeps the chest wall from expanding when breathing in; or when a patient has chronic obstructive lung disease that makes it very difficult to exhale air with its CO_2 . Many of the specific diseases and conditions that cause respiratory failure cause both too little oxygen in the blood (hypoxemia) and abnormal ventilation.

Causes and symptoms

Several different abnormalities of breathing function can cause respiratory failure. The major categories, with specific examples of each, are:

- Obstruction of the airways. Examples are chronic bronchitis with heavy secretions; emphysema; cystic fibrosis; asthma (a condition in which it is very hard to get air in and out through narrowed breathing tubes).
- Weak breathing. This can be caused by drugs or alcohol, which depress the respiratory center; extreme obesity; or sleep apnea, where patients stop breathing for long periods while sleeping.
- Muscle weakness. This can be caused by a muscle disease called myasthenia; muscular dystrophy; polio; a stroke that paralyzes the respiratory muscles; injury of the spinal cord; or Lou Gehrig’s disease.
- Lung diseases, including severe pneumonia. Pulmonary edema, or fluid in the lungs, can be the source of respiratory failure. Also, it can often be a result of heart disease; respiratory distress syndrome; pulmonary fibrosis and other scarring diseases of the lung; radiation exposure; burn injury when smoke is inhaled; and widespread lung cancer.
- An abnormal chest wall (a condition that can be caused by scoliosis or severe injury of the chest wall).

A majority of patients with respiratory failure are short of breath. Both low oxygen and high carbon dioxide can impair mental functions. Patients may

become confused and disoriented and find it impossible to carry out their normal activities or do their work. Marked CO₂ excess can cause headaches and, in time, a semi-conscious state, or even **coma**. Low blood oxygen causes the skin to take on a bluish tinge. It also can cause an abnormal heart rhythm (arrhythmia). **Physical examination** may show a patient who is breathing rapidly, is restless, and has a rapid pulse. Lung disease may cause abnormal sounds heard when listening to the chest with a stethoscope: **wheezing** in **asthma**, “crackles” in obstructive lung disease. A patient with ventilatory failure is prone to gasp for breath, and may use the neck muscles to help expand the chest.

Diagnosis

The symptoms and signs of respiratory failure are not specific. Rather, they depend on what is causing the failure and on the patient’s condition before it developed. Good general health and some degree of “reserve” lung function will help see a patient through an episode of respiratory failure. The key diagnostic determination is to measure the amount of oxygen, carbon dioxide, and acid in the blood at regular intervals. A sudden low oxygen level in the lung tissue may cause the arteries of the lungs to narrow. This, in turn, causes the resistance in these vessels to increase, which can be measured using a special catheter. A high blood level of CO₂ may cause increased pressure in the fluid surrounding the brain and spinal cord; this, too, can be measured.

Treatment

Nearly all patients are given oxygen as the first treatment. Then the underlying cause of respiratory failure must be treated. For example, **antibiotics** are used to fight a lung infection, or, for an asthmatic patient, a drug to open up the airways is commonly prescribed.

A patient whose breathing remains very poor will require a ventilator to aid breathing. A plastic tube is placed through the nose or mouth into the windpipe and is attached to a machine that forces air into the lungs. This can be a lifesaving treatment and should be continued until the patient’s own lungs can take over the work of breathing. It is very important to use no more pressure than is necessary to provide sufficient oxygen; otherwise ventilation may cause further lung damage. Drugs are given to keep the patient calm, and the amount of fluid in the body is carefully adjusted so that the heart and lungs can function as normally as possible. **Steroids**, which combat

KEY TERMS

Chronic obstructive lung disease—A common form of lung disease in which breathing, and therefore gas exchange, is labored and increasingly difficult.

Gas exchange—The process by which oxygen is extracted from inhaled air into the bloodstream, and, at the same time, carbon dioxide is eliminated from the blood and exhaled.

Hypoxemia—An abnormally low amount of oxygen in the blood, the major consequence of respiratory failure, when the lungs no longer are able to perform their chief function of gas exchange.

Pulmonary fibrosis—An end result of many forms of lung disease (especially chronic inflammatory conditions). Normal lung tissue is converted to scarred, “fibrotic” tissue that cannot carry out gas exchange.

inflammation, may sometimes be helpful but they can cause complications, including weakening the breathing muscles.

The respiratory therapist has a number of methods available to help patients overcome respiratory failure. They include:

- Suctioning the lungs through a small plastic tube passed through the nose, in order to remove secretions from the airways that the patient cannot cough up.
- Postural drainage, in which the patient is propped up at an angle or tilted to help secretions drain out of the lungs. The therapist may clap the patient on the chest or back to loosen the secretions, or a vibrator may be used for the same purpose.
- Breathing exercises often are prescribed after the patient recovers. They make the patient feel better and help to strengthen the muscles that aid breathing. One useful method is for the patient to suck on a tube attached to a clear plastic hosing containing a ball so as to keep the ball lifted. Regular deep breathing exercises are simpler and often just as helpful. Another technique is to have the patient breathe out against pursed lips to increase pressure in the airways and keep them from collapsing.

Prognosis

The outlook for patients with respiratory failure depends chiefly on its cause. If the underlying

disease can be effectively treated, with the patient's breathing supported in the meantime, the outlook is usually good.

Care is needed not to expose the patient to polluting substances in the atmosphere while recovering from respiratory failure; this could tip the balance against recovery. When respiratory failure develops slowly, pressure may build up in the lung's blood vessels, a condition called **pulmonary hypertension**. This condition may damage the vessels, worsen hypoxemia, and cause the heart to fail. If it is not possible to provide enough oxygen to the body, complications involving either the brain or the heart may prove fatal.

If the kidneys fail or the diseased lungs become infected, the prognosis is worse. In some cases, the primary disease causing the lungs to fail is irreversible. The patient, family, and physician together then must decide whether to prolong life by ventilator support. Occasionally, **lung transplantation** is a possibility, but it is a highly complex procedure and is not widely available.

Prevention

Because respiratory failure is not a disease itself, but the end result of many lung disorders, the best prevention is to treat any lung disease promptly and effectively. It is also important to make sure that any patient who has had lung disease is promptly treated for any respiratory infection (even of the upper respiratory tract). Patients with lung problems should also avoid exposure to pollutants, as much as is possible. Once respiratory failure is present, it is best for a patient to receive treatment in an intensive care unit, where specialized personnel and all the needed equipment are available. Close supervision of treatment, especially mechanical ventilation, will help minimize complications that would compound the problem.

ORGANIZATIONS

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

National Respiratory Distress Syndrome Foundation, P.O. Box 723, Montgomeryville, PA, 18936, (215) 822-3585, <http://membrane.com/phylanet/rds/>.

David A. Cramer, MD

Respiratory syncytial virus infection

Definition

Respiratory syncytial virus (RSV) is a virus that can cause severe lower respiratory infections in children under the age of two, and milder upper respiratory infections in older children and adults. RSV infection is also called **bronchiolitis**, because it is marked in young children by inflammation of the bronchioles. Bronchioles are the narrow airways that lead from the bronchi to the tiny air sacs (alveoli) in the lungs. The result is **wheezing**, difficulty breathing, and sometimes fatal **respiratory failure**.

Demographics

In the United States, RSV infections generally occur during the fall, winter, and early spring. Almost all children will be infected with RSV by their second birthday. RSV causes respiratory illness in infants and young children, and is the most important cause of bronchiolitis; it is responsible for about 120,000 hospitalizations of children every year. Currently, there is no effective vaccine against RSV; however, palivizumab, a medication that contains virus-fighting antibodies to RSV, can help prevent severe RSV disease, such as **pneumonia** or bronchiolitis in high-risk infants or young children. Certain people are more likely to have problems with RSV: Babies younger than 6 months, especially preemies; people with immune system problems, people with heart or lung problems, and adults older than 65.

Description

RSV infection is caused by a group of viruses found worldwide. There are two different subtypes of the virus with numerous different strains. Taken together, these viruses account for a significant number of deaths in infants.

RSV infection is primarily a disease of winter or early spring, with waves of illness sweeping through a community. The rate of RSV infection is estimated to be 11.4 cases in every 100 children during their first year of life. In the United States, RSV infection occurs most frequently in infants between the ages of two months and six months.

RSV infection shows distinctly different symptoms, depending on the age of the infected person. In children under two, the virus causes a serious lower respiratory infection in the lungs. In older children and healthy adults, it causes a mild upper respiratory infection often mistaken for the **common cold**.

KEY TERMS

Alveoli—Small air sacs or cavities in the lung that give the tissue a honeycomb appearance and expand its surface area for the exchange of oxygen and carbon dioxide.

Antibody—A protein produced by specialized white blood cells in response to the presence of a foreign

protein such as a virus. Antibodies help the body fight infection.

Reye's syndrome—A rare disorder in children that follows a viral infection and is associated with a reaction to aspirin. Its symptoms include vomiting, damaged liver function, and swelling of the brain.

Although anyone can get this disease, infants suffer the most serious symptoms and complications. Breast feeding seems to provide partial protection from the virus. Conditions in infants that increase their risk of infection include:

- premature birth
- lower socio-economic environment
- congenital heart disease
- chronic lung diseases, such as cystic fibrosis
- immune system deficiencies, including HIV infection
- immunosuppressive therapy given to organ transplant patients

Many older children and adults get RSV infection, but the symptoms are so similar to the common cold that the true cause is undiagnosed. People of any age with weakened immune systems, either from such diseases as **AIDS** or leukemia, or as the result of **chemotherapy** or corticosteroid medications, are more at risk for serious RSV infections. So are people with chronic lung disease.

Causes and symptoms

Respiratory syncytial virus is spread through close contact with an infected person. It has been shown that if a person with RSV infection sneezes, the virus can be carried to others within a radius of 6 f (1.8 m). This group of viruses is hardy. They can live on the hands for up to half an hour and on toys or other inanimate objects for several hours.

Scientists have yet to understand why RSV viruses attack the lower respiratory system in infants and the upper respiratory system in adults. In infants, RSV begins with such cold symptoms as a low **fever**, runny nose, and **sore throat**. Soon, other symptoms appear that suggest an infection which involves the lower airways. Some of these symptoms resemble those of **asthma**. RSV infection is suggested by:

- wheezing and high-pitched, whistling breathing
- rapid breathing (more than 40 breaths per minute)
- shortness of breath

- labored breathing out (exhalations)
- bluish tinge to the skin (cyanosis)
- croupy, seal-like, barking cough
- high fever

Breathing problems occur in RSV infections because the bronchioles swell, making it difficult for air to get in and out of the lungs. If the child is having trouble breathing, immediate medical care is needed. Breathing problems are most common in infants under one year of age; they can develop rapidly.

Diagnosis

Physical examination and imaging studies

RSV infection is usually diagnosed during a **physical examination** by the pediatrician or primary care doctor. The doctor listens with a stethoscope for wheezing and other abnormal lung sounds in the patient's chest. The doctor will also take into consideration whether there is a known outbreak of RSV infection in the area. Chest x rays give some indication of whether the lungs are hyperinflated from an effort to move air in and out. X rays may also show the presence of a secondary bacterial infection, such as pneumonia.

Laboratory tests

A blood test can also detect RSV infection. This test measures the level of antibodies the body has formed against the virus. The blood test is less reliable in infants than in older children because antibodies in the infant's blood may have come from the mother during **pregnancy**. If infants are hospitalized, other tests such as an arterial **blood gas analysis** are done to determine if the child is receiving enough oxygen.

Treatment

Home care

Home treatment for RSV infection is primarily supportive. It involves taking steps to ease the child's

breathing. **Dehydration** can be a problem, so children should be encouraged to drink plenty of fluids. **Antibiotics** have no effect on viral illnesses. In time, the body will make antibodies to fight the infection and return itself to health.

Home care for keeping a child with RSV comfortable and breathing more easily includes:

- Use a cool mist room humidifier to ease congestion and sore throat.
- Raise the baby's head by putting books under the head end of the crib.
- Give acetaminophen (Tylenol, Pandol, Tempra) for fever. Aspirin should not be given to children because of its association with Reye's syndrome, a serious disease.
- For babies too young to blow their noses, suction away any mucus with an infant nasal aspirator.

Hospital treatment

In the United States, RSV infections are responsible for 90,000 hospitalizations and 4,500 deaths each year. Children who are hospitalized receive oxygen and humidity through a mist tent or vaporizer. They also are given intravenous fluids to prevent dehydration. Mechanical ventilation may be necessary. Blood gases are monitored to assure that the child is receiving enough oxygen.

Medications

Bronchodilators, such as albuterol (Proventil, Ventolin), may be used to keep the airways open. Ribavirin (Virazole) is used for desperately ill children to stop the growth of the virus. Ribavirin is both expensive and has toxic side effects, so its use is restricted to the most severe cases.

Alternative treatment

Alternative medicine has little to say specifically about bronchiolitis, especially in very young children. Practitioners emphasize that people get viral illnesses because their immune systems are weak. Prevention focuses on strengthening the immune system by eating a healthy diet low in sugars and high in fresh fruits and vegetables, reducing **stress**, and getting regular, moderate **exercise**. Like traditional practitioners, alternative practitioners recommend **breastfeeding** infants so that the child may benefit from the positive state of health of the mother. Inhaling a steaming mixture of lemon oil, thyme oil, eucalyptus, and tea tree oil (**aromatherapy**) may make breathing easier.

Prognosis

RSV infection usually runs its course in seven to 14 days. The **cough** may linger weeks longer. There are no medications that can speed the body's production of antibodies against the virus. Opportunistic bacterial infections that take advantage of a weakened respiratory system may cause ear, sinus, and throat infections or pneumonia.

Hospitalization and **death** are much more likely to occur in children whose immune systems are weakened or who have underlying diseases of the lungs and heart. People do not gain permanent immunity to respiratory syncytial virus and can be infected many times. Children who suffer repeated infections seem to be more likely to develop asthma in later life.

Prevention

There are no vaccines against RSV. Respiratory syncytial virus infection is so common that prevention is impossible. However, steps can be taken to reduce a child's contact with the disease. People with RSV symptoms should stay at least six feet away from young children. Frequent handwashing, especially after contact with respiratory secretions, and the correct disposal of used tissues help keep the disease from spreading. Parents should try to keep their children under 18 month old away from crowded environments—for example, shopping malls during holiday seasons—where they are likely to come in contact with older people who have only mild symptoms of the disease. Child care centers should regularly disinfect surfaces that children touch.

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Restless legs syndrome

Definition

Restless legs syndrome (RLS) is a neurological disorder characterized by uncomfortable sensations in the legs and, less commonly, the arms. These sensations are exacerbated (heightened) when the person with RLS is at rest. The sensations are described as crawlly, tingly, prickly and occasionally painful. They result in a nearly insuppressible urge to move around. Symptoms are often associated with sleep disturbances.

Demographics

As much as 10% of the population of the United States and Europe may suffer from some degree of restless legs syndrome. Fewer cases are indicated in India, Japan and Singapore, suggesting racial or ethnic factors play a role in the disorder. Although the demographics can vary greatly, the majority of people suffering from RLS are female. The age of onset also varies greatly, but the number of people suffering from RLS increases with age. However, many people with RLS report that they had symptoms of the disorder in their childhood. These symptoms were often disregarded as growing pains or hyperactivity.

Description

Restless legs syndrome is a sensory-motor disorder that causes uncomfortable feelings in the legs, especially during periods of inactivity. Some people also report sensations in the arms, but this occurs much more rarely. The sensations occur deep in the legs and are usually described with terms that imply movement such as prickly, creepy-crawlly, boring, **itching**, achy, pulling, tugging and painful. The symptoms result in an irrepressible urge to move the leg and are relieved when the person suffering from RLS voluntarily moves. Symptoms tend to be worse in the evening or at night.

Restless legs syndrome is associated with another disorder called periodic limb movements in sleep (PLMS). It is estimated that four out of five patients with RLS also suffer from PLMS. PLMS is characterized by jerking leg movements while sleeping that may occur as frequently as every 20 seconds. These jerks disrupt sleep by causing continual arousals throughout the night.

People with both RLS and PLMS are prone to abnormal levels of exhaustion during the day because

they are unable to sleep properly at night. They may have trouble concentrating at work, at school or during social activities. They may also have mood swings and difficulty with interpersonal relationships. Depression and **anxiety** may also result from the lack of sleep. RLS affects people who want to travel or attend events that require sitting for long periods of time.

Causes and symptoms

Restless legs syndrome is categorized in two ways. Primary RLS occurs in the absence of other medical symptoms, while secondary RLS is usually associated with some other medical disorder. Although the cause of primary RLS is currently unknown, a large amount of research into the cause of RLS is taking place. Researchers at Johns Hopkins University published a study in July 2003 suggesting that iron deficiencies may be related to the disorder. They dissected brains from cadavers of people who suffered from RLS and found that the cells in the midbrain were not receiving enough iron. Other researchers suggest that RLS may be related to a chemical imbalance of the neurotransmitter dopamine in the brain. There is also evidence that RLS has a genetic component. RLS occurs three to five times more frequently in an immediate family member of someone who has RLS than in the general population. A site on a chromosome that may contain a gene for RLS has been identified by molecular biologists.

In many people, other medical conditions play a role in RLS and the disorder is therefore termed secondary RLS. People with peripheral neuropathies (injury to nerves in the arms and legs) may experience RLS. Such neuropathies may result from diabetes or **alcoholism**. Other chronic diseases such as kidney disorders and **rheumatoid arthritis** may result in RLS. Iron deficiencies and blood **anemias** are often associated with RLS and symptoms of the disease usually decrease once blood iron levels have been corrected. Attention deficit/hyperactivity disorder has also been implicated in RLS. Pregnant women often suffer from RLS, especially in the third trimester. Some people find that high levels of **caffeine** intake may result in RLS.

The symptoms of RLS are all associated with unpleasant feelings in the limbs. The words used to describe these feelings are various, but include such adjectives as deep-seated crawling, jittery, **tingling**, burning, aching, pulling, painful, itchy or prickly. They are usually not described as a muscle cramp or **numbness**. Most often the sensations occur during

KEY TERMS

Anemia—A condition that affects the size and number of red blood cells. It often results from lack of iron or certain B vitamins and may be treated with iron or vitamin supplements.

Insomnia—Trouble sleeping. People who suffer from RLS often lose sleep either because they spend time walking to relieve discomfort or because they have PLMS, which causes them to wake often during the night.

Periodic limb movements in sleep (PLMS)—Random movements of the arms or legs that occur at regular intervals of time during sleep.

periods of inactivity. They are characterized by an urge to get up and move. Such movements include stretching, walking, jogging or simply jiggling the legs. The feelings worsen in the evening.

A variety of symptoms are associated with RLS, but may not be characteristic of every case. Some people with RLS report involuntary arm and leg movements during the night. Others have difficulty falling asleep and are sleepy or fatigued during the day. Many people with RLS have leg discomfort that is not explained by routine medical exams.

Eighty-five percent of RLS patients either have difficulty falling asleep or wake several times during the night, and almost half experience daytime **fatigue** or sleepiness. It is common for the symptoms to be intermittent. They may disappear for several months and then return for no apparent reason. Two-thirds of patients report that their symptoms become worse with time. Some older patients claim to have had symptoms since they were in their early 20s, but were not diagnosed until their 50s. Suspected under-diagnosis of RLS may be attributed to the difficulty experienced by patients in describing their symptoms.

Diagnosis

A careful history enables the physician to distinguish RLS from similar types of disorders that cause night time discomfort in the limbs, such as **muscle cramps**, burning feet syndrome, and damage to nerves that detect sensations or cause movement (polyneuropathy).

The most important tool the doctor has in diagnosis is the history obtained from the patient. There

are several common medical conditions that are known to either cause or to be closely associated with RLS. The doctor may link the patient's symptoms to one of these conditions, which include anemia, diabetes, disease of the spinal nerve roots (lumbosacral radiculopathy), Parkinson's disease, late-stage **pregnancy**, kidney failure (uremia), and complications of stomach surgery. In order to identify or eliminate such a primary cause, blood tests may be performed to determine the presence of serum iron, ferritin, folate, vitamin B₁₂, creatinine, and thyroid-stimulating hormones. The physician may also ask if symptoms are present in any close family members, since it is common for RLS to run in families and this type is sometimes more difficult to treat.

In some cases, sleep studies such as **polysomnography** are undertaken to identify the presence of PLMS that are reported to affect 70–80% of people who suffer from RLS. The patient is often unaware of these movements, since they may not cause him to wake. However, the presence of PLMS with RLS can leave the person more tired, because it interferes with deep sleep. A patient who also displays evidence of some neurologic disease may undergo **electromyography** (EMG). During EMG, a very small, thin needle is inserted into the muscle and electrical activity of the muscle is recorded. A doctor or technician usually performs this test at a hospital outpatient department.

Treatment

The first step in treatment is to treat existing conditions that are known to be associated with RLS and that will be identified by blood tests. If the patient is anemic, iron (iron sulfate) or vitamin supplements (folate or vitamin B₁₂) will be prescribed. If **kidney disease** is identified as a cause, treatment of the kidney problem will take priority.

After treating underlying disorders, treatment for restless legs syndrome is generally two-pronged, consisting of making lifestyle changes and using medications to relieve some of the symptoms. Lifestyle changes involve making changes to the diet, exercising and performing other self-directed activities, and practicing good sleep hygiene. Although the United States Food and Drug Administration has not yet approved any drugs for treating RLS, four classes of pharmaceuticals have been found effective for treating RLS: dopaminergic agents, **benzodiazepines**, opioids and anticonvulsants.

Medications

Dopaminergic agents are the first type of drug prescribed in the treatment of RLS. Most commonly doctors prescribe dopamine-receptor agonists that are used to treat Parkinson's disease such as Mirapex (pramipexole), Permax (pergolide) and Requip (ropinirole). Sinemet (carbidopa/levodopa), which is a drug that adds dopamine to the nervous system, is also commonly prescribed. Sinemet has been used more frequently than other drugs in treating RLS, but recently a problem known as augmentation has been associated with its use. When augmentation develops, symptoms of RLS will return earlier in the day and increasing the dose will not improve the symptoms.

Antiepileptic drugs are those used for people with seizures. These are also useful in the treatment of RLS, and include Neurontin (gabapentin), Carbatrol (carbamazepine), Keppra (levetiracetam), and Topamax (topiramate).

Benzodiazepines are drugs that sedate and are typically taken before bedtime so that a patient with RLS can sleep more soundly. The most commonly prescribed sedative in RLS is Klonopin (clonazepam).

Opioids are synthetic **narcotics** that relieve **pain** and cause drowsiness. They are usually taken in the evening. The most commonly used opioids prescribed for RLS include Darvon or Darvocet (propoxyphene), Dolophine (**methadone**), Percocet (oxycodone), Ultram (Tramadol) and Vicodin (hydrocodone). One danger associated with opioids is that they can be addicting.

A few drugs have been found to worsen symptoms of RLS and they should be avoided by patients exhibiting RLS symptoms. These include anti-nausea drugs such as Antivert, Atarax, Compazine and Phenergan. **Calcium channel blockers** that are often used to treat heart conditions should be avoided. In addition, most anti-depressants tend to exacerbate symptoms of RLS. Finally, **antihistamines** such as Benadryl have been found to aggravate RLS symptoms in some people.

Lifestyle changes

Simple changes to the diet have proven effective for some people suffering from RLS. Vitamin deficiencies are a common problem in RLA patients. In patients with RLS, most physicians will check the levels of blood serum ferritin, which can indicate low iron storage. If these levels are below 50 mcg/L, then supplemental iron should be added to the diet. Other physicians have found that supplements of vitamin E, **folic acid** and **B vitamins**, and magnesium provide

relief to symptoms or RLS. Reducing or eliminating caffeine and alcohol consumption has been effective in other patients.

Alternative treatment

It is likely that the best alternative therapy will combine both conventional and alternative approaches. Levodopa may be combined with a therapy that relieves pain, relaxes muscles, or focuses in general on the nervous system and the brain. Any such combined therapy that allows a reduction in dosage of levodopa is advantageous, since this will reduce the likelihood of unacceptable levels of drug side effects. Of course, the physician who prescribes the medication should monitor any combined therapy. Alternative methods may include:

- **Acupuncture.** Patients who also suffer from rheumatoid arthritis may especially benefit from acupuncture to relieve RLS symptoms. Acupuncture is believed to be effective in arthritis treatment and may also stimulate those parts of the brain that are involved in RLS.
- **Homeopathy.** Homeopaths believe that disorders of the nervous system are especially important because the brain controls so many other bodily functions. The remedy is tailored to the individual patient and is based on individual symptoms as well as the general symptoms of RLS.
- **Reflexology.** Reflexologists claim that the brain, head, and spine all respond to indirect massage of specific parts of the feet.
- **Nutritional supplements.** Supplementation of the diet with vitamin E, calcium, magnesium, and folic acid may be helpful for people with RLS.

Some alternative methods may treat the associated condition that is suspected to cause restless legs. These include:

- Anemia or low ferritin levels. Chinese medicine will emphasize stimulation of the spleen as a means of improving blood circulation and vitamin absorption. Other treatments may include acupuncture and herbal therapies, such as ginseng (*Panax ginseng*) for anemia-related fatigue.
- Late-stage pregnancy. There are few conventional therapies available to pregnant women, since most of the drugs prescribed are not recommended for use during pregnancy. Pregnant women may benefit from alternative techniques that focus on body work, including yoga, reflexology, and acupuncture.

Prognosis

RLS usually does not indicate the onset of other neurological disease. It may remain static, although two-thirds of patients get worse with time. The symptoms usually progress gradually. Treatment with Levodopa is effective in moderate to severe cases that may include significant PLMS. However, this drug produces significant side effects, and continued successful treatment may depend on carefully monitored use of combination drug therapy. The prognosis is usually best if RLS symptoms are recent and can be traced to another treatable condition that is associated with RLS. Some associated conditions are not treatable. In these cases, such as for rheumatoid arthritis, alternative therapies such as **acupuncture** may be helpful.

Prevention

Diet is key in preventing RLS. A preventive diet will include an adequate intake of iron and the B vitamins, especially B₁₂ and folic acid. Strict vegetarians should take vitamin supplements to obtain sufficient vitamin B₁₂. Ferrous gluconate may be easier on the digestive system than ferrous sulfate, if iron supplements are prescribed. Some medications may cause symptoms of RLS. Patients should check with their doctor about these possible side effects, especially if symptoms first occur after starting a new medication. Caffeine, alcohol, and nicotine use should be minimized or eliminated. Even a hot bath before bed has been shown to prevent symptoms for some sufferers.

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- Restless Legs Syndrome Foundation, 1904 Banbury Road, Raleigh, NC, 27608-4428, (919) 781-4428, <http://www.rls.org>.

Ann M. Haren

Restrictive cardiomyopathy

Definition

Cardiomyopathy is an ongoing disease process that damages the muscle wall of the lower chambers of the heart. Restrictive cardiomyopathy is a form of cardiomyopathy in which the walls of the heart become rigid.

Description

Restrictive cardiomyopathy is the least common type of cardiomyopathy in the United States. The stiffened heart walls cannot stretch properly to allow enough blood to fill the ventricles between heartbeats. As the stiffening worsens, **heart failure** occurs. The blood backs up into the blood vessels, causing fluid buildup in tissues (congestion and **edema**).

Causes and symptoms

Restrictive cardiomyopathy can be caused by a number of diseases. Often, the cause is unknown. The rigidity of the heart walls may be caused by fibrosis, the replacement of muscle cells with tough, fibrous tissue. In some disorders, proteins and other substances are deposited in the heart wall. **Amyloidosis** is the accumulation of a protein material, called amyloid, in the tissue of the heart wall and other organs. In **hemochromatosis**, there is too much iron in the body and some of the excess iron can build up in the heart. **Sarcoidosis** causes the formation of many small lesions, called granulomas, in the heart wall and other tissues of the body. These granulomas contain inflammatory white blood cells and other cells that decrease the flexibility of the heart.

People with restrictive cardiomyopathy usually feel tired and weak, and have **shortness of breath**, especially during **exercise**. If blood is backing up in the circulation they may also experience edema (large amounts of fluid in tissues) of the legs and feet.

Diagnosis

The diagnosis is usually based on a **physical examination**, **echocardiography**, and other tests as needed. The physician listens to the heart with a stethoscope to detect abnormal heart rhythms and heart sounds.

Echocardiography uses sound waves to make images of the heart. These images provide information about the structures of the heart and its heart valves. Echocardiography can also be used to find out how much blood the heart is pumping. It determines the

KEY TERMS

Amyloidosis—Build up of amyloid, a protein substance, in tissues of the body, including the heart.

Cardiac catheterization—A diagnostic test for evaluating heart disease; a catheter is inserted into an artery and passed into the heart.

Edema—Swelling caused by fluid buildup in tissues.

Fibrosis—Replacement of normal tissue with tough, fibrous tissue.

Hemochromatosis—A disease in which there is too much iron in the body; iron deposits can build up in the heart muscle and other tissues.

Sarcoidosis—A chronic disease causing the formation of many small lesions called granulomas in the heart wall and other tissues of the body.

amount of blood in the ventricle, called the ventricular volume, and the amount of blood the ventricle pumps each time it beats, called the ejection fraction. A healthy heart pumps at least one half the amount of blood in the left ventricle with each heartbeat.

Computed tomography scan (CT scan) and **magnetic resonance imaging** (MRI) are imaging tests that can also provide information about the structure of the heart. However, these tests are rarely needed for diagnosis.

Cardiac catheterization may be needed to confirm a diagnosis or cause. In cardiac catheterization, a small tube called a catheter is inserted into an artery and passed into the heart. It is used to measure pressure in the heart and the amount of blood pumped by the heart. A small tissue sample (biopsy) of the heart muscle can be removed through the catheter for microscopic examination. Fibrous tissue or deposits in the heart muscle can be identified in this biopsy.

Treatment

There is no effective treatment for restrictive cardiomyopathy. Treatment of a causative disease may reduce or stop the damage to the heart, but existing damage cannot be reversed. Medications may be used to lessen the workload on the heart and to control the heart rhythm. Drugs normally used to treat other types of cardiomyopathy and heart failure may cause problems for patients with restrictive

cardiomyopathy. For example, medicines that reduce the heart's workload may lower blood pressure too much.

A heart transplant may be necessary for patients who develop severe heart failure.

Prognosis

The prognosis for patients with restrictive cardiomyopathy is poor. If the disease process causing the problem can be treated, the damage to the heart muscle may be stopped. Also, medicines may relieve symptoms. However, for most patients, restrictive cardiomyopathy eventually causes heart failure. A heart transplant may be necessary when heart failure becomes too severe to treat with medicines.

Prevention

Obtaining early treatment for diseases that might cause restrictive cardiomyopathy might prevent or slow the development of heart wall stiffness. Anyone experiencing symptoms of shortness of breath, tiredness, and weakness should see a physician.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, 240 629-3246, <http://www.nhlbi.nih.gov>.

Texas Heart Institute. Heart Information Service, MC 3-116, P.O. Box 20345, Houston, TX, 77225, 832 355-4011, (800) 292-2221, <http://www.texasheart.org>.

Toni Rizzo

Reticulocyte count

Definition

A reticulocyte count is a blood test performed to assess the body's production of immature red blood cells (reticulocytes). A reticulocyte count is usually performed when patients are evaluated for anemia and response to its treatment. It is sometimes called a retic count.

Purpose

Diagnosis

A reticulocyte count provides information about the rate at which the bone marrow is producing red cells. A normal count means that the production is adequate; a decreased count means it is not. This information helps determine whether a lack of red cells in an anemic person is caused by a bone marrow problem, by excessive bleeding, or by red cell destruction.

Monitoring

The test is also used to monitor the response of bone marrow response to treatment for anemia. The reticulocyte count rises within days if the treatment is successful. It is also used following bone marrow transplant to evaluate the new marrow's cell production.

Description

Reticulocytes were first described as transitional forms of red blood cells by Wilhelm H. Erb in 1865. A red cell begins in the bone marrow as a large bluish cell filled with ribonucleic acid (RNA). As the cell matures, it shrinks. Its color gradually changes from blue to pink as its load of oxygen-carrying protein (hemoglobin) increases and the RNA decreases. The center of the cell (nucleus) becomes clumped. It is expelled three days before the cell leaves the bone marrow. The cell is now a reticulocyte. On its fourth and final day of maturation, the reticulocyte enters the bloodstream. One day later, it is a mature red blood cell.

The first step in a retic count is drawing the patient's blood sample. About 17 oz (5 mL) of blood is withdrawn from a vein into a vacuum tube. The procedure, which is called a venipuncture, takes about five minutes.

After the sample is collected, the blood is mixed with a dye (methylene blue) in a test tube. The RNA remaining in the reticulocytes picks up a deep blue stain. Drops of the mixture are smeared on slides and examined under a microscope. Reticulocytes appear as cells containing dark blue granules or a blue network. The laboratory technologist counts 1,000 red cells, keeping track of the number of reticulocytes. The number of reticulocytes is reported as a percentage of the total red cells. When the red cell count is low, the percentage of reticulocytes is inaccurately high, suggesting that more reticulocytes are present than there are in reality. The percentage is mathematically corrected for greater accuracy. This figure is called the corrected reticulocyte count or reticulocyte index.

KEY TERMS

Anemia—A condition marked by a decrease in the number or size of red blood cells

Methylene blue—A dye that is used to stain the blood cells for the reticulocyte count.

Reticulocyte—An immature red blood cell.

Reticulocyte counts can also be done on automated instruments, such as flow cytometers, using fluorescent stains. These instruments are able to detect small changes in the reticulocyte count because they count a larger number of cells (10,000–50,000).

Preparation

The doctor should make a note of any prescription medications that the patient is taking. Some drugs lower the red blood cell count.

Aftercare

Aftercare consists of routine care of the area around the puncture mark. Pressure is applied for a few seconds and the wound is covered with a bandage.

Risks

The primary risk is mild **dizziness** and the possibility of a bruise or swelling in the area where the blood was drawn. The patient can apply moist warm compresses.

Normal results

Adults have reticulocyte counts of 0.5–2.5%. Women and children usually have higher reticulocyte counts than men.

Abnormal results

A low reticulocyte count indicates that the bone marrow is not producing a normal number of red blood cells. Low production may be caused by a lack of vitamin B₁₂, **folic acid**, or iron in the diet; or by an illness affecting the bone marrow (for example, **cancer**). Further tests are needed to diagnose the specific cause.

The reticulocyte count rises when the bone marrow makes more red cells in response to blood loss or treatment of anemia.

Resources

PERIODICALS

Rowan, R. M., et al. "The Reticulocyte Count: Progress Towards the Resurrection of a Useful Clinical Test." *Clinical and Laboratory Haematology* 18, no. 1 (1996): 3-8.

Nancy J. Nordenson

Retinal artery occlusion

Definition

Retinal artery occlusion refers to the closure of the central retinal artery and usually results in complete loss of vision in one eye. Occlusion of its branches causes loss of vision in only a portion of the field of vision.

Description

Retinal artery occlusion (RAO) occurs when the central retinal artery, the main source of blood supply to the retina, or one of its branches becomes blocked.

Causes and symptoms

The main causes of RAO are the following:

- embolism (the sudden obstruction of a blood vessel by a blood clot)
- atherosclerotic disease that results in the progressive narrowing of the arteries over time
- endarteritis (the chronic inflammation of the inner layer of arteries)
- angospasm (a spasmodic contraction of a blood vessel with increase in blood pressure)

The most common symptom of RAO is an acute, painless loss of vision in one eye. The degree of loss depends on the location of the occlusion. If the occlusion occurs in the central artery of the retina, damage usually results in complete loss of vision in the affected eye. If occlusion occurs in a branch artery, vision loss will be partial and may even go unnoticed if only a section of the peripheral vision is affected.

People affected by RAO typically have high blood pressure, heart disease, or diabetes as an underlying condition. Other conditions that may increase the risk of RAO include high cholesterol and glaucoma. Incidence is slightly more common in men and in people age 60 or older.

Diagnosis

RAO is diagnosed by examination of the retina with an ophthalmoscope.

Treatment

Central retinal artery occlusion (CRAO) is an emergency. If treatment begins within an hour, the patient has the highest possibility of regaining vision in the affected eye, although complete restoration is unlikely.

A common treatment is inhalation of carbon dioxide so as to dilate the retinal vessels and move the occlusion from the central retinal artery to a branch artery. This movement reduces the area of the retina affected and may restore a certain amount of vision. Eyeball massage may also be performed, also in an effort to remove the occlusion. The physician may also consider puncturing the eyeball.

Drug therapy includes the use of carbonic anhydrase inhibitors to reduce the internal eye pressure and enhance movement of the occlusion. Both of the treatments would be used within the first 24 hours of noticeable vision loss.

Alternative treatment

Hyperbaric **oxygen therapy** may be beneficial if started within 90 minutes of the onset of symptoms. Some studies indicate a 40% improvement of visual acuity using this method.

Prognosis

The prognosis for central retinal visual acuity is poor with only about one-third of patients recovering useful vision. The longest delay in getting treatment that has been associated with significant visual recovery was approximately 72 hours.

Branch retinal artery occlusions (BRAO) have a recovery rate of 80% where vision is restored to 20/40 or better.

Prevention

Individuals affected by underlying conditions such as high blood pressure, heart disease, diabetes, glaucoma, and elevated cholesterol should treat their conditions appropriately to minimize the possibility of a retinal artery occlusion.

KEY TERMS

Angiospasm—Spasmodic contraction of a blood vessel with increase in blood pressure.

Arterioles—Small blood vessels that carry arterial (oxygenated) blood.

Atherosclerotic disease—The progressive narrowing and hardening of the arteries over time.

Central retinal artery—A branch of the ophthalmic artery that supplies blood to the retina and branches to form the arterioles of the retina.

Embolism—The sudden obstruction of a blood vessel by a blood clot.

Endarteritis—Chronic inflammation of the inner layer of arteries.

Hyperbaric oxygenation—Administration of oxygen in a compression chamber at an ambient pressure greater than 1 atmosphere, in order to increase the amount of oxygen in organs and tissues.

Occlusion—Momentary complete closure of some area or channel of the body.

Ophthalmic artery—The artery supplying the eye and adjacent structures with blood.

Ophthalmoscope—An instrument used for viewing the inside of the eye that consists of a concave mirror with a hole in the middle through which the physician examines the eye, and a light source that is reflected into the eye by the mirror.

Retina—Light sensitive layer of the eye, that consists of four major layers: the outer neural layer, containing nerve cells and blood vessels, the photoreceptor layer, a single layer that contains the light sensing rods and cones, the pigmented retinal epithelium (PRE) and the choroid, consisting of connective tissue and capillaries.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, 415 561-8500, <http://www.ao.org>.

American Diabetes Association, 1701 North Beauregard Street, Alexandria, VA, 22311, (800) 342-2383, Ask ADA@diabetes.org, <http://www.diabetes.org/>.

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review. personal.info@heart.org.

Gary Gilles

thin membrane that supplies nutrients to part of the retina. The innermost layer is the retina.

The retina is the light-sensitive membrane that receives images and transmits them to the brain. It is made up of several layers. One layer contains the photoreceptors. The photoreceptors, the rods and cones, send the visual message to the brain. Between the photoreceptor layer (also called the sensory layer) and the choroid is the pigmented epithelium.

The vitreous is a clear gel-like substance that fills up most of the inner space of the eyeball. It lies behind the lens and is in contact with the retina.

A retinal detachment occurs between the two outermost layers of the retina—the photoreceptor layer and the outermost pigmented epithelium. Because the choroid supplies the photoreceptors with nutrients, a detachment can basically starve the photoreceptors. If a detachment is not repaired within 24-72 hours, permanent damage may occur.

Causes and symptoms

Several conditions may cause retinal detachment:

- Scarring or shrinkage of the vitreous can pull the retina inward.
- Small tears in the retina allow liquid to seep behind the retina and push it forward.
- Injury to the eye can simply knock the retina loose.

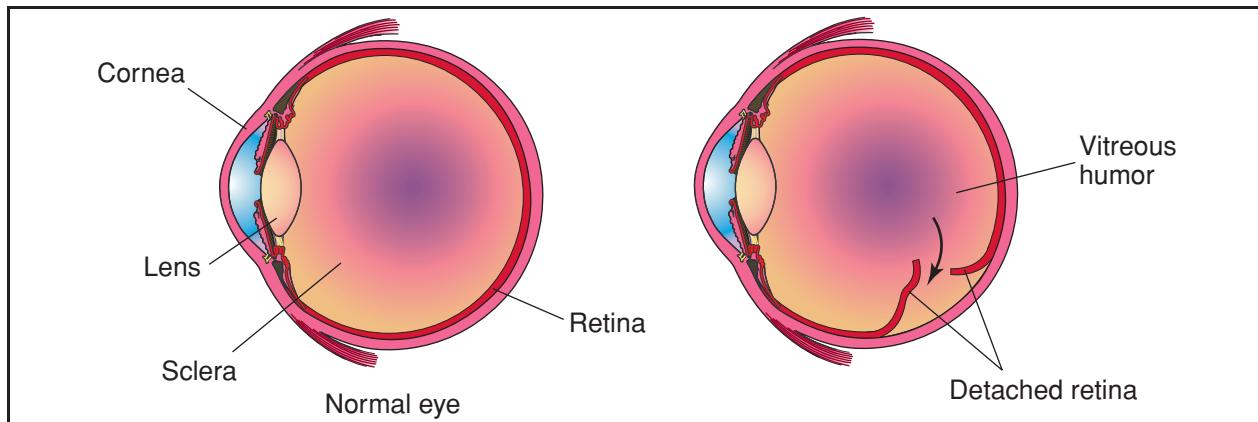
Retinal detachment

Definition

Retinal detachment is movement of the transparent sensory part of the retina away from the outer pigmented layer of the retina. In other words, the moving away of the retina from the outer wall of the eyeball.

Description

There are three layers of the eyeball. The outer, tough, white sclera. Lining the sclera is the choroid, a



Retinal detachment refers to the movement of the retina away from the inner wall of the eyeball, resulting in a sudden defect in vision. Persons suffering from diabetes have a high incidence of developing retinal disease. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

- Bleeding behind the retina, most often due to diabetic retinopathy or injury, can push it forward.
- Retinal detachment may be spontaneous. This occurs more often in the elderly or in very near-sighted (myopic) eyes.
- Cataract surgery causes retinal detachment 2% of the time.
- Tumors can cause the retina to detach.

Retinal detachment will cause a sudden defect in vision. It may look as if a curtain or shadow has just descended before the eye. If most of the retina is detached, there may be only a small hole of vision remaining. If just a part of the retina is involved, there will be a blind spot that may not even be noticed. It is often associated with *floaters*—little dark spots that float across the eye and can be mistaken for flies in the room. There may also be *flashes* of light. Anyone experiencing a sudden onset of flashes and/or floaters should contact their eye doctor immediately, as this may signal a detachment.

Diagnosis

If the eye is clear—that is, if there is no clouding of the liquids inside the eye—the detachment can be seen by looking into the eye with a hand-held instrument called an ophthalmoscope. To evaluate the blood vessels in the retina, a fluorescent dye (fluorescein) may be injected into a vein and photographed with ultraviolet light as it passes through the retina. Further studies may include computed tomography scan (CT scan), **magnetic resonance imaging** (MRI), or ultrasound study. Other lenses may be used to examine the back of the eyes. One example is binocular

indirect ophthalmoscopy. The doctor dilates the patient's eyes with eyedrops and then examines the back of the eyes with a handheld lens.

Treatment

Reattaching the retina to the inner surface of the eye requires making a scar that will hold it in place and then bringing the retina close to the scarred area. The scar can be made from the outside, through the sclera, using either a laser or a freezing cold probe (cryopexy). Bringing the retina close to the scar can be done in two ways. A tiny belt tightened around the eyeball will bring the sclera in until it reaches the retina. This procedure is called scleral buckling and may be done under **general anesthesia**. Using this procedure permits the repair of retinal detachments without entering the eyeball. Sometimes, the eye must be entered to pump in air or gas, forcing the retina outward against the sclera and its scar. This is called pneumatic retinopexy and can generally be done under **local anesthesia**.

If all else fails, and especially if there is disease in the vitreous, the vitreous may have to be removed in a procedure called **vitrectomy**. This can be done through tiny holes in the eye, through which equally tiny instruments are placed to suck out the vitreous and replace it with saline, a salt solution. The procedure must maintain pressure inside the eye so that the eye does not collapse.

Prognosis

Retinal reattachment has an 80-90% success rate.

KEY TERMS

Cauterize—To damage with heat or cold so that tissues shrink. It is an effective way to stop bleeding.

Diabetic retinopathy—Disease that damages the blood vessels in the back of the eye caused by diabetes.

Saline—A salt solution equivalent to that in the body—0.9% salt in water.

Prevention

In diseases such as diabetes, with a high incidence of retinal disease, routine eye examinations can detect early changes. Early treatment can prevent both progressing to detachment and blindness from other events like hemorrhage. The most common problem is weakness of blood vessels that causes them to break down and bleed. When enough vessels have been damaged, new vessels grow to replace them. These new vessels may grow into the vitreous, producing blind spots and scarring. The scarring can in turn pull the retina loose. Other diseases can cause the tiny holes and tears in the retina through which fluid can leak. Preventive treatment uses a laser to cauterize the blood vessels, so that they do not bleed and the holes, so they do not leak.

Good control of diabetes can help prevent diabetic eye disease. Blood pressure control can prevent **hypertension** from damaging the retinal blood vessels. Eye protection can prevent direct injury to the eyes. Regular eye exams can also detect changes that the patient may not be aware of. This is important for patients with high **myopia** who may be more prone to detachment.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, 415 561-8500, <http://www.aoa.org>.

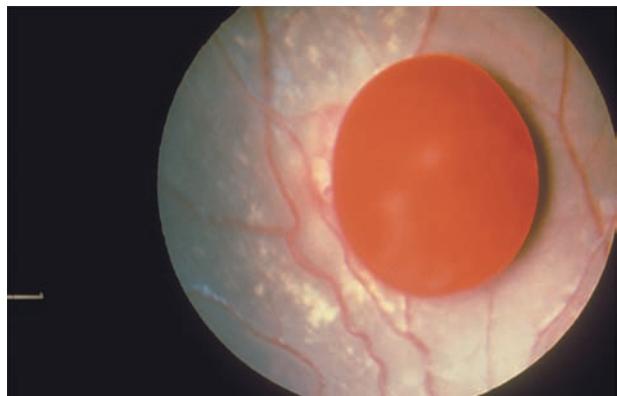
American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, 314 991-4100, (314) 991-4101, 800 365-2219, <http://www.aoa.org/>.

J. Ricker Polsdorfer, MD

Retinal hemorrhage

Definition

Retinal hemorrhage is the abnormal bleeding of the blood vessels in the retina, the membrane in the back of the eye.



A close-up view of a human eye following retinal hemorrhage.
(Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

The retina is the part of the eye that converts light into nerve signals that are processed by the brain into visual images. The retina is the inside surface of the back of the eye, consisting of millions of densely arranged, light-sensitive cells called rods and cones. Blood flow to the retina is maintained by the retinal vein and artery, and a dense network of small blood vessels (capillaries) supplies the area with circulation. These blood vessels can become damaged by injury and disease and may bleed (hemorrhage) and cause temporary or permanent loss of visual accuracy. Because the cells of the retina are so dense and sensitive, even small injuries to the blood vessels can translate into vision problems. Diseases that affect the health of the circulatory system, such as diabetes and high blood pressure, also affect the blood vessels of the eye. Damage to the blood vessels in the retina, including hemorrhage, is termed retinopathy.

Causes and symptoms

Retinal hemorrhages can be caused by injuries, usually forceful blows to the head during accidents and falls, as well as by adverse health conditions. In infants, retinal hemorrhage is frequently associated with **child abuse** and has been termed **shaken baby syndrome**. A condition called retinopathy of **prematurity** occurs in prematurely born infants or infants with low birth weights. When children are born prematurely, the blood vessels in the eye may not have had time to fully develop and may become damaged easily, leaking or hemorrhaging. The condition must be determined by an ophthalmologist, as the symptoms are not readily observable.

Diabetic retinopathy is a common eye problem associated with diabetes. Diabetes, by stressing the circulatory system, can cause damage, including hemorrhaging, to the small blood vessels of the retina. Non-proliferative retinopathy occurs when the damaged or leaking blood vessels do not spread. Symptoms of this disorder include vision spots, floaters (floating areas of blurred vision), decreased or loss of vision, or loss of fine vision for detailed activities such as reading. Proliferative retinopathy occurs when new blood vessels begin to form in damaged areas of the retina, and may lead to spots, floaters, decreased vision, or sudden loss of vision. Sudden vision loss may occur if one of the newly formed blood vessels ruptures. Due to increased pressure in the area, the retina may detach from the back of the eye, a serious condition and a cause of blindness.

People with high blood pressure (**hypertension**) may develop hypertensive retinopathy, in which blood vessels in the retina become damaged from increased blood pressure. Symptoms are typically not pronounced, but blurred or decreased vision may be caused by the disorder.

Central serous retinopathy is a condition in which the vessels behind the retina leak and cause fluid to collect in small blisters behind the retina. Symptoms include sudden blurry areas in the vision, blind spots, distorted vision areas, and loss of vision. This condition is most common in males between 20 and 50 years of age.

Diagnosis

Diagnosis of retinopathy is performed by an ophthalmologist, particularly one who specializes in disorders of the retina (retinal specialist). The ophthalmologist may perform an ophthalmoscopy, using an instrument called an ophthalmoscope to examine the inside of the eye. For a detailed view of the blood vessels of the retina, a fluorescein **angiography** test might be performed, in which a fluorescent dye is injected into the patient's bloodstream and photographs record the status of the blood vessels in the retina. Vision tests, patient history, and blood tests might also be ordered by the diagnosing physician.

Treatment

Laser surgery by an ophthalmologist is a common treatment for retinal hemorrhages, in which a laser beam is used to remove or seal off damaged or bleeding blood vessels in the retina. Some vision loss occurs with this technique. For retinal hemorrhages associated with diabetes and high blood pressure, treating the overall condition is required.

KEY TERMS

Diabetes—Disease in which the body does not properly produce or use insulin, resulting in fluctuating blood sugar levels.

Hypertension—Condition caused by high blood pressure.

Ophthalmologist—Physician specializing in the diagnosis and treatment of disorders of the eye.

Alternative treatment

Alternative treatment of retinal hemorrhages focuses on providing nutrients to strengthen and heal the injured blood vessels. **Nutritional supplements** include antioxidant **vitamins A, C, and E**; vitamin B-complex including B₆ and B₁₂; the mineral zinc; and essential fatty acids including omega-3 from fish oil and flaxseed oil. Herbal supplements include bilberry, grape seed extract, pine bark extract (pycnogenol), and lutein.

Prognosis

For retinal hemorrhages associated with retinopathy of prematurity, nearly 85 percent of cases heal without treatment. Diabetic retinopathy is the leading cause of blindness for those between 20 and 65 years old in the U.S. Diabetic retinopathy typically takes years to develop in people with diabetes, but occurs in nearly 80 percent of those with diabetes for over 20 years and who are treated with insulin. Regular monitoring and treatments can slow the degeneration of the eye, while advanced cases of the disorder lead to blindness. **Retinopathies** requiring laser treatment have a partial loss of vision due to the surgery. For hypertensive retinopathy, most vision problems go away when high blood pressure is treated and lowered. The majority of cases of central serous retinopathy disappear after three to four months, and full vision generally returns within six months, although recurrence of the disorder is common.

Prevention

The first step in sound prevention is for people with vision problems, including visual spots, flashes or floaters in the vision, and loss or distortion of visual accuracy, to see an ophthalmologist as soon as possible. To prevent complications of retinal hemorrhages in infants, the prevention includes regular prenatal care and monitoring of infants with high risks of the

disorder (born prematurely or with weight less than four pounds and six ounces). For diabetic retinopathy, control of blood sugar and blood pressure fluctuations is necessary, as well as frequently scheduled eye exams by an ophthalmologist. For retinal hemorrhages associated with hypertension, controlling high blood pressure through diet, **exercise**, and **stress reduction** is recommended. Central serous retinopathy has been associated with high **stress** levels, so preventative care for this disorder includes stress management practices.

Resources

BOOKS

Dick, Andrew D., John V. Forrester, and Annabelle Ayame Okada. *Practical Manual of Intraocular Inflammation*. New York: Informa Healthcare, 2008.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, 415 561-8500, <http://www.aao.org>.

National Eye Institute, 2020 Vision Place, Bethesda, MD, (301) 496-5248, <http://www.nei.nih.gov>.

Douglas Dupler

where the optic nerve enters the eye, by dilated retinal veins, and by retinal hemorrhages. CRVO is also called venous stasis retinopathy, or hemorrhagic retinopathy.

In BRVO, the superotemporal branch vein is the most often affected vessel. Retinal hemorrhages follows, often occurring at the crossing of two vessels near the optic disk. Initially the hemorrhage may be extensive and underlie the fovea.

The exact cause of RVO is not yet identified, but the following mechanisms been proposed:

- external compression between the central connective strand and the cribriform plate
- venous disease
- blood clot formation

Conditions associated with RVO risk include:

- hypertension
- hyperlipidemia
- diabetes mellitus
- hyperviscosity
- hypercoagulability
- glaucoma
- trauma

Diagnosis

A complete physical evaluation is recommended for CRVO and BRVO, including complete blood tests, and glucose tolerance test (for non-diabetics). In the case of a **head injury** when bleeding around the optic nerve is a possibility, an MRI may be performed.

Treatment

Following a patient with RVO is vital. Patients should be seen at least monthly for the first three months to monitor for signs of other complications, such as the abnormal formation of blood vessels (neovascularization) in the iris of the eye or glaucoma.

The treatment for retinal vein occlusion varies for each case and should be given based on the doctor's best recommendation. Although treatments for occlusion itself are limited, surgical treatment of the occlusion provides an option.

Treatments may include anticoagulants with heparin, bishydroxycoumarin, and streptokinase. When the blood is highly viscous, dilution of the blood may be useful. Ideally, an alternate pathway is needed to allow venous drainage. Recent reports published in 1999 suggest that use of a laser to create a retinal choroidal hole may be useful to treat CRVO. Laser therapy depends

Retinal vein occlusion

Definition

Retinal vein occlusion refers to the closure of the central retinal vein that drains the retina or to that of one of its branches.

Description

Retinal vein occlusion (RVO) occurs when the central retinal vein, the blood vessel that drains the retina, or one of its branches becomes blocked. RVO may be categorized by the anatomy of the occluded vein and the degree of **ischemia** produced. The two major RVO types are central retinal vein occlusion (CRVO) and branch retinal vein occlusion (BRVO). CRVO has been diagnosed in patients as young as nine months to patients of 90 years. The age of affected individuals is usually low to mid 60s. Approximately 90% of patients are over 50 at the time of diagnosis, with 57% of them being male and 43% being female. BRVO accounts for some 30% of all vein occlusions.

Causes and symptoms

CRVO is a painless loss of vision that can be caused by a swollen optic disk, the small area in the retina

KEY TERMS

Anticoagulants—Drugs that act by lowering the capacity of the blood to coagulate, thus facilitating removal of blood clots.

Central retinal vein—Central blood vessel and its branches that drains the retina.

Cribriform plate—The horizontal bone plate perforated with several holes for the passage of olfactory nerve filaments from the nasal cavity.

Fovea—A small area of the retina responsible for acute vision.

Glaucoma—A group of eye diseases characterized by an increase in eyeball pressure.

Hyperlipidemia—A general term for elevated concentrations of any or all of the lipids in the plasma.

Iris—The contractile diaphragm located in the fluid in front of the lens of the eye and is perforated by the eye pupil.

Ischemia—A state of low oxygen in a tissue usually due to organ dysfunction.

Neovascularization—Abnormal or excessive formation of blood vessels as in some retinal disorders.

Occlusion—Momentary complete closure of some area or channel of the body.

Optic disk—The small area in the retina where the optic nerve enters the eye that is not sensitive to light. Also called the blind spot.

Retina—Light sensitive layer of the eye, that consists of four major layers: the outer neural layer, containing nerve cells and blood vessels, the photoreceptor layer, a single layer that contains the light sensing rods and cones, the pigmented retinal epithelium (PRE) and the choroid, consisting of connective tissue and capillaries.

on the type of occlusion. The management of laser therapy should be controlled by an ophthalmologist.

Alternative treatment

There are no documented alternative treatment methods.

Prognosis

The outlook for people with RVO is fairly good whether it is treated early or not. With no treatment at all, approximately 60% of all patients recover 20/40 vision or better within a year.

Prevention

Retinal vein occlusion is difficult to prevent because the exact cause is still uncertain. Ethnic factors may play a role since in the UK the disease is rare in Asians and West Indians.

Resources

BOOKS

Yanoff, Myron, et al, eds. *Ophthalmology*. 3rd ed. Edinburgh: Mosby International, 2009.

Michael Sherwin Walston
Ronald Watson, PhD

Retinitis pigmentosa

Definition

Retinitis pigmentosa (RP) refers to a group of inherited disorders that slowly lead to blindness due to abnormalities of the photoreceptors (primarily the rods) in the retina.

Description

The retina lines the interior surface of the back of the eye. The retina is made up of several layers. One layer contains two types of photoreceptor cells referred to as the rods and cones. The cones are responsible for sharp, central vision and color vision and are primarily located in a small area of the retina called the fovea. The area surrounding the fovea contains the rods, which are necessary for peripheral vision and night vision (scotopic vision). The number of rods increases in the periphery. The rod and cone photoreceptors convert light into electrical impulses and send the message to the brain via the optic nerve. Another layer of the retina is called the retinal pigmented epithelium (RPE).

In RP, the photoreceptors (primarily the rods) begin to deteriorate and lose their ability to function. Because the rods are primarily affected, it becomes harder to see in dim light, thus causing a loss of night



A fundus camera image showing the degeneration of the retina due to retinitis pigmentosa. The pattern of dark spots across the retina corresponds to the extent of loss of vision.
(Custom Medical Stock Photo, Inc. Reproduced by permission.)

vision. As the condition worsens, peripheral vision disappears, which results in tunnel vision. The ability to see color is eventually lost. In the late stages of the disease, there is only a small area of central vision remaining. Ultimately, this too is lost.

There are many forms of retinitis pigmentosa. Sometimes the disorder is classified by the age of onset or the inheritance pattern. RP can also accompany other conditions. This entry discusses “non-syndromic” RP, the type that is not associated with other organ or tissue dysfunction.

Approximately 100,000 Americans have RP. It is estimated to affect about one in every 4,000 Americans and Europeans. For other parts of the world, there are no published data. Nor is there any known ethnic difference in the occurrence of RP.

Causes and symptoms

Retinitis pigmentosa is an inherited disease that has many different modes of inheritance. It is known to be caused by more than 100 different genetic mutations. RP, with any inheritance pattern, may be either familial (multiple family members affected) or isolated (only one affected person). In the non-sex-linked, or autosomal, form, it can either be a dominant or recessive trait. In the sex-linked form, called x-linked recessive, it is a recessive trait. This x-linked form is more severe than the autosomal forms. Two rare forms of RP are the digenic and mitochondrial forms.

Isolated RP cases represent 10–40% of all cases. Some of these cases may be the result of new gene mutations (changes in the genes). Other isolated cases are those in which the person has a relative

with a mutation in the gene, but the relative is not affected by the condition.

Autosomal dominant RP (AdRP) occurs in about 15–25% of affected individuals. At least 12 different genes have been identified as causing AdRP. People with AdRP will usually have an affected parent. The risk for affected siblings or children is 50%.

Autosomal recessive RP (ArRP) occurs in about 5–20% of affected individuals. More than 16 genes have been identified that cause this type of RP. In ArRP, each parent of the affected person is a carrier of an abnormal gene that causes RP. Neither of these carrier parents is affected. There is a two-thirds chance that an unaffected sibling is a carrier of RP. All of the children of an affected person would be a carrier of the ArRP gene.

Five to 15% of individuals with RP have x-linked recessive RP (XLRP). Six different genes have been identified as the cause of this type of RP. Usually in this type of inheritance, males are affected carriers, while females are unaffected carriers or have a milder form of the disease. The mother may be a carrier of the mutation on the X-chromosome. It is also possible that a new mutation can occur for the first time in an affected person. For families with one affected male, there is a mathematical formula called the Bayesian analysis that can be applied to the family history. It takes into account the number of unaffected males to determine whether a female is likely to be a carrier or not. If a mother is a carrier, her children have a 50% chance of inheriting the RP gene. For affected males, all of their daughters will be carriers but none of their sons will be affected.

The digenic form of RP occurs when the affected person has inherited one copy of an altered ROM1 gene from one parent and one copy of an altered peripherin/RDS gene from the other parent. The parents are asymptomatic. Mitochondrial inheritance occurs when the gene mutation is in a mitochondrial gene. People with this type of RP have progressive **hearing loss** and mild myopathy. Both of these types of RP are very rare.

The first symptoms, a loss of night vision followed by a loss of peripheral vision, usually begin in early adolescence or young adulthood. Occasionally, the loss of the ability to see color occurs before the loss of peripheral vision. Another possible symptom is seeing twinkling lights or small flashes of lights.

Diagnosis

When a person complains of a loss of night vision, a doctor will examine the interior of the eye with an

KEY TERMS

Ophthalmoscope—An instrument, with special lighting, designed to view structures in the back of the eye.

ophthalmoscope to determine if there are changes in the retina. For people with advanced RP, the condition is characterized by the presence of clumps of black pigment in the inner retina (intraretinal). However, the appearance of the retina is not enough for an RP diagnosis since there are other disorders that may give the retina a similar appearance. There are also other reasons someone may have night blindness. Consequently, certain electrodiagnostic tests must be performed. An electroretinogram (ERG) determines the functional status of the photoreceptors by exposing the retina to light. The ERG uses a contact lens in the eye, and the output is measured on a special instrument called an oscilloscope. The functional assessments of visual fields, visual acuity, or color vision may also be performed.

The diagnosis of RP can be established when the following criteria are met:

- rod dysfunction measured by dark adaptation test or ERG,
- progressive loss in photoreceptor function,
- loss of peripheral (side) vision,
- both eyes affected (bilateral).

Molecular **genetic testing** is available on a research basis. Prenatal diagnosis for this condition has not yet been achieved.

Treatment

There are no medications or surgery to treat RP. However, researchers continue to seek possible treatments. In 2004, scientists injected stem cells to the back of mouse eyes and stopped retinal degeneration. Scientists are also exploring the possibility of retinal transplantation. Some doctors believe **vitamins A** and E will slightly slow the progression of the disease in some people. However, large doses of certain vitamins may be toxic and affected individuals should speak to their doctors before taking supplements.

If a person with RP must be exposed to bright sunlight, some doctors recommend wearing dark sunglasses to reduce the effect on the retina. Affected people should talk to their eye doctors about the correct lenses to wear outdoors.

Because there is no cure for RP, the affected person should be monitored for visual function and counseled about low-vision aids (for example, field-expansion devices). **Genetic counseling** is also appropriate. A three-generation family history with attention to other relatives with possible RP can help to clarify the inheritance pattern. For some people however, the inheritance pattern cannot be discerned.

Prognosis

There is no known cure for RP, which will eventually lead to blindness. The more severe forms will lead to blindness sooner than milder forms.

Resources

PERIODICALS

“Grant Boosts RP Research Into Transplantation.” *Ophthalmology Times* August 1, 2004: 6.

“Stem Cells Delivered Into Back of Eye Hold Promise for People With Retinitis Pigmentosa, Other Retinal Degenerations; Potential Treatment for Untreatable Blindness Shows Promise in Mice.” *Ascribe Health News Service* September 15, 2004.

OTHER

Genetic Alliance. <http://www.geneticalliance.org>.

National Federation of the Blind. <http://www.nfb.org>.

“OMIM—Online Mendelian Inheritance in Man.” National Center for Biotechnology Information. <http://www.ncbi.nlm.nih.gov/Omim/searchomim.html>.

Retinitis Pigmentosa International. <http://www.rpinternational.org>.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, 415 561-8500, <http://www.aoa.org>.

American Association of the Deaf-Blind (AADB), 8630 Fenton Street, Suite 121, Silver Spring, MD, 20910-3803, (301) 495-4403, 301 495-4404, aadb-info@aadb.org, <http://www.aadb.org>.

American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, (314) 991-4100, 314 991-4101, 800 365-2219, <http://www.aoa.org/>.

Foundation Fighting Blindness, 7168 Columbia Gateway Drive, Suite 100, Columbia, MD, 21046, (800) 683-5555, info@FightBlindness.org, <http://www.blindness.org>.

Prevent Blindness America, 211 West Wacker Drive, Suite 1700, Chicago, IL, 60606, 636 947-7486, (800) 331-2020, <http://www.nationalshare.org>, <http://www.preventblindness.org>.

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Retinoblastoma

Definition

Retinoblastoma is a malignant tumor of the retina that occurs predominantly in young children.

Description

The eye has three layers, the sclera, the choroid, and the retina. The sclera is the outer protective white coating of the eye. The choroid is the middle layer and contains blood vessels that nourish the eye. The front portion of the choroid is colored and is called the iris. The opening in the iris is called the pupil. The pupil is responsible for allowing light into the eye and usually appears black. When the pupil is exposed to bright light it contracts (closes), and when it is exposed to low light conditions it dilates (opens) so that the appropriate amount of light enters the eye. Light that enters through the pupil hits the lens of the eye. The lens then focuses the light onto the retina, the innermost of the three layers. The job of the retina is to transform the light into information that can be transmitted to the optic nerve, which will transmit this information to the brain. It is through this process that people are able to see the world around them.

Occasionally a tumor, called a retinoblastoma, will develop in the retina of the eye. Usually this tumor forms in young children but it can occasionally occur in adults. Most people with retinoblastoma develop only one tumor (unifocal) in only one eye (unilateral). Some, however, develop multiple tumors (multifocal) in one or both eyes. When retinoblastoma occurs independently in both eyes, it is then called bilateral retinoblastoma.

Occasionally, children with retinoblastoma develop trilateral retinoblastoma. Trilateral retinoblastoma results from the development of an independent **brain tumor** that often forms in a part of the brain called the pineal gland. In order for retinoblastoma to be classified as trilateral retinoblastoma, the tumor must have developed independently and not as the result of the spread of the retinal **cancer**. The prognosis for trilateral retinoblastoma is quite poor.

The retinal tumor which characterizes retinoblastoma is malignant, meaning that it can metastasize (spread) to other parts of the eye and eventually other parts of the body. In most cases, however, retinoblastoma is diagnosed before it spreads past the eye to other parts of the body (intraocular) and the

prognosis is quite good. The prognosis is poorer if the cancer has spread beyond the eye (extraocular).

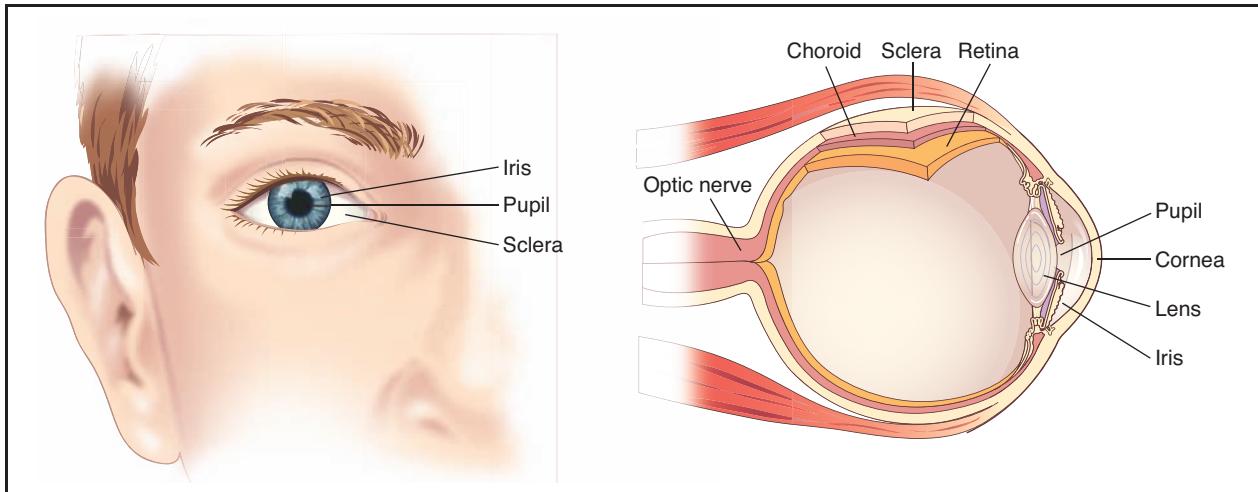
Retinoblastoma can be inherited or can arise spontaneously. Approximately 40% of people with retinoblastoma have an inherited form of the condition and approximately 60% have a sporadic (not inherited) form. Individuals with multiple independent tumors, bilateral retinoblastoma, or trilateral retinoblastoma are more likely to be affected with the inherited form of retinoblastoma.

Approximately 1 in 15,000 to 1 in 30,000 infants in Western countries are born with retinoblastoma, making it the most common childhood **eye cancer**. It is, however, a relatively rare childhood cancer and accounts for approximately 3% of childhood cancers. The American Academy of Ophthalmology estimates that 300-350 cases of retinoblastoma occur in the United States each year.

Retinoblastoma is found mainly in children under the age of five but can occasionally be seen in older children and adults. Retinoblastoma is found in individuals of all ethnic backgrounds and is found equally frequently in males and females. The incidence of bilateral retinoblastoma in the United States is thought to be slightly higher among black children than among either Caucasian or Asian American children.



This child's right eye is completely covered with a tumor associated with retinoblastoma. (Custom Medical Stock Photo, Inc. Reproduced by permission.)



An illustration of both the inner and outer eye. The illustration on the left shows the outer view of the eye, and the illustration on the right shows the inner anatomy of the eye. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Causes and symptoms

Causes

Retinoblastoma is caused by changes in or absence of a gene called RB1. RB1 is located on the long arm of chromosome 13. Cells of the body, with the exception of the egg and sperm cells, contain 23 pairs of chromosomes. All of the cells of the body excluding the egg and the sperm cells are called the somatic cells. The somatic cells contain two of each chromosome 13 and therefore two copies of the RB1 gene. Each egg and sperm cell contains only one copy of chromosome 13 and therefore only one copy of the RB1 gene.

RB1 produces a tumor suppressor protein that normally helps to regulate the cell cycle of cells such as those of the retina. A normal cell of the retina goes through a growth cycle during which it produces new cells. Genes such as tumor suppressor genes tightly regulate this growth cycle.

Cells that lose control of their cell cycle and replicate out of control are called cancer cells. These undergo many cell divisions, often at a quicker rate than normal cells, and do not have a limited lifespan. A group of adjacent cancer cells can form a mass called a tumor. Malignant (cancerous) tumors can spread to other parts of the body. A malignant tumor of the retina (retinoblastoma) can result when just one retinal cell loses control of its cell cycle and replicates out of control.

Normally the tumor suppressor protein produced by RB1 prevents a retinal cell from becoming cancerous. Each RB1 gene produces tumor suppressor protein. Only one functioning RB1 gene in a retinal cell is

necessary to prevent the cell from becoming cancerous. If both RB1 genes in a retinal cell become non-functional, then a retinal cell can become cancerous and retinoblastoma can result. An RB1 gene is non-functional when it is changed or missing (deleted) and no longer produces normal tumor suppressor protein.

Approximately 40% of people with retinoblastoma have inherited a non-functional or deleted RB1 gene from either their mother or father. Therefore, they have a changed/deleted RB1 gene in every somatic cell. A person with an inherited missing or non-functional RB1 gene will develop a retinal tumor if the remaining RB1 gene becomes changed or deleted in a retinal cell. The remaining RB1 gene can become non-functional when exposed to environmental triggers such as chemicals and radiation. In most cases, however, the triggers are unknown. Approximately 90% of people who inherit a changed or missing RB1 gene will develop retinoblastoma.

People with an inherited form of retinoblastoma are more likely to have a tumor in both eyes (bilateral) and are more likely to have more than one independent tumor (multifocal) in one or both eyes. The average age of onset for the inherited form of retinoblastoma is one year, which is earlier than the sporadic form of retinoblastoma. Although most people with the inherited form of retinoblastoma develop bilateral tumors, approximately 15% of people with a tumor in only one eye (unilateral) are affected with an inherited form of retinoblastoma.

A person with an inherited missing or non-functional RB1 gene has a 50% chance of passing on this abnormal gene to his or her offspring. The chance that their children will inherit the changed/

deleted gene and actually develop retinoblastoma is approximately 45%.

Some people with retinoblastoma have inherited a non-functioning or missing RB1 gene from either their mother or father even though their parents have never developed retinoblastoma. It is possible that one parent has a changed or missing RB1 gene in every somatic cell but has not developed retinoblastoma because their remaining RB1 gene has remained functional. It is also possible that the parent had developed a retinal tumor that was destroyed by the body. In other cases, one parent has two normal RB1 genes in every somatic cell, but some of their egg or sperm cells contain a changed or missing RB1 gene. This is called gonadal mosaicism.

Retinoblastoma can also result when both RB1 genes become spontaneously changed or deleted in a retinal cell but the RB1 genes are normal in all the other cells of the body. Approximately 60% of people with retinoblastoma have this type of disease, called sporadic retinoblastoma. A person with sporadic retinoblastoma does not have a higher chance of having children with the disease. Their relatives do not have a higher risk of developing retinoblastoma themselves or having children who develop retinoblastoma. Sporadic retinoblastoma is usually unifocal and has an average age of onset of approximately two years.

Symptoms

The most common symptom of retinoblastoma is leukocoria. Leukocoria results when the pupil reflects a white color rather than the normal black or red color that is seen on a flash photograph. It is often most obvious in flash photographs; since the pupil is exposed to a lot of light and the duration of the exposure is so short, the pupil does not have time to constrict. Children with retinoblastoma can also have problems seeing and this can cause them to appear cross-eyed (**strabismus**). People with retinoblastoma may also experience red, painful, and irritated eyes, inflamed tissue around the eye, enlarged pupils, and possibly different-colored eyes.

Diagnosis

Children who have symptoms of retinoblastoma are usually first evaluated by their pediatrician. The pediatrician will often perform a red reflex test to diagnose or confirm leukocoria. Prior to this test the doctor inserts medicated eye drops into the child's eyes so that the pupils will remain dilated and not contract when exposed to bright light. The doctor then examines the eyes with an ophthalmoscope, which shines a bright light into the eyes and allows the doctor to check for leukocoria. Leukocoria can also be diagnosed by taking

a flash Polaroid photograph of a patient who has been in a dark room for three to five minutes.

If the pediatrician suspects retinoblastoma on the basis of these evaluations, he or she will most likely refer the patient to an ophthalmologist (eye doctor) who has experience with retinoblastoma. The ophthalmologist will examine the eye using an indirect ophthalmoscope. The ophthalmoscope shines a bright light into the eye, which helps the doctor to visualize the retina. This evaluation is usually done under general anesthetic, although some very young or older patients may not require it. Prior to the examination, medicated drops are put into the eyes to dilate the pupils, and anesthetic drops may also be used. A metal clip is used to keep the eyes open during the evaluation. During the examination, a cotton swab or a metal instrument with a flattened tip is used to press on the outer lens of the eye so that a better view of the front areas of the retina can be obtained. Sketches or photographs of the tumor as seen through the ophthalmoscope are taken during the procedure.

An ultrasound evaluation is used to confirm the presence of the tumor and to evaluate its size. Computed axial tomography (CT scan) is used to determine whether the tumor has spread outside of the eye and to the brain. Sometimes **magnetic resonance imaging** (MRI) is also used to look at the eyes, eye sockets, and the brain to see if the cancer has spread.

In most cases the cancer has not spread beyond the eye, and other evaluations are unnecessary. If the cancer appears to have spread beyond the eye, then other assessments such as a blood test, spinal tap (**lumbar puncture**), and/or **bone marrow biopsy** may be recommended. During a spinal tap, a needle is inserted between the vertebrae of the spinal column and a small sample of the fluid surrounding the spinal cord is obtained. In a bone marrow biopsy, a small amount of tissue (bone marrow) is taken from inside the hip or breast bone for examination.

Genetic testing

Establishing whether someone is affected with an inherited or non-inherited form of retinoblastoma can help to ascertain whether other family members such as siblings, cousins, and offspring are at increased risk for developing retinoblastoma. It can also sometimes help guide treatment choices, since patients with an inherited form of retinoblastoma may be at increased risk for developing recurrent tumors or other types of cancers, particularly when treated with radiation. It is helpful for the families of a child diagnosed with retinoblastoma to meet with a genetic specialist such as a genetic counselor and/or geneticist. These specialists

can help to ascertain the chances that the retinoblastoma is inherited and facilitate **genetic testing** if desired.

If a patient with unilateral or bilateral retinoblastoma has a relative or relatives with retinoblastoma, it can be assumed that they have an inherited form of retinoblastoma. However, it cannot be assumed that a patient without a family history of the disease has a sporadic form.

Even when there is no family history, most cases of bilateral and trilateral retinoblastoma are inherited, as are most cases of unilateral, multifocal retinoblastoma. However, only 15% of unilateral, unifocal retinoblastoma cases are inherited.

The only way to establish whether someone has an inherited form of retinoblastoma is to see if the retinoblastoma gene is changed or deleted in the blood cells obtained from a blood sample. Approximately 5–8% of individuals with retinoblastoma possess a chromosomal abnormality involving the RB1 gene that can be detected by looking at their chromosomes under the microscope. The chromosomes can be seen by obtaining a blood sample. If this type of chromosomal abnormality is detected in a child, then analysis of the parents' chromosomes should be performed. If one of the parents possesses a chromosomal abnormality, then they are at higher risk for having other offspring with retinoblastoma. Chromosome testing would be recommended for the blood relatives of the parent with the abnormality.

Usually, however, a chromosomal abnormality is not detected in a child with retinoblastoma. In this case, specialized DNA tests that look for small RB1 gene changes need to be performed on the blood cells. DNA testing can be difficult, time consuming, and expensive, since there are many possible RB1 gene changes that can cause the gene to become nonfunctional.

If a sample of tumor is available, then it is recommended that DNA testing be performed on the tumor cells prior to DNA testing of the blood cells. This testing can usually identify the gene changes/deletions in the RB1 genes that caused the tumor to develop. In some cases, RB1 gene changes/deletions are not found in the tumor cells (approximately 20% of RB1 gene changes or deletions are not detectable). In these cases, DNA testing of the blood cells will not be able to ascertain whether someone is affected with an inherited or non-inherited form of retinoblastoma.

If the changes in both RB1 genes are detected in the tumor cell, then these same changes can be looked for in the blood cells. If an RB1 gene is deleted or changed in all of the blood cells tested, the patient can be assumed to have been born with a changed/deleted RB1 gene in all of their cells. This person has a

50% chance of passing the RB1 gene change/deletion on to his or her children. Most of the time, this change/deletion has been inherited from a parent. Occasionally the gene change/deletion occurred spontaneously in the original cell that was formed when the egg and sperm came together at conception (*de novo*).

If an RB1 gene change/deletion is found in all of the blood cells tested, both parents should undergo blood testing to check for the same RB1 gene change/deletion. If the RB1 gene change/deletion is identified in one of the parents, it can be assumed that the retinoblastoma was inherited and that siblings have a 50% chance of inheriting the altered gene. More distant blood relatives of the parent with the identified RB1 gene change/deletion may also be at risk for developing retinoblastoma. Siblings and other relatives could undergo DNA testing to see if they have inherited the RB1 gene change/deletion.

If the RB1 gene change/deletion is not identified in either parent, then the results can be more difficult to interpret. In this case, there is a 90–94% chance that the retinoblastoma was not inherited.

In some cases, a person with retinoblastoma will have an RB1 gene change/deletion detected in some of their blood cells and not others. It can be assumed that this person did not inherit the retinoblastoma from either parent. Siblings and other relatives would therefore not be at increased risk for developing retinoblastoma. Offspring would be at increased risk since some of the egg or sperm cells could have the changed/deleted RB1 gene. The risks to offspring would probably be less than 50%.

In families where there are multiple family members affected with retinoblastoma, blood samples from multiple family members are often analyzed and compared through DNA testing. Ninety-five percent of the time, this type of analysis is able to detect patterns in the DNA that are associated with a changed RB1 gene in that particular family. When a pattern is detected, at-risk relatives can be tested to establish whether they have inherited an RB1 gene change/deletion.

PRENATAL TESTING. If chromosome or DNA testing identifies an RB1 gene/deletion in someone's blood cells, then prenatal testing can be performed on this person's offspring. An **amniocentesis** or **chorionic villus sampling** can be used to obtain fetal cells which can be analyzed for the RB1 gene change/deletion or chromosomal abnormality.

Treatment

A number of different classification (staging) systems are used to establish the severity of retinoblastoma and aid in choosing an appropriate treatment plan. The most widely used staging system is the Reese-Ellsworth

system. This system is used to classify intraocular tumors and predict which tumors are favorable enough that sight can be maintained. The Reese-Ellsworth classification system is divided into:

- Group I (very favorable for maintenance of sight): small solitary or multiple tumors, less than 6.4 mm in size (1 inch = 25.4 mm), located at or below the equator of the eye
- Group II (favorable for maintenance of sight): solitary or multiple tumors, 6.4–16 mm in size, located at or behind the equator of the eye
- Group III (possible for maintenance of sight): any tumor located in front of the equator of the eye, or a solitary tumor larger than 16 mm in size and located behind the equator of the eye
- Group IV (unfavorable for maintenance of sight): multiple tumors, some larger than 16 mm in size, or any tumor extending in front of the outer rim of the retina (ora serrata)
- Group V (very unfavorable for maintenance of sight): large tumors involving more than half of the retina, or vitreous seeding, in which small pieces of tumor are broken off and floating around the inside of the eye

When choosing a treatment plan, the first important criteria to ascertain is whether the cancer is localized within the eye (intralocular) or has spread to other parts of the body (extralocular). An intraocular retinoblastoma may only involve the retina or could involve other parts of the eye. An extraocular retinoblastoma could involve only the tissues around the eye or could result from the spread of cancer to the brain or other parts of the body.

It is also important to establish whether the cancer is unilateral (one eye) or bilateral (both eyes), multifocal or unifocal. In order for the tumors to be considered multifocal, they must have arisen independently and not as the result of the spread of cancer cells. It is also important to check for trilateral retinoblastoma.

Treatments

The treatment chosen depends on the size and number of tumors, whether the cancer is unilateral or bilateral, and whether the cancer has spread to other parts of the body. The goal of treatment is to cure the cancer and prevent as much loss of vision as possible. Since the late 1990s, doctors treating patients with retinoblastoma have tended to avoid enucleation and external beam **radiation therapy** whenever possible, in favor of **chemotherapy** to reduce the tumor in addition to focal therapies. Improved methods of chemo-reduction have led to increasing success in saving patients' eyes, often with some visual function.

TREATMENT OF INTRAOCULAR TUMORS. Surgical removal of the affected eye (enucleation) is used when the tumor(s) are so large and extensive that preservation of sight is not possible. This surgery is performed under general anesthetic and usually takes less than an hour. Most children who have undergone this surgery can leave the hospital on the same day. A temporary ball is placed in the eye socket after the surgery. Approximately three weeks after the operation, a plastic artificial eye (prosthesis) that looks like the normal eye is inserted into the eye socket.

Radiation therapy is often used for treatment of large tumors when preservation of sight is possible. External beam radiation therapy involves focusing a beam of radiation on the eye. If the tumor has not spread extensively, the radiation beam can be focused on the cancerous retinal cells. If the cancer is extensive, radiation treatment of the entire eye may be necessary. External beam radiation is performed on an outpatient basis and usually occurs over a period of three to four weeks. Some children may need sedatives prior to the treatment. This type of therapy can result in a temporary loss of a patch of hair on the back of the head and a small area of "sun-burned" skin. Long-term side effects of radiation treatment can include **cataracts**, vision problems, bleeding from the retina, and decreased growth of the bones on the side of the head. People with an inherited form of retinoblastoma have an increased risk of developing other cancers as a result of this therapy. Some consideration should therefore be given to alternative treatment therapies for those with an inherited form of retinoblastoma.

Photocoagulation therapy is often used in conjunction with radiation therapy but may be used alone to treat small tumors that are located on the back of the eye. Photocoagulation involves using a laser to destroy the cancer cells. This type of treatment is done under local or **general anesthesia** and is usually not associated with post-procedural **pain**.

Thermotherapy is also often used in conjunction with radiation therapy or drug therapy (chemotherapy). Thermotherapy involves the use of heat to help shrink tumor cells. The heat is either used on the whole eye or localized to the tumor area. It is done under local or general anesthesia and is usually not painful.

Cryotherapy is a treatment often used in conjunction with radiation therapy but can also be used alone on small tumors located on the front part of the retina. Cryotherapy involves the use of intense cold to destroy cancer cells and can result in harmless, temporary swelling of the external eye and eyelids that can last for up to five days. Eye drops or ointment are sometimes provided to reduce the swelling.

KEY TERMS

Amniocentesis—Prenatal testing performed at 16 to 20 weeks of pregnancy that involves inserting a needle through the abdomen of a pregnant mother and obtaining a small sample of fluid from the amniotic sack, which contains the fetus. Often is used to obtain a sample of the fetus' cells for biochemical or DNA testing.

Benign tumor—An abnormal proliferation of cells that does not spread to other parts of the body.

Bilateral—Affecting both eyes.

Brachytherapy—Cancer treatment that involves the application of radioactive material to the site of the tumor.

Cryotherapy—Cancer treatment in which the tumor is destroyed by exposure to intense cold.

Chromosome—A microscopic structure found within each cell of the body, made of a complex of proteins and DNA.

Chorionic villus sampling (CVS)—Prenatal testing performed at 10 to 12 weeks of pregnancy, which

involves inserting a catheter through the vagina of a pregnant mother or inserting a needle through the abdomen of the mother and obtaining a sample of placenta. Often is used to obtain a sample of the fetus' cells for biochemical or DNA testing.

DNA (deoxyribonucleic acid)—The hereditary material that makes up genes; influences the development and functioning of the body.

DNA testing—Testing for a change or changes in a gene or genes.

Enucleation—Surgical removal of the eye.

Equator—Imaginary line encircling the eyeball and dividing the eye into a front and back half.

Extraocular retinoblastoma—Cancer that has spread from the eye to other parts of the body.

Gene—A building block of inheritance, made up of a compound called DNA (deoxyribonucleic acid) and containing the instructions for the production of a particular protein. Each gene is found in a specific location on a chromosome.

Brachytherapy involves the application of radioactive material to the outer surface of the eye at the base of the tumor. It is generally used for tumors of medium size. A patient undergoing this type of procedure is usually hospitalized for three to seven days. During that time, he or she undergoes one surgery to attach the radioactive material and one surgery to remove it. Eye drops are often administered for three to four weeks following the operation to prevent inflammation and infection. The long-term side effects of this treatment can include cataracts and damage to the retina, which can lead to impaired vision.

Intravenous treatment with one or more drugs (chemotherapy) is often used for treatment of both large and small tumors. Chemotherapy is sometimes used to shrink tumors prior to other treatments such as radiation therapy or brachytherapy. Occasionally, it is also used alone to treat very small tumors.

TREATMENT OF INTRAOCULAR AND UNILATERAL RETINOBLASTOMA. Often, by the time that unilateral retinoblastoma is diagnosed, the tumor is so large that useful vision cannot be preserved. In these cases removal of the eye (enucleation) is the treatment of choice. Other therapies are unnecessary if enucleation is used to treat intraocular unilateral retinoblastoma. If the tumor is small enough, other therapies such as

external beam radiation therapy, photocoagulation, cryotherapy, thermotherapy, chemotherapy, and brachytherapy may be considered.

TREATMENT OF INTRAOCULAR AND BILATERAL RETINOBLASTOMA. If vision can be preserved in both eyes, radiation therapy of both eyes may be recommended. Smaller, more localized tumors can sometimes be treated by local therapies such as cryotherapy, photocoagulation therapy, thermotherapy or brachytherapy. Some centers may use chemotherapy in place of radiation therapy when the tumors are too large to be treated by local therapies or are found over the optic nerve of the eye. Many centers are moving away from radiation treatment and toward chemotherapy because it is less likely to induce future tumors. Enucleation is performed on the more severely affected eye if sight cannot be preserved in both.

EXTRAOCULAR RETINOBLASTOMA. There is no proven effective therapy for the treatment of extraocular retinoblastomas. Commonly, radiation treatment of the eyes and chemotherapy is provided.

Alternative treatment

There are no alternative or complementary therapies specific to the treatment of retinoblastoma. Since

Intraocular retinoblastoma—Cancer that is limited to the eye and has not spread to other parts of the body.

Malignant tumor—An abnormal proliferation of cells that can spread to other sites.

Multifocal—More than one tumor present.

Ophthalmologist—Physician specializing in the diseases of the eye.

Optic nerve—The part of the eye which contains nerve fibers that transmit signals from the eye to the brain.

Oncologist—A physician specializing in the diagnosis and treatment of cancer.

Photocoagulation—Cancer treatment in which the tumor is destroyed by an intense beam of laser light.

Prenatal testing—Testing for a disease such as a genetic condition in an unborn baby.

Protein—A substance produced by a gene that is involved in creating the traits of the human body, such as hair and eye color, or is involved in controlling

the basic functions of the human body, such as control of the cell cycle.

Retina—The light-sensitive layer of the eye that receives images and sends them to the brain.

Scotoma—An area of lost or depressed vision within the visual field surrounded by an area of normal vision. Survivors of retinoblastoma frequently develop scotomas.

Somatic cells—All the cells of the body with the exception of the egg and sperm cells.

Tumor—A growth of tissue resulting from the uncontrolled proliferation of cells.

Tumor-suppressor gene—Gene involved in controlling normal cell growth and preventing cancer.

Unifocal—Only one tumor present in one eye.

Unilateral—Affecting only one eye.

Vitreous—The transparent gel that fills the back part of the eye.

Vitreous seeding—When small pieces of tumor have broken off and are floating around the vitreous.

most people diagnosed with retinoblastoma are small children, most drug-based alternative therapies designed to treat general cancer would not be recommended. Many specialists would, however, **stress** the importance of establishing a well-balanced diet, including certain fruits, vegetables, and vitamin supplements, to ensure that the body is strengthened in its fight against cancer. Some advocate the use of visualization strategies, in which patients would visualize the immune cells of their body attacking and destroying the cancer cells.

Prognosis

Individuals with intraocular retinoblastoma who do not have trilateral retinoblastoma usually have a good survival rate with a 90% chance of disease-free survival for five years. Those with extraocular retinoblastoma have less than a 10% chance of disease-free survival for the same amount of time. Trilateral retinoblastoma generally has a very poor prognosis. Patients with trilateral retinoblastoma who receive treatment have an average survival rate of approximately eight months, while those who remain untreated have an average survival rate of approximately one month. Patients with trilateral retinoblastoma who are asymptomatic at the time of diagnosis may have a better prognosis than those who experience symptoms.

Patients with an inherited form of unilateral retinoblastoma have a 70% chance of developing retinoblastoma in the other eye. Retinoblastoma reoccurs in the other eye in approximately 5% of people with a non-inherited form of retinoblastoma, so it is advisable for even these patients to be closely monitored. People with an inherited form of retinoblastoma who have not undergone radiation treatment have approximately a 26% chance of developing cancer in another part of the body within 50 years of the initial diagnosis. Those with an inherited form who have undergone radiation treatment have a 58% chance of developing a secondary cancer by 50 years after the initial diagnosis. Most of the secondary cancers are skin cancers, bone tumors (osteosarcomas), and soft-tissue **sarcomas**. Soft-tissue sarcomas are malignant tumors of the muscle, nerves, joints, blood vessels, deep skin tissues, or fat. The prognosis for retinoblastoma patients who develop secondary cancers, however, is very poor.

Survivors of retinoblastoma are likely to have visual field defects after their cancer treatment is completed, most commonly scotomas, which are areas of lost or depressed vision within an area of normal vision. The size and type of these visual defects are determined by the size and type of the original tumor and the form of therapy used to treat it.

Prevention

Although retinoblastoma cannot be prevented, appropriate screening and surveillance should be applied to all at-risk individuals to ensure that the tumor(s) are diagnosed at an early stage. The earlier the diagnosis, the more likely that an eye can be salvaged and vision maintained.

Screening of people diagnosed with retinoblastoma

Children who have been diagnosed with retinoblastoma should receive periodic dilated retinal examinations until the age of five. Young children will need to undergo these evaluations under anesthetic. After five years of age, periodic eye examinations are recommended. It may be advisable for patients with bilateral retinoblastoma or an inherited form of retinoblastoma to undergo periodic screening for the brain tumors found in trilateral retinoblastoma. There are no specific screening protocols designed to detect non-ocular tumors. All lumps and complaints of bone pain, however, should be thoroughly evaluated.

Screening of relatives

When a child is diagnosed with retinoblastoma, it is recommended that parents and siblings receive a dilated retinal examination by an ophthalmologist who is experienced in the diagnosis and treatment of the disease. It is also recommended that siblings continue to undergo periodic retinal examinations under anesthetic until they are three years of age. From three to seven years of age, periodic eye examinations are recommended. The retinal examinations can be avoided if DNA testing indicates that the patient has a non-inherited form of retinoblastoma or if the sibling has not inherited the RB1 gene change/deletion. Any relatives who are found through DNA testing to have inherited an RB1 gene change/deletion should undergo the same surveillance procedures as siblings.

The children of someone diagnosed with retinoblastoma should also undergo periodic retinal examinations under anesthetic. Retinal surveillance should be performed unless DNA testing proves that their child does not possess the RB1 gene change/deletion. If desired, prenatal detection of tumors using ultrasound may also be performed. During the ultrasound procedure, a hand-held instrument is placed on the maternal abdomen or inserted vaginally. The ultrasound produces sound waves that are reflected back from the body structures of the fetus, producing a picture that can be seen on a video screen. If a tumor is detected through this evaluation, the affected baby may be delivered a couple of weeks earlier. This can allow for earlier intervention and treatment.

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ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, 415 561-8500, <http://www.aoa.org>.

Canadian Retinoblastoma Society, 59 Bannockburn Avenue, Toronto, Canada, Ontario, M5M 2M9, (306) 642-4993, 306 642-3809, info@rbsociety.ca, <http://www.rbsociety.ca>.

Institute for Families with Blind Children, 4650 Sunset Blvd, Mail Stop 111, Los Angeles, CA, 90027, (323) 361-4649, (323) 665-7869, info@instituteforfamilies.org, http://www.instituteforfamilies.org.

Retinoblastoma International, 18030 Brookhurst Street, P.O. Box 408, Fountain Valley, CA, 92708, info@retinoblastoma.net, http://www.retinoblastoma.net.

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Retinol deficiency see **Vitamin A deficiency**

Retinopathies

Definition

Retinopathy is a noninflammatory disease of the retina. There are many causes and types of retinopathy.

Demographics

Diabetic retinopathy is the leading cause of blindness in people ages 20 to 74. Diabetic retinopathy occurs in 90% of persons with type 1 diabetes (insulin-independent, or insulin requiring) and 65% of persons with type II diabetes (non-insulin-dependent, or not requiring insulin) by about ten years after the onset of diabetes. In the United States, new cases of blindness most often are caused by diabetic retinopathy. Among these new cases of blindness, 12% are people between the ages of 20 to 44 years, and 19% are people between the ages of 45 to 64 years.



A slit lamp view of a human eye with diabetic retinopathy.
(Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

The retina is the thin membrane that lines the back of the eye and contains light-sensitive cells (photoreceptors). Light enters the eye and is focused onto the retina. The photoreceptors send a message to the brain via the optic nerve. The brain then “interprets” the electrical message sent to it, resulting in vision. The macula is a specific area of the retina responsible for central vision. The fovea is about 1.5 mm in size and located in the macula. The fovea is responsible for sharp vision. When looking at something, the fovea should be directed at the object.

Retinopathy, or damage to the retina, has various causes. A hardening or thickening of the retinal arteries is called arteriosclerotic retinopathy. High blood pressure in the arteries of the body can damage the retinal arteries and is called hypertensive retinopathy. The spreading of a **syphilis** infection to the retinal blood vessels causes syphilitic retinopathy, and diabetes damages the retinal vessels resulting in a condition called diabetic retinopathy. Sickle cell anemia also affects the blood vessels in the eyes. Exposure to the sun (or looking at the sun during an eclipse) can cause damage (solar retinopathy), as well as certain drugs (for example, chloroquine, thioridazine, and large doses of tamoxifen). The arteries and veins can become blocked, resulting in a retinal artery or vein occlusion. These are just some of the causes of the various retinopathies.

Retinopathies are divided into two broad categories, simple (or nonproliferative) retinopathies and proliferative retinopathies. The simple retinopathies include the defects identified by bulging of the vessel walls, bleeding into the eye, small clumps of dead retinal cells called cotton wool exudates, and closed vessels. This form of retinopathy is considered mild. The proliferative, or severe, forms of retinopathies include the defects identified by newly grown blood vessels, scar tissue formed within the eye, closed-off blood vessels that are badly damaged, and by the retina breaking away from the mesh of blood vessels that nourish it (**retinal detachment**).

While each disease has its own specific effect on the retina, many of the retinopathies follow the same general scenario. Blood flow to the retina is disrupted, either by blockage or breakdown of the various vessels (not all retinopathies necessarily affect the blood vessels). This can lead to bleeding (hemorrhage) and fluids, cells, and proteins leaking into the area (exudates). There can be a lack of oxygen to surrounding tissues (hypoxia) or decreased blood flow (**ischemia**). Chemicals produced by the body then cause new blood vessels to grow (neovascularization), however, these new vessels generally leak and cause more problems.

Neovascularization even can grow on the colored part of the eye (iris). The retina can swell and vision will be affected.

Causes and symptoms

There are many causes of retinopathy. Some of the more common causes are discussed here.

Diabetic retinopathy

Diabetes is a complex disorder characterized by an inability of the body to properly regulate the levels of sugar and insulin (a hormone made by the pancreas) in the blood. As diabetes progresses, the blood vessels that feed the retina become damaged in different ways. The damaged vessels can have bulges in their walls (aneurysms); they can leak blood into the surrounding jelly-like material (vitreous) that fills the inside of the eyeball; they can become completely closed; or new vessels can begin to grow where there would not normally be blood vessels. Although these new blood vessels are growing in the eye, they cannot nourish the retina and they bleed easily, releasing blood into the inner region of the eyeball, which can cause dark spots and cloudy vision.

Diabetic retinopathy begins prior to any outward signs of disease being noticed. Once symptoms are noticed, they include poorer than normal vision, fluctuating or distorted vision, cloudy vision, dark spots, episodes of temporary blindness, or permanent blindness.

Hypertensive retinopathy

High blood pressure can affect the vessels in the eyes. Some blood vessels can narrow. The blood vessels can thicken and harden (arteriosclerosis). There will be flame-shaped hemorrhages and macular swelling (**edema**). This edema may cause distorted or decreased vision.

Sickle cell retinopathy

Sickle cell anemia occurs mostly in individuals of African descent and is a hereditary disease that affects the red blood cells. The sickle-shaped blood cell reduces blood flow. People do not have visual symptoms early in the disease. However, patients need to be followed closely in case neovascularization occurs.

Retinal vein and artery occlusion

Retinal vein occlusion generally occurs in the elderly. There is usually a history of other systemic disease, such as diabetes or high blood pressure. The central retinal vein (CRV), or the retinal veins branching off of the CRV, can become compressed, stopping

KEY TERMS

Exudate—Cells, protein, fluid, or other material that passes through blood vessel walls and accumulates in the surrounding tissue.

Neovascularization—New blood vessel formation; usually leaky vessels.

Nonproliferative retinopathy—Retinopathy without the growth of new blood vessels.

Proliferative retinopathy—Retinopathy with the growth of new blood vessels (neovascularization).

the drainage of blood from the retina. This may occur if the central retinal artery hardens.

Symptoms of retinal vein occlusion include a sudden, painless loss of vision or field of vision in one eye. There may be a sudden onset of floating spots (floaters) or flashing lights. Vision may be unchanged or decrease dramatically.

Retinal artery occlusion generally is the result of an **embolism** that dislodges from somewhere else in the body and travels to the eye. Transient loss of vision may precede an occlusion. Symptoms of a central retinal artery or branch occlusion include a sudden, painless loss of vision or decrease in visual field. Ten percent of the cases of a retinal artery occlusion occur because of giant cell arteritis (a chronic **vascular disease**).

Solar retinopathy

Looking directly at the sun or watching an eclipse can cause damage. There may be a loss of the central visual field or decreased vision. The symptoms can occur hours to days after the incident.

Drug-related retinopathies

Certain medications can affect different areas of the retina. Doses of 20–40 mg a day of tamoxifen usually do not cause a problem, but much higher doses may cause irreversible damage.

Patients taking chloroquine for lupus, **rheumatoid arthritis**, or other disorders may notice a decrease in vision. If so, discontinuing medication will stop, but not reverse, any damage. Patients should never discontinue medication without the advice of their physician.

Patients taking thioridazine may notice a decrease in vision or color vision.

These drug-related retinopathies generally only affect patients taking large doses. However, patients

need to be aware if any medication they are taking will affect the eyes. Patients should inform their doctors of any visual effects.

Diagnosis

The damaged retinal blood vessels and other retinal changes are visible to an eye doctor when an examination of the retina (fundus exam) is done. This can be done using a hand-held instrument called an ophthalmoscope or another instrument called a binocular indirect ophthalmoscope. This allows the doctor to see the back of the eye. Certain retinopathies have classic signs (for example, vascular “sea fans” in sickle cell, dot and blot hemorrhages in diabetes, flame-shaped hemorrhages in high blood pressure). Patients then may be referred for other tests to confirm the underlying cause of the retinopathy. These tests include blood tests and measurement of blood pressure.

Fluorescein angiography, where a dye is injected into the patient and the back of the eyes are viewed and photographed, helps to locate leaky vessels. Sometimes patients may become nauseated from the dye.

A newer diagnostic method called digital retinal photography can be used to screen those at high risk for retinopathies, in particular, diabetics. Some researchers say the technique could lead to more cost-effective screening for people with diabetic retinopathy.

Treatment

Retinal specialists are ophthalmologists who specialize in retinal disorders. Retinopathy is a disorder of the retina that can result from different underlying systemic causes, so general physicians should be consulted as well. For drug-related retinopathies, the treatment generally is discontinuation of the drug (only under the care of a physician).

Surgery with lasers can help to prevent blindness or lessen any losses in vision. The high-energy light from a laser is aimed at the weakened blood vessels in the eye, destroying them. **Scars** will remain where the laser treatment was performed. For that reason, laser treatment cannot be performed everywhere. For example, laser photocoagulation at the fovea would destroy the area for sharp vision. In panretinal photocoagulation, a larger area of the periphery of the retina is treated in hopes of decreasing neovascularization. Prompt treatment of proliferative retinopathy may reduce the risk of severe vision loss by 50%.

Patients with retinal artery occlusion should be referred to a cardiologist. Patients with retinal vein occlusion need to be referred to a physician, as they may have an underlying systemic disorder, such as high blood pressure.

Prognosis

Nonproliferative retinopathy has a better prognosis than proliferative retinopathy. Prognosis depends on the extent of the retinopathy, the cause, and promptness of treatment.

Prevention

Complete eye examinations done regularly can help detect early signs of retinopathy. Patients on certain medications should have more frequent eye exams. They also should have a baseline eye exam when starting the drug. People with diabetes must take extra care to have thorough, periodic eye exams, especially if early signs of **visual impairment** are noticed. It is recommended that people with diabetes have re-screening eye exams every two years if their blood sugar has remained in control and more frequent exams if visual symptoms appear. Anyone experiencing a sudden loss of vision, decrease in vision or visual field, flashes of light, or floating spots should contact their eye doctor right away.

Proper medical treatment for any of the systemic diseases known to cause retinal damage will help prevent retinopathy. For diabetics, maintaining proper blood sugar and blood pressure levels is important as well; however, some form of retinopathy usually occurs in diabetics, given enough time. A proper diet, particularly for people with diabetes, and stopping **smoking** also help delay retinopathy.

Frequent, thorough eye exams and control of systemic disorders are the best prevention.

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ORGANIZATIONS

- American Academy of Ophthalmology, P.O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8549, patientinfo@aoa.org, <http://www.aoa.org>.

American Diabetes Association, 1701 North Beauregard Street, Alexandria, VA, 22311, (800) 342-2383, askada@diabetes.org, <http://www.diabetes.org>.
 American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, (800) 365-2219, <http://www.aoa.org>.
 Foundation Fighting Blindness, 7168 Columbia Gateway Dr. Suite 100, Columbia, MD, 21046, (800) 683-5555, <http://www.blindness.org>.
 National Eye Institute, 2020 Vision Place, Bethesda, MD, 20892-3655, (301) 496-5248, <http://www.nei.nih.gov/index.asp>.
 Prevent Blindness America, 211 West Wacker Dr., Suite 1700, Chicago, IL, 60606, (800) 331-2020, <http://www.preventblindness.org>.

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Retrocaval ureter see **Congenital ureter anomalies**

Retrograde cystography

Definition

A retrograde cystogram provides x-ray visualization of the bladder with injection of sterile dye.

Purpose

A retrograde cystogram is performed to evaluate the structure of the bladder and identify bladder disorders, such as tumors, or recurrent urinary tract infections. The presence of urine reflux (backward flow) into the ureters may also be visualized with this x-ray study.

Precautions

The doctor should be made aware of any previous history of reactions to shellfish, iodine, or any iodine-containing foods or dyes. Allergic reactions during previous dye studies is not necessarily a contraindication, as dye is not infused into the bloodstream for this study. Other conditions to be considered by the physician prior to proceeding with the test include active **urinary tract infection, pregnancy**, recent bladder surgery, or presence of obstruction that interferes with passage of a urinary catheter.

KEY TERMS

Bladder—A balloon-like organ located in the lower abdomen that stores urine.

Catheter—A thin tube used to inject or withdraw fluids from the body.

Stones—Also known as calculi, stones result from an excessive build-up of mineral crystals in the kidney. Symptoms of stones include intense pain in the lower back or abdomen, urinary tract infection, fever, burning sensation on urination, and/or blood in the urine.

Ureter—Tube that carries urine from the kidney to the bladder.

Urethra—Tube that empties urine from the bladder to outside the body.

Description

After administration of anesthesia, the doctor will insert a thin, tubelike instrument called a catheter through the patient's urethra and into the bladder. The contrast medium is then injected through the catheter into the bladder. X-ray pictures are taken at various stages of filling, from various angles, to visualize the bladder. Additional films are taken after drainage of the dye. The procedure takes approximately one to one and one-half hours and the patient may be asked to wait while films are developed.

Alternately, instead of a contrast dye and x-ray pictures, the test can be done with a radioactive tracer and a different camera. This is known as a "radio-nuclide" retrograde cystogram.

Preparation

The patient will be required to sign a consent form after the risks and benefits of the procedure have been explained. **Laxatives** or **enemas** may be necessary before the procedure, as the bowel must be relatively empty of stool and gas to provide visualization of the urinary tract. Immediately before the procedure, the patient should remove all clothing and jewelry and put on a surgical gown.

Aftercare

Sometimes, pulse, blood pressure, breathing status, and temperature are checked at regular intervals

after the procedure, until they are stable. The patient may have some burning on urination for a few hours after the test, due to the irritation of the urethra from the catheter. The discomfort can be reduced by liberal fluid intake, in order to dilute the urine. The appearance and amount of urine output should be noted, and the doctor should be notified if blood appears in the urine after three urinations. Also, patients should report any signs of urinary infection, including chills, **fever**, rapid pulse, and rapid breathing rate.

Normal results

A normal result would reveal no anatomical or functional abnormalities.

Abnormal results

Abnormal results may indicate:

- stones
- blood clots
- tumors
- reflex (urine passing backward from the bladder into the ureters)

ORGANIZATIONS

American Kidney Fund (AKF), 6110 Executive Boulevard, Suite 1010, Rockville, MD, 20852, (800) 638-8299, <http://www.kidneyfund.org>.

National Kidney Foundation, Inc., 30 East 33rd Street, New York, NY, 10016, (212) 889-2210, (212) 689-9261, (800) 622-9010, <http://www.kidney.org/>.

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This may occur due to poor renal function and inadequate excretion of the contrast medium (dye).

Precautions

The doctor should be made aware of any previous history of reactions to shellfish, iodine, or any iodine-containing foods or dyes. Allergic reactions during previous dye studies is not necessarily a contraindication, as dye is not infused into the bloodstream for this study. Other conditions to be considered by the physician prior to proceeding with the test include **pregnancy** and active **urinary tract infection**.

Description

After administration of anesthesia, the doctor will insert a thin, tubelike instrument (catheter) through the patient's urethra and into the bladder. A catheter is then placed into the affected ureter to instill the contrast medium. X-ray pictures are taken to visualize the ureter. If complete obstruction is found, a ureteral catheter may be left in place and secured to an indwelling urethral catheter to facilitate drainage of urine. The procedure takes approximately one hour.

Preparation

Laxatives or **enemas** may be necessary before the procedure, as the bowel must be relatively empty to provide visualization of the urinary tract. When **general anesthesia** is used for insertion of the ureteral catheter, there should be no eating and drinking after midnight prior to the procedure.

Aftercare

Even if no catheters are left in place after the procedure, the patient may have some burning on urination for a few hours after the procedure due to the irritation of the urethra. The discomfort can be reduced by liberal fluid intake, in order to dilute the urine. The appearance and amount of urine output should be noted for 24 hours after the procedure. If a stone was found, all urine should be strained to allow chemical analysis of any stones passed spontaneously. This will allow the doctor to provide advise on measures to prevent recurrent stone formation. **Antibiotics** are usually given after the procedure to prevent urinary tract infection.

Retrograde ureteropyelography

Definition

A retrograde ureteropyelogram provides x-ray visualization of the bladder, ureters, and the kidney (renal) pelvis by injection of sterile dye into the renal collecting system.

Purpose

A retrograde ureteropyelogram is performed to determine the exact location of a ureteral obstruction when it cannot be visualized on an intravenous pyelogram (a dye is injected and an x ray taken of the kidneys and the tubes that carry urine to the bladder).

Normal results

A normal result would reveal no anatomical or functional abnormalities.

Abnormal results

Abnormal results may indicate:

- congenital abnormalities
- fistulas or false passages
- renal stones
- strictures
- tumors

ORGANIZATIONS

American Kidney Fund (AKF), 6110 Executive Boulevard, Suite 1010, Rockville, MD, 20852, (800) 638-8299, <http://www.kidneyfund.org>.

National Kidney Foundation, Inc., 30 East 33rd Street, New York, NY, 10016, (212) 889-2210, (212) 689-9261, (800) 622-9010, <http://www.kidney.org/>.

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KEY TERMS

Bladder—The balloonlike organ in the lower abdomen that holds urine.

Catheter—Tube used to inject into or withdraw fluids from the bladder.

Renal—Relating to the kidneys, from the Latin word for kidneys, *renes*.

Urethra—Tube that carries the urine from the bladder out of the body.

Visualization—The process of making an internal organ visible. A radiopaque substance is introduced into the body, then an x-ray picture of the desired organ is taken.

inserted into the urethra, and dye is injected into the catheter. A clamp is applied to hold the dye in place while x-ray pictures are taken of the urethral structure. The clamp and catheter are then removed. The procedure takes approximately 15 minutes. However, the patient may be asked to wait while films are developed, which also permits the patient to be observed for any immediate side effects from the dye. The test may be performed in a hospital, doctor's office, outpatient center, or freestanding surgical facility. The time involved for reporting of test results to the doctor may vary from a few minutes to a few days.

Preparation

The patient will be asked to sign a consent form after the risks and benefits of the procedure have been explained. No diet or activity changes are necessary in preparation for the procedure. The patient will be asked to remove all clothing and put on a surgical gown before the test begins.

Normal results

The presence of no anatomical or functional abnormalities is considered a normal result.

Abnormal results

Abnormal findings may indicate:

- congenital abnormalities
- fistulas or false passages
- lacerations
- strictures
- valves, known as “posterior urethral valves”
- tumors

Description

The urethra is first visually examined by the doctor, and the opening is cleansed with an antiseptic solution. A flexible rubber or plastic catheter is then

ORGANIZATIONS

American Kidney Fund (AKF), 6110 Executive Boulevard, Suite 1010, Rockville, MD, 20852, (800) 638-8299, <http://www.kidneyfund.org>.
 National Kidney Foundation, Inc., 30 East 33rd Street, New York, NY, 10016, (212) 889-2210, (212) 689-9261, (800) 622-9010, <http://www.kidney.org/>.

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Retrograde urography see **Retrograde urethrography**

Retropharyngeal abscess see **Abscess**

Retropubic suspension

Definition

Retropubic suspension refers to the surgical procedures used to correct incontinence by supporting and stabilizing the bladder and urethra. The Burch procedure, also known as retropubic urethropexy procedure or Burch colosuspension, and **Marshall-Marchetti-Krantz procedure** (MMK) are the two primary surgeries for treating **stress** incontinence. The major difference between these procedures is the method for supporting the bladder. The Burch procedure uses sutures to attach the urethra and bladder to muscle tissue in the pelvic area. MMK uses sutures to attach these organs to the pelvic cartilage. Laparoscopic retropubic surgery can be performed with a video laparoscope through small incisions in the belly button and above the pubic hairline.

Purpose

The urinary system expels a quart and a half of urine per day. The amount of urine produced depends upon diet and medications taken, as well as **exercise** and loss of water due to sweating. The ureters, two tubes connecting the kidneys and the bladder, pass urine almost continually and when the bladder is full the brain sends a signal to the bladder to relax and let urine pass from the bladder to the urethra. People who are continent control the release of urine from the urethra via the sphincter muscles. These two sets of muscles act like rubber bands to keep the bladder closed until a conscious decision is made to urinate. The intrinsic sphincter or urethral sphincter muscles keep the bladder closed and the extrinsic sphincter muscles surround the urethra and prevent leakage.

Incontinence is common when either the urethra lacks tautness and stability (genuine stress urine incontinence, SUI) and/or the sphincter muscles are unable to keep the bladder closed (intrinsic sphincter deficiency, ISD).

Incontinence occurs in many forms with four primary types related to anatomic, neurological, and dietary causes; or disease and injury.

Stress incontinence

The most frequent form of incontinence is stress incontinence. This relates to leakage of the urethra with activity that puts stress on the abdominal muscles. The primary sign of stress incontinence is this leakage at sneezing, coughing, exercise, or other straining activities, which indicates a lack of support for the urethra due to weakened muscles, fascia, or ligaments. Pressure from the abdomen with movement, like exercising, uncompensated by tautness or stability in the urethra, causes the urethra to be displaced or mobile leading to leakage. Essentially, this hypermobility of the urethra is an indication that it is moving down or herniating through weakened pelvic structures.

To diagnose incontinence and determine treatment, three grades of severity for stress incontinence are used.

- Type I: Moderate movement of the urethra, with no hernia or cystocele.
- Type II: Severe or hypermobility in the urethra of more than 0.8 in (2 cm), with or without decent of the urethra into pelvic structures.
- Type III: Hypermobility of the urethra where the primary source of incontinence is the inability of the sphincter muscles to keep the bladder closed. This is due to weakness or deficiency in the intrinsic sphincter muscles.

Urge incontinence

Urge incontinence relates to the frequent need to urinate and may involve going to the bathroom every two hours. Accidents are common when not reaching a bathroom in time. Urge incontinence is not due to general changes in the urethra or supporting muscles. It is often linked to other disorders that produce **muscle spasms** in the bladder, such as infections. Urge incontinence can also be due to underlying illnesses like **stroke**, **spinal cord injury**, **multiple sclerosis** and **Alzheimer's disease**, which cause detrusor hyperflexia—the contracting of the bladder muscle responsible for sending urine from the bladder to the

KEY TERMS

Genuine urinary stress incontinence (USI)—Stress incontinence due to hypermobility of the urethra.

Intrinsic sphincter deficiency (ISD)—A factor in severe stress incontinence due to the inadequacy of the sphincter muscles to keep the bladder closed.

Retropubic urethropexy—A generic term for the Burch procedure and its variants that treat mild stress incontinence by stabilizing the urethra with retropubic surgery.

Stress incontinence—Leakage of urine upon movements that put pressure on the abdominal muscles such as coughing, sneezing, laughing, or exercise. One of four types of incontinence.

Urethra hypermobility—Main factor in stress urinary incontinence, with severity based upon how far the urethra has descended into the pelvic floor through herniation or cystocele.

urethra. Urge incontinence is very common in the elderly, especially those in long term care facilities.

Mixed incontinence

Mixed incontinence is a combination of stress incontinence and urge incontinence, especially in older women. Since each form of incontinence pertains to different functions or anatomy, it is very important to distinguish which part of the incontinence is to be treated by surgery.

Overflow incontinence

Overflow incontinence results in leakage from a bladder that never completely empties due to weakened bladder muscles. Overflow incontinence is involuntary and not accompanied by the urge to urinate. Many causes exist for overflow incontinence, including weak bladder muscles due to diabetes, nerve damage, or a blocked urethra. Men are more frequently affected than women.

Demographics

Over 15 million Americans have **urinary incontinence** and women comprise 85% of all cases. It affects 25% of women of reproductive age and 50% of women past **menopause**. Due to the female anatomy, women have twice the risk for stress incontinence compared to men. In addition, **childbirth** places pressure and burden on the pelvic muscles

that often weaken with age, thereby weakening urethra stability. Women are more prone to surgeries for urological changes than men and severe urinary incontinence is often associated with these surgeries as well as hysterectomies. The majority of women with incontinence have stress incontinence or mixed incontinence. Male incontinence occurs primarily in response to blockage in the prostate or after prostate surgery. It is usually treated with implants and/or an artificial sphincter insert.

Description

There are a variety of retropubic suspension surgeries available to treat stress incontinence. The variations differ by the types of structures used to support the urethra and bladder. In all procedures, parts of the pelvic anatomy (pubic bone, ligaments) serve as an anchor or wall upon which the urethra is tacked for stability. The surgery is called a suspension surgery because it stabilizes the urethra from tilting by suspending it against a part of the pelvic anatomy. The Burch procedure is often performed when other surgery is needed such as repair of the urethra for cystoceles and urethral reconstruction. However, this procedure is the most difficult of the anti-incontinent surgeries and is more common in mild forms of stress incontinence where intrinsic sphincter deficiency is not present.

The Burch procedure can be done through open abdominal surgery, which requires a long incision at the bikini line, or surgery performed through the vagina. The patient, in stirrups, receives **general anesthesia**. Within the retropubic area, the anterior vaginal wall is separated from the bladder manually. The bladder neck is identified and old **adhesions** or fatty tissues are removed. The neck of the bladder is sutured to pubic ligaments where it will form adhesions and thereby gain stability. The surgeon examines for bladder injury and the surgery is completed. Urethral position is tested by placing a cotton-tipped swab in the urethra and measuring the angle. With abdominal surgery or vaginal surgery a catheter may be put in place by the surgeon for postoperative voiding and to decrease the risks of infection. A suction drain may be placed in the retropubic space for bleeding. The drain is removed one to three days after surgery.

Recently, laparoscopic surgery has been used to perform retropubic suspensions. Laparoscopic surgery requires only three or four 0.25-inch (0.6-cm) incisions in the belly button, pubic hairline, or groin area and uses small instruments without opening the abdominal cavity. A shorter healing time is seen with

this procedure. the hospital stay is usually not more than 24 hours and recovery to normal activities takes about 7 to 14 days. However, the Burch procedure performed using laparoscopic techniques requires great skill on the part of the surgeon and research indicates that the results may not be as long lasting as those developed with abdominal or vaginal surgery.

Diagnosis/Preparation

A patient with incontinence may have multiple factors that induce transient or chronic incontinence. It is crucial that the physician obtain a complete history, physical, clinical, neurological and medication evaluation of the patient, as well as a radiographic assessment before continuing urological tests aimed at a surgical solution. The specific indications for the Burch colosuspension procedure or its variants is the correction of stress urinary incontinence. This can be a patient who also requires abdominal surgery that cannot be performed vaginally, like **hysterectomy** or sigmoid surgery, as well as patients who have SUI without ISD.

A urodynamic study with a point pressure leak test will allow a diagnosis to be made that can distinguish the patient who has a hypermobile urethra from the patient who also has ISD. The point pressure leak test, also known as the Valsalva leak test, measures the amount of abdominal pressure required to induce leakage. The patient is asked to **cough** or strain in order to encourage leakage. The point at which the patient leaks helps determine if stress incontinence with ISD contribution is present. Obese patients and patients that engage in high impact exercise regimens are not considered good candidates for retropubic suspension.

Aftercare

Patients with open retropubic procedures are given **pain** medication postoperatively that is tapered down over the next two days. A suprapubic catheter stays in place for approximately five days with voiding difficulties encountered initially in many patients. Patients with laparoscopic suspensions are reported to have less blood loss during surgery, less postoperative narcotic requirements, and shorter hospital stays. Patients are expected to refrain from strenuous activity for three months and to have a follow-up visit within three weeks after surgery.

Risks

As with any major abdominal or pelvic surgical procedures, complications that may occur after a

retropubic suspension include bleeding; injury to the bladder, urethra, and ureters; wound infection; and **blood clots**. Specific to the Burch procedure are complications that involve urethral obstruction because of urethral kinking due to elevation of the vagina or bladder base. Postoperative voiding difficulties are common and depend upon the suture tension of the urethral axis. Corrective surgery and the release of the urethra to a more anatomic position resolves voiding issues with a very high rate of success. Vaginal prolapse is also a risk of this procedure.

Normal results

The patient can expect more than 80–90% cure or great improvement in their incontinence. There is a large body of literature documenting the success of the Burch procedure. Published research shows a cure rate ranging from 63%–93%, according to the actual version of colosuspension used. Laparoscopic surgery has not produced the long term results that open surgery has and there is the possibility that the fibrosis (adhesion) necessary for a successful outcome does not occur as easily with the laparoscopic procedure. Patients not carefully screened out for ISD will not have a high level of success with the Burch procedure since the source of the incontinence will not have been treated. Sling procedures are recommended for patients with ISD instead of colosuspension surgery.

Morbidity and mortality rates

The Burch procedure may aggravate vaginal wall weakness or vaginal prolapse. This incident varies between 3% and 17%. Research on the Marshall-Marchetti-Krantz procedure pertaining to 2,712 patients found a complication rate of 21%, with wound complications and infections making up the majority, 5.5% and 3.9% respectively. Direct wound injury occurred in 1.6% and obstructions in 0.3% overall.

Alternatives

General or simple severe stress incontinence related primarily to weakening of the urethral support can be remedied with changes in diet, weight loss, and certain behavioral and rehabilitative measures. These include:

- Regular, daily exercising of the pelvic muscles called Kegel exercises, requiring 30–200 contractions a day for eight weeks.

- Biofeedback to gain awareness and control of pelvic muscles.
- Vaginal weight training in which small weights are inserted in the vagina to tighten vaginal muscles.
- Mild electrical stimulation to increase contractions in pelvic muscles.
- Bladder retraining in which the patient is taught how to resist the urge to urinate and expand the intervals between urinations.

There are also medications that can facilitate continence for those experiencing stress or urge incontinence. These include some kinds of antidepressants, although the mechanism of action is not quite understood, as well as antispasmodic medication and estrogen therapy. Finally, should behavioral, rehabilitative, and surgical procedures fail, there remain alternatives through the use of vaginal cones and urethral plugs that can be inserted and removed by the patient.

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ORGANIZATIONS

- American Foundation for Urologic Disease, 1000 Corporate Boulevard, Suite 410, Linthicum, MD, 21090, (410) 689-3990, (800) 828-7866, <http://www.afud.org>.
- National Kidney and Urologic Diseases Information Clearinghouse, 3 Information Way, Bethesda, MD, 20892-3580, (301) 654-4415, (800) 891-5390, <http://www.niddk.nih.gov>.
- Simon Foundation for Continence, P.O. Box 835, Wilmette, IL, 60091, (847) 864-3913, (800) 237-4666, <http://www.simonfoundation.org>.

Nancy McKenzie, PhD
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Reye's syndrome

Definition

Reye's syndrome or Reye syndrome (RS) is a very rare, poorly understood condition that is associated with the use of **aspirin** in children and teens following a viral infection. It primarily affects the blood, liver, and brain and is characterized by the rapid development of life-threatening neurological symptoms.

Demographics

After it was first described as a distinct condition in 1963 by the Australian pathologist R. Douglas Reye, the incidence of Reye's syndrome in the United States peaked with 555 reported cases in 1980. Thereafter the number of cases declined dramatically. There are now less than two reported cases per year in the United States. This decline was due to the association of RS with the use of aspirin to treat childhood **fever** and a corresponding drop in such use. However the true incidence of RS may be somewhat higher because the condition can sometimes be very mild and go undetected. Furthermore because of its rarity, RS may be misdiagnosed as **encephalitis**, **meningitis**, diabetes, **drug overdose**, **poisoning**, **sudden infant death syndrome**, or psychiatric illness.

Although RS can affect people of any age at any time, it primarily affects children between the ages of 4 and 14 and is most common during flu season—January, February, and March. Flu or **chickenpox** epidemics are often followed by an increase in RS. Nevertheless RS is extremely rare in adults and in the absence of aspirin use.

Description

RS develops following a viral illness such as a cold, **influenza B**, or chickenpox. Because of this it is referred to as a two-phase illness. It usually develops very rapidly and without warning, most often during recovery from the viral illness, but also within three to five days of onset of a viral infection. RA can affect most of the body's organ systems. Blood sugar levels typically drop as the ammonia levels and acidity of the blood rise. The liver may simultaneously swell, with massive accumulations of fat in both the liver and other organs. Fluid accumulation and swelling (**edema**) in the brain increases intracranial pressure. This squeezes the blood vessels, preventing blood from reaching the brain. Without treatment RA can lead to rapid brain damage and **death**.

Risk factors

The major risk factor for RA is the use of aspirin to treat fever-inducing viral illnesses—such as flu, chickenpox or an upper respiratory infection—in children under age 19 who have an underlying fatty acid oxidation disorder.

Causes and symptoms

Although the cause of Reye's syndrome is still unknown, it appears to be triggered by the use of

KEY TERMS

Aspirin—A derivative of salicylic acid used to relieve pain and fever.

Edema—Swelling; the abnormal accumulation of fluid in the interstitial spaces of tissues.

Salicylic acid; salicylate; acetylsalicylate—Aspirin; medications used as topical disinfectants and orally to relieve pain and fever.

aspirin to treat a viral illness or infection in children and teenagers who have an underlying fatty acid oxidation disorder. These are a group of inherited metabolic disorders in which the body is unable to breakdown fatty acids due to a missing or abnormal enzyme. It has been suggested that RS is an underlying metabolic condition that is unmasked by viral illness. RS has also been linked to exposure to certain toxins, such as insecticides, herbicides, and paint thinner. A “Reye’s-like” illness has been known to occur in children with certain inherited metabolic or other toxic disorders.

Symptoms of Reye’s syndrome usually develop during recovery from a viral illness. The affected child suddenly worsens and develops persistent or continuous **vomiting**. There is usually no fever. However in infants **diarrhea** is more common than **vomiting** and respiratory symptoms—such as hyperventilation or apneic episodes (breathing cessations)—are common. In addition to vomiting, stage I Reye’s syndrome includes early signs of brain dysfunction such as listlessness, lethargy, and drowsiness. Symptoms of stage II Reye’s syndrome include personality changes, such as irritability and aggressiveness, followed by disorientation—confusion, irrational behavior, combativeness, **delirium**, seizures, and **coma**.

Diagnosis

Examination

Reye’s syndrome may be suspected if a child begins vomiting, followed by neurological symptoms, during recovery from or three to six days after onset of a viral illness.

Tests

Tests for RS include blood and urine analysis, as well as tests for metabolic disorders including those of fatty acid metabolism. Blood tests may indicate:

- elevated levels of certain liver enzymes in the absence of jaundice
- increased levels of ammonia and amino acids
- low blood sugar
- increased clotting time

Procedures

Diagnostic procedures to rule out other causes of the symptoms may include:

- a liver biopsy after clotting abnormalities are corrected with vitamin K or blood products
- a skin biopsy to test for disorders of fatty acid oxidation or other metabolic functions
- lumbar puncture (spinal tap) to rule out meningitis (infection of the lining that surrounds the brain and spinal cord) or encephalitis (inflammation or infection of the brain)
- Computed tomography (CT) or magnetic resonance imaging (MRI) of the head

Treatment

Traditional

Reye’s syndrome is a life-threatening emergency that requires management in a hospital intensive-care unit. There is no cure. Treatment focuses on preventing brain damage:

- intravenous 10% glucose in an electrolyte solution to return blood sugar levels to normal
- plasma transfusions to restore normal clotting time
- monitoring of intracranial pressure and blood pressure
- intravenous mannitol and hyperventilation to lower intracranial pressure
- mechanical ventilation with a breathing machine or respirator if breathing becomes sluggish

Drugs

- vitamin K, plasma, and/or platelets for clotting abnormalities
- corticosteroids to reduce brain swelling
- diuretics to decrease intracranial pressure and increase fluid loss through urination
- barbiturates if intracranial pressure remains elevated
- anti-seizure medications
- small amounts of insulin to increase glucose metabolism

Prognosis

Sometimes RS is mild and resolves on its own. However at the time that the condition was first

recognized mortality was 80%. Earlier diagnosis and better treatment have now increased the survival rate to 80–90%. Almost all surviving children recover fully, although recovery may be slow. If Reye's syndrome progresses rapidly and the child lapses into a coma the prognosis is poorer: there may be permanent neurologic damage, requiring special physical and/or educational equipment and services.

Prevention

The best prevention for Reye's syndrome is to avoid the use of aspirin for treating fever in children and teenagers. Although aspirin is approved for use in children over age two, it should never be taken by children and teens who are recovering from flu-like symptoms or chickenpox. Teenagers who take medications without parental consultation should be warned about aspirin-containing drugs. It is also recommended that women who are **breastfeeding** not take aspirin-containing products, since salicylate can pass into breast milk. Aspirin is an ingredient in many over-the-counter and prescription drugs, including remedies for **headache**, fever, menstrual cramps, muscle **pain**, **nausea**, upset stomach, and arthritis. It is used in oral drugs, suppositories, and topical medications. It may also be an ingredient in alternative or herbal remedies. Therefore it is important to always check the list of ingredients on any medication or remedy. Aspirin can be found in unlikely products, such as Alka-Seltzer. Aspirin may be referred to by any of the following names:

- aspirin
- salicylic acid
- salicylate
- acetylsalicylate
- acetylsalicylic acid

Medications such as **acetaminophen** (Tylenol), ibuprofen (Advil, Motrin), or naproxen **sodium** (Aleve) can be used to reduce a high fever or relieve pain. However administering anti-nausea medicines can mask symptoms of RS.

Children with known fatty acid oxidation disorders should not take aspirin or aspirin-containing products. A screening test can identify a fatty acid oxidation disorder. Some hospitals and medical facilities screen newborns for fatty acid oxidation disorders to identify children who are at greater risk for developing Reye's syndrome.

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ORGANIZATIONS

National Institute of Neurological Disorders and Stroke (NINDS), NIH Neurological Institute, P.O. Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov/index.htm>.

National Reye's Syndrome Foundation, P.O. Box 829, Bryan, OH, 43506, (800) 233-7393, nrsf@reyessyndrome.org, <http://www.reyessyndrome.org/>.

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Rh disease see **Erythroblastosis fetalis**

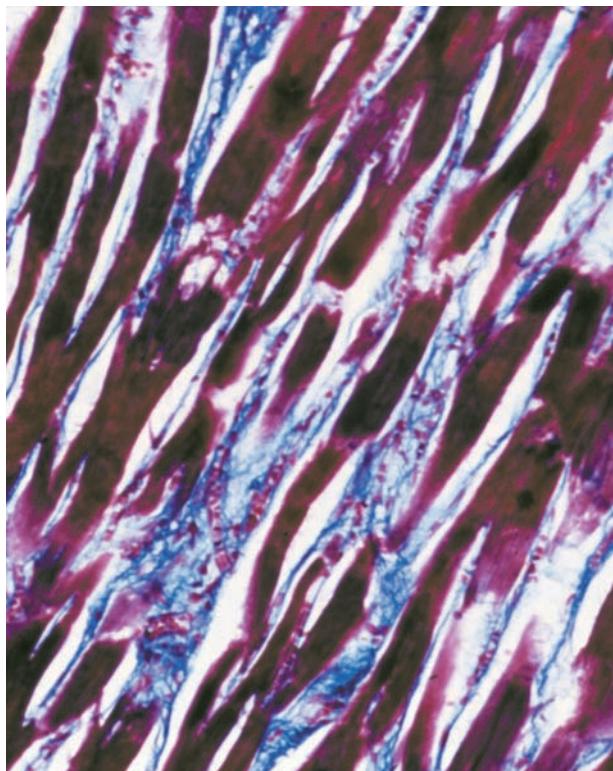
Rh incompatibility see **Erythroblastosis fetalis**

Rh typing see **Blood typing and crossmatching**

Rheumatic fever

Definition

Rheumatic fever (RF) is an illness which arises as a complication of untreated or inadequately treated **strep throat** infection. Rheumatic fever can seriously damage the valves of the heart.



A magnified image of cardiac muscle damaged by chronic myocarditis caused by rheumatic fever. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

Throat infection with a member of the Group A streptococcus (strep) bacteria is a common problem among school-aged children. It is easily treated with a 10-day course of **antibiotics** by mouth. However, when such a throat infection occurs without symptoms, or when a course of medication is not taken for the full 10 days, there is a 3% chance of that person developing rheumatic fever. Other types of strep infections (such as of the skin) do not put the patient at risk for RF.

Children between the ages of 5 and 15 are most susceptible to strep throat, and therefore most susceptible to rheumatic fever. Other risk factors include poverty, overcrowding (as in military camps), and lack of access to good medical care. Just as strep throat occurs most frequently in fall, winter, and early spring, so does rheumatic fever.

Causes and symptoms

Two different theories exist as to how a bacterial throat infection can develop into the disease called rheumatic fever. One theory, less supported by

research evidence, suggests that the bacteria produce some kind of poisonous chemical (toxin). This toxin is sent into circulation throughout the bloodstream, thus affecting other systems of the body.

Research seems to point to a different theory, however. This theory suggests that the disease is caused by the body's immune system acting inappropriately. The body produces immune cells (called antibodies), which are specifically designed to recognize and destroy invading agents; in this case, streptococcal bacteria. The antibodies are able to recognize the bacteria because the bacteria contain special markers called antigens. Due to a resemblance between Group A streptococcus bacteria's antigens and antigens present on the body's own cells, the antibodies mistakenly attack the body itself.

It is interesting to note that members of certain families seem to have a greater tendency to develop rheumatic fever than do others. This could be related to the above theory, in that these families may have cell antigens which more closely resemble streptococcal antigens than do members of other families.

In addition to fever, in about 75% of all cases of RF one of the first symptoms is arthritis. The joints (especially those of the ankles, knees, elbows, and wrists) become red, hot, swollen, shiny, and extraordinarily painful. Unlike many other forms of arthritis, the arthritis may not occur symmetrically (affecting a particular joint on both the right and left sides, simultaneously). The arthritis of RF rarely strikes the fingers, toes, or spine. The joints become so tender that even the touch of bedsheets or clothing is terribly painful.

A peculiar type of involuntary movement, coupled with emotional instability, occurs in about 10% of all RF patients (the figure used to be about 50%). The patient begins experiencing a change in coordination, often first noted by changes in handwriting. The arms or legs may flail or jerk uncontrollably. The patient seems to develop a low threshold for anger and sadness. This feature of RF is called **Sydenham's chorea** or St. Vitus' Dance.

A number of skin changes are common to RF. A rash called erythema marginatum develops (especially in those patients who will develop heart problems from their illness), composed of pink splotches, which may eventually spread into each other. It does not itch. Bumps the size of peas may occur under the skin. These are called subcutaneous nodules; they are hard to the touch, but not painful. These nodules most commonly occur over the knee and elbow joint, as well as over the spine.

The most serious problem occurring in RF is called pancarditis ("pan" means total; "carditis" refers to

KEY TERMS

Antibodies—Specialized cells of the immune system which can recognize organisms that invade the body (such as bacteria, viruses, and fungi). The antibodies are then able to set off a complex chain of events designed to kill these foreign invaders.

Antigen—A special, identifying marker on the outside of cells.

Arthritis—Inflammation of the joints.

Autoimmune disorder—A disorder in which the body's antibodies mistake the body's own tissues for foreign invaders. The immune system therefore attacks and causes damage to these tissues.

Chorea—Involuntary movements in which the arms or legs may jerk or flail uncontrollably.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body, which work together to prevent foreign invaders (bacteria, viruses, fungi, etc.) from taking hold and growing.

Inflammation—The body's response to tissue damage. Includes hotness, swelling, redness, and pain in the affected part.

Pancarditis—Inflammation of the lining of the heart, the sac around the heart, and the muscle of the heart.

inflammation of the heart). Pancarditis is an inflammation that affects all aspects of the heart, including the lining of the heart (endocardium), the sac containing the heart (pericardium), and the heart muscle itself (myocardium). About 40–80% of all RF patients develop pancarditis. This RF complication has the most serious, long-term effects. The valves within the heart (structures which allow the blood to flow only in the correct direction, and only at the correct time in the heart's pumping cycle) are frequently damaged during the course of pancarditis. This may result in blood which either leaks back in the wrong direction, or has a difficult time passing a stiff, poorly moving valve. Either way, damage to a valve can result in the heart having to work very hard in order to move the blood properly. The heart may not be able to "work around" the damaged valve, which may result in a consistently inadequate amount of blood entering the circulation.

Diagnosis

Diagnosis of RF is done by carefully examining the patient. A list of diagnostic criteria has been created. These "Jones Criteria" are divided into major and minor criteria. A patient can be diagnosed with RF if he or she has either two major criteria (conditions), or one major and two minor criteria. In either case, it must also be proved that the individual has had a previous infection with streptococcus.

The major criteria include:

- carditis
- arthritis
- chorea
- subcutaneous nodules
- erythema marginatum

The minor criteria include:

- fever
- joint pain (without actual arthritis)
- evidence of electrical changes in the heart (determined by measuring electrical characteristics of the heart's functioning during a test called an electrocardiogram, or EKG)
- evidence (through a blood test) of the presence in the blood of certain proteins, which are produced early in an inflammatory/infectious disease.

Tests are also performed to provide evidence of recent infection with group A streptococcal bacteria. A swab of the throat can be taken, and smeared on a substance in a petri dish, to see if bacteria will multiply and grow over 24–72 hours. These bacteria can then be specially processed, and examined under a microscope, to identify streptococcal bacteria. Other tests can be performed to see if the patient is producing specific antibodies; that are only made in response to a recent strep infection.

Treatment

A 10-day course of penicillin by mouth, or a single injection of penicillin G-is the first line of treatment for RF. Patients will need to remain on some regular dose of penicillin to prevent recurrence of RF. This can mean a small daily dose of penicillin by mouth, or an injection every three weeks. Some practitioners keep patients on this regimen for five years, or until they reach 18 years of age (whichever comes first). Other practitioners prefer to continue treating those patients who will be regularly exposed to streptococcal bacteria (teachers, medical workers), as well as those patients with known RF heart disease.

Arthritis quickly improves when the patient is given a preparation containing **aspirin**, or some other anti-inflammatory agent (ibuprofen). Mild carditis will also improve with such anti-inflammatory agents, although more severe cases of carditis will require steroid medications. A number of medications are available to treat the involuntary movements of chorea, including diazepam for mild cases, and haloperidol for more severe cases.

Prognosis

The long-term prognosis of an RF patient depends primarily on whether he or she develops carditis. This is the only manifestation of RF which can have permanent effects. Those patients with no or mild carditis have an excellent prognosis. Those with more severe carditis have a risk of **heart failure**, as well as a risk of future heart problems, which may lead to the need for valve replacement surgery.

Prevention

Prevention of the development of RF involves proper diagnosis of initial strep throat infections, and adequate treatment within 10 days with an appropriate antibiotic. Prevention of RF recurrence requires continued antibiotic treatment, perhaps for life. Prevention of complications of already-existing RF heart disease requires that the patient always take a special course of antibiotics when he or she undergoes any kind of procedure (even dental cleanings) that might allow bacteria to gain access to the bloodstream.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Rosalyn Carson-DeWitt, MD

Rheumatoid arthritis

Definition

Rheumatoid arthritis is an autoimmune disease that primarily damages the lining of joints. Other problems throughout the body (systemic problems) may also develop, including inflammation of blood vessels (**vasculitis**), the development of bumps (called rheumatoid nodules) in various parts of the body, lung

disease, blood disorders, and weakening of the bones (**osteoporosis**).

Description

Rheumatoid arthritis (RA) is a disease mainly characterized by chronic inflammation of the tissue lining the joints (synovium). A joint is a point of connection between two bones that allows motion. For example, an elbow joint connects an arm to the forearm allowing motion of the arm, and a knee joint connects a thigh to the lower leg, allowing the straightening and bending of the knee. RA can affect almost any joint of the body, including those of the fingers, wrists, shoulders, elbows, hips, knees, ankles, feet, and neck. It can lead to long-term joint damage, resulting in chronic **pain** and disability. RA does not only affect joints. It is a systemic disease, because it can affect other organs in the body, such as the heart, muscles, blood vessels, nervous system, and eyes.

In RA, the synovial membrane becomes severely inflamed. Usually thin and delicate, the synovium becomes thick and stiff, with numerous infoldings on its surface. The membrane is invaded by white blood cells, which produce a variety of destructive chemicals. The cartilage along the articular surfaces of the bones may be attacked and destroyed, and the bone, articular capsule, and ligaments may begin to wear away (erode). These processes severely interfere with movement in the joint.

RA is also a progressive disease. The first stage of the disease is inflammation of the synovium of the affected joint, which causes pain, warmth, stiffness, redness and swelling around the joint that can last for hours. The arthritis usually begins in the small joints of the hands and the feet, spreading later to the larger joints. In the second stage, there is an overgrowth of connective tissue on the articular surface of the affected joint resulting in a thickening of the affected synovium (pannus). Finally, as part of the autoimmune response, the inflamed cells release substances that start destroying bone and cartilage, causing joint deformity, more pain, and loss of function.

Demographics

According to the World Health Organization (WHO), RA is the most common chronic inflammatory joint disease. The incidence and prevalence of RA appear to have fallen in Europe, North America and Japan in the last 50 years. The prevalence of RA is estimated as relatively constant in many populations, at 0.5–1.0%, with low occurrences reported in

KEY TERMS

Anorexia—Loss of or markedly reduced appetite or total aversion to food.

Antibody—A protein produced by the body's immune system in response to a foreign substance.

Articular bones—Two or more bones connected to each other via a joint.

Autoimmune disease—Disease characterized by the involvement of an inappropriate immune response that leads the body to attack its own cells and tissues.

Autoimmune response—A condition in which a person's immune system fails to recognize its own cells as being "self" and attacks its own body.

Disease-modifying antirheumatic drug (DMARD)—Medication belonging to a group of medications commonly used in patients with rheumatoid arthritis that acts by lowering the autoimmune response.

Immune system—The organs and cells that defends the body against infections and other diseases.

Immunosuppressant—Medication that can block the body's immune response.

Joint—The point of connection between two bones that allows motion.

Nonsteroidal anti-inflammatory drug (NSAID)—Medication that does not contain cortisone used to reduce the symptoms of the pain and inflammation of arthritis.

Osteoarthritis—A non-inflammatory wearing away of bone and cartilage most often associated with aging.

Pannus—Overgrowth of connective tissue on the articular surface of a joint.

Pauciarticular juvenile RA—Rheumatoid arthritis found in children that affects less than four joints.

Polyarticular juvenile RA—Rheumatoid arthritis found in children that affects more than four joints.

Rheumatoid factor (RF)—An antibody present in the blood serum of many individuals affected by rheumatoid arthritis.

Synovial fluid—A lubricating fluid secreted by the synovial membrane.

Synovial membrane—A layer of connective tissue that lines the cavities of joints.

Synovium—A fibrous envelope that produces a fluid to help to reduce friction and wear in a joint.

Systemic disease—A disease that affects the entire body instead of a specific organ.

populations from China and Japan. According to the Arthritis Foundation, approximately 1.3 million Americans are afflicted by RA. The disease can affect anyone, including children, but 70% of people with RA are women. RA onset usually occurs between 30 and 50 years of age. A high prevalence of RA has been reported in the Pima (5.3%) and in the Chippewa (6.8%) Indians. Older age and female gender are risk factors both for the development of RA and for a poor outcome.

Causes and symptoms

The cause of RA remains unknown but most medical researchers believe that it is an autoimmune disease, meaning a disease characterized by the involvement of an inappropriate immune response that leads the body to attack the lining of its own joints. How this autoimmune response develops is not known but it causes the inflammation that produces the pain, swelling, and stiffness associated with RA. Other research has proposed that susceptibility to RA may be genetic or environmental.

Many researchers are examining the possibility that exposure to an organism (like a bacteria or virus) may be the first event in the development of RA. The body's normal response to such an organism is to produce cells that can attack and kill the organism, protecting the body from the foreign invader. In an autoimmune disease like RA, this immune cycle spins out of control. The body produces misdirected immune cells, which accidentally identify parts of the person's body as foreign. These immune cells then produce a variety of chemicals that injure and destroy parts of the body.

Symptoms vary from person to person and can mimic other bone and joint diseases such as **osteoarthritis**. For most people, the symptoms of rheumatoid arthritis appear gradually, although about one-third of individuals develop serious symptoms within a few months. In many people, symptoms tend to change from day to day, with periods of improvement followed by periods of worsening symptoms. In more serious cases, symptoms simply worsen progressively without periods of improvement. The wrists and hand joints are affected in more than 85% of individuals with rheumatoid arthritis. Usually if a joint on one side of the body

is inflamed, the same joint on the other side will also be affected.

The symptoms of RA are the same as for all forms of arthritis and usually include morning stiffness, lasting joint pain, joint swelling, joint stiffness, tenderness or pain when touching a joint, difficulty using or moving a joint normally, and warmth and redness in a joint.

Many patients also notice increased **fatigue**, loss of appetite, weight loss, and sometimes **fever**. Rheumatoid nodules are bumps that appear under the skin around the joints and on the top of the arms and legs. These nodules can also occur in the tissue covering the outside of the lungs and lining the chest cavity (pleura), and in the tissue covering the brain and spinal cord (meninges). Lung involvement may cause **shortness of breath** and is seen more in men. Vasculitis (inflammation of the blood vessels) may interfere with blood circulation. This can result in irritated pits (ulcers) in the skin, tissue **death (gangrene)**, and interference with nerve functioning that causes **numbness and tingling**.

Juvenile RA is a chronic inflammatory disease that affects the joints of children less than 16 years old. It is estimated to affect as many as 250,000 children in the United States alone. Most children with juvenile RA have arthritis when the illness starts, which affects multiple joints in 50% of these children, and only one joint in 30%. In all, 20% of the children affected by juvenile RA have the acute systemic form of the disease, which is characterized by fever, joint inflammation, rash, **liver disease**, and gastrointestinal disease.

Two periods of childhood are associated with an increased incidence of onset of juvenile RA. The first is from one to three years of age, and the second, from 8 to 12 years. When more than four joints are affected, the disease is described as being polyarticular. If less than four joints are affected, the disease is known as pauciarticular. Juvenile RA and this particular manifestation falls into two categories. The first occurs in girls aged one to four years old, and the onset of joint involvement is in the knees, ankles, or elbows. The second form occurs in boys aged eight years and older, and involves the larger joints, such as those of the hips and legs.

Diagnosis

The RA diagnosis may be difficult to establish, because there is no single test that can be performed to confirm RA. The diagnosis is based upon an individual's history of clinical symptoms and a complete **physical examination**. A specialized physician, often a rheumatologist, reviews all signs and symptoms experienced by a person, so as to rule out other joint diseases. This often requires various tests, which may include:

- **Rheumatoid factor (RF) test:** This diagnostic test measures the presence and amounts of rheumatoid factor in the blood. The test looks for distinctive antibodies released in the blood by people with RA to distinguish it from other forms of arthritis and other conditions that cause similar symptoms of joint pain, inflammation, and stiffness. Rheumatoid factor is an autoantibody found in about 80% of patients with RA. It is often not very specific however, because it is found in about 5% of all healthy people and in 10–20% of healthy people over the age of 65. In addition, rheumatoid factor is also positive in a large number of other autoimmune diseases and other infectious diseases, including systemic lupus erythematosus, bacterial endocarditis, malaria, and syphilis. In addition, young people who have a process called juvenile rheumatoid arthritis often have no rheumatoid factor present in their blood.

- **Antinuclear antibody (ANA) test:** This test is performed to help screen for autoimmune disorders. A small percentage of healthy people, however, have a positive ANA.

- **C-Reactive protein (CRP) test:** The CRP test is used to evaluate how active the inflammation is. CRP tests are not specific enough to diagnose RA, but provide a general marker of infection and inflammation levels.

- **Synovial fluid exam:** The clinician may examine the synovial fluid, by inserting a thin needle into a synovial joint. In RA, this fluid has certain characteristics that indicate active inflammation. The fluid is cloudy, with increased protein and decreased or normal glucose. It also contains a higher than normal number of white blood cells. While these findings suggest inflammatory arthritis, they are not specific to RA.

Other tests, including x rays and **magnetic resonance imaging (MRI)**, may be used to determine the cause of chronic back pain or examine internal organs that may be affected by RA.

The American Rheumatology Association designates that at least four of the following seven criteria must be present for at least six weeks to diagnose rheumatoid arthritis.

- morning joint stiffness lasting more than one hour
- pain simultaneously in three or more joint areas
- arthritis in the wrist or hand
- joint pain in symmetrical joint areas (e.g. both wrists, both knees)
- presence of rheumatoid nodules
- presence of serum rheumatoid factor, a protein found in blood
- x rays that show typical rheumatoid arthritis changes in the affected joints

Treatment

There is presently no cure for rheumatoid arthritis. However, treatment is available to combat the inflammation in order to prevent destruction of the joints and to prevent other complications of the disease. Efforts are also made to maintain flexibility and mobility of the joints. In addition to pain and anti-inflammatory medicines, RA is treated with **antirheumatic drugs**. Rest is prescribed for severely inflamed joints, as using them can aggravate the inflammation. Regular rest periods can often relieve pain, with short periods of bed rest considered helpful to relieve a severe flare-up in its most painful stage.

Treatment is divided into two categories: treatment of symptoms and treatment to stop or slow joint damage. Treatment to improve symptoms includes the use of various pain medications including **nonsteroidal anti-inflammatory drugs** (e.g., **aspirin**, ibuprophen, naproxen sodium) and **analgesics** (**acetaminophen**, tramadol), either alone or in combination with narcotic pain medications. **Corticosteroids** such as prednisone and cortisone are also used in the lowest effective dose to control pain and stiffness. Also beneficial is **exercise** and **physical therapy** (PT) to increase strength and flexibility.

Drugs to stop or slow joint damage are collectively called disease-modifying antirheumatic drugs (DMARDs). These drugs, especially when given early in the course of the disease, interfere with the disease process in ways that slow or stop joint damage. DMARDs are often given in combination with drugs to improve symptoms. Some common DMARDs include methotrexate (Rheumatrex, Trexall), hydroxychloroquine (Plaquenil), sulfasalazine (Azulfidine), leflunomide (Arava), D-pencillamine (Dpen, Cuprimine), azathioprine (Imuran), cyclosporine (Neoral, Sandimmune) and minocycline (Minocin, Dynacin). All these drugs have potentially serious side effects and may require regular blood or other tests.

Rheumatoid arthritis can also be treated with biologic response modifiers (BRMs). BRMs target specific proteins of the immune system that are involved in rheumatoid arthritis. They work to reduce joint inflammation by blocking a substance called tumor necrosis factor (TNF). TNF is a protein that triggers inflammation during the body's normal immune responses. When TNF production is not regulated, the excess TNF can cause inflammation. Most BRMs are approved for use in adults only. The exception is etanercept (Enbrel), which is approved for individuals over age four. Other BRMs used to treat rheumatoid arthritis include infliximab (Remicade), anakinra (Kineret), and adalimumab (Humira). BRMs interfere with and may weaken the immune system. Individuals should not receive live-

virus vaccinations while taking BRMs. Other side effects are also possible.

Hydrotherapy can help to greatly reduce pain and inflammation. Moist heat is more effective than dry heat, and cold packs are useful during acute flare-ups. Total bed rest is sometimes prescribed during the very active, painful phases of RA. Splints may be used to support and rest painful joints. Later, after inflammation has somewhat subsided, physical therapists may provide a careful exercise regimen in an attempt to maintain the maximum degree of flexibility and mobility. **Joint replacement** surgery, particularly for the knee and the hip joints, is sometimes recommended when these joints have been severely damaged.

Many complementary and alternative cures are heavily advertised for rheumatoid arthritis. The National Center for Complementary and Alternative Medicine has investigated many of these alternative cures. Most do not provide any benefit to individuals with rheumatoid arthritis. Those complementary and alternative treatments that may have possible benefit include thunder god vine (*Tripterygium wilfordii*, not available in the United States as of September 2005), gamma-linolenic acid (GLA), fish oil, glucosamine and chondroitin (effective in animals, but unproven in humans), and mind-body **stress reduction** techniques. The National Arthritis Foundation provides information on alternative and complementary therapies; persons with RA may benefit from massage, **acupuncture**, **acupressure**, and various herbs and supplements. Individuals should not replace conventional treatment with alternative therapies, and before adding any herbal or other complementary treatments should consult their physician, as some complementary therapies may interfere with the conventional treatment and/or have serious side effects.

RA patients can also undergo **occupational therapy** (OT), where they are instructed on how to protect affected joints, and how to reduce strain on the joints during daily activities. For instance, special shoes and the use of a cane can help alleviate pain in the feet, knees, and hips when walking. Occupational therapy also seeks to restore abilities that may have been lost, and to suggest approaches to maintain independence and fitness.

When treatment fails to control pain and joint damage, joint replacement surgery followed by guided **rehabilitation** may be necessary. Knee and hip replacement surgery are the most common types of surgery done on individuals with rheumatoid arthritis.

Nutrition/Dietetic concerns

There is presently no scientific evidence showing conclusively that any particular foods may have a

beneficial effect on joint inflammation, although some reports have proposed that oranges and some fish oils may reduce joint inflammation in some people with RA. A healthy, balanced diet aimed at maintaining a normal weight is important for people afflicted with RA because excess weight increases **stress** on the weight-bearing joints, contributing to joint pain, stiffness and inflammation.

Prognosis

There is no cure for rheumatoid arthritis. The course of the disease is variable. Some people have the disease for only a year or two, and then it goes away on its own without joint damage. Many other people have periods when the disease is quiet and symptoms disappear, only to flare up again for unknown reasons. For some people the disease is continuous, chronic, and progressively worsens.

A number of factors are considered to suggest the likelihood of a worse prognosis. These include:

- race and gender (female and Caucasian).
- more than 20 joints involved.
- extremely high erythrocyte sedimentation rate.
- extremely high levels of rheumatoid factor.
- consistent, lasting inflammation.
- evidence of erosion of bone, joint, or cartilage on x rays.
- poverty.
- older age at diagnosis.
- rheumatoid nodules.
- other coexisting diseases.
- certain genetic characteristics, diagnosable through testing.

In general, the long-term prognosis is poor. The irreversible destruction of joints usually begins within the first 2 years of disease onset in the majority of people with RA. Treatment can manage the pain and swelling caused by RA, and joint damage may even slow down or stop. Treatment can bring relief of symptoms to 75% of those afflicted. However, at least 1 of 10 people eventually becomes severely disabled, and the average life expectancy for a patient with RA may be shortened by 3–7 years.

Prevention

Rheumatoid arthritis cannot be prevented. Early detection and treatment can help slow the disease. Clinical trials of new medications and complementary and alternative therapies for rheumatoid arthritis are ongoing. A list of clinical trials currently enrolling patients is available at www.clinicaltrials.gov.

Health care team roles

A rheumatologist normally oversees the health care team treating an individual with rheumatoid arthritis. Nurses play an important role in patient education by teaching individuals with rheumatoid arthritis how to balance activity and rest. Physical therapists evaluate an individual's range of motion and teach appropriate exercises to promote joint mobility and muscle fitness and the appropriate use of heat and cold treatments. Physical therapists also have special equipment that can provide electrical stimulation to reduce pain and improve joint movement. Occupational therapists teach individuals how to move in ways that protect their joints and how to perform tasks of daily living in ways that reduce pain and stress on the joints. Both PT and OT are essential after surgery but may also be helpful to individuals with advanced rheumatoid arthritis undergoing non-surgical treatments.

Caregiver concerns

The prevalence of RA increases up to age 80 and represents an important cause of disability in elderly persons. In many senior patients, RA first starts during middle age. Some of these patients have secondary joint deformities and deterioration even though the inflammation may be inactive. In most patients of this age group, the arthritis is accompanied by mild or moderate generalized feelings of discomfort (malaise) and anorexia. Fever and night sweats are also occasionally reported. Elderly-onset rheumatoid arthritis (EORA), defined as RA with onset at age 60 years or over, differs slightly from RA. It is characterized by a more equal gender distribution, a higher frequency of acute systemic symptoms with involvement of the shoulder, a higher rate of disease progression, and, in later stages, more joint damage and functional disability. The efficacy and tolerability of medications is similar in both older and younger patient groups, but in the elderly, caution is required with the use of NSAIDs.

Resources

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ORGANIZATIONS

- American College of Rheumatology, 1800 Century Place, Suite 250, Atlanta, GA, 30345-4300, (404)-633-3777, (404)-633-1870, <http://www.rheumatology.org>.

Arthritis Foundation, P.O. Box 7669, Atlanta, GA, 30357-0669, (800)-283-7800, <http://www.arthritis.org>.
 National Institute of Arthritis and Musculoskeletal Diseases (NIAMS), 1 AMS Circle, Bethesda, MD, 20892-3675, (301)-495-4484, (877)-22-NIAMS, (301)-718-6366, NIAMSInfo@mail.nih.gov, <http://www.niams.nih.gov>.

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Rheumatoid spondylit see **Ankylosing spondylitis**

Rhinitis

Definition

Rhinitis is inflammation of the mucous lining of the nose.

Description

Rhinitis is a nonspecific term that covers infections, **allergies**, and other disorders whose common feature is the location of their symptoms. In rhinitis, the mucous membranes become infected or irritated, producing a discharge, congestion, and swelling of the tissues. The most widespread form of infectious rhinitis is the **common cold**.

The common cold is the most frequent viral infection in the general population, causing more absenteeism from school or work than any other illness. Colds are self-limited, lasting about 3–10 days, although they are sometimes followed by a bacterial infection. Children are more susceptible than adults; teenage boys more susceptible than teenage girls; and adult women more susceptible than adult men. In the United States, colds are most frequent during the late fall and winter.

Causes and symptoms

Colds can be caused by as many as 200 different viruses. The viruses are transmitted by sneezing and coughing, by contact with soiled tissues or handkerchiefs, or by close contact with an infected person. Colds are easily spread in schools, offices, or any place where people live or work in groups. The incubation period ranges between 24 and 72 hours.

The onset of a cold is usually sudden. The virus causes the lining of the nose to become inflamed and produce large quantities of thin, watery mucus. Children

sometimes run a **fever** with a cold. The inflammation spreads from the nasal passages to the throat and upper airway, producing a dry **cough**, **headache**, and watery eyes. Some people develop muscle or joint aches and feel generally tired or weak. After several days, the nose becomes less inflamed and the watery discharge is replaced by a thick, sticky mucus. This change in the appearance of the nasal discharge helps to distinguish rhinitis caused by a viral infection from rhinitis caused by an allergy.

Diagnosis

There is no specific test for viral rhinitis. The diagnosis is based on the symptoms. In children, the doctor will examine the child's throat and glands to rule out **measles** and other childhood illnesses that have similar early symptoms. Adults whose symptoms last longer than a week may require further testing to rule out a secondary bacterial infection, or an allergy. Bacterial infections can usually be identified from a laboratory culture of the patient's nasal discharge. Allergies can be evaluated by blood tests, skin testing for specific substances, or nasal smears.

Treatment

There is no cure for the common cold; treatment is given for symptom relief. Medications include **aspirin** or **nonsteroidal anti-inflammatory drugs** (NSAIDs) for headache and muscle **pain**, and **decongestants** to relieve stuffiness or runny nose. Patients should be warned against overusing decongestants, because they can cause a rebound effect. Over-the-counter (OTC) **antihistamines** are also available; however, most antihistamines carry warnings of drowsiness and the inability to do some tasks while medicated. Claritin is a prescription-strength OTC non-drowsy antihistamine that helps relieve symptoms of rhinitis. **Antibiotics** are not given for colds because they do not kill viruses.

Supportive care includes bed rest and drinking plenty of fluid.

Treatments under investigation include the use of ultraviolet light and injections of interferon.

Many prescription and over-the-counter drugs are available to help control the symptoms of **allergic rhinitis**. The most common class is antihistamines.

Alternative treatment

Homeopaths might prescribe any of 10 different remedies, depending on the appearance of the nasal discharge, the patient's emotional state, and the stage of infection. Naturopaths would recommend vitamin A

KEY TERMS

Interferon—A protein produced by cells infected by a virus that stimulates the body's resistance to the virus.

and zinc supplements, together with botanical preparations made from goldenseal (*Hydrastis canadensis*), licorice (*Glycyrrhiza glabra*), or astragalus (*Astragalus membranaceus*) root.

At one time, the herb (*Echinacea spp.*) was touted as a remedy to relieve cold and rhinitis symptoms. However, a study published in 2004 reported that the herb failed to relieve cold symptoms in 400 children taking it and caused skin **rashes** in some children.

Prognosis

Most colds resolve completely in about a week. Complications are unusual but may include **sinusitis** (inflammation of the nasal sinuses), bacterial infections, or infections of the middle ear.

Prevention

There is no vaccine effective against colds, and infection does not confer immunity. Prevention depends on:

- washing hands often, especially before touching the face
- minimizing contact with people already infected
- not sharing hand towels, eating utensils, or water glasses.

Resources

PERIODICALS

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Rhinoplasty

Definition

The term rhinoplasty means "nose molding" or "nose forming." It refers to a procedure in **plastic surgery** in which the structure of the nose is changed. The change can be made by adding or removing bone

or cartilage, grafting tissue from another part of the body, or implanting synthetic material to alter the shape of the nose.

Rhinoplasty is the most frequently performed cosmetic surgical procedure in the United States as of the early 2000s. According to the American Society of Plastic Surgeons (ASPS), 356,554 rhinoplasties were performed in the United States in 2003, compared to 254,140 breast augmentations and 128,667 facelifts.

Purpose

Rhinoplasty is most often performed for cosmetic reasons. A nose that is too large, crooked, misshapen, malformed at birth, or deformed by an injury or **cancer** surgery can be given a more pleasing appearance. If breathing is impaired due to the form of the nose or to an injury, it can often be improved with rhinoplasty.

Precautions

The best candidates for rhinoplasty are those with relatively minor deformities. Nasal anatomy and proportions are quite varied and the final look of any rhinoplasty operation is the result of the patient's anatomy, as well as of the surgeon's skill.

The quality of the skin plays a major role in the outcome of rhinoplasty. Patients with extremely thick skin may not see a definite change in the underlying bone structure after surgery. On the other hand, thin skin provides almost no cushion to hide the most minor of bone irregularities or imperfections.

A cosmetic change in the shape of the nose will change a person's appearance but it will not change self-image. A person who expects a different lifestyle after rhinoplasty is likely to be disappointed.

Rhinoplasty should not be performed until the pubertal growth spurt is complete, between ages 14–15 for girls and older for boys.

The cost of rhinoplasty depends on the difficulty of the work required and on the specialist chosen. Prices run from about \$3,000 to over \$6,000. If the problem was caused by an injury, insurance will usually cover the cost. A rhinoplasty done only to change a person's appearance is not usually covered by insurance.

Description

The external nose is composed of a series of interrelated parts which include the skin, the bony pyramid, cartilage, and the tip of the nose, which is both cartilage and skin. The strip of skin separating the nostrils is called the columella.

Surgical approaches to nasal reconstruction are varied. Internal rhinoplasty involves making all incisions inside the nasal cavity. The external or "open" technique involves a skin incision across the base of the nasal columella. An external incision allows the surgeon to expose the bone and cartilage more fully and is most often used for complicated procedures. During surgery, the surgeon will separate the skin from the bone and cartilage support. The framework of the nose is then reshaped in the desired form. Shape can be altered by removing bone, cartilage, or skin. The remaining skin is then replaced over the new framework. If the procedure requires adding to the structure of the nose, the donated bone, cartilage, or skin can come from the patient or from a synthetic source.

When the operation is over, the surgeon will apply a splint to help the bones maintain their new shape. The nose may also be packed or stuffed with a dressing, to help stabilize the septum.

When a local anesthetic is used, light **sedation** is usually given first, after which the operative area is numbed. It will remain insensitive to **pain** for the length of the surgery. A general anesthetic is used for lengthy or complex procedures or if the doctor and patient agree that it is the best option.

Simple rhinoplasty is usually performed in an outpatient surgery center or in the surgeon's office. Most procedures take only an hour or two, and patients may return home right away. Complex procedures may be done in the hospital and require a short stay.

Preparation

During the initial consultation, the patient and surgeon will determine what changes can be made in the shape of the nose. Most doctors take photographs at the same time. The surgeon will also explain the techniques and anesthesia options available to the patient.

For legal reasons, many plastic surgeons now screen patients for psychological stability as well as general physical fitness for surgery. When a person consults a plastic surgeon about a rhinoplasty, the doctor will spend some time talking with the patient about his or her motives for facial surgery. The following are considered psychological warning signs:

- The patient is considering surgery to please someone else, most often a spouse or partner.
- The patient expects facial surgery to guarantee career advancement.
- The patient has a history of multiple cosmetic procedures and/or complaints about previous surgeons.

- The patient thinks that the surgery will solve all his or her life problems.
- The patient has an unrealistic notion of what he or she will look like after surgery.
- The patient seems otherwise emotionally unstable.

The patient and surgeon should also discuss guidelines for eating, drinking, **smoking**, taking or avoiding certain medications, and washing of the face.

Aftercare

Patients usually feel fine immediately after surgery; however, most surgery centers do not allow patients to drive themselves home after an operation.

The first day after surgery there will be some swelling of the face. Patients should stay in bed with their heads elevated for at least a day. The nose may hurt and a **headache** is not uncommon. The surgeon will prescribe medication to relieve these conditions. Swelling and bruising around the eyes will increase for a few days but will begin to diminish after about the third day. Slight bleeding and stuffiness are normal, and vary according to the extensiveness of the surgery performed. Most people are up in two days and back to school or work in a week. No strenuous activities are allowed for two to three weeks.

Patients are given a list of postoperative instructions, which include requirements for hygiene, **exercise**, eating, and follow-up visits to the doctor. Patients should not blow their noses for the first week to avoid disruption of healing. It is extremely important to keep the surgical dressing dry. **Dressings**, splints, and stitches are removed in one to two weeks. Patients should avoid **sunburn**.

Patients should remember that it may take as long as a year for the nose to assume its final shape; the tip of the nose in particular may be mildly swollen for several months.

Risks

Any type of surgery carries a degree of risk. There is always the possibility of unexpected events, such as an infection or a reaction to the anesthesia. Some patients may have a so-called foreign body reaction to a nasal implant made from synthetic materials. In these cases the surgeon can replace the implant with a piece of cartilage from the patient's own body.

Some risks of rhinoplasty are social or psychological. The ASPS patient brochure about rhinoplasty mentions the possibility of criticism or rejection by friends or family if they feel threatened by the patient's new look. This type of reaction sometimes occurs with rhinoplasty if the friends or relatives consider the shape of the nose an important family or ethnic trait.

KEY TERMS

Cartilage—Firm supporting tissue that does not contain blood vessels.

Columella—The strip of skin running from the tip of the nose to the upper lip, which separates the nostrils.

Septum—The dividing wall in the nose.

When the nose is reshaped or repaired from inside, the **scars** are not visible, but if the surgeon needs to make the incision on the outside of the nose, there will be some slight scarring. In addition, tiny blood vessels may burst, leaving small red spots on the skin. These spots are barely visible but may be permanent.

About 10% of patients require a second procedure; however, the corrections required are usually minor.

Resources

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ORGANIZATIONS

American Academy of Facial Plastic and Reconstructive Surgery (AAFPKS), 310 South Henry Street, Alexandria, VA, 22314, (703) 299-9291, info@aafprs.org, <http://www.aafprs.org/>.

American Society of Plastic Surgeons, 444 E. Algonquin Rd, Arlington Heights, IL, 60005, (847) 228-9900, <http://www.plasticsurgery.org/>.

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Rhinovirus infection see **Common cold**

Rhytidoplasty see **Face lift**

Riboflavin deficiency

Definition

Riboflavin deficiency occurs when the chronic failure to eat sufficient amounts of foods that contain riboflavin produces lesions of the skin, lesions of smooth surfaces in the digestive tract, or nervous disorders.

Description

Riboflavin, also called vitamin B₂, is a water-soluble vitamin. The recommended dietary allowance (RDA) for riboflavin is 1.7 mg/day for an adult man and 1.3 mg/day for an adult woman. The best sources of this vitamin are meat, dairy products, and dark green vegetables, especially broccoli. Grains and legumes (beans and peas) also contribute riboflavin to the diet. Riboflavin is required for the processing of dietary fats, carbohydrates, and proteins to convert these nutrients to energy. Riboflavin is also used for the continual process of renewal and regeneration of all cells and tissues in the body.

Riboflavin is sensitive to light. For this reason, commercially available milk is sometimes supplied in cartons, rather than in clear bottles. Riboflavin is not rapidly destroyed by cooking. Milk contains about 1.7 mg riboflavin/kg. Cheese contains about 4.3 mg/kg, while beef has 2.4 mg/kg and broccoli has about 2.0 mg/kg. Apples, a food that is low in all nutrients, except water, contains only 0.1 mg riboflavin per kg.

Causes and symptoms

A deficiency only in riboflavin has never occurred in the natural environment. In contrast, diseases where people are deficient in one vitamin, such as thiamin, vitamin C, and vitamin D, for example, have been clearly documented. Poorer populations in the United States may be deficient in riboflavin, but when this happens, they are also deficient in a number of other nutrients as well. When riboflavin deficiency is actually detected, it is often associated with low consumption of milk, chronic **alcoholism**, or chronic **diarrhea**.

The symptoms of riboflavin deficiency include:

- swelling and fissuring of the lips (cheilosis)
- ulceration and cracking of the angles of the mouth (angular stomatitis)
- oily, scaly skin rashes on the scrotum, vulva, or area between the nose and lips
- inflammation of the tongue
- red, itchy eyes that are sensitive to light

KEY TERMS

Recommended dietary allowance—The recommended daily allowances (RDAs) are quantities of nutrients of the diet that are required to maintain human health. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient needed to maintain health in a population of people. The actual amounts of each nutrient required to maintain health in any specific individual differs from person to person.

Water-soluble vitamin—Water-soluble vitamins can be dissolved in water or juice. Fat-soluble vitamins can be dissolved in oil or in melted fat.

The nervous symptoms of riboflavin deficiency include:

- numbness of the hands
- decreased sensitivity to touch, temperature, and vibration

Diagnosis

Riboflavin status is diagnosed using a test conducted on red blood cells that measures the activity of an enzyme called glutathione reductase. An extract of the red blood cells is placed in two test tubes. One test tube contains no added riboflavin, while the second test tube contains a derivative of riboflavin, called flavin adenine dinucleotide. The added riboflavin derivative results in little or no stimulation of enzyme activity in patients with normal riboflavin levels. A stimulation of 20% or less is considered normal. A stimulation of over 20% means that the patient is deficient in riboflavin.

Treatment

Riboflavin deficiency can be treated with supplemental riboflavin (0.5 mg/kg body weight per day) until the symptoms disappear.

Prognosis

The prognosis for correcting riboflavin deficiency is excellent.

Prevention

Riboflavin deficiency can be prevented by including milk, cheese, yogurt, meat, and/or certain vegetables in the daily diet. Of the vegetables, broccoli, asparagus, and spinach are highest in riboflavin. These vegetables have a riboflavin content that is similar to that of milk, yogurt, or meat.

Resources

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Tom Brody, PhD

can block the ultraviolet rays. Vitamin D is found naturally in the foods listed above, but more often children receive vitamin D supplements through foods which have had the vitamin added, as in milk or infant formula.

Vitamin D is necessary in the body, because it can be converted into a hormone which stimulates **calcium** intake by the intestines. This conversion begins in the liver, where vitamin D becomes a hormone called 25-OH-D, and is completed when the kidneys convert 25-OH-D into a hormone called 1,25-diOH-D. This is the hormone that causes the intestines to absorb calcium from the person's diet. Without proper levels of vitamin D, there is not enough 1,25-diOH-D produced, which results in lower levels of calcium in the body. Adequate calcium is needed by the bones for both development and maintenance.

Rickets

Definition

Rickets is a childhood condition caused by serious **vitamin D deficiency**. This lacking in vitamin D results in weak, soft bones, along with slowed growth and skeletal development. Rickets is, by definition, a disorder which begins in childhood. If this problem occurs only later in life it is known as osteomalacia.

Description

Rickets occurs when the body has a severe lack of vitamin D during the developmental years. Vitamin D is essential to the development of strong, healthy bones. A child with rickets can experience stunted growth and will most likely be short in stature as an adult. This is because, without proper vitamin D levels, decreased mineralization of the bones at the growth plate level affects the strength, size and shape of the bones. A related condition called osteomalacia can occur in adults with the same sort of vitamin D deficiency, but osteomalacia occurs only in adulthood after the growth plates of the bones have closed.

Most vitamin D is produced by the body, although some can be directly supplied by diet. In order to accomplish production of vitamin D, the body requires both cholesterol and ultraviolet light. Most often, the cholesterol comes from digesting animal tissue, oils, fats, and egg yolks. The ultraviolet light is usually supplied by direct sunlight. Only when this light is available can the skin alter the cholesterol molecule to make vitamin D. Children who do not receive enough sunlight are at greater risk of developing rickets, as are children with darker skin, which

Causes and symptoms

Rickets is directly caused by insufficient calcium for bone mineralization during growth and development. This is caused by vitamin D deficiency which can be a result of too little cholesterol, ultraviolet light, or vitamin D supplement. During the Industrial Revolution, rickets was quite common in cities because pollution in the air blocked much of the sunlight needed for vitamin D production in the body. There is also a hereditary type of rickets, called X-linked hypophosphatemia, that causes the kidneys to be unable to retain phosphate.

The most commonly recognized symptoms of rickets occur in the arms and legs, where **stress** on the underdeveloped bones can cause bowing. Children with rickets may feel **pain** or tenderness in the bones of their arms, legs, spine, pelvis, and ribs. The skull may develop an odd or asymmetrical shape. Calcium levels in the blood will be low and overall growth is often impaired.

Diagnosis

The initial approach to diagnosing rickets involves a musculoskeletal examination followed by an x ray if often. Affected children may have obviously widened spaces between their joints or bowing of the bones in their arms and legs. Some children may not experience normal dental development as well. A doctor may also assess levels of serum calcium, alkaline phosphatase and other indicator chemicals by using a blood test. While calcium levels can be normal or slightly low, alkaline phosphatase levels in a child with rickets can be high even compared to a normal

KEY TERMS

25-hydroxy-vitamin D—This is the form of vitamin D that is measured in order to assess vitamin D deficiency.

Cholesterol—A fat-soluble steroid alcohol (sterol) found in animal fats and oils, and in egg yolks. The human body needs cholesterol to produce vitamin D.

Growth plate—The place in long bones where growth occurs during childhood.

International unit (IU)—A measurement of biological activity in which one IU is equal to one mg (milligram).

Mineralization—The process by which the body uses minerals to build bone structure.

X-linked hypophosphatemia—A type of rickets caused by genetic factors which prevent the kidneys from retaining phosphate.

adult. While x rays can prove misleading, diagnosis by chemical analysis is highly accurate.

Treatment

The treatment for rickets primarily involves corrections of the conditions which led to the disorder. This can be as simple as a change in diet to include foods high in vitamin D such as milk, fish, or liver. Treatment might also mean a gradual increase in the amount sunlight received by the child. In more severe cases, bracing or surgery may be necessary to aid in the correction and repair of bones. Treatment is usually mild and bone deformities usually reduce over time.

Alternative Treatment

There is currently little known about any alternative method for treating rickets. Treatments which involve raising vitamin D levels and ultraviolet light exposure are usually simple and effective.

Prognosis

Children with rickets are likely to suffer from stunted growth, bone abnormalities and bone pain, however these symptoms often disappear with treatment. In women, deformation of the pelvic bone structure can prevent vaginal **childbirth** later in life. Most deformities correct with growth when proper levels of vitamin D are restored and normal bone calcification is maintained.

Prevention

Rickets caused by vitamin D deficiency is simple to prevent. Commercially available infant formula is usually fortified with more than enough vitamin D for infants. For parents who breastfeed their children, it is recommended by the U.S. Department of Health and Human Services that children also receive 400 international units (10 micrograms) of vitamin D supplement. This is because human breast milk contains little

vitamin D. It is also important that children are allowed decent amounts of sunlight. As little as twenty minutes each day can be sufficient. For children living in cities, where pollution is likely to block ultraviolet light, and children with dark skin, which can block ultraviolet light, vitamin D supplement is especially important.

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Tish Davidson, A.M.

Rickets see **Vitamin D deficiency**

Rickettsia rickettsii infection see **Rocky Mountain spotted fever**

Rickettsialpox

Definition

Rickettsialpox is a relatively mild disease caused by a member of the bacterial family called Rickettsia. Rickettsialpox causes rash, **fever**, chills, heavy sweating, **headache**, eye **pain** (especially when exposed to light), weakness, and achy muscles.

Description

Like other members of the family of Rickettsia, the bacteria causing rickettsialpox live in mice. Tiny mites feed on these infected mice, thus acquiring the organism. When these mites feed on humans, the bacteria can be transmitted.

Rickettsialpox occurs mostly within cities. In the United States, the disease has cropped up in such places as New York City, Boston, Philadelphia, Pittsburgh, and Cleveland. It has also been identified in Russia, Korea, and Africa.

Causes and symptoms

The specific bacteria responsible for rickettsialpox is called *Rickettsia akari*. A person contracts this bacteria through the bite of an infected mite. After a person has been bitten by an infected mite, there is a delay of about 10 days to three weeks prior to the onset of symptoms.

The first symptom is a bump which appears at the site of the original bite. The bump (papule) develops a tiny, fluid-filled head (vesicle). The vesicle sloughs away, leaving a crusty black scab in its place (eschar). In about a week, the patient develops a fever, chills, heavy sweating, headache, eye pain (especially when exposed to light), weakness, and achy muscles. The fever rises and falls over the course of about a week. A bumpy rash spreads across the body. Each individual papule follows the same progression: papule, then vesicle, then eschar. The rash does not affect the palms of the hands or the soles of the feet.

Diagnosis

Most practitioners are able to diagnose rickettsialpox simply on the basis of its rising and falling fever, and its characteristic rash. Occasionally, blood will be drawn and tests performed to demonstrate the presence of antibodies (immune cells directed against specific bacterial agents) which would confirm a diagnosis of rickettsialpox.

Treatment

Because rickettsialpox is such a mild illness, some practitioners choose to simply treat the symptoms (giving **acetaminophen** for fever and achiness, pushing fluids to avoid **dehydration**). Others will give their patients a course of the antibiotic tetracycline, which will shorten the course of the illness to about one to two days.

KEY TERMS

Eschar—A crusty, blackish scab.

Papule—A bump on the skin.

Vesicle—A fluid-filled head on a papule.

Prognosis

Prognosis for full recovery from rickettsialpox is excellent. No deaths have ever been reported from this illness, and even the skin rash heals without scarring.

Prevention

As with all mite- or tick-borne illnesses, prevention includes avoidance of areas known to harbor the insects, and/or careful application of insect repellents. Furthermore, because mice pass the bacteria on to the mites, it is important to keep mice from nesting in or around residences.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Rosalyn Carson-DeWitt, MD

Rifampin see **Antituberculosis drugs**

Ringing ears see **Tinnitus**

Ringworm

Definition

Ringworm is a common fungal infection of the skin. The name is a misnomer, however, since the disease is not caused by a worm. Ringworm may also be referred to as dermatophyte infection or dermatophytosis. Dermatophytes are parasitic fungi that live on keratin, a fibrous structural protein found in hair, nails, and the outer layer of skin.

Ringworm is often classified as a **zoonosis** because humans can contract it from infected animals (as well as from other humans).



Ringworm on a man's chin. These infections are most common on the feet, scalp, or in toenails, but they can infect any part of the skin. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Demographics

Ringworm is a common fungal infection found in all countries around the world. It is estimated that between 14% and 20% of the world's population has some form of ringworm at any given time. There are an estimated 4.3 million outpatient visits in the United States each year for treatment of ringworm infections; 22% of these visits are for infections of the nails, 20% for body ringworm, 19% for athlete's foot, 15% for ringworm of the scalp, and nine percent for ringworm of the groin (**jock itch**).

The single most common type of ringworm worldwide is infection of the feet, sometimes called tinea pedis or athlete's foot. This type of ringworm is characterized by **itching**, scaly patches, and a burning sensation between the toes; itchy blisters; or unusually dry skin on the sides and soles of the feet.

Some forms of ringworm are more common in certain age groups than in others. As noted below, ringworm of the scalp and hair commonly affects children but is rare after **puberty**. One study of schoolchildren in the American Midwest reported in 2010

that rates of scalp ringworm in schoolchildren range from one percent in some schools to as high as 30% in others. African American children are at particularly high risk of this type of ringworm. One reason why younger children are more susceptible to ringworm of the scalp is that sebum, a waxy substance secreted by skin glands that protects the skin and hair against fungi and some bacteria, is not produced in humans in significant quantities until puberty. Tinea pedis, tinea cruris (jock itch), and nail infections are much more common in adults. About 3% of adult males worldwide and 1.4% of females have fungal nail infections.

Description

Ringworm is characterized by itchy patches of rough, reddened skin. Raised eruptions usually form the circular pattern that gives the condition its name. As the lesions grow, the centers start to heal. The inflamed borders expand and spread the infection. Ringworm is usually a superficial infection confined to the upper layers of the skin or scalp. In some cases, however, it can invade deeper layers of tissue, producing raised areas of boggy and reddened skin known as kerion.

Types of ringworm

Ringworm is a term that is commonly used to encompass several types of fungal infection. Sometimes, however, only body ringworm is classified as true ringworm.

Body ringworm (*tinea corporis*) can affect any part of the body except the scalp, feet, and facial area where a man's beard grows (*tinea barbae*). The well-defined flaky sores can be dry and scaly or moist and crusty.

Scalp ringworm (*tinea capitis*) is most common in children. It causes scaly, swollen blisters or a rash that looks like black dots. Sometimes inflamed and filled with pus, scalp ringworm lesions can cause crusting, flaking, and round bald patches. Most common in black children, scalp ringworm can cause scarring and permanent hair loss.

Ringworm of the groin (*tinea cruris* or jock itch) produces raised red sores with well-marked edges. It can spread to the buttocks, inner thighs, and external genitalia.

Ringworm of the nails (*tinea unguium*) generally starts at the tip of one or more toenails, which gradually thicken and discolor. The nail may deteriorate or pull away from the nail bed. Fingernail infection is far less common than toenail infection.

Risk factors

Risk factors for ringworm infections include:

- Living in a hot, humid climate or having a personal tendency to heavy sweating.
- Participating in contact sports, particularly wrestling, football, or rugby.
- Wearing closely fitted clothes or clothing made from synthetic fabrics that do not "breathe."
- Pet ownership. According to the American Academy of Family Physicians (AAFP), there are about two million cases each year in the United States of ringworm acquired from a pet cat or dog.
- Living in a college dormitory, military barracks, or other group housing situation.
- Having AIDS or any other disorder that weakens the immune system.
- Having diabetes mellitus.
- Having cuts, scrapes, or minor breaks in the skin.
- Using greasy hair gels or oils to groom the hair.
- Male sex. Men are more likely than women to contract ringworm infections, particularly athlete's foot and jock itch. Fungal nail infections are twice as common in men as in women.

Causes and symptoms

Causes

Ringworm is caused by parasitic fungi belonging to one of three genera: *Trichophyton*, *Microsporum*, or *Epidermophyton*. Humans can acquire the parasites through any of three routes of transmission: person-to-person contact, including contact with sheets, towels, sports equipment, or other personal items used by an infected person; contact with an infected animal; or contact with contaminated soil, including garden soil. *Trichophyton rubrum* and *Trichophyton tonsurans* are most commonly spread from person to person, while *Microsporum canis* is most commonly transmitted to humans from infected household pets. Cats are the most common carriers of *Microsporum canis*, but the fungus is also frequently carried by dogs, horses, pet mice and rabbits, and farm animals.

When dermatophytes are transmitted to a human or animal's skin, fur, nails, or hair, they obtain nutrients from keratin, a protein found in these tissues. The rash and other symptoms of ringworm are caused by the immune system's reaction to the metabolic byproducts of the fungi.

Symptoms

The symptoms of ringworm typically begin between 4 and 14 days after exposure and include one or more of the following:

- Itchy red patches of scaly skin that may also blister and ooze tissue fluid. The patches are often ring-shaped, with normal-appearing skin in the center of the ring.
- Nearby skin may appear darker or lighter than normal.
- Ringworm infections of the scalp or beard area (in adult males) typically produce bald spots or areas of hair loss. In some cases the hair loss may be permanent. Severe cases of ringworm on the scalp may be marked by raised nodules or pustules.
- Infections of the nails may produce thickened, discolored, and crumbly nails. In some cases the entire nail may detach from the underlying nail bed. Infections of the toenails are more common than infections of the fingernails.
- It is possible for a secondary bacterial infection to develop in areas of the body infected by the ringworm fungi, often as a result of scratching itchy areas. The person may then develop a fever along with increased reddening of the affected area, a discharge of pus, and swelling. *A doctor should be*

KEY TERMS

Dermatologist—A doctor who specializes in diagnosing and treating diseases of the skin.

Dermatophyte—The medical name for three genera of fungi that cause ringworm in humans and domestic pets. The name is derived from two Greek words that mean “skin” and “plant.”

Keratin—A type of protein that provides structure to the nails, hair, and outer layer of skin.

Kerion—A raised boggy or swollen patch of reddened skin that develops as a complication of ringworm.

Pustule—A small elevation of the skin containing pus or cloudy tissue fluid.

Sebum—An oily or waxy substance secreted by certain glands in the skin that protects hair and skin against fungi and some bacteria.

Tinea—The general medical term for a fungal infection of the skin. It is often used as a synonym for ringworm.

Wood's lamp—A special ultraviolet lamp used by dermatologists to diagnose ringworm and other skin disorders.

Zoonosis (plural, zoonoses)—Any disease that can be transmitted to humans by animals. Ringworm is a zoonosis caused by fungi.

contacted at once if these symptoms of bacterial infection develop.

Household pets carrying dermatophytes often do not have any noticeable symptoms, although they may develop circular bare patches of skin.

Diagnosis

Diagnosis of ringworm is based on a combination of patient history, an office examination, and laboratory tests.

Examination

In many cases the diagnosis of ringworm can be made by a primary care physician, but the patient may also be referred to a dermatologist (a doctor who specializes in treating skin disorders). The doctor will usually ask some questions about the patient’s living situation (including pets); school or work history; participation in sports or other outdoor activities; and any history of immune disorders.

Tests

The doctor will begin with a visual examination of the patient’s skin or other affected areas of the body. He or she may also use a Wood’s lamp, which is a special kind of ultraviolet lamp named for the doctor who invented it in 1903. The patient sits in a darkened room while the doctor shines the ultraviolet light about four or five inches away from the affected area. Normal skin or hair will not change color under the lamp. While ringworm caused by *Trichophyton tonsurans* will not fluoresce under a Wood’s lamp, ringworm caused by *Microsporum canis* will appear as blue-green or greenish patches.

Procedures

The doctor may also take a scraping of material from the affected area and dissolve it in a solution of potassium hydroxide. The resultant mixture can be examined under a microscope. When dermatophytes are present, the doctor will be able to see their spores or other characteristic structures. If it is important to identify the particular species of fungus, the doctor can use a special medium called dermatophyte test medium or DTM. A scraping of material from the patient’s hair or skin is embedded in the DTM and cultured at room temperature for 10–14 days. If dermatophytes are present, the DTM will turn bright red. Other fungi will not cause a color change.

The tests used by veterinarians to diagnose ringworm in pets are the same as those used in humans.

Treatment

Traditional

A person with body ringworm should wear loose clothing and check daily for raw, open sores. Wet dressings applied to moist sores two or three times a day can lessen inflammation and loosen scales. The doctor may suggest placing special pads between folds of infected skin, and anything the patient has touched or worn should be sterilized in boiling water. Patients should see their doctor if symptoms do not improve after four weeks of self-care.

Infected nails should be cut short and straight and carefully cleared of dead cells with an emery board.

Patients with jock itch should:

- wear cotton underwear and change it more than once a day

- keep the infected area dry
- apply antifungal ointment over a thin film of anti-fungal powder

Patients should wash their sheets, pillowcases, and pajamas every day while infected.

Drugs

Some ringworm infections disappear without treatment. Others respond to such topical antifungal medications as naftifine (Caldesene Medicated Powder) or tinactin (Desenex). Ringworm that covers large areas of the body is usually treated with either prescription topical or oral medications. Topical prescription drugs include butenafine (Mentax), ciclopirox (Loprox), miconazole (Monistat-Derm), oxiconazole (Oxistat), or terbinafine (Lamisil). Oral medications for ringworm include traconazole (Sporanox), fluconazole (Diflucan), and ketoconazole (Nizoral). Medications should be continued for two weeks after lesions disappear.

Oral medications for ringworm do have side effects, the most common of which are digestive upsets, abnormal liver functioning, and skin **rashes**. In addition, people taking these drugs should avoid taking **antacids** for **indigestion** or peptic ulcer disease during treatment for ringworm, as antacids interfere with the effectiveness of oral antifungal drugs.

Shampoo containing selenium sulfide can help prevent spread of scalp ringworm, but prescription shampoo or oral medication is usually needed to cure hair or scalp infections. Ketoconazole is particularly effective in treating ringworm of the hair or scalp.

The doctor will also prescribe oral **antibiotics** if the patient has developed a secondary bacterial infection.

Pets with ringworm are treated with many of the same medications used to treat the fungi in humans, particularly terbinafine and fluconazole. The veterinarian will also often recommend trimming or clipping the pet's fur during treatment. Close shaving is not recommended, however, because of the risk of causing breaks or small cuts in the cat or dog's skin. Another treatment for infected pets is twice-weekly dips in a diluted solution of lime sulfur over a three- to eight-week period to eliminate the fungal spores.

Alternative

The fungal infection ringworm can be treated with homeopathic remedies. Among the homeopathic remedies recommended are:

- *sepia* for brown, scaly patches
- *tellurium* for prominent, well-defined, reddish sores

- *graphites* for thick scales or heavy discharge
- *sulphur* for excessive itching.

Topical applications of antifungal herbs and essential oils also can help resolve ringworm. Tea tree oil (*Melaleuca spp.*), thuja (*Thuja occidentalis*), and lavender (*Lavandula officinalis*) are the most common. Two drops of essential oil in 1/4 oz of carrier oil is the dose recommended for topical application. Essential oils should not be applied to the skin undiluted. Botanical medicine can be taken internally to enhance the body's immune response. A person must be susceptible to exhibit this overgrowth of fungus on the skin. Echinacea (*Echinacea spp.*) and astragalus (*Astragalus membranaceus*) are the two most common immune-enhancing herbs. A well-balanced diet, including protein, complex carbohydrates, fresh fruits and vegetables, and good quality fats, is also important in maintaining optimal immune function. Alternative treatments should be used with care, as the benefits of many such treatments have not been confirmed by scientific research.

Prognosis

Ringworm can usually be cured but recurrence is common. Chronic infection develops in one patient in five. Patients with weakened immune systems may develop invasive dermatophyte infections that are difficult to treat.

It can take six to 12 months for new hair to cover bald patches, and three to 12 months to cure infected fingernails. Toenail infections do not always respond to treatment.

Prevention

The following precautions may help to lower the risk of dermatophyte infections:

- Maintain good personal hygiene, including frequent handwashing.
- Do not share towels, bedding, sports equipment, hair brushes, or other similar items.
- Stay cool and dry during hot, sticky weather or when traveling to tropical climates. Wear cotton, linen, or other natural fabrics that absorb perspiration rather than holding it against the body, and wear loose rather than closely fitted garments.
- Make sure household pets have regular veterinary checkups, and vacuum the household regularly so that shed fur does not accumulate.
- Keep common areas in the house or school clean; be particularly careful about the cleanliness of locker rooms, gyms, and swimming pools.

- Notify local public health authorities if there is an outbreak of ringworm in your child's school or day-care center.
- Use a dilute solution of chlorine bleach (1/4 cup per gallon of water) to disinfect counter tops and other hard surfaces that are safe to bleach.

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ORGANIZATIONS

- American Academy of Dermatology (AAD), P.O. Box 4014, Schaumburg, IL, 60168, (847) 330-0230, (866) 503-SKIN, (847) 240-1859, <http://www.aad.org/>.
- American College of Sports Medicine (ACSM), P.O. Box 1440, Indianapolis, IN, 46206, (317) 637-9200, (317) 634-7817, <http://www.acsm.org/>.
- American Veterinary Medical Association (AVMA), 1931 North Meacham Rd., Suite 100, Schaumburg, IL, 60173-4360, (847) 925-8070, (847) 925-1329, avmainfo@avma.org, <http://www.avma.org/>.
- Centers for Disease Control and Prevention (CDC), 1600 Clifton Rd., Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.
- National Institute of Allergy and Infectious Diseases (NIAID), 6610 Rockledge Dr., MSC 6612, Bethesda, MD, 20892-6612, (301) 496-5717, (866) 284-4107, (301) 402-3573, <http://www3.niaid.nih.gov>.

Maureen Haggerty
Rebecca J. Frey, PhD

Rinne test see **Hearing tests with a tuning fork**

Ritonavir see **Protease inhibitors**

River blindness see **Filariasis**

RMSF see **Rocky Mountain spotted fever**

Rocky Mountain spotted fever

Definition

Rocky Mountain spotted fever (RMSF) is a tick-borne illness caused by a bacteria, resulting in a high fever and a characteristic rash.

Description

The bacteria causing RMSF is passed to humans through the bite of an infected tick. The illness begins within about two weeks of such a bite. RMSF is the most widespread tick-borne illness in the United States, occurring in every state except Alaska and Hawaii. The states in the mid-Atlantic region, the Carolinas, and the Virginias have a great deal of tick activity during the spring and summer months, and the largest number of RMSF cases come from those states. About 5% of all ticks carry the causative bacteria. Children under the age of 15 years have the majority of RMSF infections.

Causes and symptoms

The bacterial culprit in RMSF is called *Rickettsia rickettsii*. It causes no illness in the tick carrying it, and can be passed on to the tick's offspring. When a tick attaches to a human, the bacteria is passed. The tick must be attached to the human for about six hours for this passage to occur. Although prompt tick removal will cut down on the chance of contracting RMSF, removal requires great care. If the tick's head and body are squashed during the course of removal, the bacteria can be inadvertently rubbed into the tiny bite wound.

Symptoms of RMSF begin within two weeks of the bite of the infected tick. Symptoms usually begin suddenly, with high fever, chills, **headache**, severe weakness, and muscle **pain**. Pain in the large muscle of the calf is very common, and may be particularly severe. The patient may be somewhat confused and delirious. Without treatment, these symptoms may last two weeks or more.

The rash of RMSF is quite characteristic. It usually begins on the fourth day of the illness, and occurs in at least 90% of all patients with RMSF. It starts around the wrists and ankles, as flat pink marks (called macules). The rash spreads up the arms and legs, toward the chest, abdomen, and back. Unlike **rashes** which accompany various viral infections, the rash of RMSF does spread to the palms of the hands and the soles of the feet. Over a couple of days, the macules turn a reddish-purple color. They are now called petechiae, which are tiny areas of bleeding under the skin (pinpoint hemorrhages). This signifies a new phase of the illness. Over the next several days, the individual petechiae may spread into each other, resulting in larger patches of hemorrhage.

The most severe effects of RMSF occur due to damage to the blood vessels, which become leaky. This accounts for the production of petechiae. As blood and fluid leak out of the injured blood vessels, other tissues and organs may swell and become damaged, and:

- breathing difficulties may arise as the lungs are affected.
- heart rhythms may become abnormal
- kidney failure occurs in very ill patients
- liver function drops
- the patient may experience nausea, vomiting, abdominal pain, and diarrhea
- the brain may swell (encephalitis) in about 25% of all RMSF patients (brain injury can result in seizures, changes in consciousness, actual coma, loss of coordination, imbalance on walking, muscle

spasms, loss of bladder control, and various degrees of paralysis)

- the clotting system becomes impaired, and blood may be evident in the stools or vomit

Diagnosis

Diagnosis of RMSF is almost always made on the basis of the characteristic symptoms, coupled with either a known tick bite (noted by about 60–70% of patients) or exposure to an area known to harbor ticks. Complex tests exist to nail down a diagnosis of RMSF, but these are performed in only a few laboratories. Because the results of these tests take so long to obtain, they are seldom used. This is because delaying treatment is the main cause of **death** in patients with RMSF.

Treatment

It is essential to begin treatment absolutely as soon as RMSF is seriously suspected. Delaying treatment can result in death.

Antibiotics are used to treat RMSF. The first choice is a form of tetracycline; the second choice (used in young children and pregnant women) is chloramphenicol. If the patient is well enough, treatment by oral intake of medicine is perfectly effective. Sicker patients will need to be given the medication through a needle in the vein (intravenously). Penicillin and sulfa drugs are not suitable for treatment of RMSF, and their use may increase the death rate by delaying the use of truly effective medications.

Very ill patients will need to be hospitalized in an intensive care unit. Depending on the types of complications a particular patient experiences, a variety of treatments may be necessary, including intravenous fluids, blood transfusions, anti-seizure medications, **kidney dialysis**, and mechanical ventilation (a breathing machine).

Alternative treatment

Although alternative treatments should never be used in place of conventional treatment with antibiotics, they can be useful adjuncts to antibiotic therapy. The use of *Lactobacillus acidophilus* and *L. bifidus* supplementaion during and after antibiotic treatment can help rebalance the intestinal flora. **Acupuncture**, homeopathy, and botanical medicine can all be beneficial supportive therapies during recovery from this disease.

KEY TERMS

Encephalitis—Inflammation of the tissues of the brain.

Macule—A flat, discolored area on the skin.

Petechia—A small, round, reddish purple spot on the skin, representing a tiny area of bleeding under the skin.

Prognosis

Prior to the regular use of antibiotics to treat RMSF, the death rate was about 25%. Although the death rate from RMSF has improved greatly with an understanding of the importance of early use of antibiotics, there is still a 5% death rate. This rate is believed to be due to delays in the administration of appropriate medications.

Certain risk factors suggest a worse outcome in RMSF. Death rates are higher in males and increase as people age. It is considered a bad prognostic sign to develop symptoms of RMSF within only two to five days of a tick bite.

Prevention

The mainstay of prevention involves avoiding areas known to harbor ticks. However, because many people enjoy recreational activities in just such areas, the following steps can be taken:

- Wear light colored clothing (so that attached ticks are more easily noticed).
- Wear long sleeved shirts and long pants; tuck the pants legs into socks.
- Spray clothing with appropriate tick repellents.
- Examine. Anybody who has been outside for any amount of time in an area known to have a population of ticks should examine his or her body carefully for ticks. Parents should examine their children at the end of the day.
- Remove any ticks using tweezers, so that infection doesn't occur due to handling the tick. Grasp the tick's head with the tweezers, and pull gently but firmly so that the head and body are entirely removed.
- Keep areas around homes clear of brush, which may serve to harbor ticks.

ORGANIZATIONS

Centers for Disease Control and Prevention, 1600 Clifton Rd., NE, Atlanta, GA, 30333, (404) 639-3311, (800) 311-3435, <http://www.cdc.gov>.

Rosalyn Carson-DeWitt, MD

Rogaine see **Minoxidil**

Rolfing

Definition

Rolfing, also called Rolf therapy or structural integration, is a holistic system of bodywork that uses deep manipulation of the body's soft tissue to realign and balance the body's myofascial structure. Rolfing improves posture, relieves chronic **pain**, and reduces **stress**.

Purpose

Rolfing helps to improve posture and bring the body's natural structure into proper balance and alignment. This can bring relief from general aches and pains, improve breathing, increase energy, improve self-confidence, and relieve physical and mental stress. Rolfing has also been used to treat such specific physical problems as chronic back, neck, shoulder, and joint pain, and repetitive stress injuries, including **carpal tunnel syndrome**. Many amateur and professional athletes, including Olympic skaters and skiers, use Rolfing to keep in top condition, to prevent injuries, and to more quickly recover from injuries.

Description

Origins

Ida Pauline Rolf (1896–1979) was a biochemist from New York who developed structural integration over the course of many years after an accident as a young woman. She was kicked by a horse's hoof on a trip out West and developed symptoms resembling those of acute **pneumonia**. She made her way to a hospital in Montana, where she was treated by a physician who called in an osteopath to assist in her treatment. After the osteopath treated her, she was able to breathe normally. After her return to New York, her mother took her to a blind osteopath for further treatment. He taught her about the body's structure and function, after which Rolf became

IDA P. ROLF, PH.D. (1896–1979)

Born in New York City and raised in the Bronx, Ida P. Rolf attended school in the New York area, graduating from Barnard College in 1916. In 1920, she graduated from the Columbia University College of Physicians and Surgeons with a doctorate in biological chemistry. For the next 12 years, she worked in the departments of chemotherapy and organic chemistry at the Rockefeller Institute. During an extended leave of absence, she studied atomic physics and mathematics at the Swiss Technical University in Zurich and homeopathic medicine in Geneva. During the 1930s, she studied osteopathy, chiropractic medicine, tantric yoga, the Alexander Technique of tension reduction through body movement, and the philosophy of altered states of consciousness of Alfred H.S. Korzybski.

Her interest in body structure, movement, and manipulation began after being kicked by a horse shortly after graduating from Barnard. The accident left her with acute pneumonia. Dissatisfied with conventional medical treatment, she began her quest for more natural and effective ways of treating the body.

By 1940, Dr. Rolf had developed a technique of body movement she called structural integration, also known today as Rolfing. The therapy reshapes the body's muscular structure by applying pressure and energy, freeing the body from physical and emotional traumas. In 1977, she authored *Rolfing: The Integration of Human Structures*. She continued to teach and refine her therapy until her death in 1979. Dr. Rolf's desire to teach her work to others led to her establishing the Guild for Structural Engineering, now known as the Rolf Institute of Structural Integration: <http://www.rolf.org/>

dissatisfied with conventional medical treatment. Following completion of a doctorate in biochemistry from Columbia University in 1920, Rolf studied atomic physics, mathematics, and **homeopathic medicine** in Europe. After 1928, when her father died and left her an inheritance that allowed her to pursue her own studies, she explored various forms of alternative treatment, including **osteopathy**, **chiropractic** medicine, tantric **yoga**, the **Alexander technique** of tension reduction through body movement, and Alfred Korzybski's philosophy of altered states of consciousness.

By 1940, Rolf had synthesized what she had learned from these various disciplines into her own technique of body movement that she called structural integration, which later became known as

Rolfing. During the Second World War, Rolf continued to study with an osteopath in California named Amy Cochran. In the mid-1960s, Gestalt therapist Fritz Perls invited Rolf to Esalen, where she began to develop a following among people involved in the human potential movement. In 1977, she published *Rolfing: The Integration of Human Structures*, the definitive book on structural integration bodywork. She continued to refine the therapy until her **death** in 1979. Rolf's work is carried on through her Guild for Structural Integration, now known as the Rolf Institute of Structural Integration, which she founded in 1971 in Boulder, Colo.

Rolfing is more than just a massage of the body's surface. It is a system that reshapes the body's myofascial structure by applying pressure and energy, thereby freeing the body from the effects of physical and emotional traumas. Although Rolfing is used extensively to treat **sports injuries** and back pain, it is not designed as a therapy for any particular condition. Rather, it is a systematic approach to overall wellness. It works by counteracting the effects of gravity, which over time pulls the body out of alignment. This pull causes the body's connective tissue to become harder and stiffer, and the muscles to atrophy. Signs of this stiffening and contraction include slouching or an overly erect posture.

Rolfing identifies the vertical line as the ideal that the body should approximate. The mission statement of the Guild for Structural Integration describes Rolfing as "a method and a philosophy of personal growth and integrity...The vertical line is our fundamental concept. The physical and psychological embodiment of the vertical line is a way of Being in the physical world [that] forms a basis for personal growth and integrity."

The basic ten

Basic Rolfing treatment consists of 10 sessions, each lasting 60–90 minutes and costing about \$100 each. The sessions are spaced a week or longer apart. After a period of integration, specialized or advanced treatment sessions are available. A "tuneup" session is recommended every six months. In each session, the Rolfer uses his or her fingers, hands, knuckles, and elbows to rework the connective tissue over the entire body. The tissues are worked until they become pliable, allowing the muscles to lengthen and return to their normal alignment. The deep tissue manipulation improves posture and agility, and increases the body's range

KEY TERMS

Atrophy—A progressive wasting and loss of function of any part of the body.

Carpal tunnel syndrome—A condition caused by compression of the median nerve in the carpal tunnel of the hand, characterized by pain.

Fascia—The sheet of connective tissue that covers the body under the skin and envelops every muscle, bone, nerve, gland, organ, and blood vessel. Fascia helps the body to retain its basic shape.

Osteopathy—A system of medical practice that believes that the human body can make its own

remedies to heal infection. It originally used manipulative techniques but also added surgical, hygienic, and medicinal methods when needed.

Parasympathetic nervous system—A part of the autonomic nervous system that is concerned with conserving and restoring energy. It is the part of the nervous system that predominates in a state of relaxation.

Structural integration—The term used to describe the method and philosophy of life associated with Rolfing. Its fundamental concept is the vertical line.

of movement. Rolfers also believe that the blocked energy accumulated in the tissue from emotional tension is released through Rolfing treatment, causing the patient to feel more energetic and have a more positive frame of mind.

Clients are asked to wait for a period of six to 12 months before scheduling advanced work, known as the PostTen/Advanced Series. This period allows the body to integrate the work done in the “Basic Ten.”

Rolfing movement integration

Rolfing movement integration, or RMI, is intended to help clients develop better awareness of their vertical alignment and customary movement patterns. They learn to release tension and discover better ways to use body movement effectively.

Rolfing rhythms

Rolfing rhythms are a series of exercises intended to remind participants of the basic principles of Rolfing: ease, length, balance, and harmony with gravity. In addition, Rolfing rhythms improve the client's flexibility as well as muscle tone and coordination.

Preparations

No pre-procedure preparations are needed to begin Rolfing treatment. The treatment is usually done on a massage table with the patient wearing only undergarments. Prior to the first session, however, the client is asked to complete a health questionnaire, and photographs are taken to assist with evaluation of his or her progress.

Precautions

Since Rolfing involves vigorous deep tissue manipulation, it is often described as uncomfortable and sometimes painful, especially during the first several sessions. In the past decade, however, Rolfers have developed newer techniques that cause less discomfort to participants. Since Rolfing is a bodywork treatment that requires the use of hands, it may be a problem for people who do not like or are afraid of being touched. It is not recommended as a treatment for any disease or a chronic inflammatory condition such as arthritis, and can worsen such a condition. Anyone with a serious medical condition, including heart disease, diabetes, or respiratory problems, should consult with a medical practitioner before undergoing Rolfing.

Side effects

There are no reported serious side effects associated with Rolfing when delivered by a certified practitioner to adults and juveniles.

Research and general acceptance

There is a growing amount of mainstream scientific research documenting the effectiveness of Rolf therapy. A 1988 study published in the *Journal of the American Physical Therapy Association* indicated that Rolfing stimulates the parasympathetic nervous system, which can help speed the recovery of damaged tissue. Other studies done in the 1980s concerned the effectiveness of Rolfing in treating figure skaters and children with **cerebral palsy**. In 1992 a presentation was made to the National Center of Medical **Rehabilitation** Research regarding Rolfing in the treatment of degenerative joint disease. A 1997 article in *The Journal of Orthopedic*

and Sports Physical Therapy reported that Rolfing can provide effective and sustained pain relief from lower back problems.

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ORGANIZATIONS

Rolf Institute of Structural Integration, 5055 Chaparral Ct. Suite 103, Boulder, CO, 80301, (303) 449-5903, (303) 449-5978, (800) 530-8875, <http://www.rolf.org>.

Ken R. Wells

Purpose

An inflamed or infected pulp is called pulpitis. It is the most common cause of a **toothache**. To relieve the **pain** and prevent further complications, the tooth may be extracted (surgically removed) or saved by root canal treatment.

Demographics

Root canal treatment has become a common dental procedure. According to the American Association of Endodontists, more than 15 million root canal treatments are performed every year in North America as of 2010, with a 97% success rate.

Description

Inside the tooth, the pulp of a tooth is comprised of soft tissue that contains the blood supply, by which the tooth receives its nutrients; and the nerve, by which the tooth senses hot and cold. This tissue is vulnerable to damage from deep dental decay, accidental injury, tooth fracture, or trauma from repeated dental procedures such as multiple fillings or restorations over time. If a tooth becomes diseased or injured, bacteria may build up inside the pulp, spreading infection from the natural crown of the tooth to the root tips in the jawbone. Pus

Root canal treatment

Definition

Root canal treatment, also known as endodontic treatment, is a dental procedure in which the diseased or damaged pulp (central core) of a tooth is removed and the inside areas (the pulp chamber and root canals) are filled and sealed.

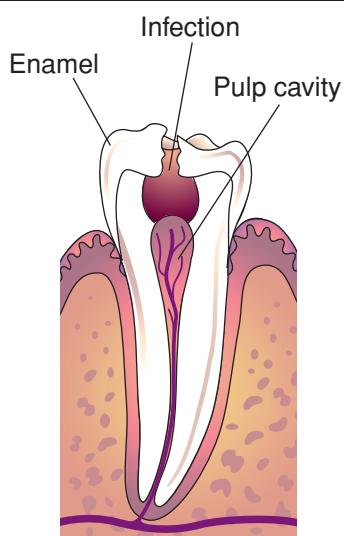


Figure A



Figure B



Figure C

Root canal treatment is a dental procedure in which the diseased pulp of a tooth is removed and the inside areas are filled and sealed. In figure A, the infection can be seen above the pulp cavity. The dentist drills into the enamel and the pulp cavity is extracted (figure B). Finally, the dentist fills the pulp cavity with antibiotic paste and a temporary filling (figure C). (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

KEY TERMS

Abscess—A cavity or space in tooth or gum tissue filled with pus as the result of infection. Its swelling exerts pressure on the surrounding tissues, causing pain.

Apicoectomy—Also called root resectioning. The root tip of a tooth is accessed in the bone and a small amount is shaved away. The diseased tissue is removed and a filling is placed to reseal the canal.

Crown—The natural crown of a tooth is that part of the tooth covered by enamel. Also, a restorative crown is a protective shell that fits over a tooth.

Endodontic—Pertaining to the inside structures of the tooth, including the dental pulp and tooth root, and the periapical tissue surrounding the root.

Endodontist—A dentist who specializes in the diagnosis and treatment of disorders affecting the inside structures of teeth.

Extraction—The surgical removal of a tooth from its socket in a bone.

Gutta percha—An inert, latex-like substance used for filling root canals.

Pulp—The soft innermost layer of a tooth, containing blood vessels and nerves.

Pulp chamber—The area within the tooth occupied by dental pulp.

Pulpitis—Inflammation of the pulp of a tooth involving the blood vessels and nerves.

Root canal—The space within a tooth that runs from the pulp chamber to the tip of the root.

Root canal treatment—The process of removing diseased or damaged pulp from a tooth, then filling and sealing the pulp chamber and root canals.

Smear layer—A layer of organic and inorganic material produced on teeth by dental instrumentation that may also contain bacteria and their by-products.

accumulating at the ends of the roots can form a painful **abscess** that can damage the bone supporting the teeth. Such an infection may produce pain that is severe, constant, or throbbing. It can also result in prolonged sensitivity to heat or cold, swelling, and tenderness in the surrounding gums, facial swelling, or discoloration of the tooth. In some cases, however, the pulp may die so gradually that there is little noticeable pain.

Root canal treatment is performed under **local anesthesia**. A thin sheet of rubber called a rubber dam is placed in the mouth and around the base of the tooth to isolate the tooth and help to keep the operative field dry. The dentist removes any **tooth decay** and makes an opening through the natural crown of the tooth into the pulp chamber. Creating this access also relieves the pressure inside the tooth and can dramatically ease pain.

The dentist determines the length of the root canals, usually with a series of x rays. Small wire-like files are then used to clean the entire canal space of diseased pulp tissue and bacteria. The debris is flushed out with large amounts of water (irrigation). The canals are also slightly enlarged and shaped to receive an inert (non-reactive) filling material called gutta percha. However, the tooth is not filled and permanently sealed until it is completely free of active infection. The dentist may place a temporary seal, or leave the tooth open to drain, and prescribe an antibiotic to counter any spread of infection from the tooth. This is why root canal treatment may require several visits to the dentist.

Once the canals are completely clean, they are filled with gutta percha and a sealer cement to prevent bacteria from entering the tooth in the future. A metal post may be placed in the pulp chamber for added structural support and better retention of the crown restoration. The tooth is protected by a temporary filling or crown until a permanent restoration may be made. This restoration is usually a gold or porcelain crown, although it may be a gold inlay, or an amalgam or composite filling (paste fillings that harden).

The use of lasers to perform root canal therapy is a recent but controversial innovation, although it has been approved by the Food and Drug Administration (FDA). In theory, the beam of intense light from the erbium laser that the dentist uses melts the debris (also called the smear layer) inside the tooth, cleansing it completely. It is possible, however, for a laser beam to miss some of the infection within the tooth, particularly if the root canal itself is unusually shaped or has a number of small crevices. In addition, the use of lasers in root canal treatment has been reported to occasionally damage the tooth.

Diagnosis/Preparation

Signs that a root canal treatment is necessary include severe pain while chewing, prolonged sensitivity

to heat or cold, or a darkening of the tooth. Swelling and tenderness of the gums or pimples appearing on the gums are also common symptoms. However, it is also possible that no symptoms will be noticed. The dentist will take an x ray of the tooth to determine if there is any sign of infection in the surrounding bone.

Aftercare

Once a root canal treatment is performed, the recipient must have a crown placed over the tooth to protect it. The cost of the treatment and the crown may be expensive. However, replacing an extracted tooth with a fixed bridge, a removable partial denture, or an implant to maintain the space and restore the chewing function is typically even more expensive.

During the time when **antibiotics** are being used, care should be taken to avoid using the tooth to chew food. The tooth has been structurally weakened and may break, or there is a possibility of the interior of the tooth becoming reinfected.

If the tooth feels sensitive following the procedure, a standard over-the-counter pain medication such as ibuprofen or naproxen may be taken. This sensitivity will fade after a few days. In most cases the patient can resume regular activity the following day.

Risks

There is a possibility that a root canal treatment will not be successful the first time. If infection and inflammation recur and an x ray indicates a repeat treatment is feasible, the old filling material is removed and the canals are thoroughly cleaned out. The dentist will try to identify and correct problems with the first root canal treatment before filling and sealing the tooth a second time.

In cases where an x ray indicates that another root canal treatment cannot correct the problem, endodontic surgery may be performed. In a procedure called an apicoectomy, or root resectioning, the root end of the tooth is accessed in the bone, and a small amount is shaved away. The area is cleaned of diseased tissue and a filling is placed to reseal the canal.

Normal results

With successful root canal treatment, the tooth will no longer cause pain. However, because it does not contain an internal nerve, it no longer has sensitivity to hot, cold, or sweets. Because these are signs

of dental decay, the root canal recipient must receive regular dental check-ups with periodic x rays to avoid further disease in the tooth. The restored tooth may last a lifetime. However, with routine wear, the filling or crown may eventually need to be replaced.

Morbidity and mortality rates

About 5% of patients will experience persistent pain after root canal therapy. In some cases, despite proper root canal treatment and endodontic surgery, the tooth dies and must be extracted. This outcome, however, is relatively uncommon.

Alternatives

The only alternative to performing a root canal procedure is to extract the diseased tooth. After restoration or extraction, the two main goals are to allow normal chewing and to maintain proper alignment and spacing between teeth. A fixed bridge, a removable partial denture or an implant will accomplish both goals. However, these are usually more expensive than a root canal treatment.

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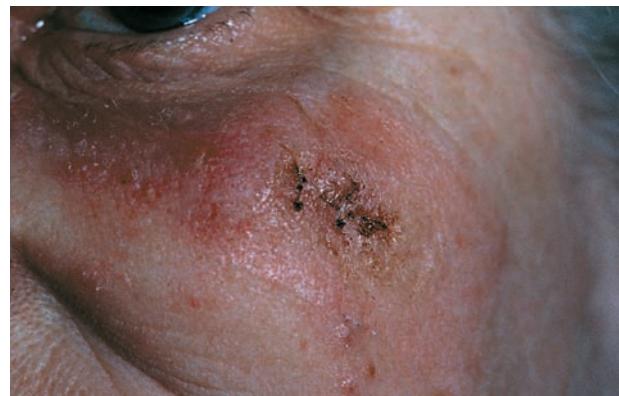
Academy of General Dentistry, 211 East Chicago Avenue, Chicago, IL, 60611, (312) 440-4300, <http://www.agd.org>.

American Academy of Pediatric Dentistry, 211 East Chicago Avenue, #700, Chicago, IL, 60611-2663, (312) 337-2169, (312) 337-6329, <http://www.aapd.org>.

American Association of Endodontists, 211 E. Chicago Ave., Suite 1100, Chicago, IL, 60611-2691, (312) 266-7255, (312) 266-9867, (866) 451-9020, (800) 872-3636, info@aae.org, <http://www.aae.org>.

American Dental Association, 211 E. Chicago Avenue, Chicago, IL, 60611, (312) 440-2500, (312) 440-7494, <http://www.ada.org>.

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Rosacea on a woman's cheek. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

The similarity in appearance of rosacea to **acne** led people in the past to erroneously call the disease acne rosacea or adult acne. Like acne, the skin can have pimples and papules. Unlike acne, however, people with rosacea do not have blackheads.

In early stages of rosacea, people typically experience repeated episodes of flushing. Later, areas of the face are persistently red, telangiectasia appear on the nose and cheeks, as well as inflamed papules and pustules. Over time, the skin may take on a roughened, orange peel texture. Very late in the disorder, a small group of patients with rosacea will develop rhinophyma, which can give the nose a bulb-like look.

Up to one half of patients with rosacea may experience symptoms related to their eyes. Ocular rosacea, as it is called, frequently precedes the other manifestations on the skin. Most of these eye symptoms do not threaten sight, however. Telangiectasia may appear around the borders of the eyelid, the eyelids may be chronically inflamed, and small lumps called chalazions may develop. The cornea of the eye, the transparent covering over the lens, can also be affected, and in some cases vision will be affected.

Causes and symptoms

There is no known specific cause of rosacea. A history of redness and flushing precedes the disease in most patients. The consensus among many experts is that multiple factors may lead to an overreaction of the facial blood vessels, which triggers flushing. Over time, persistent episodes of redness and flushing leave the face continually inflamed. Pimples and blood-vessel changes follow.

Certain genetic factors may also come into play, although these have not been fully described. The

Rosacea

Definition

Rosacea is a skin disease typically appearing in people during their 30s and 40s. It is marked by redness (erythema) of the face, flushing of the skin, and the presence of hard pimples (papules) or pus-filled pimples (pustules), and small visible spider-like veins called telangiectasias. In later stages of the disease, the face may swell and the nose may take on a bulb-like appearance called rhinophyma.

Description

Rosacea produces redness and flushing of the skin, as well as pustules and papules. Areas of the face, including the nose, cheeks, forehead, and chin, are the primary sites, but some people experience symptoms on their necks, backs, scalp, arms, and legs.

disease is more common in women and light-skinned, fair-haired people. It may be more common in people of Celtic background, although this is an area of disagreement among experts.

Certain **antibiotics** are useful in the treatment of rosacea, leading some researchers to suspect a bacterium or other infectious agent may be the cause. One of the newest suspects is a bacterium called *Helicobacter pylori*, which has been implicated in causing many cases of stomach ulcers but the evidence here is mixed.

Other investigators have observed that a particular parasite, the mite *Demodex folliculorum*, can be found in areas of the skin affected by rosacea. The mite can also be detected, however, in the skin of people who do not have the disease. It is likely that the mite does not cause rosacea, but merely aggravates it.

Diagnosis

Diagnosis of rosacea is made by the presence of clinical symptoms. There is no specific test for the disease. Episodes of persistent flushing, redness (erythema) of the nose, cheeks, chin, and forehead, accompanied by pustules and papules are hallmarks of the disease. A dermatologist will attempt to rule out a number of other diseases that have similar symptoms. Acne vulgaris is perhaps the disorder most commonly mistaken for rosacea, but redness and spider-like veins are not observed in patients with acne. Blackheads and cysts, however, are seen in acne patients, but not in those with rosacea.

Other diseases that produce some of the same symptoms as rosacea include perioral **dermatitis** and **systemic lupus erythematosus**.

Treatment

The mainstay of treatment for rosacea is oral antibiotics. These appear to work by reducing inflammation in the small blood vessels and structure of the skin, not by destroying bacteria that are present. Among the more widely used oral antibiotics is tetracycline. In many patients, antibiotics are effective against the papules and pustules that can appear on the face, but they appear less effective against the background redness, and they have no effect on telangiectasia. Patients frequently take a relatively high dose of antibiotics until their symptoms are controlled, and then they slowly reduce their daily dose to a level that just keeps their symptoms in check. Other oral antibiotics used include erythromycin and minocycline.

Some patients are concerned about long-term use of oral antibiotics. For them, a topical agent applied directly to the face may be tried in addition to an oral antibiotic, or in its place. **Topical antibiotics** are also useful for controlling the papules and pustules of rosacea, but do not control the redness, flushing, and telangiectasias. The newest of these topical agents is metronidazole gel, which can be applied twice daily. Like the oral antibiotics, topical preparations appear to work by reducing inflammation, not by killing bacteria.

Vitamin A derivatives, called retinoids, also appear useful in the treatment of rosacea. An oral retinoid, called isotretinoin, which is used in severe cases of acne also reduces the pustules and papules in severe cases of rosacea that do not respond to antibiotics. Isotretinoin must be taken with care, however, particularly in women of childbearing age. They must agree to a reliable form of **contraception**, because the drug is known to cause **birth defects**.

Topical vitamin A derivatives that are used in the treatment of acne also may have a role in the treatment of rosacea. Accumulating evidence suggests that topical isotretinoin and topical azelaic acid can reduce the redness and pimples. Some patients who use these medications experience skin irritation that tends to resolve with time.

For later stages of the disorder, a surgical procedure may be needed to improve the appearance of the skin. To remove the telangiectasias, a dermatologist may use an electrocautery device to apply a current to the blood vessel in order to destroy it. Special lasers, called tunable dye lasers, can also be adjusted to selectively destroy these tiny blood vessels.

A variety of surgical techniques can be used to improve the shape and appearance of a bulbous nose in the later stages of the disease. Surgeons may use a scalpel or laser to remove excess tissue from the nose and restore a more natural appearance.

Alternative treatment

Alternative treatments have not been extensively studied in rosacea. Some reports advocate gentle circular massage for several minutes daily to the nose, cheeks, and forehead. Scientifically controlled studies are lacking, however.

Many people are able to avoid outbreaks by reducing things that trigger flushing. Alcoholic beverages, hot beverages, and spicy foods are among the more common factors in the diet that can provoke flushing. Reducing or eliminating these items in the diet can help limit rosacea outbreaks in many people.

KEY TERMS

Blackhead—A plug of fatty cells capped with a blackened mass.

Erythema—A diffuse red and inflamed area of the skin.

Papule—A small hard elevation of the skin.

Pustule—A small pus-filled elevation of the skin.

Retinoid—A synthetic vitamin A derivative used in the treatment of a variety of skin disorders.

Rhinophyma—Long-term swelling and overgrowth in skin tissue of the nose that leaves it with a knobby bulb-like look.

Telangiectasia—Small blood veins visible at the surface of the skin of the nose and cheeks.

Exposure to heat, cold, and sunlight are also known triggers of flushing. The specific things that provoke flushing vary considerably from person to person, however. It usually takes some trial and error to figure these out.

A deficiency in hydrochloric acid (HCl) in the stomach may be a cause of rosacea, and supplementation with HCl capsules may bring relief in some cases.

Prognosis

The prognosis for controlling symptoms of rosacea and improving the appearance of the face is good. Many people require life-long treatment and achieve good results. There is no known cure for the disorder.

Prevention

Rosacea cannot be prevented but once correctly diagnosed, outbreaks can be treated and repeated episodes can be limited.

Use mild soaps

Avoiding anything that irritates the skin is a good preventive measure for people with rosacea. Mild soaps and cleansers are recommended. Astringents and alcohol should be avoided.

Learn what triggers flushing

Reducing factors in the diet and environment that cause flushing of the face is another good preventive strategy. Alcoholic and hot beverages, and spicy foods are among the more common triggers.

Use sunscreen

Limiting exposure of the face to excesses of heat and cold can also help. A sunscreen with a skin protection factor (SPF) of 15 or greater used daily can limit the damage to the skin and small blood vessels caused by the sun, and reduce outbreaks.

ORGANIZATIONS

American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168-4014, (847) 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

National Rosacea Society, 198 James St/, Barrington, IL, 60010, (888) 662-5874, rosaceas@aol.com, <http://www.rosacea.org>.

Richard H. Camer

Rosary bead esophagus see **Diffuse esophageal spasm**

Roseola

Definition

Roseola is a common disease of babies or young children, in which several days of very high **fever** are followed by a rash.

Description

Roseola is an extraordinarily common infection, caused by a virus. About 90% of all children have been exposed to the virus, with about 33% actually demonstrating the syndrome of fever followed by rash.

The most common age for a child to contract roseola is between six and twelve months. Roseola infection strikes boys and girls equally. The infection may occur at any time of year, although late spring and early summer seem to be peak times for it.

Causes and symptoms

About 85% of the time, roseola is caused by a virus called Human Herpesvirus 6, or HHV-6. Although the virus is related to those herpesviruses known to cause



Roseola rash on infant's back. (© PHOTOTAKE Inc./Alamy.)

sores on the lips or genitalia, HHV-6 causes a very different type of infection. HHV-6 is believed to be passed between people via infected saliva. A few other viruses (called enteroviruses) can produce a similar fever-then-rash illness, which is usually also called roseola.

Researchers believe that it takes about 5–15 days to develop illness after having been infected by HHV-6. Roseola strikes suddenly, when a previously-well child spikes an impressively high fever. The temperature may reach 106°F. As is always the case with sudden fever spikes, the extreme change in temperature may cause certain children to have seizures. About 5–35% of all children with roseola will have these “febrile seizures.”

The most notable thing about this early phase of roseola is the absence of symptoms, other than the high fever. Although some children have a slightly reddened throat, or a slightly runny nose, most children have no symptoms whatsoever, other than the sudden development of high fever. This fever lasts for between three and five days.

KEY TERMS

Jaundice—The development of a yellowish tone to the skin and the whites of the eyes, caused by poor liver function.

Mononucleosis—An infection which causes swelling of lymph nodes, spleen, and liver, usually accompanied by extremely sore throat, fever, headache, and intense long-lasting fatigue.

Somewhere around the fifth day, a rash begins on the body. The rash is usually composed of flat pink patches or spots, although there may be some raised patches as well. The rash usually starts on the chest, back, and abdomen, and then spreads out to the arms and neck. It may or may not reach the legs and face. The rash lasts for about three days, then fades.

Very rarely, roseola will cause more serious disease. Patients so afflicted will experience significant swelling of the lymph nodes, the liver, and the spleen. The liver may become sufficiently inflamed to interfere with its functioning, resulting in a yellowish color to the whites of the eyes and the skin (**jaundice**). This syndrome (called a mononucleosis-like syndrome, after the disease called mononucleosis that causes many of the same symptoms) has occurred in both infants and adults.

Diagnosis

The diagnosis of roseola is often made by carefully examining the feverish child to make sure that other illnesses are not causing the temperature spike. Once it is clear that no **pneumonia**, ear infection, **strep throat**, or other common childhood illness is present, the practitioner usually feels comfortable waiting to see if the characteristic rash of roseola begins.

Treatment

There are no treatments available to stop the course of roseola. **Acetaminophen** or ibuprofen is usually given to try to lower the fever. Children who are susceptible to seizures may be given a sedative medication when the fever first spikes, in an attempt to prevent such a seizure.

Prognosis

Children recover quickly and completely from roseola. The only complications are those associated with seizures, or the rare mononucleosis-like syndrome.

Prevention

Other than the usual good hygiene practices always recommended to decrease the spread of viral illness, no methods are available to specifically prevent roseola.

Resources

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Rosalyn Carson-DeWitt, MD

Roseola infantum see **Roseola**

Ross River Virus

Definition

Ross River Virus (RRV) is Australia's most common and widespread mosquito-borne pathogen. Also known as RRV disease, it can cause debilitating polyarthritides, rash, **fever**, and constitutional symptoms.

Description

Originally known as epidemic polyarthritides, RRV is a member of the *Togaviridae* family of arboviruses. RRV is transmitted in an animal host-vector-human cycle, where the vector is the mosquito. Serological investigations have indicated that native macropods are the main vertebrate hosts of RRV, although other animals can become infected as well. The RRV lives in the blood stream of an infected animal. When a mosquito feeds on the infected animal, the virus is transmitted to the insect where it rapidly multiplies. The virus is then passed onto the next animal or person the mosquito **bites**. It has been proposed that human-mosquito-human transmission can occur during RRV epidemics. One-third of all humans bitten by an infected mosquito will develop the RRV disease.

The RRV disease occurs throughout continental Australia. However, the majority of RRV infections occur in the northern states and along coastal areas; in particular, the state of Queensland. Of the 4,800 cases reported annually in Australia, approximately 2,700 of these occur in Queensland. In addition to these cases, many more go unreported. Infection can occur year round, but outbreaks typically coincide with the increased mosquito activity of the wet season (between late November and the end of April). Also, areas with

intensive irrigation and those near salt marches have higher mosquito populations, and, thus, tend to exhibit a higher number of RRV cases.

In addition to continental Australia, RRV is endemic to the Solomon Islands, East Timor, Papua New Guinea, and the adjacent islands of Indonesia. Epidemics have also been reported in the Cook Islands, Fiji, French Polynesia, New Caledonia, and Western Samoa.

Causes and symptoms

Many people that are infected with RRV will never develop symptoms. However, 25% to 45% of cases will develop symptoms within three days to three weeks (averaging nine days) of the infection. Symptoms will vary between patients, but typically include arthralgia, arthritis, myalgia, skin rash, fever, **fatigue**, **headache**, and swollen lymph nodes. **Tingling** and **pain** in the palms of the hands and soles of the feet can accompany these symptoms. Other, less frequent, symptoms can include general malaise, **nausea**, sore eyes, and **sore throat**.

Most patients with RRV disease (83% to 98%) experience symptoms of polyarthritides involving the wrists, knees, ankles, and small joints of the extremities. Less frequently affected joints include the elbows, toes, tarsal joints, vertebral joints, shoulders, and hips. Symptoms can range from restricted joint movement to prominent swell and severe pain. Although severe joint pain can last for only 2 to 6 weeks, over half of patients will continue to experience joint pain for 6 to 12 months after RRV infection. Symptoms typically diminish over time, but relapses are common and have been known to persist for several years. This persistent polyarthritides can lead to fatigue and myalgia, contributing to RRV diseases high morbidity.

Diagnosis

Diagnosis usually consists of serological tests to determine the presence and increase of RRV antibodies. Samples should be taken during the acute or convalescent stages of the illness. Testing will help clinicians differentiate between RRV disease and Barham Forest virus disease, a very similar arbovirus. Virological tests can help distinguish between RRV disease and other causes of arthritis.

Treatment

No cure for RRV disease currently exists, so only its symptoms can be treated. In one of the few studies

KEY TERMS

Arthralgia—Sharp, severe pain, extending along a nerve or group of nerves, experienced in a joint and/or joints.

Astrovirus viruses—Also known as arthropod-borne viruses, these viruses are maintained in nature through biological transmission between vertebrate hosts and blood-feeding arthropods. Infection occurs when an infected arthropod, such as a mosquito, feeds off a vertebrate, such as a human.

Macropods—Derived from the Greek, macropod literally means “large footed.” Macropods are marsupials belonging to the family Macropodidae, which includes kangaroos, wallabies, tree kangaroos, pademelons, and several others.

Myalgia—Muscular pain or tenderness, typically of a diffuse and/or nonspecific nature.

Polyarthritis—A nonspecific term for arthritis involving two or more joints, typically associated with autoimmune forms of arthritis. Symptoms usually include pain, inflammation, and/or swelling in multiple joints.

on RRV disease treatment, Cordon and Rouse (1995) found that roughly one-third of patients (36.4%) reported that **nonsteroidal anti-inflammatory drugs** (NSAIDS) provided them with the best symptomatic relief. In addition to NSAIDS, others patients found that rest (24.1%), **aspirin** and paracetamol (16.4%), or physical therapies (10.3%), such as **hydrotherapy**, massage, and physiotherapy, were their only source of symptom relief. Unfortunately, 20% of patients found none of these interventions effective. Health providers typically use one or a combination of these treatments for their patients. In particular, paracetamol has been found to be effective for treating the pain and fever associated with RRV disease.

Although some clinicians have found the use of oral corticosteroid useful and effective, this practice is considered unwise and unnecessary. The adverse effects associated with **corticosteroids** may outweigh their benefits, and may even worsen the RRV disease. More study is required on this and other treatment interventions of RRV disease.

Prognosis

Patients infected with RRV disease will fully recover within four to seven months. Although milder cases can recover within a few weeks, many cases have persisted for several years. Only the symptoms can be treated during this time, not the disease. Fortunately, RRV infection usually provides the patient with life-long immunity to future infection.

Prevention

Prevention techniques of RRV typically coincide with measures used to avoid mosquito bites; the

primary source of the virus. These include the use of insect repellent (with 5% to 20% DEET) on exposed body parts, wearing loose-fitting clothes over the limbs and torso while outdoors, using mosquito coils and/or citronella candles outdoors, and limiting outdoor activities during peak biting periods and/or in areas with high mosquito density. While camping outdoors, knockdown spray or bed netting with pyrethrum is suggested. Additional steps for reducing risk of being bitten include using screens in homes and removing mosquito-breeding areas near the home, such as uncovered water containers and old tires. Mosquito eradication programs can assist in reducing insect populations. An RRV vaccine is currently being developed.

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Jason Fryer

Rotator cuff injury

Definition

A rotator cuff injury is a tear or inflammation of the rotator cuff tendons in the shoulder.

Description

Rotator cuff injury is known by several names, including pitcher's shoulder, swimmer's shoulder, and tennis shoulder. As these names imply, the injury occurs most frequently in athletes practicing sports that require the arm to be moved over the head repeatedly, such as pitching, swimming, tennis, and weight lifting. Rotator cuff tendonitis is an inflammation of the shoulder tendons while a rotator cuff tear is a ripping of one or more of the tendons.

The tendons of four muscles make up the rotator cuff. The muscles are the supraspinatus, infraspinatus, teres minor, and subscapularis. The tendons attach the muscles to four shoulder bones: the shoulder blade (scapula), the upper arm bone (humerus), and the collarbone (clavicle.) The rotator cuff tendons can also degenerate due to age, usually starting around age 40. Rotator cuff injury may also be caused by falling on the outstretched arm or joint of the elbow. Either of these may produce enough force to drive the humerus into the shoulder socket.

Causes and symptoms

Some areas of the rotator cuff tendons have poor blood supply. Thus, the tissue is very slow to heal and maintain itself during normal use. Tearing and inflammation in athletes is usually due to hard and repetitive use, especially in baseball pitchers. In non-athletes over age 40, the injuries usually occur as a result of lifting heavy objects. The two primary symptoms are **pain** and weakness in the shoulder or arm, especially with arm movement or at night. A partial tear may cause pain but still allow normal arm movement. A complete tear usually leaves the injured person unable to raise the arm away from the side.

Diagnosis

Diagnosis is usually made after a **physical examination**, often by a sports medicine physician. X rays are also sometimes used in diagnosis as well as an arthrogram. However, the arthrogram is an invasive procedure and may be painful afterwards. For this reason, **magnetic resonance imaging (MRI)** is

KEY TERMS

Arthrogram—A test done by injecting dye into the shoulder joint and then taking x-rays. Areas where the dye leaks out indicate a tear in the tendons.

Arthroscope—An instrument for the visual examination of the interior of a joint.

Arthroscopy—Examination of a joint with an arthroscope or joint surgery using an arthroscope.

Cortisone—A hormone produced naturally by the adrenal glands or made synthetically.

Magnetic resonance imaging (MRI) scan—A special radiological test that uses magnetic waves to create pictures of an area, including bones, muscles, and tendons.

Spur—Any projection from a bone.

preferred to determine tendon tears as it also show greater detail than the arthrogram.

Treatment

The primary treatment is resting the shoulder and, for minor tears and inflammation, applying ice packs. Anti-inflammatory medications may also be prescribed. As soon as pain decreases, **physical therapy** is usually started to help regain normal motion. If pain persists after several weeks, the physician may inject cortisone into the affected area.

Serious tears to the rotator cuff tendons usually require surgery to repair. An instrument called an arthroscope is used to view the shoulder joint and confirm the presence of a tear. The arthroscope can also be used to remove any bone spurs that may be present in the shoulder area. Current arthroscopic procedures usually involve a 2 in (5.1 cm) incision in the outer shoulder. Through this incision the torn rotator edge may be reattached to the humerus with stitches.

Alternative treatment

There are no effective alternative medicine treatments for rotator cuff injuries.

Prognosis

The prognosis for recovery from minor rotator cuff injuries is excellent. For serious injuries, the prognosis is usually good, some six weeks of physical therapy being required following surgery. Full recovery

may take several more months. In some cases, the injury is so severe that it requires tendon grafts and muscle transfers. In rare cases, a severe injury is not repairable, usually because the tendon has been torn for too long a time.

Prevention

The best prevention is to avoid repetitive overhead arm movements and to develop shoulder strength.

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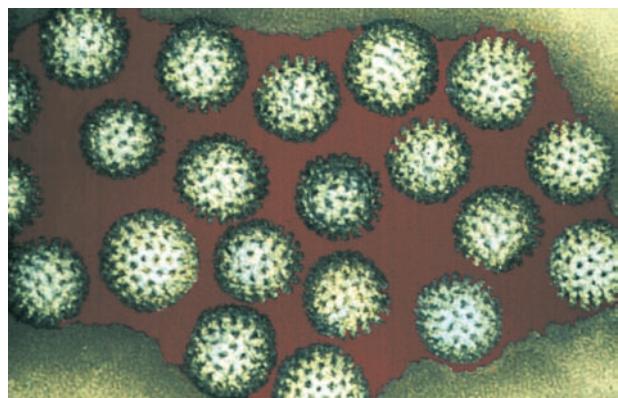
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ORGANIZATIONS

American Academy of Orthopaedic Surgeons, 6300 North River Road, Rosemont, IL, 60018-4262, (847) 823-7186, (847) 823-8125, pemr@aaos.org, <http://www.aaos.org>.

American Orthopaedic Society for Sports Medicine, 6300 N. River Road, Ste. 500, Rosemont, IL, 60018, (847) 292-4900, <http://www.sportsmed.org>.

Ken R. Wells



Rotaviruses are probably the most common viruses to infect humans and animals. These viruses are associated with gastroenteritis and diarrhea in humans and other animals.

(Dr. Linda Stannard/SPL/Photo Researchers, Inc.)

it mainly targets infants and young children. The outbreaks are usually in the cooler months of winter.

The virus is classified into different groups (Group A through group G), depending on the type of protein marker (antigen) that is present on its surface. The diarrheal infection of children is caused by the Group A rotaviruses. Group B rotaviruses have caused major epidemics of adult diarrhea in China. Group C rotavirus has been associated with rare cases of diarrheal outbreaks in Japan and England. Groups D through G have not been detected in humans.

Rotavirus infections

Definition

Rotavirus is the major cause of **diarrhea** and **vomiting** in young children worldwide. The infection is highly contagious and may lead to severe **dehydration** (loss of body fluids) and even **death**. In the United States, more than 50,000 children are hospitalized and up to 125 die each year as a result of rotavirus infection.

Description

Gastroenteritis, or inflammation of the stomach and the intestine, is the second most common illness in the United States, after the **common cold**. More than one-third of such cases are caused by viruses. Many different viruses can cause gastroenteritis, but the most common ones are the rotavirus and the Norwalk virus.

The name rotavirus comes from the Latin word "rota" for wheel and is given because the viruses have a distinct wheel-like shape. Rotavirus infection is also known as infantile diarrhea, or winter diarrhea, because

Causes and symptoms

The main symptoms of the rotavirus infection are **fever**, stomach cramps, **vomiting**, and diarrhea (this could lead to severe dehydration). The symptoms last anywhere from four to six days. If a child has dry lips and tongue, dry skin, sunken eyes, and wets fewer than six diapers a day, it is a sign of dehydration and a physician needs to be notified. Because of the excellence of healthcare in this country, rotavirus is rarely fatal to American children. However, it causes deaths of up to a million children in the third world countries, every year.

The virus is usually spread by the "fecal-oral route." In other words, a child can catch a rotavirus infection if she puts her finger in her mouth after touching toys or things that have been contaminated by the stool of another infected child. This usually happens when children do not wash their hands after using the toilet, or before eating food.

The viruses can also spread by way of contaminated food and drinking water. Infected food handlers who prepare salads, sandwiches, and other foods that require no cooking can spread the disease. Generally,

symptoms appear within 4–48 hours after exposure to the contaminated food or water.

Children between the ages of six months and two years, especially in a daycare setting, are the most susceptible to this infection. Breastfed babies may be less likely to become infected, because breast milk contains antibodies (proteins produced by the white blood cells of the immune system) that fight the illness. Nearly every child by the age of four has been infected by this virus, and has rotavirus antibodies in their body. The disease also targets the elderly and people who have weak immune systems.

Children who have been infected once can be infected again. However, second infections are less severe than the first infections. By the time a child has had two infections, the chances of subsequent severe infection is remote.

Diagnosis

The rotavirus infection is diagnosed by identifying the virus in the patient's stool. This is done using electron microscopy. Immunological tests such as ELISA (Enzyme-linked immunosorbent assay) are also widely used for diagnosis, and several commercial kits are available.

Treatment

“Oral rehydration therapy,” or drinking enough fluids to replace those lost through bowel movements and vomiting, is the primary aim of the treatment. Electrolyte and fluid replacement solutions are available over the counter in food and drug stores. Dehydration is one of the greatest dangers for infants and young children. If the diarrhea becomes severe, it may be necessary to hospitalize the patient so that fluids can be administered intravenously.

Anti-diarrheal medication should not be given to children unless directed to do so by the physician. Antibiotic therapy is not useful in viral illness. Specific drugs for the virus are not available.

Prognosis

Most of the infections resolve spontaneously. Dehydration due to severe diarrhea is one of the major complications.

Prevention

The best way to prevent the disease is by proper food handling and thorough hand washing, after using the toilet and whenever hands are soiled. In child care centers and hospital settings, the staff should be

educated about personal and environmental hygiene. All dirty diapers should be regarded as infectious and disposed of in a sanitary manner.

Vaccines that prevent rotavirus in young children have been tested in nationwide trials. Researchers report that the vaccines appear to prevent the infection in 80% of the tested children. The vaccine is intended to be given orally (by mouth) at two, four, and six months of age. The only side effect of the vaccine is a low-grade fever in a small percentage of the children, three to four days after the **vaccination**. Within the next few years, a rotavirus vaccine may become part of every child's immunization schedule.

Resources

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Lata Cherath, PhD

Roundworm infections

Definition

Roundworm infections are diseases of the digestive tract and other organ systems caused by nematodes. Nematodes are parasitic worms with long, cylindrical bodies.

Description

Roundworm infections are widespread throughout the world, with some regional differences. Ascariasis and trichuriasis are more common in warm, moist climates where people use human or animal feces for fertilizer. Anisakiasis is most common in countries where raw or pickled fish or squid is a popular food item.

Causes and symptoms

The causes and symptoms of roundworm infection vary according to the species. Humans acquire most types of roundworm infection from contaminated food or by touching the mouth with unwashed hands.

Anisakiasis

Anisakiasis is caused by anisakid roundworms. Humans are not the primary host for these parasites. Anisakid roundworms infest whales, seals, and

dolphins; crabs then ingest roundworm eggs from the feces of these animals. In the crabs, the eggs hatch into larvae that can infect fish. The larvae enter the muscles of marine animals further up the food chain, including squid, mackerel, herring, cod, salmon, tuna, and halibut. Humans become accidental hosts when they eat raw or undercooked fish containing anisakid larvae. The larvae attach themselves to the tissues lining the stomach and intestine, and eventually die inside the inflamed tissue.

In humans, anisakiasis can produce a severe syndrome that affects the stomach and intestines, or a mild chronic disease that may last for weeks or years. In acute anisakiasis, symptoms begin within one to seven hours after the patient eats infected seafood. Patients are often violently sick, with **nausea**, **vomiting**, **diarrhea**, and severe abdominal **pain** that may resemble **appendicitis**. In chronic anisakiasis, the patient has milder forms of stomach or intestinal irritation that resemble stomach ulcers or **irritable bowel syndrome**. In some cases, the acute form of the disease is followed by chronic infestation.

Ascariasis

Ascariasis, which is caused by *Ascaris lumbricoides*, is one of the most widespread parasitic infections in humans, affecting over 1.3 billion people worldwide. Ascarid roundworms cause a larger burden on the human host than any other parasite; adult worms can grow as long as 12 or 14 inches, and release 200,000 eggs per day. The eggs infect people who eat unwashed vegetables from contaminated soil or touch their mouths with unwashed hands. Once inside the digestive tract, the eggs release larvae that penetrate the intestinal wall and migrate to the lungs through the liver and the bloodstream. After about 10 days in the lungs, the larvae migrate further into the patient's upper lung passages and airway, where they are swallowed. When they return to the intestine, they mature into adults and reproduce. The time period from the beginning of the infection to egg production is 60–75 days.

The first symptoms of infection may occur when the larvae reach the lungs. The patient may develop chest pain, coughing, difficulty breathing, and inflammation of the lungs. In some cases, the patient's sputum is streaked with blood. This phase of the disease is sometimes called Loeffler's syndrome. It is marked by an accumulation of parasites in the lung tissue and by eosinophilia (an abnormal increase in the number of a specific type of white blood cell). The intestinal phase of ascariasis is marked by stomach pain, cramping, nausea, and intestinal blockage in severe cases.

Toxocariasis

Toxocariasis is sometimes called visceral larva migrans (VLM) because the larval form of the organism hatches inside the intestines and migrates throughout the body to other organs (viscera). The disease is caused by *Toxocara canis* and *T. cati*, which live within the intestines of dogs and cats. Most human patients are children between the ages of two and four years, who become infected after playing in sandboxes or soil contaminated by pet feces, although adults are also susceptible. The eggs can survive in soil for as long as seven years.

The organism's eggs hatch inside the human intestine and release larvae that are carried in the bloodstream to all parts of the body, including the eyes, liver, lungs, heart, and brain. The patient usually has a **fever**, with coughing or **wheezing** and a swollen liver. Some patients develop skin **rashes** and inflammation of the lungs. The larvae may survive inside the body for months, producing allergic reactions and small granulomas, which are tissue swellings or growths produced in response to inflammation. Infection of the eye can produce ocular larva migrans (OLM), which is the first symptom of toxocariasis in some patients.

Trichuriasis

Trichuriasis, caused by *Trichuris trichiura*, is sometimes called whipworm because the organism has a long, slender, whiplike front end. The adult worm is slightly less than an inch long. Trichuriasis is most common in warm, humid climates, including the southeastern United States. The number of people with trichuriasis may be as high as 800 million worldwide.

Whipworm larvae hatch from swallowed eggs in the small intestine and move on to the upper part of the large intestine, where they attach themselves to the lining. The adult worms produce eggs that are passed in the feces and mature in the soil. Patients with mild infections may have few or no symptoms. In cases of heavy infestation, the patient may have abdominal cramps and other symptoms resembling amebic **dysentery**. In children, severe trichuriasis may cause anemia and developmental retardation.

Diagnosis

Since the first symptoms of roundworm infection are common to a number of illnesses, a doctor is most likely to consider the possibility of a parasitic disease on the basis of the patient's history—especially in children. The definite diagnosis is based on the results of

KEY TERMS

Eosinophilia—An abnormal increase in the number of a specific type of white blood cell. Eosinophilia is a characteristic of all types of roundworm infections.

Granuloma—A tissue swelling produced in response to inflammation. Granulomas are important in diagnosing toxocariasis.

Loeffler's syndrome—The respiratory phase of ascariasis, marked by inflammation of the lungs and eosinophilia.

Nematode—A parasitic roundworm with a long, cylindrical body.

Ocular larva migrans (OLM)—A syndrome associated with toxocariasis, in which the eye is invaded by migrating larvae.

Visceral larva migrans (VLM)—Another name for toxocariasis. The name is derived from the life cycle of the organism.

Whipworm—Another name for trichuriasis. The name comes from the organism's long whiplike front end.

stool or tissue tests. In trichuriasis, adult worms may also be visible in the lining of the patient's rectum. In ascariasis, adult worms may appear in the patient's feces or vomit; they can also be detected by x ray and ultrasound. In toxocariasis, larvae are sometimes found in tissue samples taken from a granuloma. If a patient with toxocariasis develops OLM, it is important to obtain a granuloma sample in order to distinguish between OLM and **retinoblastoma** (a type of eye tumor).

Anisakiasis is one of two roundworm infections that cannot be diagnosed from stool specimens. Instead, the diagnosis is made by x rays of the patient's stomach and small intestine. The larvae may appear as small threads when double contrast x rays are used. In acute cases, the doctor may use an endoscope (an instrument for examining the interior of a body cavity) to look for or remove larvae.

Blood tests cannot be used to differentiate among different types of roundworm infections, but the presence of eosinophilia can help to confirm the diagnosis.

Patients with trichuriasis or ascariasis should be examined for signs of infection by other roundworm species; many patients are infected by several parasites at the same time.

Treatment

Trichuriasis, ascariasis, and toxocariasis are treated with anthelmintic medications. These are drugs that destroy roundworms either by paralyzing them or by blocking them from feeding. Anthelmintic drugs include pyrantel pamoate, piperazine, albendazole, and mebendazole. Mebendazole cannot be given to pregnant women because it may harm the fetus. Treatment with anthelmintic drugs does not prevent reinfection.

There is no drug treatment for anisakiasis; however, symptoms usually resolve in one to two weeks when the larvae die. In some cases, the larvae are removed with an endoscope or by surgery.

Patients with an intestinal obstruction caused by ascariasis may be given **nasogastric suction**, followed by anthelmintic drugs, in order to avoid surgery. If suction fails, the worms must be removed surgically to prevent intestinal rupture or blockage.

Prognosis

The prognosis for recovery from roundworm infections is good for most patients. The severity of infection, however, varies considerably from person to person. Children are more likely to have heavy infestations and are also more likely to suffer from malabsorption and **malnutrition** than adults.

Ascariasis is the only roundworm infection with a significant mortality rate. *A. lumbricoides* grows large enough to perforate the bile or pancreatic ducts; in addition, a mass of worms in the digestive tract can cause rupture or blockage of the intestines. It is estimated that 20,000 children die every year from intestinal ascariasis.

Prevention

There are no effective vaccines against any of the soil-transmitted roundworms, nor does infection confer immunity. Prevention of infection or reinfection requires adequate hygiene and sanitation measures, including regular and careful handwashing before eating or touching the mouth with the hands.

With respect to specific infections, anisakiasis can be prevented by avoiding raw or improperly prepared fish or squid. Trichuriasis, ascariasis, and toxocariasis can be prevented by keeping children from playing in

soil contaminated by human or animal feces; by teaching children to wash their hands before eating; and by having pets dewormed regularly by a veterinarian.

Resources

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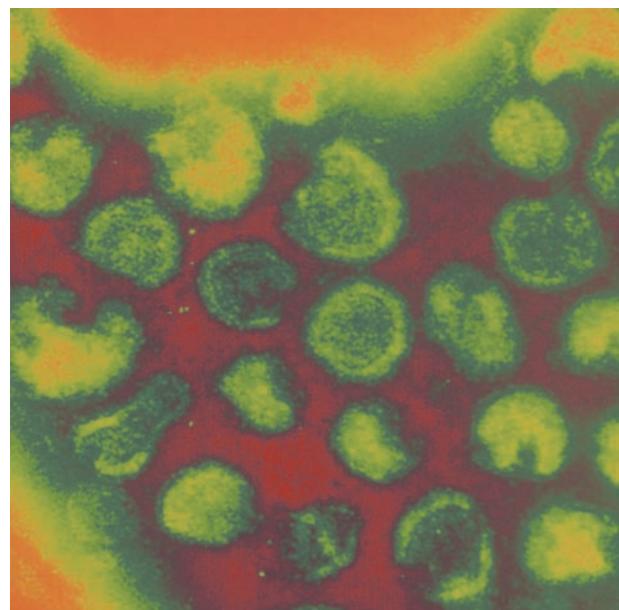
Rebecca J. Frey, PhD

Routine urinalysis see **Urinalysis**

RSV see **Respiratory syncytial virus infection**

RTA see **Renal tubular acidosis**

RU-486 see **Mifepristone**



A digitized image of rubella virus particles. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Rubella

Definition

Rubella is a highly contagious viral disease, spread through contact with discharges from the nose and throat of an infected person. Although rubella causes only mild symptoms of low **fever**, **swollen glands**, joint **pain**, and a fine red rash in most children and adults, it can have severe complications for women in their first trimester of **pregnancy**. These complications include severe **birth defects** or **death** of the fetus.

Description

Rubella is also called German **measles** or three-day measles. This disease was once a common childhood illness, but its occurrence has been drastically reduced since vaccine against rubella became available in 1969. In the 20 years following the introduction of the vaccine, reported rubella cases dropped 99.6%. Only 229 cases of rubella were reported in the United States in 1996.

Rubella is spread through contact with fluid droplets expelled from the nose or throat of an infected person. A person infected with the rubella virus is contagious for about seven days before any symptoms appear and continues to be able to spread the disease for about four days after the appearance of symptoms. Rubella has an incubation period of 12–23 days.

Although rubella is generally considered a childhood illness, people of any age who have not been vaccinated or previously caught the disease can become

infected. Having rubella once or being immunized against rubella normally gives lifetime immunity. This is why **vaccination** is so effective in reducing the number of rubella cases.

Women of childbearing age who do not have immunity against rubella should be the most concerned about getting the disease. Rubella infection during the first three months of pregnancy can cause a woman to miscarry or cause her baby to be born with birth defects. Although it has been practically eradicated in the United States, rubella is still common in less developed countries because of poor immunization penetration, creating a risk to susceptible travelers. Some countries have chosen to target rubella vaccination to females only and outbreaks in foreign-born males have occurred on cruise ships and at U.S. summer camps.

Causes and symptoms

Rubella is caused by the rubella virus (*Rubivirus*). Symptoms are generally mild, and complications are rare in anyone who is not pregnant.

The first visible sign of rubella is a fine red rash that begins on the face and rapidly moves downward to cover the whole body within 24 hours. The rash lasts about three days, which is why rubella is sometimes called the three-day measles. A low fever and swollen glands, especially in the head (around the ears) and neck, often accompany the rash. Joint pain and sometimes joint



A red rash is one characteristic of rubella, or German measles, as seen on this man's arm. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)

swelling can occur, more often in women. It is quite common to get rubella and not show any symptoms (subclinical infection).

Symptoms disappear within three to four days, except for joint pain, which may linger for a week or two. Most people recover fully with no complications. However, severe complications may arise in the unborn children of women who get rubella during the first three months of their pregnancy. These babies may be miscarried or stillborn. A high percentage are born with birth defects. Birth defects are reported to occur in 50% of women who contract the disease during the first month of pregnancy, 20% of those who contract it in the second month, and 10% of those who contract it in the third month.

The most common birth defects resulting from congenital rubella infection are eye defects such as **cataracts**, glaucoma, and blindness; deafness; congenital

heart defects; and **mental retardation**. Taken together, these conditions are called congenital rubella syndrome (CRS). The risk of birth defects drops after the first trimester, and by the 20th week there are rarely any complications.

Diagnosis

The rash caused by the rubella virus and the accompanying symptoms are so similar to other viral infections that it is impossible for a physician to make a confirmed diagnosis on visual examination alone. The only sure way to confirm a case of rubella is by isolating the virus with a blood test or in a laboratory culture.

A blood test is done to check for rubella antibodies. When the body is infected with the rubella virus, it produces both immunoglobulin G (IgG) and immunoglobulin M (IgM) antibodies to fight the infection. Once IgG exists, it persists for a lifetime, but the special IgM antibody usually wanes over six months. A blood test can be used either to confirm a recent infection (IgG and IgM) or determine whether a person has immunity to rubella (IgG only). The lack of antibodies indicates that a person is susceptible to rubella.

All pregnant women should be tested for rubella early in pregnancy, whether or not they have a history of vaccination. If the woman lacks immunity, she is counseled to avoid anyone with the disease and to be vaccinated after giving birth.

Treatment

There is no drug treatment for rubella. Bed rest, fluids, and **acetaminophen** for pain and temperatures over 102°F (38.9°C) are usually all that is necessary.

Babies born with suspected CRS are isolated and cared for only by people who are sure they are immune to rubella. Congenital heart defects are treated with surgery.

Alternative treatment

Rather than vaccinating a healthy child against rubella, many alternative practitioners recommend allowing the child to contract the disease naturally at the age of five or six years, since the immunity conferred by contracting the disease naturally lasts a lifetime. It is, however, difficult for a child to contract rubella naturally when everyone around him or her has been vaccinated.

Ayurvedic practitioners recommend making the patient comfortable and giving the patient ginger or clove tea to hasten the progress of the disease.

KEY TERMS

Incubation period—The time it takes for a person to become sick after being exposed to a disease.

Trimester—The first third, or thirteen weeks, of pregnancy.

Traditional Chinese medicine uses a similar approach. Believing that inducing the skin rash associated with rubella hastens the progress of the disease, traditional Chinese practitioners prescribe herbs such as peppermint (*Mentha piperita*) and *chai-hu* (*Bupleurum chinense*). Cicada is often prescribed as well. Western herbal remedies may be used to alleviate rubella symptoms. Distilled witch hazel (*Hamamelis virginiana*) helps calm the **itching** associated with the skin rash and an eyewash made from a filtered diffusion of eyebright (*Euphrasia officinalis*) can relieve eye discomfort. Antiviral western herbal or Chinese remedies can be used to assist the immune system in establishing equilibrium during the healing process. Depending on the patient's symptoms, among the remedies a homeopath may prescribe are *Belladonna*, *Pulsatilla*, or *Phytolacca*.

Prognosis

Complications from rubella infection are rare in children, pregnant women past the 20th week of pregnancy, and other adults. For women in the first trimester of pregnancy, there is a high likelihood of the child being born with one or more birth defect. Unborn children exposed to rubella early in pregnancy are also more likely to be miscarried, stillborn, or have a low birthweight. Although the symptoms of rubella pass quickly for the mother, the consequences to the unborn child can last a lifetime.

Prevention

Vaccination is the best way to prevent rubella and is normally required by law for children entering school. Rubella vaccine is usually given in conjunction with measles and **mumps** vaccines in a shot referred to as MMR (mumps, measles, and rubella). Children receive one dose of MMR vaccine at 12-15 months and another dose at four to six years.

Pregnant women should not be vaccinated, and women who are not pregnant should avoid conceiving for at least three months following vaccination. To date, however, accidental rubella vaccinations during

pregnancy have not clearly been associated with the same risk as the natural infection itself. Women may be vaccinated while they are **breastfeeding**. People whose immune systems are compromised, either by the use of drugs such as **steroids** or by disease, should discuss possible complications with their doctor before being vaccinated.

ORGANIZATIONS

March of Dimes Birth Defects Foundation, 1275 Mamaroneck Ave., White Plains, NY, 10605, (914) 997-4488, <http://www.modimes.org>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Tish Davidson, A.M.

Rubella test

Definition

The **rubella** test is a routine blood test performed as part of prenatal care of pregnant women. It is sometimes also used to screen women of childbearing age before the first **pregnancy**.

Purpose

The test is given to evaluate whether a woman is immune to rubella (German **measles**) as a result of childhood exposure or immunization, or whether she may be presently infected with the disease. The question of a current infection is particularly urgent for pregnant women. Although the disease itself is not serious in adults, it can cause **miscarriage**, **stillbirth**, or damage to the fetus during the first trimester (three months) of pregnancy. The rubella test is regarded as a more reliable indication of the patient's immune status than her history because reinfection with rubella is possible even after immunization. The results of the test may influence decisions to terminate a pregnancy.

Description

The rubella test belongs to a category of blood tests called hemagglutination inhibition (HI) tests. Hemagglutination refers to the clumping or clustering of red blood cells caused by a disease antibody, virus, or certain other substances. Inhibition refers to interference with the clumping process. The presence of rubella antibodies inhibits the cell clumping caused

by the rubella virus. Thus, the addition of the virus to a sample of the patient's blood allows a doctor to determine the presence and concentration of rubella antibodies and the patient's immunity to the disease.

When a person is infected with the rubella virus, the body produces both immunoglobulin G (IgG) and immunoglobulin M (IgM) antibodies to fight the infection. Once IgG exists, it persists for a lifetime, but the special IgM antibody usually wanes over six months. The rubella test can either confirm that a recent infection has occurred (both IgG and IgM are present) or that a patient has immunity to rubella (IgG only is present).

When the test is performed to confirm the diagnosis of rubella in a woman already pregnant, two blood samples are drawn. One is drawn during the acute phase of the illness about three days after the rash breaks out, and the second is drawn during the convalescent phase about three weeks later. The specimens are then tested simultaneously by a single laboratory. Alternatively, a pregnant woman with a rash suspected to be rubella can be tested for IgM antibody. If the test shows that IgM antibody is present, then a recent rubella infection has occurred.

Because there have been cases of children born with rubella syndrome even though the mother's blood test indicated that she was sufficiently immune to rubella, some researchers are presently recommending a second test, known as a synthetic peptide enzyme-linked immunosorbent assay (ELISA). This test screens for the presence of rubella virus neutralizing (RVN) antibodies in the mother's blood.

Normal results

If the patient has been successfully immunized against rubella or has had the disease, the HI antibody titer (concentration) will be greater than 1:10–1:20. The red blood cells will fail to clump when the rubella virus is added to the blood serum.

In the case of paired testing for pregnant women, a fourfold rise in antibody titer between the first and second blood samples indicates the suspicious rash was caused by rubella. The alternative test for IgM antibody confirms recent rubella infection if IgM is found in the patient's blood.

Abnormal results

If the patient has little or no immunity to rubella, her HI antibody titer will be 1:8 or less. Women without

KEY TERMS

Antibody—A protein molecule produced by the immune system that is specific to a virus, such as the rubella virus. The antibody combines with the virus and disables it.

Hemagglutination—The clumping or clustering of red blood cells caused by certain viruses, antibodies, or other substances.

Inhibition—Restraint of or interference with a biological process, such as the clumping of blood cells.

Titer—The concentration of a substance in a given sample of blood or other tissue fluid.

immunity should receive immunization against rubella provided that they avoid pregnancy for a period of three months following immunization. Women with disease of the immune system or who are taking corticosteroid medications should receive immune serum globulin rather than rubella vaccine to prevent infection.

Resources

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ORGANIZATIONS

American Academy of Family Physicians (AAFP), 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2680, (913) 906-6000, (913) 906-6075, (800) 271-2237, <http://www.aafp.org/>.

Rebecca J. Frey, PhD

Rubeola see **Measles**

Ruptured disk see **Herniated disk**

RVT see **Renal vein thrombosis**

S

Sacroiliac disease

Definition

Sacroiliac disease is high-impact trauma to the sacroiliac joint that can cause **death**, or bone, and nerve damage.

Description

The sacroiliac joint is a strong, weight bearing synovial joint between the ilium and sacrum bones of the pelvis. The bones are held in place and allowed limited movements by a system of sacroiliac ligaments. Relaxation of this and other joints and ligaments is important during **pregnancy** and is accomplished by a special hormone called relaxin. Usually the sacroiliac is damaged by high-impact injuries. These injuries may be life threatening and mortality is approximately 20% if neighboring structures are also damaged. Injuries to this area often includes neurological deficits. Dislocation and nerve damage are frequently missed in the diagnosis.

Causes and symptoms

The primary cause of **dislocations**, **fractures**, and accompanying damage is usually a traumatic accident. Patients receiving such injuries require emergency medical attention. There may be severe blood loss due to breakage of large bones and resuscitative measures may be required for stabilization.

Diagnosis

The diagnosis can be difficult since nerve damage can mimic other conditions with similar symptoms (i.e., **low back pain** in persons with **sciatica**). Additionally imaging studies and **physical examination** maneuvers will miss the diagnosis. The definitive method for diagnosing sacroiliac

pathology would be injection of local anesthetic in the correct area of the affected sacroiliac joint. This procedure is usually performed using advance guidance systems (CT or fluoroscopic assisted guidance). If the **pain** is relieved by anesthetic injection, then the diagnosis is confirmed. There are three typical patterns of pain: pain directly over the joint, pain in the groin extending down the affected leg that can mimic the signs associated with a herniated lumbar disc, and pain widely dispersed in the affected leg.

Treatment

Treatment initially can include emergency interventions, but usually is conservative. Treatment includes **physical therapy**, manipulation, and medications for pain control. In some cases a sacroiliac belt can help with symptoms. In sacroiliac joint disease that has already progressed and is chronic and severe, corrective joint fusion may be indicated.

Prognosis

Outcome is variable and takes into account the extent of injuries, early diagnosis, and responsiveness to conservative treatment.

Prevention

There is no known prevention since the disease is secondary to an accident.

Resources

BOOKS

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Firestein, Gary S., et al. *Kelley's Textbook of Rheumatology*. Philadelphia: Saunders/Elsevier, 2009.

KEY TERMS

Herniated disk—A protrusion in a disk located in the spinal column.

Ligament—Fibrous tissue which connect bones.

Synovial joint—A joint that allows for bone movement.

ORGANIZATIONS

American Academy of Orthopaedic Surgeons, 6300 North River Road, Rosemont, IL 60018-4262, (847) 823-7186, (847) 823-8125, <http://www.aaos.org>.

Laith Farid Gulli, M.D.

SAD see **Seasonal affective disorder**

Salivary gland scan

Definition

A salivary gland scan is a nuclear medicine test that examines the uptake and secretion in the salivary glands of a radioactively labeled marker substance. The pattern of uptake and secretion shows if these glands are functioning normally.

Purpose

A salivary gland scan is done to help diagnose the cause of **dry mouth**. It is a test that is done when Sjogren's syndrome, salivary duct obstruction, asymmetric hypertrophy, or growths such as Warthin's tumors are suspected.

Precautions

Salivary gland scans are a safe and effective way to diagnose problems associated with dry mouth. The level of radioactivity in the marker substance is low and poses no threat to health. The only people who should not undergo this test are pregnant women.

Other recent nuclear medicine tests may affect the results of this scan. It may be necessary to wait until earlier radiopharmaceuticals have been cleared from the body before undergoing this scan.

Description

A salivary gland scan, also called a parotid gland scan, is a noninvasive test. The patient is positioned under a gamma scintillation camera that detects radiation. The patient then is injected with a low-level radioactive marker, usually technetium-99m or technetium pertechnetate.

Immediately after the injection, imaging begins. For accurate results, the patient must stay still during imaging. After several images, the patient is given lemon drop candies to suck on, which stimulate the salivary glands. Another set of images is made for comparison purposes. The entire process takes about ten minutes for the injection and 30–45 minutes for the scan.

Preparation

No special preparations are needed for this test. It is not necessary to fast or to restrict medications before testing. Any blood that needs to be drawn for other tests should be taken before the radiopharmaceutical is injected.

Aftercare

Patients can return to normal activities immediately.

Risks

A salivary gland scan is a safe test. The only risk is to the fetus of a pregnant woman. Women who are pregnant should discuss the risks and benefits of this procedure with their doctor.

Normal results

Normally functioning salivary glands take up the radiopharmaceutical then secrete it when stimulated by the lemon drops.

Abnormal results

Abnormally functioning salivary glands fail to exhibit a normal uptake and secretion pattern. This test does not differentiate between benign and malignant lesions.

Resources

BOOKS

Beers, Mark H., Robert S. Porter, and Thomas V. Jones, eds. *The Merck Manual of Diagnosis and Therapy*. 18th ed. Whitehouse Station, NJ: Merck Research Laboratories, 2006.

KEY TERMS

Hypertrophy—Overgrowth of tissue not due to a tumor.

Parotid gland—The salivary gland that lies below and in front of each ear.

Radiopharmaceutical—A radioactive pharmaceutical or chemical (usually radioactive iodine or cobalt) used for diagnostic or therapeutic purposes.

Sjögren's syndrome—A disease often associated with rheumatoid arthritis, that causes dry mouth, lesions on the skin, and enlargement of the parotid glands. It is often seen in menopausal women.

Technetium—A synthetic element used in nuclear medicine; it is obtained from the fission of uranium.

Warthin's tumor—A benign tumor of the parotid gland.

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ORGANIZATIONS

- Society of Nuclear Medicine (SNM), 1850 Samuel Morse Dr., Reston, VA, 20190, (703) 708-9000, (703) 708-9015, <http://www.snm.org>.

Tish Davidson, AM
Rebecca J. Frey, PhD

Salivary gland tumors

Definition

A salivary gland tumor is an uncontrolled growth of cells that originates in one of the many saliva-producing glands in the mouth.

Description

The tongue, cheeks, and palate (the hard and soft areas at the roof of the mouth) contain many glands that produce saliva. In saliva there are enzymes, or

catalysts, that begin the breakdown (digestion) of food while it is still in the mouth. The glands are called salivary glands because of their function.

There are three big pairs of salivary glands in addition to many smaller ones. The parotid glands, submandibular glands and sublingual glands are the large, paired salivary glands. The parotids are located inside the cheeks, one below each ear. The submandibular glands are located on the floor of the mouth, with one on the inner side of each part of the lower jaw, or mandible. The sublingual glands are also in the floor of the mouth, but they are under the tongue.

The parotids are the salivary glands most often affected by tumors. Yet most of the tumors that grow in the parotid glands are benign, or not cancerous. Approximately 8 out of 10 salivary tumors diagnosed are in a parotid gland. One in 10 diagnosed is in a submandibular gland. The remaining 10% are diagnosed in other salivary glands.

In general, glands more likely to show tumor growth are also glands least likely to show malignant tumor growth. Thus, although tumors of the sublingual glands are rare, almost all of them are malignant. In contrast, about one in four tumors of the parotid glands is malignant.

Cancers of the salivary glands begin to grow in epithelial cells, or the flat cells that cover body surfaces. Thus, they are called carcinomas, cancers that by definition begin in epithelial cells.

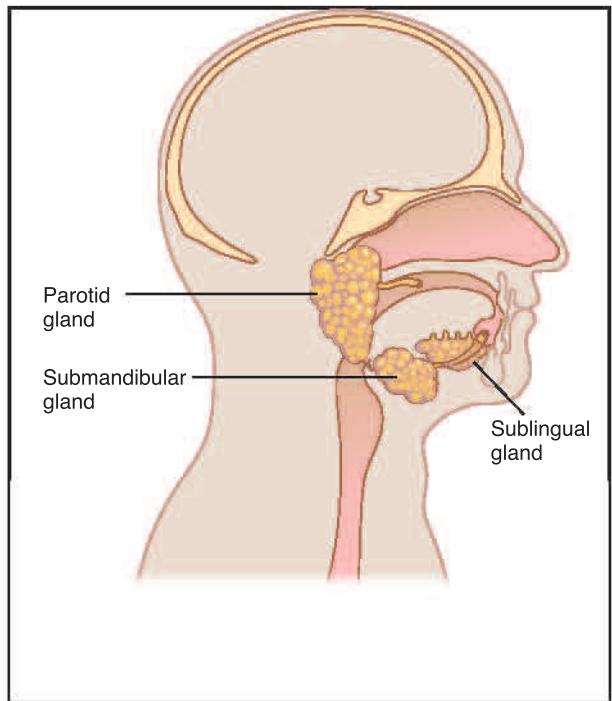
Demographics

Cancers in the mouth account for fewer than 2% of all cases of **cancer** and about 1.5% of cancer deaths. About 7% of all cancers diagnosed in the head and neck region are diagnosed in a salivary gland. Men and women are at equal risk.

Mortality from salivary gland tumors in the United States is higher among male African Americans below the age of 50 than among older workers of any race or either sex.

Causes and symptoms

When survivors of the 1945 atomic bombings of Nagasaki and Hiroshima began to develop salivary gland tumors at a high rate, radiation was suspected as a cause. Ionizing radiation, particularly gamma radiation, is a factor that contributes to tumor development. So is **radiation therapy**. Adults who received radiation therapy for enlarged adenoids or tonsils when they were children are at greater risk for salivary gland tumors.



Location of the three main salivary glands. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Another reported risk factor is an association between wood dust inhalation and adenocarcinoma of the minor salivary glands of the nose and paranasal sinuses. There is also evidence that people infected with herpes viruses may be at greater risk for salivary gland tumors. And individuals infected with human **immunodeficiency** virus (HIV) have more salivary gland disease in general, and may be at greater risk for salivary gland tumors.

Although there has been speculation that the electromagnetic fields generated by cell phones increase the risk of salivary gland tumors, a recent study done in Denmark has concluded that the use of cell phones, pagers, and similar devices is not a risk factor.

There seems to be some link between **breast cancer** and salivary gland tumors. Women with breast cancer are more likely to be diagnosed with salivary gland tumors. Also linked to salivary gland tumors is alcohol use, exposure to sunlight (ultraviolet radiation) and hair dye use. There is evidence that people infected with herpes viruses may be at greater risk for salivary gland tumors. Individuals infected with human immunodeficiency virus (HIV) have more salivary gland disease in general, and may be at greater risk for salivary gland tumors.

Symptoms are often absent until the tumor is large or has metastasized (spread to other sites). In many cases, the tumor is first discovered by the patient's dentist. During regular dental examinations, the dentist looks for masses on the palate or under the tongue or in the cheeks, and such checkups are a good way to detect tumors early. Some symptoms are:

- a lump or mass in the mouth
- swelling in the face
- pain in the jaw or the side of the face
- difficulty swallowing
- difficulty breathing
- difficulty speaking

Diagnosis

A tissue sample will be taken for study via a biopsy. Usually an incision is necessary to take the tissue sample. Sometimes it is possible to take a tissue sample with a needle.

Magnetic resonance imaging (MRI) and computed tomography (CT) scans are also used to evaluate the tumor. They help determine whether the cancer has spread to sites adjacent to the salivary gland where it is found. MRI offers a good way to examine the tonsils and the back of the tongue, which are soft tissues. CT is tapped as a way of studying the jaw, which is bone.

Treatment

To assess the stage of growth of a salivary gland tumor, many features are examined, including how big it is and the type of abnormal cell growth. Analysis of the types of abnormal cell growth in tissue is so specific that many salivary gland tumors are given unique names.

In stage I cancer the tumor is less than one inch in size and it has not spread. Stage II salivary gland cancers are larger than one inch and smaller than two and one-half inches, but they have not spread. Stage III cancers are smaller than one inch, but they have spread to a lymph node. Stage IV cancers have spread to adjacent sites in the head, which may include the base of the skull and nearby nerves, or they are larger than two and one-half inches and have invaded a lymph node.

Surgical removal (excision) of the tumor is the most common treatment. **Chemotherapy** and radiation therapy may be part of the treatment, particularly if the cancer has metastasized, or spread to other sites; chemotherapy of salivary gland cancers, however, does not appear to extend survival or improve the patient's quality of life. Because there are many nerves

KEY TERMS

Adenoids—Common name for the pharyngeal tonsils, which are lymph masses in the wall of the air passageway (pharynx) just behind the nose.

Biopsy—Tissue sample is taken from the body for examination.

Computed tomography (CT)—X rays are aimed at slices of the body (by rotating equipment) and results are assembled with a computer to give a three-dimensional picture of a structure.

Lymph—Tissue that is part of the lymphatic system, the system that collects and returns fluid to the blood vessels and produces substances that fight infection.

Magnetic resonance imaging (MRI)—Magnetic fields and radio frequency waves are used to take pictures of the inside of the body.

Tonsils—Common name for the palatine tonsils, which are lymph masses in the back of the mouth, on either side of the tongue.

and blood vessels near the three major pairs of salivary glands, particularly the parotids, the surgery can be quite complicated. A complex surgery is especially true if the tumor has spread.

A promising form of treatment for patients at high risk of tumor recurrence in the salivary glands near the base of the skull is **gamma knife surgery**. Used as a booster treatment following standard neutron radiotherapy, gamma knife surgery appears to be well tolerated by the patients and to have minimal side effects.

Alternative treatment

Any technique, such as **yoga**, **meditation** or **bio-feedback**, that helps a patient cope with **anxiety** over the condition and discomfort from treatment is useful and should be explored as an option.

Prognosis

Tumors in small salivary glands that are localized can usually be removed without much difficulty. The outlook for survival once the tumor is removed is very good if it has not metastasized.

For parotid cancers, the five-year survival rate is more than 85% whether or not a lymph node is involved at diagnosis. Ten-year survival rate is just under 50%.

Most early stage salivary gland tumors are removed, and they do not return. Those that do return, or recur, are the most troublesome and reduce the chance an individual will remain cancer-free.

Prevention

Minimizing intake of beverages containing alcohol may be important. Avoiding unnecessary exposure of the head to radiation and to sunlight may also be considered preventative. Anything that reduces

the risk of contracting a sexually transmitted disease, such as the use of **condoms**, also may lower the risk of salivary gland cancer.

Resources

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OTHER

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ORGANIZATIONS

SPOHNC, Support for People with Oral and Head and Neck Cancer, PO Box 53, Locust Valley, NY, 11560-0053, (516) 671-8794, (800) 377-0928, info@spohnc.org, <http://www.spohnc.org>.

Diane M. Calabrese
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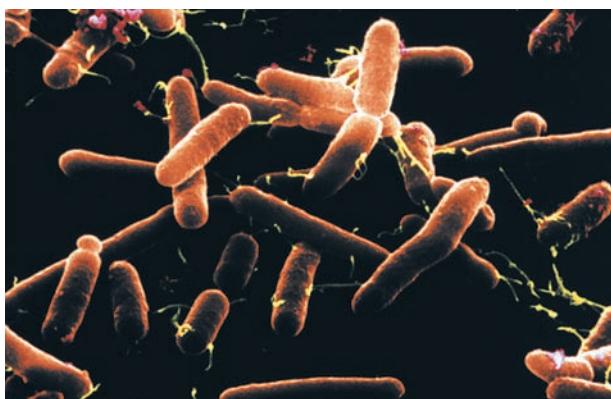
Salmonella food poisoning

Definition

Salmonella **food poisoning** is a bacterial food poisoning caused by the *Salmonella* bacterium. It results in the swelling of the lining of the stomach and intestines (**gastroenteritis**). While domestic and wild animals, including poultry, pigs, cattle, and pets such as turtles, iguanas, chicks, dogs, and cats can transmit this illness, most people become infected by ingesting foods contaminated with significant amounts of *Salmonella*.

Demographics

Salmonella food poisoning occurs worldwide, however it is most frequently reported in North America and Europe. Only a small proportion of infected people are tested and diagnosed, and as few as 1% of cases are actually reported. While the infection rate may seem relatively low, even an attack rate of less than 0.5% in



Salmonella enteritidis. Exposure to this bacterium usually occurs by contact with contaminated food. (© Oliver Meckes/Photo Researchers, Inc.)

such a large number of exposures results in many infected individuals. The poisoning typically occurs in small, localized outbreaks in the general population or in large outbreaks in hospitals, restaurants, or institutions for children or the elderly. In the United States, *Salmonella* is responsible for about 15% of all cases of food poisoning.

Description

Improperly handled or undercooked poultry and eggs are the foods which most frequently cause *Salmonella* food poisoning. Chickens are a major carrier of *Salmonella* bacteria, which accounts for its prominence in poultry products. However, identifying foods which may be contaminated with *Salmonella* is particularly difficult because infected chickens typically show no signs or symptoms. Since infected chickens have no identifying characteristics, these chickens go on to lay eggs or to be used as meat.

At one time, it was thought that *Salmonella* bacteria were only found in eggs which had cracked, thus allowing the bacteria to enter. Ultimately, it was learned that, because the egg shell has tiny pores, even uncracked eggs which sat for a time on a surface (nest) contaminated with *Salmonella* could themselves become contaminated. It is known also that the bacteria can be passed from the infected female chicken directly into the substance of the egg before the shell has formed around it.

Anyone may contract *Salmonella* food poisoning, but the disease is most serious in infants, the elderly, and individuals with weakened immune systems. In these individuals, the infection may spread from the intestines to the blood stream, and then to other body sites, causing **death** unless the person is treated promptly with **antibiotics**. In addition, people who have had part or all of their stomach or their spleens removed, or who have sickle cell anemia, **cirrhosis** of the liver, leukemia, lymphoma, **malaria**, louse-borne **relapsing fever**, or Acquired **Immunodeficiency Syndrome (AIDS)** are particularly susceptible to *Salmonella* food poisoning.

Causes and symptoms

Salmonella food poisoning can occur when someone drinks unpasteurized milk or eats undercooked chicken or eggs, or salad **dressings** or desserts which contain raw eggs. Even if *Salmonella*-containing foods such as chicken are thoroughly cooked, any food can become contaminated during preparation if conditions and equipment for food preparation are unsanitary.

Other foods can then be accidentally contaminated if they come into contact with infected surfaces. In addition, children have become ill after playing with turtles or iguanas, and then eating without washing their hands. Because the bacteria are shed in the feces for weeks after infection with *Salmonella*, poor hygiene can allow such a carrier to spread the infection to others.

Symptoms appear about one-two days after infection, and include **fever** (in 50% of patients), **nausea and vomiting**, **diarrhea**, and abdominal cramps and **pain**. The diarrhea is usually very liquid, and rarely contains mucus or blood. Diarrhea usually lasts for about four days. The illness usually ends in about five-seven days.

Serious complications are rare, occurring most often in individuals with other medical illnesses. Complications occur when the *Salmonella* bacteria make their way into the bloodstream (**bacteremia**). Once in the bloodstream, the bacteria can enter any organ system throughout the body, causing disease. Other infections which can be caused by *Salmonella* include:

- bone infections (osteomyelitis)
- joint infections (arthritis)
- infection of the sac containing the heart (pericarditis)
- infection of the tissues which cover the brain and spinal cord (meningitis)
- infection of the liver (hepatitis)
- lung infections (pneumonia)
- infection of aneurysms (aneurysms are abnormal outpouchings which occur in weak areas of the walls of blood vessels)
- infections in the center of already-existing tumors or cysts.

Diagnosis

Under appropriate laboratory conditions, *Salmonella* can be grown and then viewed under a microscope for identification. Early in the infection, the blood is far more likely to positively show a presence of the *Salmonella* bacterium when a sample is grown on a nutrient substance (culture) for identification purposes. Eventually, however, positive cultures can be obtained from the stool and in some cases from a **urine culture**.

Treatment

Even though Salmonella food poisoning is a bacterial infection, most practitioners do not treat simple cases with antibiotics. Studies have shown that using

KEY TERMS

Carrier—Someone who has an organism (bacteria, virus, fungi) in his or her body, without signs of illness. The individual may therefore pass the organism on to others.

Gastroenteritis—Inflammation of the stomach and intestines. Usually causes nausea, vomiting, diarrhea, abdominal pain and cramps.

antibiotics does not usually reduce the length of time that the patient is ill. Paradoxically, it appears that antibiotics do, however, cause the patient to shed bacteria in their feces for a *longer* period of time. In order to decrease the length of time that a particular individual is a carrier who can spread the disease, antibiotics are generally not given.

In situations where an individual has a more severe type of infection with *Salmonella* bacteria, a number of antibiotics may be used. Chloramphenicol was the first antibiotic successfully used to treat *Salmonella* food poisoning. It is still a drug of choice in developing countries because it is so inexpensive, although some resistance has developed to it. Ampicillin and trimethoprim-sulfonamide have been used successfully in the treatment of infections caused by chloramphenicol-resistant strains. Newer types of antibiotics, such as cephalosporin or quinolone, are also effective. These drugs can be given by mouth or through a needle in the vein (intravenously) for very ill patients. With effective antibiotic therapy, patients feel better in 24–48 hours, the temperature returns to normal in three-five days, and the patient is generally recovered by 10–14 days.

Alternative treatment

A number of alternative treatments have been recommended for food poisoning. One very effective treatment that is strongly recommended is supplementation with *Lactobacillus acidophilus*, *L. bulgaricus*, and/or *Bifidobacterium* to restore essential bacteria in the digestive tract. These preparations are available as powders, tablets, or capsules from health food stores; yogurt with live *L. acidophilus* cultures can also be eaten.

Fasting or a liquid-only diet is often used for food poisoning. Homeopathic treatment can work very effectively in the treatment of *Salmonella* food

poisoning. The appropriate remedy for the individual and his/her symptoms must be used to get the desired results. Some examples of remedies commonly used are *Chamomilla*, *Nux vomica*, *Ipecac*, and *Colchicum*.

Juice therapy, including carrot, beet, and garlic juices, is sometimes recommended, although it can cause discomfort for some people. Charcoal tablets can help absorb toxins and remove them from the digestive tract through bowel elimination. A variety of herbs with antibiotic action, including citrus seed extract, goldenseal (*Hydrastis canadensis*), and Oregon grape (*Mahonia aquifolium*), may also be effective in helping to resolve cases of food poisoning.

Prognosis

The prognosis for uncomplicated cases of *Salmonella* food poisoning is excellent. Most people recover completely within a week's time. In cases where other medical problems complicate the illness, prognosis depends on the severity of the other medical conditions, as well as the specific organ system infected with *Salmonella*.

Prevention

Prevention of *Salmonella* food poisoning involves the proper handling and cooking of foods likely to carry the bacteria. This means that recipes utilizing uncooked eggs (Caesar salad dressing, meringue toppings, mousses) need to be modified to eliminate the raw eggs. Not only should chicken be cooked thoroughly, until no pink juices flow, but all surfaces and utensils used on raw chicken must be carefully cleaned to prevent *Salmonella* from contaminating other foods. Careful handwashing is a must before, during, and after all food preparation involving eggs and poultry. Handwashing is also important after handling and playing with pets such as turtles, iguanas, chicks, dogs and cats.

Resources

BOOKS

- Forsythe, Stephen J. *The Microbiology of Safe Food*, 2nd ed. New York, NY: Wiley-Blackwell, 2010.
- Hwang, Andy., and Lihan Huand., eds. *Ready-to-Eat Foods: Microbial Concerns and Control Measures*. Boca Raton, FL: CRC Press, 2010.
- Landau, Elaine. *Food Poisoning and Foodborne Diseases (USA Today Health Reports: Diseases and Disorders.)* Minneapolis MN: Twenty-First Century Books, 2010.

ORGANIZATIONS

Centers for Disease Control and Prevention, 1600 Clifton Rd., NE, Atlanta, GA, 30333, (404) 639-3311, (800) 311-3435, <http://www.cdc.gov>.

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Laura Jean Cataldo, RN, Ed.D.

Salmonella paratyphi infection see
Paratyphoid fever

Salmonella typhi infection see **Typhoid fever**

Salpingectomy

Definition

Salpingectomy is the removal of one or both of a woman's fallopian tubes, the tubes through which an egg travels from the ovary to the uterus.

Purpose

A salpingectomy may be performed for several different reasons. Removal of one tube (unilateral salpingectomy) is usually performed if the tube has become infected (a condition known as salpingitis).

Salpingectomy is also used to treat an **ectopic pregnancy**, a condition in which a fertilized egg has implanted in the tube instead of inside the uterus. In most cases, the tube is removed only after drug treatments designed to save the structure have failed. (Women with one remaining fallopian tube are still able to get pregnant and carry a **pregnancy** to term.) The other alternative to salpingectomy is surgery to remove the fetus from the fallopian tube, followed by surgery to repair the tube.

A bilateral salpingectomy (removal of both the tubes) is usually done if the ovaries and uterus are also going to be removed. If the fallopian tubes and the ovaries are both removed at the same time, this is called a **salpingo-oophorectomy**. A salpingo-oophorectomy is necessary when treating ovarian and **endometrial cancer** because the fallopian tubes and ovaries are the most common sites to which **cancer** may spread.

Description

Regional or **general anesthesia** may be used. Often a laparoscope (a hollow tube with a light on one end) is used in this type of operation, which means

KEY TERMS

Ectopic pregnancy—The development of a fetus at a site other than the inside of the uterus; most commonly, the egg implants itself in the fallopian tube.

Laparoscope—A surgical instrument with a light attached that is inserted through the abdominal wall to allow the surgeon to see the organs in the abdomen.

that the incision can be much smaller and the recovery time much shorter.

In this procedure, the surgeon makes a small incision just beneath the navel. The surgeon inserts a short hollow tube into the abdomen and, if necessary, pumps in carbon dioxide gas in order to move intestines out of the way and better view the organs. After a wider double tube is inserted on one side for the laparoscope, another small incision is made on the other side through which other instruments can be inserted. After the operation is completed, the tubes and instruments are withdrawn. The tiny incisions are sutured and there is very little scarring.

In the case of a pelvic infection, the surgeon makes a horizontal (bikini) incision 4-6 in (10-15 cm) long in the abdomen right above the pubic hairline. This allows the doctor to remove the scar tissue. (Alternatively, a surgeon may use a vertical incision from the pubic bone toward the navel, although this is less common.)

Preparation

The patient is given an injection an hour before surgery to encourage drowsiness.

Aftercare

Aftercare varies depending on whether the tube was removed by **laparoscopy** or through an abdominal incision. Even when major surgery is performed, most women are out of bed and walking around within three days. Within a month or two, a woman can slowly return to normal activities such as driving, exercising, and working.

Risks

All surgery, especially under general anesthesia, carries certain risks, such as the risk of scarring, hemorrhaging, infection, and reactions to the anesthesia. Pelvic surgery can also cause internal scarring which can lead to discomfort years afterward.

ORGANIZATIONS

American College of Obstetricians and Gynecologists, PO Box 96920, Washington, DC, 20090-6920, (202) 638-5577, <http://www.acog.org>.

Carol A. Turkington

Salpingitis see **Pelvic inflammatory disease**

Salpingo-oophorectomy

Definition

Unilateral salpingo-oophorectomy is the surgical removal of a fallopian tube and an ovary. If both sets of fallopian tubes and ovaries are removed, the procedure is called a bilateral salpingo-oophorectomy.

Purpose

This surgery is performed to treat ovarian or other gynecological cancers, or infections caused by **pelvic inflammatory disease**. Occasionally, removal of one or both ovaries may be done to treat **endometriosis**, a condition in which the lining of the uterus (the endometrium) grows outside of the uterus (usually on and around the pelvic organs). The procedure may also be performed if a woman has been diagnosed with an **ectopic pregnancy** in a fallopian tube and a salpingostomy (an incision into the fallopian tube to remove the **pregnancy**) cannot be done. If only one fallopian tube and ovary are removed, the woman may still be able to conceive and carry a pregnancy to term. If both are removed, however, the woman is rendered permanently infertile. This procedure is commonly combined with a **hysterectomy** (surgical removal of the uterus); the ovaries and fallopian tubes are removed in about one-third of hysterectomies.

Until the 1980s, women over age 40 having hysterectomies routinely had healthy ovaries and fallopian tubes removed at the same time. Many physicians reasoned that a woman over 40 was approaching **menopause** and soon her ovaries would stop secreting estrogen and releasing eggs. Removing the ovaries would eliminate the risk of **ovarian cancer** and only accelerate menopause by a few years.

In the 1990s, the approach to routine salpingo-oophorectomy began to change. The risk of ovarian **cancer** in women who have no family history of the disease is less than 1%. Moreover, removing the ovaries increases the risk of cardiovascular disease and

KEY TERMS

BRCA1 or BRCA2 genetic mutation—A genetic mutation that predisposes otherwise healthy women to breast cancer.

Endometriosis—A painful disease in which cells from the lining of the uterus (endometrium) become attached to other organs in the pelvic cavity. The condition is hard to diagnose and often causes severe pain as well as infertility.

Fallopian tubes—Tubes that extend from either end of the uterus that convey the egg from the ovary to the uterus during each monthly cycle.

Hysterectomy—The surgical removal of the uterus.

Ureter—The tube that carries urine from the bladder to the kidneys.

accelerates **osteoporosis** unless a woman takes prescribed hormone replacements.

Demographics

Overall, ovarian cancer accounts for only 4% of all cancers in women. For women at increased risk, **oophorectomy** may be considered after the age of 35 if childbearing is complete. Factors that increase a woman's risk of developing ovarian cancer include age (most ovarian cancers occur after menopause), the presence of a mutation in the BRCA1 or BRCA2 gene, the number of menstrual periods a woman has had (affected by age of onset, pregnancy, **breastfeeding**, and oral contraceptive use), history of **breast cancer**, diet, and family history. The incidence of ovarian cancer is highest among Native American (17.5 cases per 100,000 population), Caucasian (15.8 per 100,000), Vietnamese (13.8 per 100,000), Hispanic (12.1 per 100,000), and Hawaiian (11.8 per 100,000) women; it is lowest among Korean (7.0 per 100,000) and Chinese (9.3 per 100,000) women. African American women have an ovarian cancer incidence of 10.2 per 100,000 population.

Endometriosis, another reason why salpingo-oophorectomy may be performed, has been estimated to affect up to 10% of women. Approximately four out of every 1,000 women are hospitalized as a result of endometriosis each year. Women 25–35 years of age are affected most, with 27 being the average age of diagnosis.

Description

General or regional anesthesia will be given to the patient before the procedure begins. If the procedure is

performed through a laparoscope, the surgeon can avoid a large abdominal incision and can shorten recovery. With this technique, the surgeon makes a small cut through the abdominal wall just below the navel. A tube containing a tiny lens and light source (a laparoscope) is then inserted through the incision. A camera can be attached that allows the surgeon to see the abdominal cavity on a video monitor. When the ovaries and fallopian tubes are detached, they are removed through a small incision at the top of the vagina. The organs can also be cut into smaller sections and removed. When the laparoscope is used, the patient can be given either regional or **general anesthesia**; if there are no complications, the patient can leave the hospital in a day or two.

If a laparoscope is not used, the surgery involves an incision 4–6 in (10–15 cm) long into the abdomen extending either vertically up from the pubic bone toward the navel, or horizontally (the “bikini incision”) across the pubic hairline. The scar from a bikini incision is less noticeable, but some surgeons prefer the vertical incision because it provides greater visibility while operating. A disadvantage to abdominal salpingo-oophorectomy is that bleeding is more likely to be a complication of this type of operation. The procedure is more painful than a laparoscopic operation and the recovery period is longer. A woman can expect to be in the hospital two to five days and will need three to six weeks to return to normal activities.

Diagnosis/preparation

Before surgery, the doctor will order blood and urine tests, and any additional tests such as ultrasound or x rays to help the surgeon visualize the woman's condition. The woman may also meet with the anesthesiologist to evaluate any special conditions that might affect the administration of anesthesia. A colon preparation may be done, if extensive surgery is anticipated.

On the evening before the operation, the woman should eat a light dinner, then take nothing by mouth, including water or other liquids, after midnight.

Aftercare

If performed through an abdominal incision, salpingo-oophorectomy is major surgery that requires three to six weeks for full recovery. However, if performed laparoscopically, the recovery time can be much shorter. There may be some discomfort around the incision for the first few days after surgery, but most women are walking around by the third day. Within a month or so, patients can gradually resume normal activities such as driving, exercising, and working.

Immediately following the operation, the patient should avoid sharply flexing the thighs or the knees. Persistent back **pain** or bloody or scanty urine indicates that a ureter may have been injured during surgery.

If both ovaries are removed in a premenopausal woman as part of the operation, the sudden loss of estrogen will trigger an abrupt **premature menopause** that may involve severe symptoms of hot flashes, vaginal dryness, painful intercourse, and loss of sex drive. (This is also called “surgical menopause.”) In addition to these symptoms, women who lose both ovaries also lose the protection these hormones provide against heart disease and osteoporosis many years earlier than if they had experienced natural menopause. Women who have had their ovaries removed are seven times more likely to develop coronary heart disease and much more likely to develop bone problems at an early age than are premenopausal women whose ovaries are intact. For these reasons, some form of **hormone replacement therapy** (HRT) may be prescribed to relieve the symptoms of surgical menopause and to help prevent heart and bone disease.

Reaction to the removal of fallopian tubes and ovaries depends on a wide variety of factors, including the woman’s age, the condition that required the surgery, her reproductive history, how much social support she has, and any previous history of depression. Women who have had many gynecological surgeries or chronic pelvic pain seem to have a higher tendency to develop psychological problems after the surgery.

Risks

Major surgery always involves some risk, including infection, reactions to the anesthesia, hemorrhage, and **scars** at the incision site. Almost all pelvic surgery causes some internal scars, which in some cases can cause discomfort years after surgery.

Potential complications after a salpingo-oophorectomy include changes in sex drive, hot flashes, and other symptoms of menopause if both ovaries are removed. Women who have both ovaries removed and who do not take estrogen replacement therapy run an increased risk for cardiovascular disease and osteoporosis. Women with a history of psychological and emotional problems before an oophorectomy are more likely to experience psychological difficulties after the operation.

Results

If the surgery is successful, the fallopian tubes and ovaries will be removed without complication, and the underlying problem resolved. In the case of cancer, all the cancer will be removed. A woman will become infertile following a bilateral salpingo-oophorectomy.

Morbidity and mortality rates

Studies have shown that the complication rate following salpingo-oophorectomy is essentially the same as that following hysterectomy. The rate of complications differs by the type of hysterectomy performed. Abdominal hysterectomy is associated with a higher rate of complications (9.3%), while the overall complication rate for vaginal hysterectomy is 5.3%, and 3.6% for laparoscopic vaginal hysterectomy. The risk of **death** is about one in every 1,000 (1/1,000) women having a hysterectomy. The rates of some of the more commonly reported complications are:

- excessive bleeding (hemorrhaging): 1.8–3.4%
- fever or infection: 0.8–4.0%
- accidental injury to another organ or structure: 1.5–1.8%

Because of the cessation of hormone production that occurs with a bilateral oophorectomy, women who lose both ovaries also prematurely lose the protection these hormones provide against heart disease and osteoporosis. Women who have undergone bilateral oophorectomy are seven times more likely to develop coronary heart disease and much more likely to develop bone problems at an early age than are premenopausal women whose ovaries are intact.

Alternatives

Depending on the specific condition that warrants an oophorectomy, it may be possible to modify the surgery so at least a portion of one ovary remains, allowing the woman to avoid early menopause. In the case of endometriosis, there are a number of alternative treatments that are usually pursued before a salpingo-oophorectomy (with or without hysterectomy) is performed. These include excising the growths without removing any organs, blocking or destroying the nerves that provide sensation to some of the pelvic structures, or prescribing drugs that decrease estrogen levels.

Resources

OTHER

- “BRCA1 and BRCA2: Cancer Risk and Genetic Testing.” National Cancer Institute (accessed February 8, 2010). <http://www.cancer.gov/cancertopics/factsheet/Risk/BRCA>
- “Oophorectomy (ovary removal surgery).” Mayo Clinic, April 16, 2009. <http://www.mayoclinic.com/health/oophorectomy/MY00554/METHOD=print>
- “Ovarian Cancer.” Medline Plus, November 5, 2009. <http://www.nlm.nih.gov/medlineplus/ency/article/000889.htm>

ORGANIZATIONS

- American Cancer Society, 1599 Clifton Road NE, Atlanta, GA, 30329-4251, (800) 227-2345, <http://www.cancer.org>.
- American College of Obstetricians and Gynecologists, 409 Twelfth Street SW, P.O. Box 96920, Washington, DC, 20090-6920, (202) 638-5577, <http://www.acog.org>.
- National Cancer Institute, Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD, 20892-2580, (800) 422-6237, <http://www.nci.nih.gov>.

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San Joaquin fever see **Coccidioidomycosis**

Sanfilippo's syndrome see

Mucopolysaccharidoses

Saquinavir see **Protease inhibitors**

The prevalence figures for sarcoidosis are very unusual. In the United States, prevalence figures range from five (5/100,000 in the United States) for whites to 40 for blacks. In Europe, prevalence ranges from three in Poland, to 10 in France, to 64 in Sweden, to 200 for Irish women living in London. Furthermore, a person from a group with very low prevalence who leaves his or her native land for a second location with a higher prevalence will then have the same risk as anyone living in that second location.

Sarcoidosis affects both men and women, although women are more likely to have the disorder. The average age for diagnosis is around 20–40 years.

Causes and symptoms

The cause of sarcoidosis is not known. Because the granulomas are primarily made up of cells from the immune system (macrophages and lymphocytes), an immune connection is strongly suspected. One of the theories which has been put forth suggests that exposure to some toxic or infectious material starts up an immune response. For some reason, the body is unable to stop the response, and it spreads from the original organ to other organs.

Because sarcoidosis has been noted to occur in family groups, a genetic cause has also been suggested. Research shows that identical twins are more likely to both have sarcoidosis than are nonidentical twins or other siblings.

Some cases of sarcoidosis occur without the patient even noting any symptoms. These cases are often discovered by chance during routine chest x rays. Most cases of sarcoidosis, however, begin with very nonspecific symptoms, such as decreased energy, weakness, and a dry **cough**. Occasionally, the cough is accompanied by some mild **pain** in the breastbone (sternum). Some patients note that they are having unusual **shortness of breath** while exercising. Some patients develop **fever**, decreased appetite, and weight loss.

Virtually every system of the body has the potential to suffer the effects of sarcoidosis:

- tender reddish bumps (nodules) or patches often appear on the skin
- the eyes may become red and teary, and the vision blurry
- the joints may become swollen and painful (arthritis)
- lymph nodes in the neck, armpits, and groin become enlarged and tender, lymph nodes within the chest, around the lungs, also become enlarged
- fluid may accumulate around the lungs (pleural effusion), making breathing increasingly difficult

Sarcoidosis

Definition

Sarcoidosis is a disease which can affect many organs within the body. It causes the development of granulomas. Granulomas are masses resembling little tumors. They are made up of clumps of cells from the immune system.

Description

Sarcoidosis is a very puzzling disorder. In addition to having no clear-cut understanding of the cause of sarcoidosis, researchers are also puzzled by its distribution in the world population. In the United States, for example, 10–17 times as many African-Americans are affected as white Americans. In Europe, whites are primarily affected.

Prevalence is a way of measuring the number of people affected per 100,000 people in a given population.

- nasal stuffiness is common, as well as a hoarse sound to the voice
- cysts in the bone may cause pain in the hands and feet, or in other bony areas
- the bone marrow may decrease the production of all blood cells; decreased number of red blood cells causes anemia, fewer white blood cells increases the chance of infections, fewer platelets can increase the chance of bleeding
- the body's ability to process calcium often becomes abnormal, so that excess calcium passes through the kidneys and into the urine; this may cause kidney stones to form
- the liver may become enlarged
- the heart may suffer a variety of complications, including abnormal or missed beats (arrhythmias), inflammation of the covering of the heart (pericarditis), and an increasing tendency toward weak, ineffective pumping of the blood (heart failure)
- the nervous system may display the effects of sarcoidosis by hearing loss, chronic inflammation of the coverings of the brain and spinal cord (meningitis), abnormalities of the nerve that is involved in vision (optic nerve dysfunction), seizures, and the development of psychiatric disorders

Any, all, or even none of the above symptoms may be present in sarcoidosis.

Diagnosis

Diagnosis depends on information from a number of sources, including the patient's symptoms, the **physical examination**, x-ray pictures of the chest, and a number of other laboratory examinations of blood or other tissue. None of these categories of information are sufficient to make the diagnosis of sarcoidosis. There is no one test or sign or symptom which clearly points to sarcoidosis, excluding all other types of diseases. This is because nearly all of the symptoms and laboratory results in sarcoidosis also occur in other diseases. Diagnosis, then, requires careful consideration of many facts.

The physical examination in sarcoidosis may reveal the characteristic **skin lesions**. Wheezes may be heard throughout the lungs. The liver may be enlarged. Examination of the eyes using a special light called a slit-lamp may reveal changes indicative of sarcoidosis.

The **chest x ray** will show some pattern of abnormalities, which may include enlargement of the lymph nodes which drain the lung, scarring and abnormalities to the tissue of the lungs, and fluid accumulation around the lungs.

KEY TERMS

Granuloma—Masses made up of a variety of immune cells, as well as fibroblasts (cells which make up connective tissue).

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body which work together to prevent foreign invaders (bacteria, viruses, fungi, etc.) from taking hold and growing.

Lung function tests measure such things as the amount of air an individual can breathe in and breathe out, the speed at which the air flows in and out, and the amount of air left in the lung after blowing out as much as possible in one second. A variety of lung function tests may show abnormal results in sarcoidosis.

Other types of tests may be abnormal in sarcoidosis. The abnormal test results may also indicate other diseases. They include an elevation of a substance called angiotensin-converting enzyme in the blood, and an increased amount of **calcium** present in 24 hours worth of urine.

Bronchoscopy is a very helpful diagnostic test. This involves passing a tiny tube (bronchoscope) through the nose or mouth, down the trachea, and into the airways (bronchial tubes). The bronchial tubes can be inspected through the bronchoscope. The bronchoscope is also designed in such a way as to allow biopsies to be obtained. Bronchoalveolar lavage involves washing the surfaces with a sterile saltwater (saline) solution. The saline is then retrieved and examined in a laboratory. Cells and debris from within the bronchial tubes and the tiny sacs of the lung (the alveoli) will be obtained in this way, and can be studied for the presence of an abnormally large number of white blood cells. A tiny piece of the lung tissue can also be obtained through the bronchoscope. This can be studied under a microscope to look for the characteristic granulomas and inflammation of sarcoidosis.

A gallium 67 scan involves the injection of a radioactive material called gallium 67. In sarcoidosis, areas of the body which are inflamed will retain the gallium 67. These areas will then show up on the scan.

Treatment

Many cases of sarcoidosis resolve without treatment. If treatment is needed, the most effective one for sarcoidosis is the administration of steroid medications. These medications work to decrease

inflammation throughout the body. The long-term use of steroid medications has serious potential side-effects. Patients are only treated with **steroids** when the problems caused by sarcoidosis are particularly serious. Many cases of sarcoidosis resolve without treatment.

Prognosis

The prognosis for sarcoidosis is quite good. About 60–70% of the time, sarcoidosis cures itself within a year or two. In about 20–30% of patients, permanent damage occurs to the lungs. About 15–20% of all patients go on to develop a chronic, relapsing form of sarcoidosis. **Death** can be blamed on sarcoidosis in about 10% of all sarcoidosis cases.

Prevention

Until researchers are able to pinpoint the cause of sarcoidosis, there will be no available recommendations for how to prevent it.

Resources

PERIODICALS

Zitkus, Bruce S. "Sarcoidosis: Varied Symptoms Often Impede Diagnosis of this Multisystem Disorder." *American Journal of Nursing* 97, no. 10 (October 1997): 40+.

Rosalyn Carson-DeWitt, MD



A specimen of a femur bone indicating the cancerous growth around the knee. Osteosarcoma is the most common primary cancer of the bone. (SPL/Photo Researchers, Inc.)

- bone cancers which originate in the hard material of the bone.
- soft-tissue sarcomas which begin in blood vessels, nerves, or tissues containing muscles, fat, or fiber.

Types of bone tumors

Osteogenic sarcoma, or osteosarcoma, is the most common form of bone cancer, accounts for 6% of all instances of the disease, and for about 5% of all cancers that occur in children. Nine hundred new cases of osteosarcoma are diagnosed in the United States every year. The disease usually affects teenagers, and is almost twice as common in boys as in girls.

Osteosarcomas, which grow very rapidly, can develop in any bone but most often occur along the edge or on the end of one of the fast-growing long bones that support the arms and legs. About 80% of all osteosarcomas develop in the parts of the upper and lower leg nearest the knee (the distal femur or in the proximal tibia). The next likely location for an osteosarcoma is the bone of the upper arm closest to the shoulder (the proximal humerus).

Ewing's sarcoma is the second most common form of childhood bone cancer. Accounting for fewer than 5% of bone tumors in children, Ewing's sarcoma usually begins in the soft tissue (the marrow) inside bones of the leg, hips, ribs, and arms. It rapidly infiltrates the lungs, and may metastasize to bones in other parts of the body.

More than 80% of patients who have Ewing's sarcoma are white, and the disease most frequently affects children between ages 5–9, and young adults between ages 20–30. About 27% of all cases of Ewing's

Sarcomas

Definition

A sarcoma is a bone tumor that contains **cancer** (malignant) cells. A benign bone tumor is an abnormal growth of noncancerous cells.

Description

A primary bone tumor originates in or near a bone. Most primary bone tumors are benign, and the cells that compose them do not spread (metastasize) to nearby tissue or to other parts of the body.

Malignant primary bone tumors account for fewer than 1% of all cancers diagnosed in the United States. They can infiltrate nearby tissues, enter the bloodstream, and metastasize to bones, tissues, and organs far from the original malignancy. Malignant primary bone tumors are characterized as either:

sarcoma occur in children under age 10, and 64% occur in adolescents between ages 10–20.

Chondrosarcomas are cancerous bone tumors that most often appear in middle age. Usually originating in strong connective tissue (cartilage) in ribs or leg or hip bones, chondrosarcomas grow slowly. They rarely spread to the lungs. It takes years for a chondrosarcoma to metastasize to other parts of the body, and some of these tumors never spread.

Parosteal osteogenic sarcomas, fibrosarcomas, and chordomas are rare. Parosteal osteosarcomas generally involve both the bone and the membrane that covers it. Fibrosarcomas originate in the ends of the bones in the arm or leg, and then spread to soft tissue. Chordomas develop on the skull or spinal cord.

Osteochondromas, which usually develop between age 10–20, are the most common noncancerous primary bone tumors. Giant cell tumors generally develop in a section of the thigh bone near the knee. Giant cell tumors are originally benign but sometimes become malignant.

Causes and symptoms

The cause of bone cancer is unknown, but the tendency to develop it may be inherited. Children who have bone tumors are often tall for their age, and the disease seems to be associated with growth spurts that occur during childhood and adolescence. Injuries can make the presence of tumors more apparent but do not cause them.

A bone that has been broken or exposed to high doses of radiation used to treat other cancers is more likely than other bones to develop osteosarcoma. A history of noncancerous bone disease also increases bone-cancer risk.

The amount of radiation in diagnostic x rays poses little or no danger of bone-cancer development, but children who have a family history of the most common childhood cancer of the eye (**retinoblastoma**), or who have inherited rare cancer syndromes have a greater-than-average risk of developing bone cancer. Exposure to chemicals found in some paints and dyes can slightly raise the risk.

Both benign and malignant bone tumors can distort and weaken bone and cause **pain**, but benign tumors are generally painless and asymptomatic.

It is sometimes possible to feel a lump or mass, but pain in the affected area is the most common early symptom of bone cancer. Pain is not constant in the initial stages of the disease, but it is aggravated by activity and may be worse at night. If the tumor is

located on a leg bone, the patient may limp. Swelling and weakness of the limb may not be noticed until weeks after the pain began.

Other symptoms of bone cancer include:

- a bone that breaks for no apparent reason
- difficulty moving the affected part of the body
- fatigue
- fever
- a lump on the trunk, an arm or leg, or another bone
- persistent, unexplained back pain
- weight loss

Diagnosis

Physical examination and routine x rays may yield enough evidence to diagnose benign bone tumors, but removal of tumor tissue for microscopic analysis (biopsy) is the only sure way to rule out malignancy.

A needle biopsy involves using a fine, thin needle to remove small bits of tumor, or a thick needle to extract tissue samples from the innermost part (the core) of the growth. An excisional biopsy is the surgical removal of a small, accessible tumor. An incisional biopsy is performed on tumors too large or inaccessible to be completely removed. The surgeon performing an incisional biopsy cuts into the patient's skin and removes a portion of the exposed tumor. Performed under local or general anesthetic, biopsy reveals whether a tumor is benign or malignant and identifies the type of cancer cells the malignant tumor contains.

Bone cancer is usually diagnosed about three months after symptoms first appear, and 20% of malignant tumors have metastasized to the lungs or other parts of the body by that time.

Imaging techniques

The following procedures are used, in conjunction with biopsy, to diagnose bone cancer:

- Bone x rays. These x rays usually provide a clear image of osteosarcomas.
- Computerized axial tomography (CAT scan) is a specialized x ray that uses a rotating beam to obtain detailed information about an abnormality and its physical relationship to other parts of the body. A CAT scan can differentiate between osteosarcomas and other types of bone tumors, illustrate how tumor cells have infiltrated other tissues, and help surgeons decide which portion of a growth would be best to biopsy. Because more than four of every five malignant bone tumors metastasize to the lungs, a CAT scan of the chest is performed to see if these

organs have been affected. Chest and abdominal CAT scans are used to determine whether Ewing's sarcoma has spread to the lungs, liver, or lymph nodes.

- Magnetic resonance imaging (MRI) is a specialized scan that relies on radio waves and powerful magnets to reflect energy patterns created by tissue abnormalities and specific diseases. An MRI provides more detailed information than does a CAT scan about tumors and marrow cavities of the bone, and can sometimes detect clusters of cancerous cells that have separated from the original tumor. This valuable information helps surgeons select the most appropriate approach for treatment.
- Radionuclide bone scans. These scans involve injecting a small amount of radioactive material into a vein. Primary tumors or cells that have metastasized absorb the radioactive material and show up as dark spots on the scan.

Cytogenetic and molecular genetic studies, which assess the structure and composition of chromosomes and genes, may also be used to diagnose osteosarcoma. These tests can sometimes indicate what form of treatment is most appropriate.

Laboratory studies

A **complete blood count** (CBC) reveals abnormalities in the blood, and may indicate whether bone marrow has been affected. A blood test that measures levels of the enzyme lactate dehydrogenase (LDH) can predict the likelihood of a specific patient's survival.

Immunohistochemistry involves adding special antibodies and chemicals, or stains, to tumor samples. This technique is effective in identifying cells that are found in Ewing's sarcoma but are not present in other malignant tumors.

Reverse transcription polymerase chain reaction (RT-PCR) relies on chemical analysis of the substance in the body that transmits genetic information (RNA) to:

- evaluate the effectiveness of cancer therapies
- identify mutations consistent with the presence of Ewing's sarcoma
- reveal cancer that recurs after treatment has been completed

Staging

Once bone cancer has been diagnosed, the tumor is staged. This process indicates how far the tumor has spread from its original location. The stage of a tumor suggests which form of treatment is most appropriate, and predicts how the condition will probably respond to therapy.

An osteosarcoma may be localized or metastatic. A localized osteosarcoma has not spread beyond the bone where it arose or beyond nearby muscles, tendons, and other tissues. A metastatic osteosarcoma has spread to the lungs, to bones not directly connected to the bone in which the tumor originated, or to other tissues or organs.

Treatment

Since the 1960s, when **amputation** was the only treatment for bone cancer, new **chemotherapy** drugs and innovative surgical techniques have improved survival with intact limbs. Because osteosarcoma is so rare, patients should consider undergoing treatment at a major cancer center staffed by specialists familiar with the disease.

A treatment plan for bone cancer, developed after the tumor has been diagnosed and staged, may include:

- Amputation. Amputation may be the only therapeutic option for large tumors involving nerves or blood vessels that have not responded to chemotherapy. MRI scans indicate how much of the diseased limb must be removed, and surgery is planned to create a cuff, formed of muscles and skin, around the amputated bone. Following surgery, an artificial (prosthetic) leg is fitted over the cuff. A patient who actively participates in the rehabilitation process may be walking independently as soon as three months after the amputation.
- Chemotherapy. Chemotherapy is usually administered in addition to surgery, to kill cancer cells that have separated from the original tumor and spread to other parts of the body. Although chemotherapy can increase the likelihood of later development of another form of cancer, the American Cancer Society maintains that the need for chemotherapeutic bone-cancer treatment is much greater than the potential risk.
- Surgery. Surgery, coordinated with diagnostic biopsy, enhances the probability that limb-salvage surgery can be used to remove the cancer while preserving nearby blood vessels and bones. A metal rod or bone graft is used to replace the area of bone removed, and subsequent surgery may be needed to repair or replace rods that have loosened or broken. Patients who have undergone limb-salvage surgery need intensive rehabilitation. It may take as long as a year for a patient to regain full use of a leg following limb-salvage surgery, and patients who have this operation may eventually have to undergo amputation.

- Radiation therapy. Radiation therapy is used often to treat Ewing's sarcoma.
- Rotationplasty. Rotationplasty, sometimes performed after a leg amputation, involves attaching the lower leg and foot to the thigh bone, so that the ankle replaces the knee. A prosthetic is later added to make the leg as long as it should be. Prosthetic devices are not used to lengthen limbs that remain functional after amputation to remove osteosarcomas located on the upper arm. When an osteosarcoma develops in the jaw bone, the entire lower jaw is removed. Bones from other parts of the body are later grafted on remaining bone to create a new jaw.

Follow-up treatments

After a patient completes the final course of chemotherapy, CAT scans, bone scans, x rays, and other diagnostic tests may be repeated to determine if any traces of tumor remain. If none are found, treatment is discontinued, but patients are advised to see their oncologist and orthopedic surgeon every two or three months for the next year. X rays of the chest and affected bone are taken every four months. An annual echocardiogram is recommended to evaluate any adverse effect chemotherapy may have had on the heart, and CT scans are performed every six months.

Patients who have received treatment for Ewing's sarcoma are examined often - at gradually lengthening intervals - after completing therapy. Accurate growth measurements are taken during each visit and blood is drawn to be tested for side effects of treatment. X rays, CT scans, bone scans, and other imaging studies are generally performed every three months during the first year. If no evidence of tumor growth or recurrence is indicated, these tests are performed less frequently in the following years.

Some benign bone tumors shrink or disappear without treatment. However, regular examinations are recommended to determine whether these tumors have changed in any way.

Alternative treatment

Alternative treatments should never be substituted for conventional bone-cancer treatments or used without the approval of a physician. However, some alternative treatments can be used as adjunctive and supportive therapies during and following conventional treatments.

Dietary adjustments can be very helpful for patients with cancer. Whole foods, including grains, beans, fresh fruits and vegetables, and high quality fats, should be emphasized in the diet, while processed foods should be avoided. Increased consumption of

fish, especially cold water fish like salmon, mackerel, halibut, and tuna, provides a good source of **omega-3 fatty acids**. **Nutritional supplements** can build strength and help maintain it during and following chemotherapy, radiation, or surgery. These supplements should be individually prescribed by an alternative practitioner who has experience working with cancer patients.

Many cancer patients claim that **acupuncture** alleviates pain, **nausea**, and **vomiting**. It can also be effective in helping to maintain energy and relative wellness during surgery, chemotherapy, and radiation. Massage, **reflexology**, and relaxation techniques are said to relieve pain, tension, **anxiety**, and depression. **Exercise** can be an effective means of reducing mental and emotional **stress**, while increasing physical strength. **Guided imagery**, **biofeedback**, hypnosis, body work, and progressive relaxation can also enhance quality of life.

Claims of effectiveness in fighting cancer have been made for a variety of herbal medicines. These botanical remedies work on an individual basis and should only be used when prescribed by a practitioner familiar with cancer treatment.

Treating cancer is a complex and individual task. It should be undertaken by a team of support practitioners with varying specialities who can work together for healing the person with cancer.

Prognosis

Benign brain tumors rarely recur, but sarcomas can reappear after treatment was believed to have eliminated every cell.

Likelihood of long-term survival depends on:

- the type and location of the tumor
- how much the tumor has metastasized, and on what organs, bones, or tissues have been affected

More than 85% of patients survive for more than five years after complete surgical removal of low-grade osteosarcomas (tumors that arise in mature tissue and contain a small number of cancerous cells). About 25–30% of patients diagnosed with high-grade osteosarcomas (tumors that develop in immature tissue and contain a large number of cancer cells) will die of the disease.

Two-thirds of all children diagnosed with Ewing's sarcoma will live for more than five years after the disease is detected. The outlook is most favorable for children under age 10, and least favorable in patients whose cancer is not diagnosed until after it has metastasized: fewer than three of every 10 of these patients remain alive five years later. More than 80% of patients whose Ewing's sarcoma is confined to a small area and surgically removed live, for at least

five years. Postsurgical radiation and chemotherapy add years to their lives. More than 70% of patients live five years or more with a small Ewing's sarcoma that cannot be removed, but only three out of five patients with large, unremovable tumors survive that long.

Prevention

There is no known way to prevent bone cancer.

ORGANIZATIONS

American Cancer Society, 1599 Clifton Rd. NE, Atlanta, GA, 30329 (800) 227-2345, <http://www.cancer.org>.

CancerCare, 275 Seventh Ave. Floor 22, New York, NY, 10001, (212) 712-8400, (212) 712-8495, (800) 813-4673, info@cancercare.org, <http://www.cancercare.org>.

National Cancer Institute (National Institutes of Health), NCI Office of Communications and Education, 6116 Executive Blvd. Suite 300, Bethesda, MD, 20892-8322, (800) 4-CANCER (422-6237), cancergovstaff@mail.nih.gov, <http://www.cancer.gov/>.

Maureen Haggerty

Saw palmetto

Definition

Saw palmetto, also known as Cabbage Palm and American dwarf palm, is a scrubby palm plant found in coastal regions of the southern United States and southern California.

Purpose

Saw palmetto is widely used in Europe to treat benign prostatic hypertrophy, and is the most popularly used herbal treatment for that condition in the United States. Some natural health practitioners use it to treat coughs and respiratory congestion.

In controlled clinical studies, the preponderance of evidence indicates that Saw palmetto is equally as effective, and better tolerated, at a tiny fraction of the cost, than brand drugs Proscar and Avodart for treating mild to moderate benign prostatic hypertrophy.

Saw palmetto does not act by reducing the size of the prostate gland. Like Proscar and Avodart, it



Saw palmetto leaves. (Photo Researchers, Inc.)

interferes with enzymes that transform testosterone, the male hormone, into a form that maintains and increases prostate size.

Preparations

Ripe Saw palmetto plant berries are used whole, as liquid extracts, dried and ground and placed in tablet or capsule form, or as a tea.

A typical dose is 320 mg per day.

Precautions

Though used as medicines, herbal products are regulated like dietary supplements in the United States. Manufacturers are responsible only for their production processes. Manufacturing standards, and combinations of herbs within herbal products, may vary.

Many herbal products sold in stores vary from stated label potency.

Side effects

Side effects of Saw palmetto are dose related and vary from mild abdominal discomfort to cramps, **nausea, vomiting** and **diarrhea**.

Interactions

Saw palmetto can increase the blood-thinning effects actions of Ginko, warfarin, **aspirin**, non steroid anti inflammatory drugs like ibuprofen, and Plavix.

Saw palmetto may alter the effectiveness of testosterone-replacement therapy and reduce the effectiveness of female hormone therapies like birth control.

Drug-herbal and herbal-herbal interactions are not well understood and have not been thoroughly tested. People must be careful observers of themselves as they take new drugs or herbs, or as they take these products regularly over many months.

Resources

OTHER

“Saw Palmetto.” *Herbs at a Glance*. National Center for Complementary and Alternative Medicine. <http://nccam.nih.gov/health/palmetto>.

James Waun, MD, RPh

Scabies

Definition

Scabies is a relatively contagious infection caused by a tiny mite (*Sarcoptes scabiei*).

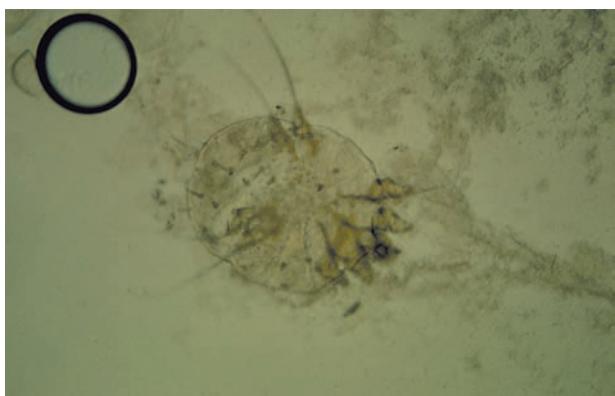
Demographics

Scabies is most common among people who live in overcrowded conditions, and whose ability to practice good hygiene is limited. Scabies can be passed between people by close skin contact. Although the mites can live only away from human skin for about three days, sharing clothing or bedclothes can pass scabies among family members or close contacts. In May 2002, the Centers for Disease Control (CDC) included scabies in its updated guidelines for the treatment of sexually transmitted infections (STI).

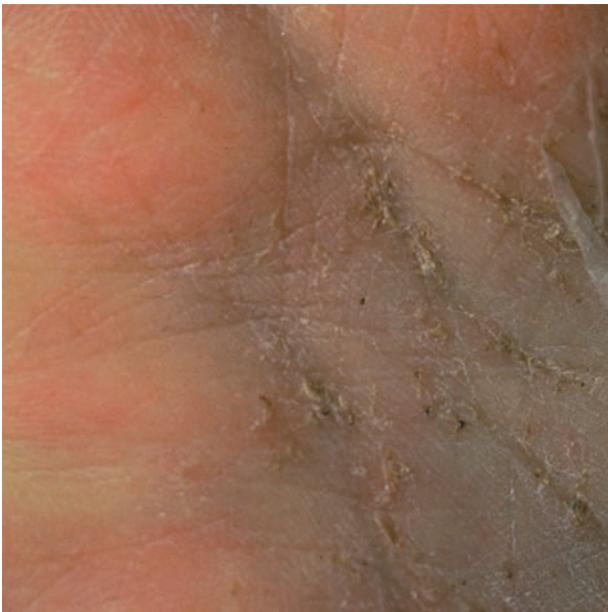
Description

Scabies is caused by a tiny insect about 0.3 mm long called a mite. When a human comes in contact with the female mite, the mite burrows under the skin, laying eggs along the line of its burrow. These eggs hatch, and the resulting offspring rise to the surface of the skin, mate, and repeat the cycle either within the skin of the original host, or within the skin of its next victim.

The intense **itching** almost always caused by scabies is due to a reaction within the skin to the feces of the mite. The first time someone is infected with scabies, he or she may not notice any itching for a number of weeks (four to six weeks). With subsequent infections, the itchiness will begin within hours of picking up the first mite.



An enhanced image of a scab mite. (Custom Medical Stock Photo, Inc. Reproduced by permission.)



Scab mites have penetrated under the skin of this person's hand. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Causes and symptoms

The itching, or pruritus, from scabies is worse after a hot shower and at night. Burrows are seen as winding, slightly raised gray lines along the skin. The female mite may be seen at one end of the burrow, as a tiny pearl-like bump underneath the skin. Because of the intense itching, burrows may be obscured by scratch marks left by the patient. The most common locations for burrows include the sides of the fingers, between the fingers, the top of the wrists, around the elbows and armpits, around the nipples of the breasts in women, in the genitalia of men, around the waist (beltline), and on the lower part of the buttocks. Babies may have burrows on the soles of their feet, palms of their hands, and faces.

Scratching seems to serve some purpose in scabies, as the mites are apparently often inadvertently removed. Most infestations with scabies are caused by no more than 15 mites altogether.

Infestation with huge numbers of mites (on the order of thousands to millions) occurs when an individual does not scratch, or when an individual has a weakened immune system. These patients include those who live in institutions; are mentally retarded, or physically infirm; have other diseases which affect the amount of sensation they have in their skin (**leprosy** or **syringomyelia**); have leukemia or diabetes; are taking medications which lower their immune

response (**cancerchemotherapy**, drugs given after organ transplantation); or have other diseases which lower their immune response (such as acquired **immunodeficiency** syndrome, or **AIDS**). This form of scabies, with its major infestation, is referred to as crusted scabies or Norwegian scabies. Infected patients have thickened, crusty areas all over their bodies, including over the scalp. Their skin is scaly. Their fingernails may be thickened and horny.

Diagnosis

Diagnosis can be made simply by observing the characteristic burrows of the mites causing scabies. A sterilized needle can be used to explore the pearly bump at the end of a burrow, remove its contents, and place it on a slide to be examined. The mite itself may then be identified under a microscope.

Occasionally, a type of mite carried on dogs (*Sarcoptes scabiei var. canis*) may infect humans. These mites cannot survive for very long on humans, and so the infection is very light.

Treatment

Several types of lotions (usually containing five percent permethrin) can be applied to the body, and left on for 12–24 hours. One topical application is usually sufficient, although the scabicide may be reapplied after a week if mites remain. Preparations containing lindane are no longer recommended for treating scabies because of the potential for damage to the nervous system by lindane. Itching can be lessened by the use of calamine lotion or antihistamine medications.

In addition to topical medications, the doctor may prescribe oral ivermectin. Ivermectin is a drug that was originally developed for veterinary practice as a broad-spectrum antiparasite agent. Studies done in humans, however, have found that ivermectin is as safe and effective as topical medications for treating scabies. A study published in 2003 reported that ivermectin is safe for people in high-risk categories, including those with compromised immune systems.

Prognosis

The prognosis for complete recovery from scabies infestation is excellent. In patients with weak immune systems, the biggest danger is that the areas of skin involved with scabies will become secondarily infected with bacteria.

KEY TERMS

- Mite**—An insect parasite belonging to the order Acarina. The organism that causes scabies is a mite.
- Pruritus**—An unpleasant itching sensation. Scabies is characterized by intense pruritus.
- Topical**—A type of medication applied to the skin or body surface.

Prevention

Good hygiene is essential in the prevention of scabies. When a member of a household is diagnosed with scabies, all that person's recently-worn clothing and bedding should be washed in very hot water.

Resources

BOOKS

Rainwater, Don. *Unexplained Skin Problems: Home Treatment And Precautions*. Charleston, SC: CreateSpace, 2009.

PERIODICALS

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- Burstein, G. R., and K. A. Workowski. "Sexually Transmitted Diseases Treatment Guidelines." *Current Opinion in Pediatrics* 15 (August 2003): 391–397.
- Fawcett, R. S. "Ivermectin Use in Scabies." *American Family Physician* 68 (September 15, 2003): 1089–1092.
- Santoro, A. F., M. A. Rezac, and J. B. Lee. "Current Trend in Ivermectin Usage for Scabies." *Journal of Drugs in Dermatology* 2 (August 2003): 397–401.

ORGANIZATIONS

American Academy of Dermatology (AAD), 930 East Woodfield Rd., Schaumburg, IL, 60173, (847) 330–0230, <http://www.aad.org>.

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Scarlatina see **Scarlet fever**

Scarlet fever

Definition

Scarlet fever is an infection that is caused by a bacteria called streptococcus. The disease is characterized by a **sore throat**, fever, and a sandpaper-like rash

on reddened skin. It is primarily a childhood disease. If scarlet fever is untreated, serious complications such as **rheumatic fever** (a heart disease) or kidney inflammation (**glomerulonephritis**) can develop.

Description

Scarlet fever, also known as scarlatina, gets its name from the fact that the patient's skin, especially on the cheeks, is flushed. A sore throat and raised rash over much of the body are accompanied by fever and sluggishness (lethargy). The fever usually subsides within a few days and recovery is complete by two weeks. After the fever is gone, the skin on the face and body flakes; the skin on the palms of the hands and soles of the feet peels more dramatically.

This disease primarily affects children ages two to ten. It is highly contagious and is spread by sneezing, coughing, or direct contact. The incubation period is three to five days, with symptoms usually beginning on the second day of the disease, and lasting from four to ten days.

Early in the twentieth century, severe scarlet fever epidemics were common. Today, the disease is rare. Although this decline is due in part to the availability of **antibiotics**, that is not the entire reason since the decline began before the widespread use of antibiotics. One theory is that the strain of bacteria that causes scarlet fever has become weaker with time.

Causes and symptoms

Scarlet fever is caused by Group A streptococcal bacteria (*S. pyogenes*). Group A streptococci can be highly toxic microbes that can cause **strep throat**, wound or skin infections, **pneumonia**, and serious kidney infections, as well as scarlet fever. The Group A streptococci are hemolytic bacteria, which means that the bacteria have the ability to lyse or break red blood cells. The strain of streptococcus that causes scarlet fever is slightly different from the strain that causes most strep throats. The scarlet fever strain of bacteria produces a toxin, called an erythrogenic toxin. This toxin is what causes the skin to flush.

The main symptoms and signs of scarlet fever are fever, lethargy, sore throat, and a bumpy rash that blanches under pressure. The rash appears first on the upper chest and spreads to the neck, abdomen, legs, arms, and in folds of skin such as under the arm or groin. In scarlet fever, the skin around the mouth tends to be pale, while the cheeks are flushed. The patient usually has a "strawberry tongue," in which inflamed bumps on the tongue rise above a bright red

GLADYS DICK (1881–1963)

Before 1922, not much was known about the then-endemic disease of scarlet fever, which primarily affected children in Europe and North America, killing about 25% of the children who contracted it. Additionally, scarlet fever had many complications, some of which were severe and could be crippling. Gladys Dick, with her husband, George Dick, successfully isolated the bacteria which caused scarlet fever, developed a test for human vulnerability to the disease, and devised preventive methods. The couple patented their findings, specifically the way their scarlet fever toxin and antitoxin were prepared, although this decision was controversial at the time.

In 1923, the Dicks published papers in which they proved that scarlet fever was caused by hemolytic streptococcus. Within a few years, the Dicks also published papers on how to prevent, test, diagnose, and treat scarlet fever. Their groundbreaking work ensured that the disease was finally understood and brought under control.

Dick and her husband announced the development of what came to be known as the Dick test in 1924. This skin test showed whether the patient was susceptible or immune to scarlet fever. The test involved injecting a toxin-containing substance in the arm and determining if the skin around the area became inflamed. If it did, the patient was vulnerable to scarlet fever. This test was also useful in predicting if pregnant women would develop puerperal infection during childbirth.

coating. Finally, dark red lines (called Pastia's lines) may appear in the creases of skin folds.

Diagnosis

Cases of scarlet fever are usually diagnosed and treated by pediatricians or family medicine practitioners. The chief diagnostic signs of scarlet fever are the characteristic rash, which spares the palms and soles of the feet, and the presence of a strawberry tongue in children. Strawberry tongue is rarely seen in adults.

The doctor will take note of the signs and symptoms to eliminate the possibility of other diseases. Scarlet fever can be distinguished from **measles**, a viral infection that is also associated with a fever and rash, by the quality of the rash, the presence of a sore throat in scarlet fever, and the absence of the severe eye inflammation and severe runny nose that usually accompany measles.

The doctor will also distinguish between a strep throat, a viral infection of the throat, and scarlet fever. With a strep infection, the throat is sore and appears beefy and red. White spots appear on the tonsils.

Lymph nodes under the jawline may swell and become tender. However, none of these symptoms are specific for strep throat and may also occur with a viral infection. Other signs are more characteristic of bacterial infections. For example, inflammation of the lymph nodes in the neck is typical in strep infections, but not viral infections. On the other hand, **cough**, **laryngitis**, and stuffy nose tend to be associated with viral infections rather than strep infections. The main feature that distinguishes scarlet fever from a mere strep throat is the presence of the sandpaper-red rash.

Laboratory tests are needed to make a definitive diagnosis of a strep infection and to distinguish a strep throat from a viral sore throat. One test that can be performed is a blood cell count. Bacterial infections are associated with an elevated **white blood cell count**. In viral infections, the white blood cell count is generally below normal.

A **throat culture** can distinguish between a strep infection and a viral infection. A throat swab from the infected person is brushed over a nutrient gel (a sheep blood agar plate) and incubated overnight to detect the presence of hemolytic bacteria. In a positive culture, a clear zone will appear in the gel surrounding the bacterium, indicating that a strep infection is present.

Treatment

Although scarlet fever will often clear up spontaneously within a few days, antibiotic treatment with either oral or injectable penicillin is usually recommended to reduce the severity of symptoms, prevent complications, and prevent spread to others. Antibiotic treatment will shorten the course of the illness in small children but may not do so in adolescents or adults. Nevertheless, treatment with antibiotics is important to prevent complications.

Since penicillin injections are painful, oral penicillin may be preferable. If the patient is unable to tolerate penicillin, alternative antibiotics such as erythromycin or clindamycin may be used. However, the entire course of antibiotics, usually 10 days, will need to be followed for the therapy to be effective. Because symptoms subside quickly, there is a temptation to stop therapy prematurely. It is important to take all of the pills in order to kill the bacteria. Not completing the course of therapy increases the risk of developing rheumatic fever and kidney inflammation.

If the patient is considered too unreliable to take all of the pills or is unable to take oral medication, daily injections of procaine penicillin can be given in the hip or thigh muscle. Procaine is an anesthetic that makes the injections less painful.

KEY TERMS

Clindamycin—An antibiotic that can be used instead of penicillin.

Erythrogenic toxin—A toxin or agent produced by the scarlet fever-causing bacteria that causes the skin to turn red.

Erythromycin—An antibiotic that can be used instead of penicillin.

Glomerulonephritis—A serious inflammation of the kidneys that can be caused by streptococcal bacteria; a potential complication of untreated scarlet fever.

Hemolytic bacteria—Bacteria that are able to burst red blood cells.

Lethargy—The state of being sluggish.

Pastia's lines—Red lines in the folds of the skin, especially in the armpit and groin, that are characteristic of scarlet fever.

Penicillin—An antibiotic that is used to treat bacterial infections.

Procaine penicillin—An injectable form of penicillin that contains an anesthetic to reduce the pain of the injection.

Rheumatic fever—A heart disease that is a complication of a strep infection.

Sheep blood agar plate—A petri dish filled with a nutrient gel containing red blood cells that is used to detect the presence of streptococcal bacteria in a throat culture. Streptococcal bacteria will lyse or break the red blood cells, leaving a clear spot around the bacterial colony.

Strawberry tongue—A sign of scarlet fever in which the tongue appears to have a red coating with large raised bumps.

Bed rest is not necessary, nor is **isolation** of the patient. **Aspirin** or Tylenol (**acetaminophen**) may be given for fever or relief of **pain**.

Prognosis

If treated promptly with antibiotics, full recovery is expected. Once a patient has had scarlet fever, they develop immunity and cannot develop it again.

Prevention

Avoiding exposure to children who have the disease will help prevent the spread of scarlet fever.

Resources

BOOKS

Cecil, Russell L., Lee Goldman, and D. A. Ausiello. *Cecil Medicine*. 23rd ed. Philadelphia: Saunders Elsevier, 2008.

Sally J. Jacobs, EdD

Description

A scar is a manifestation of the skin's healing process. After skin or tissue is wounded, the body releases collagen to mend the damage. Collagen, a protein, reattaches the damaged skin. As the wound heals, a temporary crust forms and covers it. The crust is a scab that protects the damaged area.

Causes of scars include cuts, sores, surgery, and **burns**. Severe **acne** and chicken pox may also scar skin. The degree that skin scars depends on more than the size and depth of the wound. Age also affects the process. The healing process is stronger in younger skin. That results in scars that are thicker than those of older people. Other factors affecting the type of scar are ethnicity, heredity, and the location of the injury.

Children are active and susceptible to cuts and injuries. They and people with fair complexions tend to get hypertrophic scars. While Asians and blacks are likely to have keloid scars, people from other ethnic groups also experience this form of scarring.

Keloid and hypertrophic scars have similar appearances. However, the keloid scar expands beyond the original wound.

The location of the wound also has an effect on its size. If the scar is located on places like the knee or shoulder, it will eventually widen because these areas are in motion.

Treatment could minimize a scar but will not erase the mark.

Scars

Definition

Scars are marks created during the healing of damage to the skin or tissues.

Causes and symptoms

Scarring is the natural process of repairing an open wound, injury, surgical incision, or other conditions like acne. Initially, a scar is red because blood vessels are created while the body forms scar tissue. The damaged area is covered by a protective scab that eventually falls off. The scar may become brown or pink. It generally fades over time and becomes less visible.

The healing process takes from one year to 18 months. Some scars heal naturally. Other scars require additional treatment.

Hypertrophic scars and keloids

Hypertrophic scars and **keloids** are caused by an over-active healing process. This produces an excessive amount of collagen at the wound site. Both types of scars are red, thick, and raised above the wound.

Hypertrophic scars do not extend beyond the wound site. The scar may itch and usually heals without professional treatment in about a year.

Keloids are large scars that could form after surgery, an injury, burn, or body **piercing**. This scarring often occurs on the ear lobe or chest. Sometimes keloids develop spontaneously.

The keloid is raised, rigid, and grows beyond the wound. The keloid can continue to grow. Scars are generally harmless, but may itch or feel tender. In addition, a person may feel self-conscious about the scar's appearance.

Contracture scars

Contracture scars are caused by the loss of a large section of skin due to burns or other injury. The scar contracts or tightens around the wound. This contraction could impact a person's mobility. If the scar deepens, it could affect muscles and nerves.

Acne scars

Acne scars may appear after the severe stage of acne, a skin condition usually caused by hormonal changes. The inflammatory condition is seen in adolescence, but acne can occur later in life.

Severe acne is triggered by clogged pores that cause bacteria to multiply. It occurs more frequently in adolescent boys than girls. If the acne is not treated, there could be scarring. The types of scars include pit-like pockmarks.

Diagnosis

Since visible scars could make people self-conscious, they will probably seek treatment rather than a diagnosis. Medical professionals who treat scars include dermatologists and plastic surgeons. Dermatologists are physicians who care for the skin. Their expertise includes three or more years of medical and surgical training.

Scar treatment is usually not covered by insurance. Cosmetic procedures, those done to improve a person's appearance, are considered elective surgery and are paid for by the patient. However, if scars cause a physical impairment, coverage may be issued. Examples of impairments include burn scars and keloids that restrict motion. For coverage to be approved, it is helpful for the primary care doctor to document the patient's case in writing.

Treatment

A scar is permanent and cannot be completely removed. However, treatment can alter a scar's appearance. These procedures range from the application of over-the-counter ointment to surgery. Scar treatment should start after an injury because wound care affects scarring. The wound should be cleaned and covered. Picking at the scab breaks the collagen and allows germs to enter the wound. Time also helps with healing. Scars become smaller, and the color fades.

However, additional treatment is required for some scars. While some procedures are more effective for keloids and hypertrophic scars, the procedure for acne scars is based on the type of scarring. Treatment for burn scars may include skin grafts surgery.

Surgery

The surgical procedure for scars is referred to as scar revision because the procedure modifies the scar's appearance. The cost of scar revision averages \$500–3,000 in 2010, according to <http://www.plasticsurgeons.com>.

This procedure works well on scars that are wide or long. Other treatments may be recommended for keloids because a surgical incision could cause a new scar and create another keloid. To reduce the risk of another scar, surgery may be followed by the injection of cortisone **steroids**.

Steroid Injections

Steroid injection is a singular form of treatment for scars, particularly keloid and hypertrophic scars.

Corticosteroids are an anti-inflammatory drug that helps to lessen the scar's red color and thickness. The treatment flattens the scar and helps with **itching**. Injection costs vary and could cost \$150 per scar, according to a member of the American Academy of Dermatologists (AAD).

Cryosurgery

Cryosurgery involves the freezing of tissue with a probe containing nitrous oxide. It is used to modify scars, especially keloid and hypertrophic scars. Treatment could cost \$175 per lesion, according to the AAD member.

Dermabrasion

Dermabrasion is the removal of a layer of the skin's surface. Scars including those caused by acne are smoothed or sanded by an instrument. The procedure costs approximately \$150 per treatment.

Silicone gel sheets

Silicone gel sheets can be purchased over-the-counter. The sheets are worn over the scar area to seal moisture. The treatment helps with itching and to reduce scar thickness and color. Cost of sheets for small **wounds** ranges from \$30–50.

Alternative treatment

Alternate methods of treating scars range from applying Vitamin E to massaging the skin. People should consult with a doctor or other health care professional before starting treatment involving contact with the scarred area.

These procedures include applying obtained Vitamin E, aloe vera, or cocoa butter to the scar. Vitamin E is sold as an oil or obtained by opening a vitamin capsule. Aloe is an African plant and is sold in capsule form and as a skin care product. Cocoa butter is a fat made from cacao seeds.

Those items are thought to help with healing so that a scar is less visible. However, time also helps to lessen the scar's appearance. Those substances should be applied only after a scar is well-healed.

Massaging mild scars is done to relax rigid scar tissue. The scar is massaged for about two minutes. Afterwards, Vitamin E oil is applied to the skin. The process should be discontinued if the area becomes sore or red.

Prognosis

The prognosis for scar treatment depends on factors including the type and severity of the scar. Keloids may return, and all scars are permanent. If treatment does not completely minimize a scar to the patient's satisfaction, the person can apply make-up to the scarred area.

Prevention

The primary way to prevent scarring is to avoid injuries. People should wear protective gear when participating in sports. Furthermore, acne should be treated before the condition reaches the severe stage.

If injured, a person should immediately treat the wound because this reduces the risk of scarring. The wound should be cleaned and covered. If stitches aren't needed, a butterfly bandage is effective at keeping the wound closed. Moreover, a balanced diet also helps with the healing process.

Picking at the scab should be avoided because this interferes with the healing process and raises the risk of scarring.

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ORGANIZATIONS

American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168-4014, (847) 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

American Academy of Facial Plastic and Reconstructive Surgery (AAFPRS), 310 South Henry Street, Alexandria, VA, 22314, (703) 299-9291, info@aafprs.org, <http://www.aafprs.org/>.

American Society of Plastic Surgeons, 444 E. Algonquin Rd, Arlington Heights, IL, 60005, (847) 228-9900, <http://www.plasticsurgery.org/>.

Liz Swain

Schatzki's ring see **Lower esophageal ring**

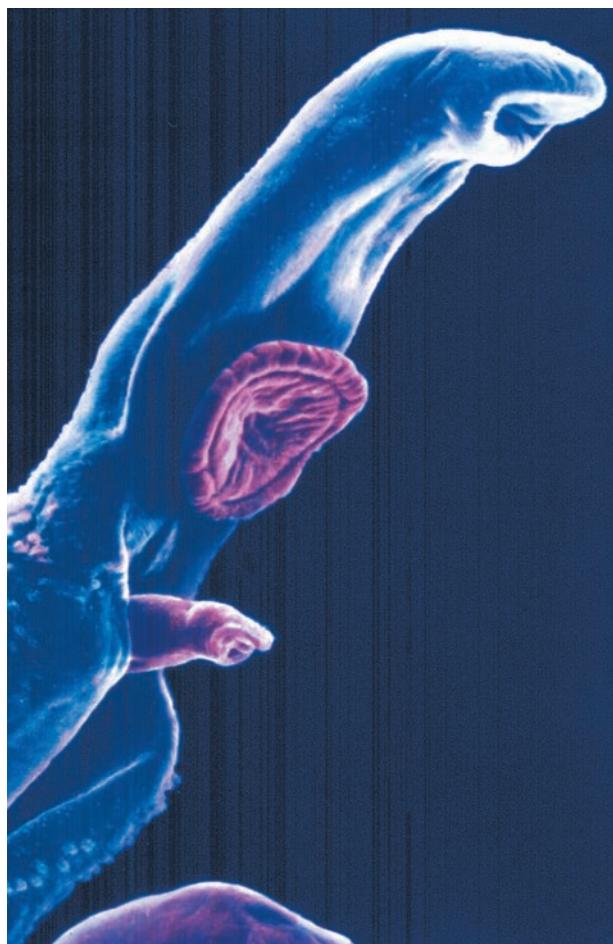
Schistosomiasis

Definition

Schistosomiasis, also known as bilharziasis or snail **fever**, is a primarily tropical parasitic disease caused by the larvae of one or more of five types of flatworms or blood flukes known as schistosomes. The name bilharziasis comes from Theodor Bilharz, a German pathologist, who identified the worms in 1851.

Description

Infections associated with worms present some of the most universal health problems in the world. In fact, only **malaria** accounts for more diseases than schistosomiasis. The World Health Organization (WHO)



A scanning electron microscopy (SEM) of the head region of the male and female adult flukes of *Schistosoma* sp. These worms cause schistosomiasis (bilharziasis) in humans. Flukes live in human blood vessels and their eggs contaminate freshwater. (Photo Researchers, Inc.)

estimates that 200 million people are infected and 120 million display symptoms. Another 600 million people are at risk of infection. Schistosomes are prevalent in rural and outlying city areas of 74 countries in Africa, Asia, and Latin America. In Central China and Egypt, the disease poses a major health risk.

There are five species of schistosomes that are prevalent in different areas of the world and produce somewhat different symptoms:

- *Schistosoma mansoni* is widespread in Africa, the Eastern-Mediterranean, the Caribbean, and South America and can only infect humans and rodents.
- *S. mekongi* is prevalent only in the Mekong river basin in Asia.
- *S. japonicum* is limited to China and the Philippines and can infect other mammals, in addition to humans, such as pigs, dogs, and water buffalos. As a result, it can be harder to control disease caused by this species.
- *S. intercalatum* is found in central Africa.
- *S. haematobium* occurs predominantly in Africa and the Eastern Mediterranean.

Intestinal schistosomiasis, caused by *Schistosoma japonicum*, *S. mekongi*, *S. mansoni*, and *S. intercalatum*, can lead to serious complications of the liver and spleen. Urinary schistosomiasis is caused by *S. haematobium*.

It is difficult to know how many individuals die of schistomiasis each year because **death** certificates and patient records seldom identify schistosomiasis as the primary cause of death. Mortality estimates vary related to the type of schistosome infection but is generally low, for example, 2.4 of 100,000 die each year from infection with *S. mansoni*.

Causes and symptoms

All five species are contracted in the same way, through direct contact with fresh water infested with the free-living form of the parasite known as cercariae. The building of dams, irrigation systems, and reservoirs, and the movements of refugee groups introduce and spread schistosomiasis.

Eggs are excreted in human urine and feces and, in areas with poor sanitation, contaminate freshwater sources. The eggs break open to release a form of the parasite called miracidium. Freshwater snails become infested with the miracidium, which multiply inside the snail and mature into multiple cercariae that the snail ejects into the water. The cercariae, which survive outside a host for 48 hours, quickly penetrate unbroken skin, the lining of the mouth, or the gastrointestinal tract. Once inside the human body, the worms penetrate the wall of the nearest vein and travel to

the liver where they grow and sexually mature. Mature male and female worms pair and migrate either to the intestines or the bladder where egg production occurs. One female worm may lay an average of 200 to 2,000 eggs per day for up to twenty years. Most eggs leave the blood stream and body through the intestines. Some of the eggs are not excreted, however, and can lodge in the tissues. It is the presence of these eggs, rather than the worms themselves, that causes the disease.

Early symptoms of infection

Many individuals do not experience symptoms. If present, it usually takes four to six weeks for symptoms to appear. The first symptom of the disease may be a general ill feeling. Within twelve hours of infection, an individual may complain of a **tingling** sensation or light rash, commonly referred to as "swimmer's itch," due to irritation at the point of entrance. The rash that may develop can mimic **scabies** and other types of **rashes**. Other symptoms can occur two to ten weeks later and can include fever, aching, **cough**, **diarrhea**, or gland enlargement. These symptoms can also be related to avian schistosomiasis, which does not cause any further symptoms in humans.

Katayama fever

Another primary condition, called Katayama fever, may also develop from infection with these worms, and it can be very difficult to recognize. Symptoms include fever, lethargy, the eruption of pale temporary bumps associated with severe **itching** (urticarial) rash, liver and spleen enlargement, and bronchospasm.

Intestinal schistosomiasis

In intestinal schistosomiasis, eggs become lodged in the intestinal wall and cause an immune system reaction called a granulomatous reaction. This immune response can lead to obstruction of the colon and blood loss. The infected individual may have what appears to be a pot-belly. Eggs can also become lodged in the liver, leading to high blood pressure through the liver, enlarged spleen, the build-up of fluid in the abdomen (**ascites**), and potentially life-threatening dilations or swollen areas in the esophagus or gastrointestinal tract that can tear and bleed profusely (esophageal varices). Rarely, the central nervous system may be affected. Individuals with chronic active schistosomiasis may not complain of typical symptoms.

Urinary tract schistosomiasis

Urinary tract schistosomiasis is characterized by blood in the urine, **pain** or difficulty urinating, and

frequent urination and are associated with *S. haematobium*. The loss of blood can lead to **iron deficiency anemia**. A large percentage of persons, especially children, who are moderately to heavily infected experience urinary tract damage that can lead to blocking of the urinary tract and **bladder cancer**.

Diagnosis

Proper diagnosis and treatment may require a tropical disease specialist because the disease can be confused with malaria or typhoid in the early stages. The healthcare provider should do a thorough history of travel in endemic areas. The rash, if present, can mimic scabies or other rashes, and the gastrointestinal symptoms may be confused with those caused by bacterial illnesses or other intestinal parasites. These other conditions will need to be excluded before an accurate diagnosis can be made. As a result, clinical evidence of exposure to infected water along with physical findings, a negative test for malaria, and an increased number of one type of immune cell, called an eosinophil, are necessary to diagnose acute schistosomiasis.

Eggs may be detected in the feces or urine. Repeated stool tests may be required to concentrate and identify the eggs. Blood tests may be used to detect a particular antigen or particle associated with the schistosome that induces an immune response. Persons infected with schistosomiasis may not test positive for six months, and as a result, tests may need to be repeated to obtain an accurate diagnosis. Blood can be detected visually in the urine or with chemical strips that react to small amounts of blood.

Sophisticated imaging techniques, such as ultrasound, computed tomography scan (CT scan), and **magnetic resonance imaging** (MRI), can detect damage to the blood vessels in the liver and visualize polyps and ulcers of the urinary tract, for example, that occur in the more advanced stages. *S. haematobium* is difficult to diagnose with ultrasound in pregnant women.

Treatment

The use of medications against schistosomiasis, such as praziquantel (Biltricide), oxamniquine, and metrifonate, have been shown to be safe and effective. Praziquantel is effective against all forms of schistosomiasis and has few side effects. This drug is given in either two or three doses over the course of a single day. Oxamniquine is typically used in Africa and South America to treat intestinal schistosomiasis. Metrifonate has been found to be safe and effective in the treatment of urinary schistosomiasis. Patients are typically checked for the presence of living eggs at

KEY TERMS

Ascites—The condition that occurs when the liver and kidneys are not functioning properly and a clear, straw-colored fluid is excreted by the membrane that lines the abdominal cavity (peritoneum).

Cercariae—The free-living form of the schistosome worm that has a tail, swims, and has suckers on its head for penetration into a host.

Miracidium—The form of the schistosome worm that infects freshwater snails.

three and six months after treatment. If the number of eggs excreted has not significantly decreased, the patient may require another course of medication.

Prognosis

If treated early, prognosis is very good and complete recovery is expected. The illness is treatable, but people can die from the effects of untreated schistosomiasis. The severity of the disease depends on the number of worms, or worm load, in addition to how long the person has been infected. With treatment, the number of worms can be substantially reduced, and the secondary conditions can be treated. The goal of the World Health Organization is to reduce the severity of the disease rather than to completely stop transmission of the disease. There is, however, little natural immunity to reinfection. Treated individuals do not usually require retreatment for two to five years in areas of low transmission. The World Health Organization has made research to develop a vaccine against the disease one of its priorities.

Prevention

Prevention of the disease involves several targets and requires long term community commitment. Infected patients require diagnosis, treatment, and education about how to avoid reinfecting themselves and others. Adequate healthcare facilities need to be available, water systems must be treated to kill the worms and control snail populations, and sanitation must be improved to prevent the spread of the disease.

To avoid schistosomiasis in endemic areas:

- contact the CDC for current health information on travel destinations.
- upon arrival, ask an informed local authority about the infestation of schistosomiasis before being exposed

to freshwater in countries that are likely to have the disease.

- do not swim, stand, wade, or take baths in untreated water.
- treat all water used for drinking or bathing. Water can be treated by letting it stand for three days, heating it for five minutes to around 122°F (around 50°C), or filtering or treating water chemically, with chlorine or iodine, as with drinking water.
- Should accidental exposure occur, infection can be prevented by hastily drying off or applying rubbing alcohol to the exposed area.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Ruth E. Mawyer, RN

Schizencephaly see **Congenital brain defects**

Schizoaffective disorder

Definition

Schizoaffective disorder is a mental illness that shares the psychotic symptoms of **schizophrenia** and the mood disturbances of depression or **bipolar disorder**.

Demographics

Females tend to suffer from schizoaffective disorder more so than men. However, due to the broad clinical manifestations associated with this mental illness, the actual rate of schizoaffective disorder in adults is unknown.

Description

The term schizoaffective disorder was first used in the 1930s to describe patients with acute psychotic symptoms such as **hallucinations** and **delusions** along with disturbed mood. These patients tended to function well before becoming psychotic; their psychotic symptoms lasted relatively briefly; and they tended to do well afterward. Over the years, however, the term schizoaffective disorder has been applied to a variety of patient groups. The current definition contained in the American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders IV (DSM-IV)* recognizes patients with schizoaffective disorder as those whose mood symptoms are

sufficiently severe to warrant a diagnosis of depression or other full-blown mood disorder and whose mood symptoms overlap at some period with psychotic symptoms that satisfy the diagnosis of schizophrenia (e.g. hallucinations, delusions, or thought process disorder).

Causes and symptoms

The cause of schizoaffective disorder remains unknown and subject to continuing speculation. Some investigators believe schizoaffective disorder is associated with schizophrenia and may be caused by a similar biological predisposition. Others disagree, stressing the disorder's similarities to **mood disorders** such as depression and bipolar disorder (manic depression). They believe its more favorable course and less intense psychotic episodes are evidence that schizoaffective disorder and mood disorders share a similar cause.

Many researchers, however, believe schizoaffective disorder may owe its existence to both disorders. These researchers believe that some people have a biologic predisposition to symptoms of schizophrenia that varies along a continuum of severity. On one end of the continuum are people who are predisposed to psychotic symptoms but never display them. On the other end of the continuum are people who are destined to develop outright schizophrenia. In the middle are those who may at some time show symptoms of schizophrenia, but require some other major trauma to set the progression of the disease into motion. It may be an early brain injury, either through a complicated delivery, prenatal exposure to the flu virus or illicit drugs; or it may be emotional, nutritional, or other type of deprivation in early childhood. In this view, major life stresses, or a mood disorder like depression or bipolar disorder, may be sufficient to trigger the psychotic symptoms. In fact, patients with schizoaffective disorder frequently experience depressed mood or **mania** within days of the appearance of psychotic symptoms. Some clinicians believe that "schizomanic" patients are fundamentally different from "schizodepressed" types; the former are similar to bipolar patients, while the latter are a very heterogeneous group.

Symptoms of schizoaffective disorder vary considerably from patient to patient. Delusions, hallucinations, and evidence of disturbances in thinking—as observed in full-blown schizophrenia—may be seen. Similarly, mood fluctuations such as those observed in major depression or bipolar disorder may also be seen. These symptoms tend to appear in distinct episodes

that impair the individual's ability to function well in daily life. But between episodes, some patients with schizoaffective disorder remain chronically impaired while some may do quite well in day-to-day living.

Diagnosis

There are no accepted tissue or brain imaging tests or techniques to diagnose schizophrenia, mood disorders, or schizoaffective disorder. Instead, physicians look for the hallmark signs and symptoms of schizoaffective disorder described above, and they attempt to rule out other illnesses or conditions that may produce similar symptoms. These include:

- Mania. True manic patients can experience episodes of hallucinations and delusions similar to those seen in schizoaffective disorder; but these episodes do not persist for long periods after the mania recedes, as they do in schizoaffective disorder.
- Psychotic depression. Patients with psychotic depression experience hallucinations and delusions similar to those seen in schizoaffective disorder; but these symptoms do not persist after the depressive symptoms recede, as they do in schizoaffective disorder.
- Schizophrenia. Depressed mood, mania, or other symptoms may be present in patients with schizophrenia, but patients with schizoaffective disorder meet all the criteria set out for a full-blown mood disorder.
- Medical and neurological disorders that mimic psychotic/affective disorders.

Treatment

Antipsychotic medications used to treat schizophrenia and the **antidepressant drugs** and mood stabilizers used in depression and bipolar disorder are the primary treatments for schizoaffective disorder.

These treatments have not been well studied in controlled investigations. Studies suggest that traditional antipsychotics such as haloperidol are effective in treating psychotic symptoms. Newer generation antipsychotics, such as clozaril and risperidone, have not been as well studied, but also appear effective. For patients with symptoms of bipolar disorder, lithium is often the mood stabilizer of choice, and it is often augmented with an anticonvulsant such as valproate. For those with depressive symptoms, the evidence supporting the use of antidepressant medications in addition to antipsychotic medications is more mixed. **Electroconvulsive therapy** (electric shock) is frequently tried in patients who otherwise do not respond to antidepressant or mood stabilizing drugs.

KEY TERMS

Bipolar disorder—A mood disorder marked by alternating episodes of extremely low mood (depression) and exuberant highs (mania). Also known as manic-depression disorder.

Mood disorder—A collection of disorders that includes major depression and bipolar disorder. They are all characterized by major disruptions in patients' moods and emotions.

Schizophrenia—A major mental illness marked by psychotic symptoms, including hallucinations, delusions, and severe disruptions in thinking.

While the mainstay of treatment for schizoaffective disorder is antipsychotic medications and mood stabilizers, certain forms of **psychotherapy** for both patients and family members can be useful. Therapy designed to provide structure and help augment patients' ability to solve problems may aid in improving patients' ability to function in the day-to-day world, reducing **stress** and the risk of recurrence. Vocational and other rehabilitative training can help patients to work on skills they need to develop. Whereas hospitalization may be necessary for acute psychotic episodes, halfway houses and day hospitals can provide needed treatment while serving as a bridge for patients to reenter the community.

Alternative treatment

While alternative therapies should never be considered a replacement for medication, these treatments can help support people with schizoaffective disorder and other mental illnesses. Dietary modifications that eliminate processed foods and emphasize whole foods, along with nutritional supplementation, may be helpful. **Acupuncture**, homeopathy, and botanical medicine can support many aspects of the person's life and may help decrease the side effects of any medications prescribed.

Prognosis

In general, patients with schizoaffective disorder have a more favorable prognosis than do those with schizophrenia, but a less favorable course than those with a pure mood disorder. Medication and other interventions can help quell psychotic symptoms and

stabilize mood in many patients, but there is great variability in outcome from patient to patient.

Prevention

There is no known way to prevent schizoaffective disorder. Treatment with antipsychotic and mood stabilizing drugs may prevent recurrences. Some researchers believe prompt treatment can prevent the development of full-blown schizophrenia, but this hypothesis remains the subject of some disagreement.

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ORGANIZATIONS

American Psychiatric Association (APA), 1000 Wilson Blvd., Suite 1825, Arlington, VA, 22209, (703) 907-7300, apa@psych.org, <http://www.psych.org/>.

American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (202) 336-5700, <http://www.apa.org>.

National Alliance for Research on Schizophrenia and Depression (NARSAD), 60 Cutter Mill Rd., Suite 404, Great Neck, NY, 11021, (516) 829-0091, <http://www.mhsource.com>.

National Alliance on Mental Illness (NAMI), Colonial Place Three, 2107 Wilson Blvd., Suite 300, Arlington, VA, 22201, (703) 524-7600, (800) 950-NAMI (6264), (703) 524-9094, <http://www.nami.org/Hometemplate.cfm>.

National Institute of Mental Health (NIMH), 6001 Executive Blvd., Room 8184, MSC 9663, Bethesda, MD, 20892, (301) 443-4513, (866) 615-6464, (301) 443-4279, nimhinfo@nih.gov, <http://www.nimh.nih.gov/index.shtml>.

National Mental Health Association (NMHA), 2000 N. Beauregard St., 6th Floor, Alexandria, VA, 22311, (703) 684-7722, (800) 969-NMHA, (703) 684-5968, <http://www1.nmha.org/>.

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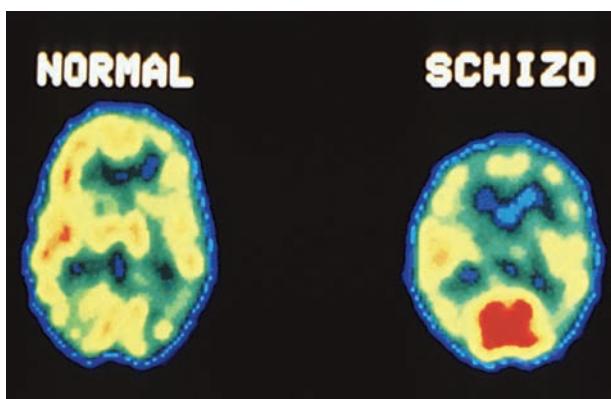
Schizophrenia

Definition

Schizophrenia is a psychotic disorder (or a group of disorders) marked by severely impaired thinking, emotions, and behaviors. Schizophrenic patients are typically unable to filter sensory stimuli and may have enhanced perceptions of sounds, colors, and other features of their environment. Most schizophrenics, if untreated, gradually withdraw from interactions with other people and lose their ability to take care of personal needs and grooming.

Demographics

According to the World Health Organization (WHO), schizophrenia is estimated to afflict about 24 million people worldwide, mostly in the 15–35 year age group. Though the incidence is low (3 in 10,000), the prevalence is high due to chronicity. It is also estimated that more than 50% of persons with schizophrenia are not receiving appropriate care and that 90% of people with untreated schizophrenia live in developing countries. Schizophrenia affects 2.2 million people in the United States, some 280,000 in



Positron emission tomography (PET) scans comparing a normal brain (left) with the brain of a schizophrenic. (Photo Researchers, Inc.)

Canada, 285,000 in Australia, and 250,000 in the United Kingdom. It ranks among the top 10 causes of disability in developed countries worldwide.

The disease typically begins in early adulthood between the ages of 15 and 25. Men tend to develop schizophrenia slightly earlier than women: most men become ill between 16 and 25 years old, while most women develop symptoms several years later. The average age of onset is 18 in men and 25 in women. Schizophrenia onset is quite rare in children under the age of 10, and in people over 40 years of age.

Description

The course of schizophrenia in adults can be divided into three phases or stages. In the acute phase, the patient has an overt loss of contact with reality (psychotic episode) that requires intervention and treatment. In the second or stabilization phase, the initial psychotic symptoms have been brought under control but the patient is at risk for relapse if treatment is interrupted. In the third or maintenance phase, the patient is relatively stable and can be kept indefinitely on antipsychotic medications. Even in the maintenance phase, however, relapses are not unusual and patients do not always return to full functioning.

The term schizophrenia comes from two Greek words that mean “split mind.” It was observed around 1908, by a Swiss doctor named Eugen Bleuler, to describe the splitting apart of mental functions that he regarded as the central characteristic of schizophrenia.

Recently, some psychotherapists have begun to use a classification of schizophrenia based on two main types. People with Type I, or positive schizophrenia, have a rapid (acute) onset of symptoms and tend to respond well to drugs. They also tend to suffer more from the “positive” symptoms, such as **delusions** and **hallucinations**. People with Type II, or negative schizophrenia, are usually described as poorly adjusted before their schizophrenia slowly overtakes them. They have predominantly “negative” symptoms, such as withdrawal from others and a slowing of mental and physical reactions (psychomotor retardation).

The fourth (1994) edition of the *Diagnostic and Statistical Manual of Mental Disorders (DSM-IV)* specifies five subtypes of schizophrenia.

Paranoid

The key feature of this subtype of schizophrenia is the combination of false beliefs (delusions) and hearing voices (auditory hallucinations), with more nearly normal emotions and cognitive functioning (cognitive functions include reasoning, judgment, and memory).

The delusions of paranoid schizophrenics usually involve thoughts of being persecuted or harmed by others or exaggerated opinions of their own importance, but may also reflect feelings of jealousy or excessive religiosity. The delusions are typically organized into a coherent framework. Paranoid schizophrenics function at a higher level than other subtypes, but are at risk for suicidal or violent behavior under the influence of their delusions.

Disorganized

Disorganized schizophrenia (formerly called hebephrenic schizophrenia) is marked by disorganized speech, thinking, and behavior on the patient's part, coupled with flat or inappropriate emotional responses to a situation (affect). The patient may act silly or withdraw socially to an extreme extent. Most patients in this category have weak personality structures prior to their initial acute psychotic episode.

Catatonic

Catatonic schizophrenia is characterized by disturbances of movement that may include rigidity, stupor, agitation, bizarre posturing, and repetitive imitations of the movements or speech of other people. These patients are at risk for **malnutrition**, exhaustion, or self-injury. This subtype is presently uncommon in Europe and the United States. **Catatonia** as a symptom is most commonly associated with **mood disorders**.

Undifferentiated

Patients in this category have the characteristic positive and negative symptoms of schizophrenia but do not meet the specific criteria for the paranoid, disorganized, or catatonic subtypes.

Residual

This category is used for patients who have had at least one acute schizophrenic episode but do not presently have strong positive psychotic symptoms, such as delusions and hallucinations. They may have negative symptoms, such as withdrawal from others, or mild forms of positive symptoms, which indicate that the disorder has not completely resolved.

Risk factors

The risk of schizophrenia among first-degree biological relatives is ten times greater than that observed in the general population. The incidence of schizophrenia in most developed countries also appears to be higher among people born in cities than among those born in rural areas.

Causes and symptoms

One of the reasons for the ongoing difficulty in classifying schizophrenic disorders is incomplete understanding of their causes. As of 2009, it is thought that these disorders are the end result of a combination of genetic, neurobiological, and environmental causes. A leading neurobiological hypothesis looks at the connection between the disease and excessive levels of dopamine, a chemical that transmits signals in the brain (neurotransmitter). The genetic factor in schizophrenia has been underscored by recent findings that first-degree biological relatives of schizophrenics are ten times as likely to develop the disorder as are members of the general population.

Prior to recent findings of abnormalities in the brain structure of schizophrenic patients, several generations of psychotherapists advanced a number of psychoanalytic and sociological theories about the origins of schizophrenia. These theories ranged from hypotheses about the patient's problems with **anxiety** or aggression to theories about **stress** reactions or interactions with disturbed parents. Psychosocial factors are now thought to influence the expression or severity of schizophrenia, rather than cause it directly.

Another hypothesis suggests that schizophrenia may be caused by a virus that attacks the hippocampus, a part of the brain that processes sense perceptions. Damage to the hippocampus would account for schizophrenic patients' vulnerability to sensory overload.

Symptoms of schizophrenia

Patients with a possible diagnosis of schizophrenia are evaluated on the basis of a set or constellation of symptoms; there is no single symptom that is unique to schizophrenia. In 1959, the German psychiatrist Kurt Schneider proposed a list of so-called first-rank symptoms, which he regarded as diagnostic of the disorder.

These symptoms include:

- delusions
- somatic hallucinations
- hearing voices commenting on the patient's behavior
- thought insertion or thought withdrawal

Somatic hallucinations refer to sensations or perceptions concerning body organs that have no known medical cause or reason, such as the notion that one's brain is radioactive. Thought insertion and/or withdrawal refers to delusions that an outside force (for example, the FBI, the CIA, Martians, etc.) has the power to put thoughts into one's mind or remove them.

Positive symptoms

The positive symptoms of schizophrenia are those that represent an excessive or distorted version of normal functions. Positive symptoms include Schneider's first-rank symptoms as well as disorganized thought processes (reflected mainly in speech) and disorganized or catatonic behavior. Disorganized thought processes are marked by such characteristics as looseness of associations, in which the patient rambles from topic to topic in a disconnected way; tangentially, which means that the patient gives unrelated answers to questions; and "word salad," in which the patient's speech is so incoherent that it makes no grammatical or linguistic sense. Disorganized behavior means that the patient has difficulty with any type of purposeful or goal-oriented behavior, including personal self-care or preparing meals. Other forms of disorganized behavior may include dressing in odd or inappropriate ways, sexual self-stimulation in public, or agitated shouting or cursing.

Negative symptoms

The *DSM-IV* definition of schizophrenia includes three so-called negative symptoms. They are called negative because they represent the lack or absence of behaviors. The negative symptoms that are considered diagnostic of schizophrenia are a lack of emotional response (affective flattening), poverty of speech, and absence of volition or will. In general, the negative symptoms are more difficult for doctors to evaluate than the positive symptoms.

Diagnosis

Examination

A doctor must make a diagnosis of schizophrenia on the basis of a standardized list of outwardly observable symptoms, not on the basis of internal psychological processes. There are no specific laboratory tests that can be used to diagnose schizophrenia. Researchers have, however, discovered that patients with schizophrenia have certain abnormalities in the structure and functioning of the brain compared to normal test subjects. These discoveries have been made with the help of imaging techniques such as **computed tomography scans** (CT scans).

When a psychiatrist assesses a patient for schizophrenia, he or she will begin by excluding physical conditions that can cause abnormal thinking and some other behaviors associated with schizophrenia. These conditions include organic brain disorders (including traumatic injuries of the brain) temporal lobe **epilepsy**, **Wilson disease**, Huntington's chorea, and **encephalitis**.

The doctor will also need to rule out **substance abuse** disorders, especially amphetamine use.

After ruling out organic disorders, the clinician will consider other psychiatric conditions that may include psychotic symptoms or symptoms resembling **psychosis**. These disorders include mood disorders with psychotic features; delusional disorder; dissociative disorder not otherwise specified (DDNOS) or **multiple personality disorder**; schizotypal, schizoid, or paranoid **personality disorders**; and atypical reactive disorders. In the past, many individuals were incorrectly diagnosed as schizophrenic. Some patients who were diagnosed prior to the changes in categorization introduced by *DSM-IV* should have their diagnoses, and treatment, reevaluated. In children, the doctor must distinguish between psychotic symptoms and a vivid fantasy life, and also identify learning problems or disorders. After other conditions have been ruled out, the patient must meet a set of criteria specified by *DSM-IV*:

- **Characteristic symptoms.** The patient must have two (or more) of the following symptoms during a one-month period: delusions; hallucinations; disorganized speech; disorganized or catatonic behavior; negative symptoms.
- Decline in social, interpersonal, or occupational functioning, including self-care.
- **Duration.** The disturbed behavior must last for at least six months.
- **Diagnostic exclusions.** Mood disorders, substance abuse disorders, medical conditions, and developmental disorders have been ruled out.

Treatment

Traditional

The treatment of schizophrenia depends in part on the patient's stage or phase. Patients in the acute phase are hospitalized in most cases, to prevent harm to the patient or others and to begin treatment with antipsychotic medications. A patient having a first psychotic episode may be given a CT or MRI (**magnetic resonance imaging**) scan to rule out structural brain disease.

Most schizophrenics can benefit from **psychotherapy** once their acute symptoms have been brought under control by antipsychotic medication. Psychoanalytic approaches are not recommended. Behavior therapy, however, is often helpful in assisting patients to acquire skills for daily living and social interaction. It can be combined with **occupational therapy** to prepare the patient for eventual employment.

KEY TERMS

Affective flattening—A loss or lack of emotional expressiveness. It is sometimes called blunted or restricted affect.

Akathisia—Agitated or restless movement, usually affecting the legs and accompanied by a sense of discomfort. It is a common side effect of neuroleptic medications.

Catatonic behavior—Behavior characterized by muscular tightness or rigidity and lack of response to the environment. In some patients rigidity alternates with excited or hyperactive behavior.

Delusion—A fixed, false belief that is resistant to reason or factual disproof.

Depot dosage—A form of medication that can be stored in the patient's body tissues for several days or weeks, thus minimizing the risk of the patient forgetting daily doses. Haloperidol and fluphenazine can be given in depot form.

Dopamine receptor antagonists (DAs)—The older class of antipsychotic medications, also called neuroleptics. These primarily block the site on nerve cells that normally receive the brain chemical dopamine.

Dystonia—Painful involuntary muscle cramps or spasms.

Extrapyramidal symptoms (EPS)—A group of side effects associated with antipsychotic medications. EPS include parkinsonism, akathisia, dystonia, and tardive dyskinesia.

First-rank symptoms—A set of symptoms designated by Kurt Schneider in 1959 as the most important diagnostic indicators of schizophrenia. These symptoms include delusions, hallucinations, thought insertion or removal, and thought broadcasting. First-rank symptoms are sometimes referred to as Schneiderian symptoms.

Hallucination—A sensory experience of something that does not exist outside the mind. A person can experience a hallucination in any of the five senses. Auditory hallucinations are a common symptom of schizophrenia.

Huntington's chorea—A hereditary disease that typically appears in midlife, marked by gradual

loss of brain function and voluntary movement. Some of its symptoms resemble those of schizophrenia.

Negative symptoms—Symptoms of schizophrenia characterized by the absence or elimination of certain behaviors. DSM-IV specifies three negative symptoms: affective flattening, poverty of speech, and loss of will or initiative.

Neuroleptic—Another name for the older type of antipsychotic medications given to schizophrenic patients.

Parkinsonism—A set of symptoms originally associated with Parkinson's disease that can occur as side effects of neuroleptic medications. The symptoms include trembling of the fingers or hands, a shuffling gait, and tight or rigid muscles.

Positive symptoms—Symptoms of schizophrenia that are characterized by the production or presence of behaviors that are grossly abnormal or excessive, including hallucinations and thought-process disorder. DSM-IV subdivides positive symptoms into psychotic and disorganized.

Poverty of speech—A negative symptom of schizophrenia, characterized by brief and empty replies to questions. It should not be confused with shyness or reluctance to talk.

Psychotic disorder—A mental disorder characterized by delusions, hallucinations, or other symptoms of lack of contact with reality. The schizophrenias are psychotic disorders.

Serotonin dopamine antagonist (SDA)—The newer second-generation antipsychotic drugs, also called atypical antipsychotics. SDAs include clozapine (Clozaril), risperidone (Risperdal), and olanzapine (Zyprexa).

Wilson disease—A rare hereditary disease marked by high levels of copper deposits in the brain and liver. It can cause psychiatric symptoms resembling schizophrenia.

Word salad—Speech that is so disorganized that it makes no linguistic or grammatical sense.

Family therapy is often recommended for the families of schizophrenic patients, to relieve the feelings of guilt that they often have as well as to help them understand the patient's disorder. The family's

attitude and behaviors toward the patient are key factors in minimizing relapses (for example, by reducing stress in the patient's life), and family therapy can often strengthen the family's ability to cope with

the stresses caused by the schizophrenic's illness. Family therapy focused on communication skills and problem-solving strategies is particularly helpful. In addition to formal treatment, many families benefit from support groups and similar mutual help organizations for relatives of schizophrenics.

Drugs

The primary form of treatment of schizophrenia is antipsychotic medication. **Antipsychotic drugs** help to control almost all the positive symptoms of the disorder. They have minimal effects on disorganized behavior and negative symptoms. Between 60–70% of schizophrenics will respond to antipsychotics. In the acute phase of the illness, patients are usually given medications by mouth or by intramuscular injection. After the patient has been stabilized, the antipsychotic drug may be given in a long-acting form called a depot dose. Depot medications last two to four weeks; they have the advantage of protecting the patient against the consequences of forgetting or skipping daily doses. In addition, some patients who do not respond to oral neuroleptics have better results with depot form. Patients whose long-term treatment includes depot medications are introduced to the depot form gradually during their stabilization period. Most people with schizophrenia are kept on antipsychotic medications indefinitely during the maintenance phase of their disorder to minimize the possibility of relapse.

The most frequently used antipsychotics fall into two classes: the older dopamine receptor antagonists, or DAs, and the newer serotonin dopamine antagonists, or SDAs. (Antagonists block the action of some other substance; for example, dopamine antagonists counteract the action of dopamine.) The exact mechanisms of action of these medications are not known, but it is thought that they lower the patient's sensitivity to sensory stimuli and so indirectly improve the patient's ability to interact with others.

DOPAMINE RECEPTOR ANTAGONIST. The dopamine antagonists include the older antipsychotic (also called neuroleptic) drugs, such as haloperidol (Haldol), chlorpromazine (Thorazine), and fluphenazine (Prolixin). These drugs have two major drawbacks: it is often difficult to find the best dosage level for the individual patient, and a dosage level high enough to control psychotic symptoms frequently produces extrapyramidal side effects, or EPS. EPSs include parkinsonism, in which the patient cannot walk normally and usually develops a tremor; dystonia, or painful **muscle spasms** of the head, tongue, or neck; and akathisia, or restlessness. A type of long-term EPS is called **tardive dyskinesia**, which features

slow, rhythmic, automatic movements. Schizophrenics with **AIDS** are especially vulnerable to developing EPS.

SEROTONIN DOPAMINE ANTAGONISTS. The serotonin dopamine antagonists, also called atypical antipsychotics, are newer medications that include clozapine (Clozaril), risperidone (Risperdal), and olanzapine (Zyprexa). The SDAs have a better effect on the negative symptoms of schizophrenia than do the older drugs and are less likely to produce EPS than the older compounds. The newer drugs are significantly more expensive in the short term, although the SDAs may reduce long-term costs by reducing the need for hospitalization. They are also presently unavailable in injectable forms. The SDAs are commonly used to treat patients who respond poorly to the DAs. However, many psychotherapists now regard the use of these atypical antipsychotics as the treatment of first choice.

ANTIDEPRESSANTS. Patients with schizophrenia have a lifetime prevalence of 80% for major depression; others suffer from **phobias** or other **anxiety disorders**. These patients may be prescribed antidepressants or a short course of **benzodiazepines** along with their antipsychotic medications.

Alternative

Alternative and complementary therapies that are being investigated for the treatment of schizophrenia include gingko biloba, an Asian shrub, and vitamin therapy. One Chinese study reported that a group of patients who had not responded to conventional antipsychotic medications benefited from a thirteen-week trial of gingko extract, with significantly fewer side effects. Vitamin therapy is recommended by naturopathic practitioners on the grounds that many hospitalized patients with schizophrenia suffer from nutritional deficiencies. The supplements recommended include **folic acid**, niacin, vitamin B₆, and vitamin C.

Many clinical trials for the treatment of schizophrenia are currently sponsored by the National Institutes of Health (NIH) and other agencies. In 2009, NIH reported 400 ongoing or recently completed studies.

A few examples include:

- The safety and effectiveness of an investigational drug (MK0557) for the treatment of cognitive impairment in patients with schizophrenia. (NCT00482430)
- The efficacy of preventative treatment with sarcosine to reduce symptoms and delay or avoid disease progression in individuals defined as being at high risk for schizophrenia. (NCT00276263)

- The role of genetics in the development of schizophrenia by studying heritable traits in families where at least one member has schizophrenia. (NCT00001486)
- The effectiveness of D-cycloserine and glycine for treating negative symptoms (such as loss of interest, loss of energy, loss of warmth, and loss of humor) which occur between phases of positive symptoms (marked by hallucinations, delusions, and thought confusions) in schizophrenics. (NCT00000372)
- Study of the biological basis of schizophrenia to determine which symptoms are related to the illness itself and which are related to medications used to treat the illness.(NCT00001247)
- The use of single photon emission computed tomography (SPECT) to study brain nicotine receptors (proteins on the surface of brain cells) in healthy subjects and in patients with schizophrenia. (NCT00061789)

Clinical trial information is constantly updated by NIH and the most recent information on schizophrenia trials can be found at: <http://clinicaltrials.gov/search/open/condition=%22Schizophrenia%22>

Prognosis

One important prognostic sign is the patient's age at onset of psychotic symptoms. Patients with early onset of schizophrenia are more often male, have a lower level of functioning prior to onset, a higher rate of brain abnormalities, more noticeable negative symptoms, and worse outcomes. Patients with later onset are more likely to be female, with fewer brain abnormalities and thought impairment, and more hopeful prognoses.

The average course and outcome for schizophrenics are less favorable than those for most other mental disorders, although as many as 30% of patients diagnosed with schizophrenia recover completely and the majority experience some improvement. Two factors that influence outcomes are stressful life events and a hostile or emotionally intense family environment. Schizophrenics with a high number of stressful changes in their lives, or who have frequent contacts with critical or emotionally over-involved family members, are more likely to relapse. Overall, the most important component of long-term care of schizophrenic patients is complying with their regimen of antipsychotic medications.

Prevention

There is no proven way to prevent onset of schizophrenia. Researchers have investigated the possibility of treating schizophrenia before symptoms start (such as when the likelihood of hereditary transmission is high). Other areas of research include the links

between schizophrenia and family stress, drug use, and exposure to certain infectious agents.

Resources

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ORGANIZATIONS

- American Psychiatric Association (APA), 1000 Wilson Blvd., Suite 1825, Arlington, VA, 22209, (888) 357-7924, apa@psych.org, <http://www.psych.org>.
- National Alliance on Mental Health (NAMI), 3803 N. Fairfax Dr., Suite 100, Arlington, VA, 22203, (703) 524-7600, (888) 999-NAMI (6264), (703) 524-9094, <http://www.nami.org>.
- National Institute of Mental Health (NIMH), 6001 Executive Blvd., Rm 8184, MSC 9663, Rockville, MD, 20892-9663, (301) 443-4513, nimhinfo@nih.gov, <http://www.nimh.nih.gov>.
- National Mental Health Association (NMHA), 2001 North Beauregard St., 12th Floor, Alexandria, VA, 22311, (703) 684-7722, (800) 969-6642, infoctr@nmha.org, <http://www.nmha.org>.
- Schizophrenia Society of Canada, 100-4 Fort Street, Winnipeg, MB, Canada, R3C1C4, (204) 786-1616, (800) 263-5545, (204) 783-4898, info@schizophrenia.ca, <http://www.schizophrenia.ca>.

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Schwannoma see **Brain tumor**

Sciatic nerve pain see **Sciatica**

Sciatica

Definition

Sciatica refers to **pain** or discomfort associated with the sciatic nerve. This nerve runs from the lower part of the spinal cord, down the back of the leg, to the foot. Injury to or pressure on the sciatic nerve can cause the characteristic pain of sciatica: a sharp or burning pain that radiates from the lower back or hip, possibly following the path of the sciatic nerve to the foot.

Description

The sciatic nerve is the largest and longest nerve in the body. About the thickness of a person’s thumb, it spans from the lower back to the foot. The nerve originates in the lower part of the spinal cord, the so-called lumbar region. As it branches off from the spinal cord, it passes between the bony vertebrae (the component bones of the spine) and runs through the pelvic girdle, or hip bones. The nerve passes through the hip joint and continues down the back of the leg to the foot.

Sciatica is a fairly common disorder and approximately 40% of the population experiences it at some point in their lives. However, only about 1% have coexisting sensory or motor deficits. Sciatic pain has several root causes and treatment may hinge upon the underlying problem.

Of the identifiable causes of sciatic pain, lumbosacral radiculopathy and back strain are the most frequently suspected. The term lumbosacral refers to the lower part of the spine, and radiculopathy describes a problem with the spinal nerve roots that pass between the vertebrae and give rise to the sciatic nerve. This area between the vertebrae is cushioned with a disk of shock-absorbing tissue. If this disk shifts or is damaged through injury or disease, the spinal nerve root may be compressed by the shifted tissue or the vertebrae.

This compression of the nerve roots sends a pain signal to the brain. Although the actual injury is to the nerve roots, the pain may be perceived as coming from anywhere along the sciatic nerve.

The sciatic nerve can be compressed in other ways. Back strain may cause **muscle spasms** in the lower back, placing pressure on the sciatic nerve. In rare cases, infection, **cancer**, bone inflammation, or other diseases may be causing the pressure. More likely, but often overlooked, is the piriformis syndrome. As the sciatic nerve passes through the hip joint, it shares the space with several muscles. One of these muscles, the piriformis muscle, is closely associated with the sciatic nerve. In some people, the nerve actually runs through the muscle. If this muscle is injured or has a spasm, it places pressure on the sciatic nerve, in effect, compressing it.

In many sciatica cases, the specific cause is never identified. About half of affected individuals recover from an episode within a month. Some cases can linger a few weeks longer and may require aggressive treatment. In some cases, the pain may return or potentially become chronic.

Causes and symptoms

Individuals with sciatica may experience some lower back pain, but the most common symptom is pain that radiates through one buttock and down the back of that leg. The most identified cause of the pain is compression or pressure on the sciatic nerve. The extent of the pain varies between individuals. Some people describe pain that centers in the area of the hip, and others perceive discomfort all the way to the foot. The quality of the pain also varies; it may be described as **tingling**, burning, prickly, aching, or stabbing.

Onset of sciatica can be sudden, but it can also develop gradually. The pain may be intermittent or continuous, and certain activities, such as bending, coughing, sneezing, or sitting, may make the pain worse.

Chronic pain may arise from more than just compression on the nerve. According to some pain researchers, physical damage to a nerve is only half of the equation. A developing theory proposes that some nerve injuries result in a release of neurotransmitters and immune system chemicals that enhance and sustain a pain message. Even after the injury has healed, or the damage has been repaired, the pain continues. Control of this abnormal type of pain is difficult.

Diagnosis

Before treating sciatic pain, as much information as possible is collected. The individual is asked to recount the location and nature of the pain, how long it has continued, and any accidents or unusual activities prior to its onset. This information provides clues that may point to back strain or injury to a specific location. Back pain from disk disease, piriformis syndrome, and back strain must be differentiated from more serious conditions such as cancer or infection. Lumbar stenosis, an overgrowth of the covering layers of the vertebrae that narrows the spinal canal, must also be considered. The possibility that a difference in leg lengths is causing the pain should be evaluated; the problem can be easily be treated with a foot orthotic or built-up shoe.

Often, a straight-leg-raising test is done, in which the person lies face upward and the health-care provider raises the affected leg to various heights. This test pinpoints the location of the pain and may reveal whether it is caused by a disk problem. Other tests, such as having the individual rotate the hip joint, assess the hip muscles. Any pain caused by these movements may provide information about involvement of the piriformis muscle, and piriformis weakness is tested with additional leg-strength maneuvers.

Further tests may be done depending on the results of the **physical examination** and initial pain treatment. Such tests might include **magnetic resonance imaging** (MRI) and **computed tomography scans** (CT scans). Other tests examine the conduction of electricity through nerve tissues, and include studies of the electrical activity generated as muscles contract (**electromyography**), nerve conduction velocity, and evoked potential testing. A more invasive test involves injecting a contrast substance into the space between the vertebrae and making x-ray images of the spinal cord (**myelography**), but this procedure is usually done only if surgery is being considered. All of these tests can reveal problems with the vertebrae, the disk, or the nerve itself.

Treatment

Initial treatment for sciatica focuses on pain relief. For acute or very painful flare-ups, bed rest is advised for up to a week in conjunction with medication for the pain. Pain medication includes **acetaminophen**, **non-steroidal anti-inflammatory drugs** (NSAIDs), such as **aspirin**, or **muscle relaxants**. If the pain is unremitting, opioids may be prescribed for short-term use or a local anesthetic will be injected directly into the lower back. Massage and heat application may be suggested as adjuncts.

If the pain is chronic, different pain relief medications are used to avoid long-term dosing of NSAIDs, muscle relaxants, and opioids. **Antidepressant drugs**, which have been shown to be effective in treating pain, may be prescribed alongside short-term use of muscle relaxants or NSAIDs. Local anesthetic injections or epidural **steroids** are used in selected cases.

As the pain allows, **physical therapy** is introduced into the treatment regime. Stretching exercises that focus on the lower back, buttock, and hamstring muscles are suggested. The exercises also include finding comfortable, pain-reducing positions. Corsets and braces may be useful in some cases, but evidence for their general effectiveness is lacking. However, they may be helpful to prevent exacerbations related to certain activities.

With less pain and the success of early therapy, the individual is encouraged to follow a long-term program to maintain a healthy back and prevent re-injury. A physical therapist may suggest exercises and regular activity, such as water **exercise** or walking. Patients are instructed in proper body mechanics to minimize symptoms during light lifting or other activities.

If the pain is chronic and conservative treatment fails, surgery to repair a **herniated disk** or cut out part or all of the piriformis muscle may be suggested,

KEY TERMS

Disk—Dense tissue between the vertebrae that acts as a shock absorber and prevents damage to nerves and blood vessels along the spine.

Electromyography—A medical test in which a nerve's ability to conduct an impulse is measured.

Lumbosacral—Referring to the lower part of the backbone or spine.

Myelography—A medical test in which a special dye is injected into a nerve to make it visible on an x-ray.

Piriformis—A muscle in the pelvic girdle that is closely associated with the sciatic nerve.

Radiculopathy—A condition in which the spinal nerve root of a nerve has been injured or damaged.

Spasm—Involuntary contraction of a muscle.

Vertebrae—The component bones of the spine.

particularly if there is neurologic evidence of nerve or nerve-root damage.

Alternative treatment

Massage is a recommended form of therapy, especially if the sciatic pain arises from muscle spasm. Symptoms may also be relieved by icing the painful area as soon as the pain occurs. Ice should be left on the area for 30–60 minutes several times a day. After 2–3 days, a hot water bottle or heating pad can replace the ice. **Chiropractic** or **osteopathy** may offer possible solutions for relieving pressure on the sciatic nerve and the accompanying pain. **Acupuncture** and **biofeedback** may also be useful as pain control methods. Body work, such as the **Alexander technique**, can assist an individual in improving posture and preventing further episodes of sciatic pain.

Prognosis

Most cases of sciatica are treatable with pain medication and physical therapy. After 4–6 weeks of treatment, an individual should be able to resume normal activities.

Prevention

Some sources of sciatica are not preventable, such as disk degeneration, back strain due to **pregnancy**, or accidental falls. Other sources of back strain, such as poor posture, overexertion, being overweight, or wearing high heels, can be corrected or avoided.

Cigarette **smoking** may also predispose people to pain, and should be discontinued.

General suggestions for avoiding sciatica, or preventing a repeat episode, include sleeping on a firm mattress, using chairs with firm back support, and sitting with both feet flat on the floor. Habitually crossing the legs while sitting can place excess pressure on the sciatic nerve. Sitting a lot can also place pressure on the sciatic nerves, so it's a good idea to take short breaks and move around during the work day, long trips, or any other situation that requires sitting for an extended length of time. If lifting is required, the back should be kept straight and the legs should provide the lift. Regular exercise, such as swimming and walking, can strengthen back muscles and improve posture. Exercise can also help maintain a healthy weight and lessen the likelihood of back strain.

Resources

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Julia Barrett

SCID see **Severe combined immunodeficiency**

Scleral buckling see **Retinal detachment**

Scleroderma

Definition

Scleroderma is a progressive disease that affects the skin and connective tissue (including cartilage, bone, fat, and the tissue that supports the nerves and blood vessels throughout the body). There are two major forms of the disorder. The type known as localized scleroderma affects the skin mainly. Systemic scleroderma, which is also called systemic sclerosis, affects the smaller blood vessels and internal organs of the body.

Demographics

Scleroderma occurs in all races of people all over the world, but it affects about four females for every male. Among children, localized scleroderma is more common, and systemic sclerosis is comparatively rare. Most patients with systemic sclerosis are diagnosed between ages 30 and 50. In the United States, about 300,000



Scleroderma is a serious, progressive disease caused by the overproduction and accumulation of collagen throughout the body, resulting in hardening (sclerosis) and scarring (fibrosis) of the skin and connective tissue. (Photo Researchers, Inc.)

people have scleroderma. Young African-American women and Native Americans of the Choctaw tribe have especially high rates of the disease. In 2003, researchers reported that they had identified 12 different genetic markers associated with scleroderma in the Choctaw population.

Description

Scleroderma is an autoimmune disorder, which means that the body's immune system turns against itself. In scleroderma, there is an overproduction of abnormal collagen (a type of protein fiber present in connective tissue). This collagen accumulates throughout the body, causing hardening (sclerosis), scarring (fibrosis), and other damage. The damage may affect the appearance of the skin, or it may involve only the internal organs. The symptoms and severity of scleroderma vary from person to person.

Causes and symptoms

The cause of scleroderma is still a puzzle. Although the accumulation of collagen appears to be a hallmark of the disease, researchers do not know why it occurs. Some theories suggest that damage to blood vessels may cause the tissues of the body to receive an inadequate amount of oxygen, a condition called **ischemia**. Some researchers believe that the resulting damage causes the immune system to overreact, producing an autoimmune disorder. According to this theory of scleroderma, the immune system gears up to fight an invader, but no invader is actually present. Cells in the immune system called antibodies react to the body's own tissues as if they were foreign. The antibodies

turn against the already damaged blood vessels and the vessels' supporting tissues. These immune cells are designed to deliver potent chemicals in order to kill foreign invaders. Some of these cells dump these chemicals on the body's own tissues instead, causing inflammation, swelling, damage, and scarring.

Most cases of scleroderma have no recognizable triggering event. Some cases, however, have been traced to exposure to toxic (poisonous) substances. For example, coal miners and gold miners, who are exposed to high levels of silica dust, have above-average rates of scleroderma. Other chemicals associated with the disease include polyvinyl chloride, benzene, toluene, and epoxy resins. In 1981, 20,000 people in Spain were stricken with a syndrome similar to scleroderma when their cooking oil was accidentally contaminated. Certain medications, especially a drug used in **cancer** treatment called bleomycin (Blenoxane), may lead to scleroderma. Some claims of a scleroderma-like illness have been made by women with silicone **breast implants**, but a link has not been proven in numerous studies.

Symptoms of systemic scleroderma

A condition called Raynaud's phenomenon is the first symptom in about 95% of all patients with systemic scleroderma. In Raynaud's phenomenon, blood vessels of the fingers and/or toes (the digits) react to cold in an abnormal way. The vessels clamp down, preventing blood flow to the tip of the digit. Eventually, the flow is cut off to the entire finger or toe. Over time, oxygen deprivation may result in open ulcers on the skin surface. These ulcers can lead to tissue **death (gangrene)** and loss of the digit. When Raynaud's phenomenon is the first sign of scleroderma, the next symptoms usually appear within two years.

SKIN AND EXTREMITIES. Involvement of the skin leads to swelling underneath the skin of the hands, feet, legs, arms, and face. Swelling is followed by thickening and tightening of the skin, which becomes taut and shiny. Severe tightening may lead to abnormalities. For example, tightening of the skin on the hands may cause the fingers to become permanently curled (flexed). Structures within the skin are damaged (including those producing hair, oil, and sweat), and the skin becomes dry and scaly. Ulcers may form, with the danger of infection. **Calcium** deposits often appear under the skin.

In systemic scleroderma, the mouth and nose may become smaller as the skin on the face tightens. The small mouth may interfere with eating and dental hygiene. Blood vessels under the skin may become enlarged and show through the skin, appearing as purplish marks or

red spots. This chronic dilation of the small blood vessels is called telangiectasis.

Muscle weakness, joint **pain** and stiffness, and **carpal tunnel syndrome** are common in scleroderma. Carpal tunnel syndrome involves scarring in the wrist, which puts pressure on the median nerve running through that area. Pressure on the nerve causes **numbness**, **tingling**, and weakness in some of the fingers.

DIGESTIVE TRACT. The tube leading from the mouth to the stomach (the esophagus) becomes stiff and scarred. Patients may have trouble swallowing food. The acid contents of the stomach may start to flow backward into the esophagus (esophageal reflux), causing a very uncomfortable condition known as **heartburn**. The esophagus may also become inflamed.

The intestine becomes sluggish in processing food, causing bloating and pain. Foods are not digested properly, resulting in **diarrhea**, weight loss, and anemia. Telangiectasis in the stomach or intestine may cause rupture and bleeding.

RESPIRATORY AND CIRCULATORY SYSTEMS. The lungs are affected in about 66% of all people with systemic scleroderma. Complications include **shortness of breath**, coughing, difficulty breathing due to tightening of the tissue around the chest, inflammation of the air sacs in the lungs (alveolitis), increased risk of **pneumonia**, and an increased risk of cancer. For these reasons, lung disease is the most likely cause of death associated with scleroderma.

The lining around the heart (pericardium) may become inflamed. The heart may have greater difficulty pumping blood effectively (**heart failure**). Irregular heart rhythms and enlargement of the heart also occur in scleroderma.

Kidney disease is another common complication. Damage to blood vessels in the kidneys often causes a major rise in the person's blood pressure. The blood pressure may be so high that there is swelling of the brain, causing severe headaches, damage to the retinas of the eyes, seizures, and failure of the heart to pump blood into the body's circulatory system. The kidneys may also stop filtering blood and go into failure. Treatments for high blood pressure have greatly improved these kidney complications. Before these treatments were available, kidney problems were the most common cause of death for people with scleroderma.

Other problems associated with scleroderma include painful dryness of the eyes and mouth, enlargement and destruction of the liver, and a low-functioning thyroid gland.

Diagnosis

Diagnosis of scleroderma is complicated by the fact that some of its symptoms can accompany other connective-tissue diseases. The most important symptom is thickened or hardened skin on the fingers, hands, forearms, or face. This symptom is found in 98% of people with scleroderma. It can be detected in the course of a **physical examination**. The person's medical history may also contain important clues, such as exposure to toxic substances on the job. There are a number of nonspecific laboratory tests on blood samples that may indicate the presence of an inflammatory disorder (but not specifically scleroderma). The antinuclear antibody (ANA) test is positive in more than 95% of people with scleroderma.

Other tests can be performed to evaluate the extent of the disease. These include a test of the electrical system of the heart (an electrocardiogram), lung-function tests, and x-ray studies of the gastrointestinal tract. Various blood tests can be given to study kidney function.

Treatment

Mainstream treatments

As of 2010, there is no cure for scleroderma. A drug called D-penicillamine has been used to interfere with the production of abnormal collagen. Experts believe the drug helps decrease the degree of skin thickening and tightening, and slow the progress of the disease in other organs. Taking vitamin D and using ultraviolet light may be helpful for localized scleroderma. One group of British researchers reported in 2003 that long-wavelength ultraviolet A light is particularly effective in treating localized scleroderma. **Corticosteroids** have been used to treat joint pain, **muscle cramps**, and other symptoms of inflammation. Other drugs have been studied that reduce the activity of the immune system (immunosuppressants). Because these medications can have serious side effects, they are used in only the most severe cases of scleroderma.

The various complications of scleroderma are treated individually. Raynaud's phenomenon requires that people try to keep their hands and feet warm constantly. Nifedipine is a medication that is sometimes given to help control Raynaud's. Thick ointments and creams are used to treat dry skin. **Exercise** and massage may help joint involvement. They may also help people retain more movement despite skin tightening. An exercise regimen for stretching the mouth opening has been reported to be a helpful alternative to surgery in managing this condition. Skin ulcers need prompt attention and may require **antibiotics**. People with esophageal reflux

KEY TERMS

Autoimmune disorder—A disorder in which the body's immune cells mistake the body's own tissues as foreign invaders. The immune cells then work to destroy tissues in the body.

Collagen—The main supportive protein of cartilage, connective tissue, tendon, skin, and bone.

Connective tissue—A group of tissues responsible for support throughout the body, including cartilage, bone, fat, tissue underlying skin, and tissues that support organs, blood vessels, and nerves throughout the body.

Fibrosis—The abnormal development of fibrous tissue; scarring.

Limited scleroderma—A subtype of systemic scleroderma with limited skin involvement. It is sometimes called the CREST form of scleroderma, after the initials of its five major symptoms.

Localized scleroderma—Thickening of the skin from overproduction of collagen.

Morphea—The most common form of localized scleroderma.

Raynaud phenomenon/Raynaud disease—A condition in which blood flow to the body's tissues is reduced by a malfunction of the nerves that regulate the constriction of blood vessels. When attacks of Raynaud's occur in the absence of other medical conditions, it is called Raynaud disease. When attacks occur as part of a disease (as in scleroderma), it is called Raynaud phenomenon.

Sclerosis—Hardening.

Systemic sclerosis—A rare disorder that causes thickening and scarring of multiple organ systems.

Telangiectasias—Very small arteriovenous malformations, or connections between the arteries and veins. The result is small red spots on the skin known as "spider veins."

will be advised to eat small amounts more often, rather than several large meals a day. They should also avoid spicy foods and items containing **caffeine**. Some patients with esophageal reflux have been successfully treated with surgery. Acid-reducing medications may be given for heartburn. People must be monitored for the development of high blood pressure. If found, they should be promptly treated with appropriate medications, usually angiotensin-converting enzyme inhibitors (ACE inhibitors) or other **vasodilators**. When fluid accumulates due to heart failure, **diuretics** can be given to get rid of the excess fluid.

Patients with scleroderma may also benefit from some form of counseling or **psychotherapy**, as they are at increased risk of depression. One study found that 46% of patients in its sample met the criteria for a depressive disorder.

Alternative treatments

One alternative therapy that some naturopaths have used in treating patients with scleroderma is superoxide dismutase (SOD), an antioxidant enzyme used in its injectable form. More research, however, needs to be done on the benefits of this treatment.

Prognosis

The prognosis for people with scleroderma varies. Some have a very limited form of the disease called

morphea, which affects only the skin. These individuals have a very good prognosis. Other people have a subtype of systemic scleroderma called limited scleroderma. For them, the prognosis is relatively good. Limited scleroderma is characterized by limited involvement of the patient's skin and a cluster of five symptoms called the CREST syndrome. CREST stands for:

- C = Calcinosis
- R = Raynaud's disease (phenomenon)
- E = Esophageal dysmotility (stiffness and malfunctioning of the esophagus)
- S = Sclerodactyly (thick, hard, rigid skin over the fingers)
- T = Telangiectasias

In general, people with very widespread skin involvement have the worst prognosis. This level of disease is usually accompanied by involvement of other organs and the most severe complications. Although women are more commonly stricken with scleroderma, men more often die of the disease. The two factors that negatively affect survival are male sex and older age at diagnosis. The most common causes of death include heart, kidney, and lung diseases. About 65% of all patients survive 11 years or more following a diagnosis of scleroderma.

Prevention

There are no known ways to prevent scleroderma. People can try to decrease occupational exposure to high-risk substances.

Resources

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ORGANIZATIONS

- American Academy of Family Physicians (AAFP), 114 Tomahawk Creek Parkway, Leawood, KS, 66211-2672, (800) 274-2237, (913) 906-6269, fp@aafp.org, www.familydoctor.org.
 American College of Rheumatology (ACR), 2200 Lake Blvd. NE, Atlanta, GA, 30319, (404) 633-3777, http://www.rheumatology.org.
 National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMSD), 1 AMS Circle, Bethesda, MD, 20892–3675, (301) 495-4484, (877) 226-4267, (301) 718-6366, NIAMSinfo@mail.nih.gov, http://www.niams.nih.gov.
 National Institutes of Health (NIH), 9000 Rockville Pike, Bethesda, MD, 20892, (301) 496-4000, http://www.nih.gov/index.html.
 National Library of Medicine (NLM), 8600 Rockville Pike, Bethesda, MD, 20894, http://www.nlm.nih.gov/.

National Organization for Rare Disorders, Inc. (NORD), 55 Kenosia Ave., PO Box 1968, Danbury, CT, 06813, (203) 744-0100, (800) 999-6673, http://www.rarediseases.org.

Scleroderma Foundation, 300 Rosewood Dr., Suite 105, Danvers, MA, 01923, (978) 463-5843, (800) 722-HOPE, (978) 463-5809, http://www.scleroderma.org.

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Sclerotherapy for esophageal varices

Definition

Sclerotherapy for esophageal varices (also called endoscopic sclerotherapy) is a treatment for esophageal bleeding that involves the use of an endoscope and the injection of a sclerosing solution into veins.

Purpose

In most hospitals, sclerotherapy for esophageal varices is the treatment of choice to stop esophageal bleeding during acute episodes, and to prevent further incidences of bleeding. Emergency sclerotherapy is often followed by preventive treatments to eradicate distended esophageal veins.

Precautions

Sclerotherapy for esophageal varices cannot be performed on an uncooperative patient, since movement during the procedure could cause the vein to tear or the esophagus to perforate and bleed. It should not be performed on a patient with a perforated gastrointestinal tract.

Description

Esophageal varices are enlarged or swollen veins on the lining of the esophagus which are prone to bleeding. They are life-threatening, and can be fatal in up to 50% of patients. They usually appear in patients with severe **liver disease**. Sclerotherapy for esophageal varices involves injecting a strong and irritating solution (a sclerosant) into the veins and/or the area beside the distended vein. The sclerosant injected into the vein causes **blood clots** to form and stops the bleeding. The sclerosant injected into the area beside the distended vein stops the bleeding by thickening and swelling the vein to compress the blood vessel.

KEY TERMS

Endoscope—An instrument used to examine the inside of a canal or hollow organ. Endoscopic surgery is less invasive than traditional surgery.

Esophagus—The part of the digestive canal located between the pharynx (part of the digestive tube) and the stomach.

Sclerosant—An irritating solution that stops bleeding by hardening the blood or vein it is injected into.

Varices—Swollen or enlarged veins, in this case on the lining of the esophagus.

Most physicians inject the sclerosant directly into the vein, although injections into the vein and the surrounding area are both effective. Once bleeding has been stopped, the treatment can be used to significantly reduce or destroy the varices.

Sclerotherapy for esophageal varices is performed by a physician in a hospital, with the patient awake but sedated. Hyoscine butylbromide (Buscopan) may be administered to freeze the esophagus, making injection of the sclerosant easier. During the procedure, an endoscope is passed through the patient's mouth to the esophagus to view the inside. The branches of the blood vessels at or just above where the stomach and esophagus come together, the usual site of variceal bleeding, are located. After the bleeding vein is identified, a long, flexible sclerotherapy needle is passed through the endoscope. When the tip of the needle's sheath is in place, the needle is advanced, and the sclerosant is injected into the vein or the surrounding area. The most commonly used sclerosants are ethanolamine and **sodium tetradecyl sulfate**. The needle is withdrawn. The procedure is repeated as many times as necessary to eradicate all distended veins.

Sclerotherapy for esophageal varices controls acute bleeding in about 90% of patients, but it may have to be repeated within the first 48 hours to achieve this success rate. During the initial hospitalization, sclerotherapy is usually performed two or three times. Preventive treatments are scheduled every few weeks or so, depending on the patient's risk level and healing rate. Several studies have shown that the risk of recurrent bleeding is much lower in patients treated with sclerotherapy: 30–50%, as opposed to 70–80% for patients not treated with sclerotherapy.

Preparation

Before sclerotherapy for esophageal varices, the patient's vital signs and other pertinent data are recorded, an intravenous line is inserted to administer fluid or blood, and a sedative is prescribed.

Aftercare

After sclerotherapy for esophageal varices, the patient will be observed for signs of blood loss, lung complications, **fever**, a perforated esophagus, or other complications. Vital signs are monitored, and the intravenous line maintained. **Pain** medication is usually prescribed. After leaving the hospital, the patient follows a diet prescribed by the physician, and, if appropriate, can take mild pain relievers.

Risks

Sclerotherapy for esophageal varices has a 20–40% incidence of complications, and a 1–2% percent mortality rate. Complications can arise from the sclerosant or the endoscopic procedure. Minor complications, which are uncomfortable but do not require active treatment or prolonged hospitalization, include transient chest pain, difficulty swallowing, and fever, which usually go away after a few days. Some people have allergic reactions to the solution. Infection occurs in up to 50% of cases. In 2–10% of patients, the esophagus tightens, but this can usually be treated with dilatation. More serious complications may occur in 10–15% of patients treated with sclerotherapy. These include perforation or bleeding of the esophagus and lung problems, such as aspiration **pneumonia**. Long-term sclerotherapy can damage the esophagus, and increase the patient's risk of developing **cancer**.

Patients with advanced liver disease complicated by bleeding are very poor risks for this procedure. The surgery, premedications, and anesthesia may be sufficient to tip the patient into protein intoxication and hepatic **coma**. The blood in the bowels acts like a high protein meal; therefore, protein intoxication may be induced.

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Lori De Milto

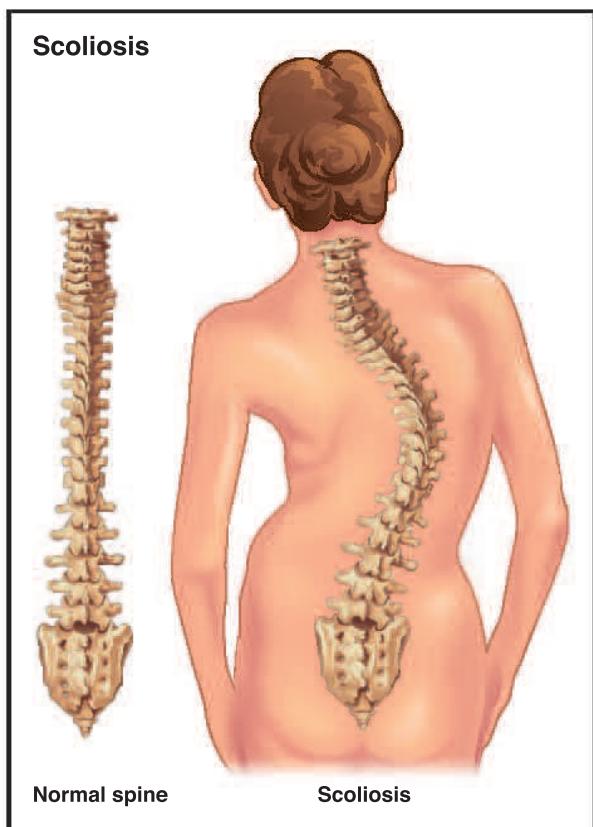
Scoliosis

Definition

Scoliosis is a S- or C-shaped sideways curvature of the spine of 10 degrees or greater.

Demographics

According to the American Academy of Orthopaedic Surgeons (AAOS), in 2009, scoliosis was estimated to affect approximately 2% of the population in the United States. The incidence is much higher (approximately 20%) if a family member has curvature of the spine. The National Scoliosis Foundation reports that scoliosis affects infants, adolescents, and adults worldwide irrespective of race or socio-economic status. The primary age of onset for scoliosis is 10–15 years old, and it occurs equally among both genders. However, females are eight times more likely to progress to a curve magnitude that requires treatment.



Normal spine compared to a spine affected by scoliosis.
(Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Description

When viewed from the rear, the spine usually appears to form a straight vertical line. Scoliosis is a lateral (side-to-side) curve in the spine, usually combined with a rotation of the vertebrae. (The lateral curvature of scoliosis should not be confused with the normal set of front-to-back spinal curves visible from the side.) While a small degree of lateral curvature does not cause any medical problems, larger curves can cause postural imbalance and lead to muscle **fatigue** and **pain**. More severe scoliosis can interfere with breathing and lead to arthritis of the spine (spondylosis).

Four out of five cases of scoliosis are *idiopathic*, meaning the cause is unknown. Children with idiopathic scoliosis appear to be otherwise entirely healthy, and have not had any bone or joint disease early in life. Scoliosis is not caused by poor posture, diet, or carrying a heavy book bag exclusively on one shoulder.

Idiopathic scoliosis is further classified according to age of onset:

- Infantile. Curvature appears before age three. This type is quite rare in the United States, but is more common in Europe.
- Juvenile. Curvature appears between ages three and 10. This type may be equivalent to the adolescent type, except for the age of onset.
- Adolescent. Curvature appears between ages of 10 and 13, near the beginning of puberty. This is the most common type of idiopathic scoliosis.
- Adult. Curvature begins after physical maturation is completed.

Three other types of scoliosis can also occur:

- Congenital scoliosis, due to congenital birth defects in the spine, and often associated with other structural abnormalities.
- Neuromuscular scoliosis, due to loss of control of the nerves or muscles that support the spine. The most common causes of this type of scoliosis are cerebral palsy and muscular dystrophy.
- Degenerative scoliosis, typically caused by degeneration of the discs that separate the vertebrae or arthritis in the joints that link them.

Risk factors

Scoliosis curves are more likely to worsen in girls than in boys. The younger the child when scoliosis appears, the greater the chance the curve will worsen. Children who are born with scoliosis are also at greater risk of worsening of the curve. A number of medical conditions are also known to predispose children to

scoliosis, such as Turner's syndrome, **muscular dystrophy**, **cerebral palsy**, **Marfan syndrome**, **Friedreich's ataxia**, **rheumatoid arthritis**, osteogenesis imperfect, and **spina bifida**.

Causes and symptoms

As mentioned above, the cause of scoliosis is unknown in 80–85% of cases (idiopathic scoliosis). For this reason, causes of curves are typically classified as either nonstructural or structural.

In nonstructural scoliosis, the spine is structurally normal but appears curved. This is a temporary, changing curve, caused by an underlying condition, such as unequal leg length, **muscle spasms**, or inflammatory conditions.

In structural scoliosis, the curve is fixed. Sometimes structural scoliosis is one part of a syndrome or disease, such as Marfan syndrome, an inherited connective tissue disorder. In some cases, it occurs by itself. Structural scoliosis can be caused by neuromuscular diseases (such as cerebral palsy, poliomyelitis, or muscular dystrophy), **birth defects** (such as hemivertebra), injury, certain infections, tumors, metabolic diseases, connective tissue disorders, or rheumatic diseases.

Idiopathic scoliosis has long been observed to run in families. Twin and family studies have consistently indicated a genetic contribution to the condition. However, no consistent pattern of transmission has been observed in familial cases. As of 2009, no genes have been identified that specifically cause the idiopathic form of scoliosis.

Most researchers have concluded that scoliosis is a complex trait. As such, there are likely to be multiple genetic, environmental, and potentially additional factors that contribute to the etiology of the condition. Complex traits are difficult to study due to the difficulty in identifying and isolating these multiple factors.

Scoliosis causes a noticeable asymmetry in the torso when viewed from the front or back. The first sign of scoliosis is often seen when a child is wearing a bathing suit or underwear. A child may appear to be standing with one shoulder higher than the other, or to have a tilt in the waistline. One shoulder blade may appear more prominent than the other due to rotation. In girls, one breast may appear higher than the other, or larger if rotation pushes that side forward.

Curve progression is greatest near the adolescent growth spurt. Scoliosis that begins early on is more likely to progress significantly than scoliosis that begins later in **puberty**.

KEY TERMS

Cobb angle—A measure of the curvature of scoliosis, determined by measurements made on x rays.

Hemivertebra—Condition in which one side of a vertebra fails to form normally before birth.

Marfan syndrome—Inherited connective tissue disorder.

Muscular dystrophy—Group of inherited disorders in which strength and muscle bulk gradually decline.

Narcotic—Medication derived from opium or synthetic opium.

Scoliometer—A tool for measuring trunk asymmetry; it includes a bubble level and angle measure.

Spondylosis—Arthritis of the spine.

More than 30 states have screening programs in schools for adolescent scoliosis, usually conducted by trained school nurses or gym teachers.

Diagnosis

Examination

Diagnosis for scoliosis is done by an orthopedist. A complete medical history is taken, including questions about family history of scoliosis. The **physical examination** includes determination of pubertal development in adolescents, a **neurological exam** that may reveal a neuromuscular cause, and measurements of trunk asymmetry. Examination of the trunk is done while the patient is standing, bending over, and lying down, and involves both visual inspection and use of a simple mechanical device called a scoliometer.

Tests

If a curve is detected, one or more x rays will usually be taken to define the curve or curves more precisely. An x ray is used to document spinal maturity, any pelvic tilt or hip asymmetry, and the location, extent, and degree of curvature. The curve is defined in terms of where it begins and ends, in which direction it bends, and by an angle measure known as the Cobb angle. The Cobb angle is found by projecting lines parallel to the vertebrae tops at the extremes of the curve; projecting perpendiculars from these lines; and measuring the angle of intersection. To properly track the progress of scoliosis, it is important to project from the same points of the spine each time.

Occasionally, **magnetic resonance imaging** (MRI) is used, primarily to look more closely at the condition of the spinal cord and nerve roots extending from it if neurological problems are suspected.

Treatment

Treatment decisions for scoliosis are based on the degree of curvature, the likelihood of significant progression, and the presence of pain, if any.

Traditional

Curves less than 20 degrees are not usually treated, except by regular follow-up for children who are still growing. Watchful waiting is usually all that is required in adolescents with curves of 20–25 degrees, or adults with curves up to 40 degrees or slightly more, as long as there is no pain.

For children or adolescents whose curves progress to 25 degrees, and who have a year or more of growth left, bracing may be required. Bracing cannot correct curvature, but may be effective in halting or slowing progression. Bracing is rarely used in adults, except where pain is significant and surgery is not an option, as in some elderly patients.

There are two different categories of braces, those designed for nearly 24 hour per day use and those designed for night use. The full-time brace styles are designed to hold the spine in a vertical position, while the night use braces are designed to bend the spine in the direction opposite the curve.

The Milwaukee brace is a full-time brace which consists of metal uprights attached to pads at the hips, rib cage, and neck. Other types of full-time braces, such as the Boston brace, involve underarm rigid plastic molding to encircle the lower rib cage, abdomen, and hips. Because they can be worn out of sight beneath clothing, the underarm braces are better tolerated and often leads to better compliance. The Boston brace is currently the most commonly used. Full-time braces are often prescribed to be worn for 22–23 hours per day, though some clinicians believe that recommending brace use of 16 hours leads to better compliance and results.

Night use braces bend the patient's scoliosis into a correct angle, and are prescribed for 8 hours of use during sleep. Some investigators have found that night use braces are not as effective as the day use types.

Bracing may be appropriate for scoliosis due to some types of neuromuscular disease, including spinal muscular atrophy, before growth is finished. Duchenne muscular dystrophy is not treated by bracing,

since surgery is likely to be required, and since later surgery is complicated by loss of respiratory capacity.

Surgery for idiopathic scoliosis is usually recommended if:

- the curve has progressed despite bracing
- the curve is greater than 40–50 degrees before growth has stopped in an adolescent
- the curve is greater than 50 degrees and continues to increase in an adult
- there is significant pain

Orthopedic surgery for neuromuscular scoliosis is often done earlier. The goals of surgery are to correct the deformity as much as possible, to prevent further deformity, and to eliminate pain as much as possible. Surgery can usually correct 40–50% of the curve, and sometimes as much as 80%. Surgery cannot always completely remove pain.

The surgical procedure for scoliosis is called *spinal fusion*, because the goal is to straighten the spine as much as possible, and then to fuse the vertebrae together to prevent further curvature. To achieve fusion, the involved vertebra are first exposed, and then scraped to promote regrowth. Bone chips are usually used to splint together the vertebrae to increase the likelihood of fusion. To maintain the proper spinal posture before fusion occurs, metal rods are inserted alongside the spine, and are attached to the vertebrae by hooks, screws, or wires. Fusion of the spine makes it rigid and resistant to further curvature. The metal rods are no longer needed once fusion is complete, but are rarely removed unless their presence leads to complications.

Spinal fusion leaves the involved portion of the spine permanently stiff and inflexible. While this leads to some loss of normal motion, most functional activities are not strongly affected, unless the very lowest portion of the spine (the lumbar region) is fused. Normal mobility, **exercise**, and even contact sports are usually all possible after spinal fusion. Full recovery takes approximately six months.

Drugs

Pain relievers such as ibuprofen (Tylenol), **sodium**, and **acetaminophen** are used to relieve pain. Non-steroidal anti-inflammatory medications (NSAID) such as **aspirin** are also used to treat both pain and treat inflammation. For severe pain, a narcotic pain medication may be prescribed. In some cases, a nerve block may be performed, involving the injection of pain-relieving medications into the tissues around an affected nerve. The block numbs the nerves and surrounding area and removes the pain sensation.

Alternative

Clinical trials for the treatment of scoliosis are currently sponsored by the National Institutes of Health (NIH) and other agencies. In 2009, NIH reported 25 ongoing or recently completed studies. Some examples include the following:

- A pilot study to determine the presence, frequency, and severity of mental health disorders amongst adolescents undergoing spinal surgery for scoliosis. (NCT00445393)
- The evaluation of the amount of radiographic correction obtained using different spinal instrumentation rods in use for the surgical correction of juvenile and adolescent idiopathic scoliosis. (NCT00510575)
- The evaluation of the risk of curve progression in adolescents with scoliosis who wear a brace versus those who do not and the study of whether there are reliable factors that can predict the usefulness of bracing for a particular individual with scoliosis. (NCT00448448)
- The evaluation of gabapentin to improve postoperative analgesia and reduce narcotic consumption and side effects in children undergoing corrective spinal surgery for idiopathic scoliosis. (NCT00684112)

Clinical trial information is constantly updated by NIH and the most recent information on scoliosis trials can be found at: <http://clinicaltrials.gov>.

Prognosis

The prognosis for a person with scoliosis depends on many factors, including the age at which scoliosis begins and the treatment received. Most cases of mild adolescent idiopathic scoliosis need no treatment, do not progress, and do not cause pain or functional limitations. Untreated severe scoliosis often leads to spondylosis, and may impair breathing.

Prevention

There is no known way to prevent scoliosis. The progression of scoliosis, however, may be prevented through bracing or surgery.

Resources

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American Academy of Orthopaedic Surgeons (AAOS), 317 Massachusetts Ave NE, 1st Floor, Washington, DC, 20002, (202) 546-4430, (202) 546-5051, <http://www.aaos.org>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), 31 Center Dr., Rm. 4C02, MSC 2350, Bethesda, MD, 20892-2350, (301) 496-8190, (877) 22-NIAMS, NIAMInfo@mail.nih.gov, <http://www.niams.nih.gov>.

National Scoliosis Foundation, 5 Cabot Place, Stoughton, MA, 02072, (781) 341-8333, (800) NSF-MYBACK (673-6922), nsf@scoliosis.org, <http://www.scoliosis.org>.

Scoliosis Research Society, 555 East Wells Street, Suite 1100, Milwaukee, WI, 53202-3823, (414) 289-9107, (414) 276-3349, info@srs.org, <http://www.srs.org>.

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KEY TERMS

Radioisotope—An unstable form of an element that gives off radiation to become stable.

Scrotum—The bag of skin below the penis that contains the testes.

Description

A radioisotope, technetium-99, combined in a chemical (pertechnate) is injected intravenously while the patient is under a special machine that detects radiation. This radiation detector, called a gamma camera, scans the scrotum at one minute intervals for about five minutes, then less often for another 10 or 15 minutes. It then creates pictures (either x ray or polaroid) that reveal where the isotope went in the scrotum. Since both sides are scanned, even greater accuracy is obtained by comparison.

Preparation

This procedure is usually done as an emergency to determine the need for immediate surgery.

Risks

The amount of radiation is so slight that even the sensitive testicular tissue is at minimum risk.

Normal results

Blood flow appears unobstructed.

Abnormal results

Three possible possible images appear. They are:

- Increased blood flow indicating infection
- No blood flow indicating testicular torsion
- Blood flow illuminated in a “donut” shaped pattern that indicates torsion that has resolved itself within the last few days.

Resources

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J. Ricker Polsdorfer, MD

Scrotal sonogram see **Scrotal ultrasound**

Scrotal ultrasound

Definition

Scrotal ultrasound is an imaging technique used for the diagnosis of suspected abnormalities of the scrotum. It uses harmless, high-frequency sound waves to form an image. The sound waves are reflected by scrotal tissue to form a picture of internal structures. It is not invasive and involves no radiation.

Purpose

Ultrasound of the scrotum is the primary imaging method used to evaluate disorders of the testicles and surrounding tissues. It is used when a patient has acute **pain** in the scrotum. Some of the problems for which the use of scrotal ultrasound is valuable include an absent or undescended testicle, an inflammation problem, **testicular torsion**, a fluid collection, abnormal blood vessels, or a mass (lump or tumor).

A sudden onset of pain in the scrotum is considered a serious problem, as delay in diagnosis and treatment can lead to loss of function. **Epididymitis** is the most common cause of this type of pain. Epididymitis is an inflammation of the epididymis, a tubular structure that transports sperm from the testes. It is most often caused by bacterial infection, but may occur after injury, or arise from an unknown cause. Epididymitis is treatable with **antibiotics**, which usually resolves pain quickly. Left untreated, this condition can lead to **abscess** formation or loss of blood supply to the testicle.

Testicular torsion is the twisting of the spermatic cord that contains the blood vessels which supply the testicles. It is caused by abnormally loose attachments of tissues that are formed during fetal development. Torsion can be complete, incomplete, or intermittent. Spontaneous detorsion, or untwisting, can occur, making diagnosis difficult. Testicular torsion arises most commonly during adolescence, and is acutely painful. Scrotal ultrasound is used to distinguish this condition from inflammatory problems, such as epididymitis. Testicular torsion is a surgical emergency; it should be operated on as soon as possible to avoid permanent damage to the testes.

A scrotal sac with an absent testicle may be the result of a congenital anomaly (an abnormality present at birth), where a testicle fails to develop. More often, it is due to an undescended testicle. In the fetus, the testicles normally develop just outside the abdomen and descend into the scrotum during the seventh month. Approximately 3% of full-term baby boys have undescended testicles. It is important to distinguish between an

KEY TERMS

Hydrocele—A collection of fluid between two layers of tissue surrounding the testicle; the most common cause of painless scrotal swelling.

Varicocele—An abnormal enlargement of the veins which drain the testicles.

undescended testicle and an absent testicle, as an undescended testicle has a very high probability of developing **cancer**.

Ultrasound can be used to locate and evaluate masses in the scrotum. Most masses within the testicle are malignant or cancerous, and most outside the testicle are benign. Primary cancer of the testicles is the most common malignancy in men between the ages of 15–35. Fluid collections and abnormalities of the blood vessels in the scrotum may appear to the physician as masses and need evaluation by ultrasound. A hydrocele, the most common cause of painless scrotal swelling, is a collection of fluid between two layers of tissue surrounding the testicle. An abnormal enlargement of the veins which drain the testicles is called a varicocele. It can cause discomfort and swelling, which can be examined by touch (palpated). Varicocele is a common cause of male **infertility**.

Precautions

Clear scrotal ultrasound images are difficult to obtain if a patient is unable to remain still.

Description

The patient lies on his back on an examining table. The technologist will usually take a history of the problem, then gently palpate the scrotum. A rolled towel is placed between the patient's legs to support the scrotum. The penis is lifted up onto the abdomen and covered. A gel that enhances sound transmission is put directly on the scrotum. The technologist then gently places a transducer (an electronic imaging device) against the skin. It is moved over the area creating images from reflected sound waves, which appear on a monitor screen. There is no discomfort from the study itself. However, if the scrotum is very tender, even the slight pressure involved may be painful.

Normal results

A normal study would reveal testicles of normal size and shape, with no masses.

Abnormal results

An abnormal result of an ultrasound of the scrotum may reveal an absent or undescended testicle, an inflammation problem, testicular torsion, a fluid collection, abnormal blood vessels, or a mass.

Resources

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Scrub typhus

Definition

Scrub typhus is an **infectious disease** that is transmitted to humans from field mice and rats through the bite of mites that live on the animals. The main symptoms of the disease are **fever**, a wound at the site of the bite, a spotted rash on the trunk, and swelling of the lymph glands.

Description

Scrub typhus is caused by *Rickettsia tsutsugamushi*, a tiny parasite about the size of bacteria that belongs to the family Rickettsiaceae. Under the microscope, rickettsiae are either rod-like (bacilli) or spherical (coccii) in shape. Because they are intracellular parasites, they can live only within the cells of other animals.

R. tsutsugamushi lives primarily in mites that belong to the species *Leptotrombidium (Trombicula) akamushi* and *Leptotrombidium deliense*. In Japan, some cases of scrub typhus have been reportedly transmitted by mites of the species *Leptotrombidium scutellare* and *Leptotrombidium pallidum*. The mites have four-stage life cycles: egg, larva, nymph, and adult. The larva is the only stage that can transmit the disease to humans and other vertebrates.

The tiny chiggers (mite larvae) attach themselves to the skin. During the process of obtaining a meal, they may either acquire the infection from the host or transmit the rickettsiae to other mammals or humans. In regions where scrub typhus is a constant threat, a natural cycle of *R. tsutsugamushi* transmission occurs between mite larvae and small mammals (e.g., field mice and rats). Humans enter a cycle of rickettsial infection only accidentally.

Scrub typhus is also known as *tsutsugamushi disease*.

The name tsutsugamushi is derived from two Japanese words: tsutsuga, meaning something small and dangerous, and mushi, meaning creature. The infection is called scrub typhus because it generally occurs after exposure to areas with secondary (scrub) vegetation. It has recently been found, however, that the disease can also be prevalent in such areas as sandy beaches, mountain deserts, and equatorial rain forests. Therefore, it has been suggested that the names mite-borne typhus, or chigger-borne typhus, are more appropriate. Since the disease is limited to eastern and southeastern Asia, India, northern Australia and the adjacent islands, it is also commonly referred to as tropical typhus.

The seasonal occurrence of scrub typhus varies with the climate in different countries. It occurs more frequently during the rainy season. Certain areas such as forest clearings, riverbanks, and grassy regions provide optimal conditions for the infected mites to thrive. These small geographic regions are high-risk areas for humans and have been called scrub-typhus islands.

Causes and symptoms

The incubation period of scrub typhus is about 10 to 12 days after the initial bite. The illness begins rather suddenly with shaking chills, fever, severe **headache**, infection of the mucous membrane lining the eyes (the conjunctiva), and swelling of the lymph nodes (lymphadenopathy). A wound (lesion) is often seen at the site of the chigger bite. Bite **wounds** are common in whites but rare in Asians.

The initial lesion, which is about 1 cm (0.4 in) in diameter and flat, eventually becomes elevated and filled with fluid. After it ruptures, it becomes covered with a black scab (eschar). The patient's fever rises during the first week, generally reaching 40–40.5°C (104–105°F). About the fifth day of fever, a red spotted rash develops on the trunk, often extending to the arms and legs. It may either fade away in a few days or may become spotted and elevated (maculopapular) and brightly colored. **Cough** is present during the first week of the fever. An infection of the lung (pneumonitis) may develop during the second week.

In severe cases, the patient's pulse rate increases and blood pressure drops. The patient may become delirious and lose consciousness. Muscular twitching may develop. Enlargement of the spleen is observed. Inflammation of the heart muscle (interstitial **myocarditis**) is more common in scrub typhus than in other rickettsial diseases. In untreated patients, high fever may last for more than two weeks. With specific therapy, however,

the fever breaks within 36 hours. The patient's recovery is prompt and uneventful.

Diagnosis

Patient history and physical examination

Differentiating scrub typhus from other forms of typhus as well as from fever, typhoid and meningococcal infections is often difficult during the first several days before the initial rash appears. The geographical location of scrub typhus, the initial sore caused by the chigger bite, and the occurrence of specific proteins capable of destroying the organism (antibodies) in the blood, provide helpful clues and are useful in establishing the diagnosis.

Laboratory tests

Diagnostic procedures involving the actual isolation of rickettsiae from the blood or other body tissues are usually expensive, time-consuming, and hazardous to laboratory workers. As a result, several types of tests known as serological (immunological) tests are used widely to confirm the clinical diagnosis in the laboratory.

Specific antibodies develop in the body in response to an infection. The development of antibodies during the recovery period indicates that an immune response is present. The formation of antibodies is the basic principle of a serological test. Three different tests are available to diagnose rickettsial infections. The most widely used is the Weil-Felix test. This test is based on the fact that some of the antibodies that are formed in the body during a rickettsial infection can react with certain strains (OX-2 and OX-19) of *Proteus* bacteria and cause them to clump (agglutinate). The clumping is easily seen under the microscope. The Weil-Felix test is easy and inexpensive to perform, with the result that it is widely used. The Weil-Felix test, however, is not very specific. In addition, the clumping is not detectable until the second week of the illness, which limits the test's usefulness in early diagnosis.

A second test known as a complement fixation (CF) test is based on the principle that if antibodies are formed in the body in response to the illness, then the antigen and the antibody will form complexes. These antigen-antibody complexes have the ability to inactivate, or fix, a protein that is found in blood serum (serum complement). The serum complement fixation can be measured using standardized biochemical tests and confirms the presence of antibodies. A third test known as the fluorescent antibody test uses fluorescent tags that are attached to antibodies for easy detection. This test has been developed using three strains of

KEY TERMS

Agglutinin—An antibody that causes particulate antigens such as bacteria or other cells to clump together.

Endemic area—A geographical region where a particular disease is prevalent.

Eschar—A hard crust or scab. In scrub typhus, an eschar forms over the initial sore from the chigger bite.

Intracellular parasite—An organism which can only feed and live within the cell of a different animal.

Maculopapular rash—A rash characterized by raised, spotted lesions.

Prophylactic dosage—Giving medications to prevent or protect against diseases.

Rickettsia—A rod-shaped infectious microorganism that can reproduce only inside a living cell. Scrub typhus is a rickettsial disease.

Serological tests—Tests of immune function that are performed using the clear yellow liquid part of blood.

Rickettsia tsutsugamushi and has proven to be the most specific for diagnosis.

Treatment

Scrub typhus is treated with **antibiotics**. Chloramphenicol (Chloromycetin, Fenicol) and tetracycline (Achromycin, Tetracyn) are the drugs of choice. They bring about prompt disappearance of the fever and dramatic clinical improvement. If the antibiotic treatment is discontinued too quickly, especially in patients treated within the first few days of the fever, relapses may occur. In patients treated in the second week of illness, the antibiotics may be stopped one to two days after the fever disappears.

Antibiotics are given intravenously to patients too sick to take them by mouth. Patients who are severely ill and whose treatment was delayed may be given **corticosteroids** in combination with antibiotics for three days.

Prognosis

Before the use of antibiotics, the mortality rate for scrub typhus varied from 1–60%, depending on the geographic area and the rickettsial strain. Recovery also

took a long time. With modern treatment methods, however, deaths are rare and the recovery period is short.

Prevention

General precautions

There are no effective vaccines for scrub typhus. In endemic areas, precautions include wearing protective clothing. Insect repellents containing dibutyl phthalate, benzyl benzoate, diethyl toluamide, and other substances can be applied to the skin and clothing to prevent chigger bites. Clearing of vegetation and chemical treatment of the soil may help to break up the cycle of transmission from chiggers to humans to other chiggers.

Prophylactic antibiotic dosage

It has been shown that a single oral dose of chloramphenicol or tetracycline given every 5 days for a total of 35 days, with 5-day nontreatment intervals, actually produces active immunity to scrub typhus. This procedure is recommended under special circumstances in certain areas where the disease is endemic.

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ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

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Scurvy

Definition

Scurvy is a condition caused by a lack of vitamin C (ascorbic acid) in the diet. Signs of scurvy include tiredness, muscle weakness, joint and muscle aches, a rash on the legs, and bleeding gums. In the past, scurvy was common among sailors and other people deprived of fresh fruits and vegetables for long periods of time.

Description

Scurvy is very rare in countries where fresh fruits and vegetables are readily available and where processed foods have vitamin C added. Vitamin C is an important antioxidant vitamin involved in the development of connective tissues, lipid and vitamin metabolism, biosynthesis of neurotransmitters, immune function, and wound healing. It is found in fruits, especially citrus fruits like oranges, lemons, and grapefruit, and in green leafy vegetables like broccoli and spinach. In adults, it may take several months of vitamin C deficiency before symptoms of scurvy develop.

Currently, the recommended dietary allowance (RDA) for vitamin C is 50–60 mg/day for adults; 35 mg/day for infants; 40–45 mg/day for children 1–14; 70 mg/day during pregnancy; and 90–95 mg/day during lactation. The body's need for vitamin C



An x-ray image of an infant suffering from scurvy. (© Lester V. Bergman/Corbis.)

KEY TERMS

Ascorbic acid—Another term for vitamin C, a nutrient found in fresh fruits and vegetables. Good sources of vitamin C in the diet are citrus fruits like oranges, lemons, limes, and grapefruits, berries, tomatoes, green peppers, cabbage, broccoli, and spinach.

Recommended dietary allowance (RDA)—The daily amount of a vitamin the average person needs to maintain good health.

increases when a person is under **stress**, **smoking**, or taking certain medications.

Causes and symptoms

A lack of vitamin C in the diet is the primary cause of scurvy. This can occur in people on very restricted **diets**, who are under extreme physiological stress (for example, during an infection or after an injury), and in chronic alcoholics. Infants can develop scurvy if they are weaned from breast milk and switched to cow's milk without an additional supplement of vitamin C. Babies of mothers who took extremely high doses of vitamin C during pregnancy can develop infantile scurvy. In children, the deficiency can cause painful swelling of the legs along with **fever**, **diarrhea**, and **vomiting**. In adults, early signs of scurvy include feeling weak, tired, and achy. The appearance of tiny red blood-blisters to larger purplish blotches on the skin of the legs is a common symptom. Wound healing may be delayed and **scars** that had healed may start to break down. The gums swell and bleed easily, eventually leading to loosened teeth. Muscle and joint **pain** may also occur.

Diagnosis

Scurvy is often diagnosed based on the symptoms present. A dietary history showing little or no fresh fruits or vegetables are eaten may help to diagnose vitamin C deficiency. A blood test can also be used to check the level of ascorbic acid in the body.

Treatment

Adult treatment is usually 300–1,000 mg of ascorbic acid per day. Infants should be treated with 50 mg of ascorbic acid up to four times per day.

Prognosis

Treatment with vitamin C is usually successful, if the deficiency is recognized early enough. Left untreated, the condition can cause **death**.

Prevention

Eating foods rich in vitamin C every day prevents scurvy. A supplement containing the RDA of vitamin C will also prevent a deficiency. Infants who are being weaned from breast milk to cow's milk need a supplement containing vitamin C.

Resources

BOOKS

Frankenburg, Frances Rachel. *Vitamin Discoveries and Disasters: History, Science, and Controversies*. Westport, CT: Praeger, 2009.

Altha Roberts Edgren

Seafood poisoning see **Fish and shellfish poisoning**

Seasonal affective disorder

Definition

Seasonal affective disorder (SAD) is a mood disorder in which major depressive episodes and/or manic episodes occur at predictable times of the year, with depressive episodes typically occurring during the fall and winter months. The term SAD can also be applied to depressive episodes with a seasonal pattern that do not meet the criteria for major depressive disorder or a **bipolar disorder** (i.e., subsyndromal). SAD is also sometimes called seasonal mood disorder.

Demographics

SAD is more likely to occur in higher latitudes where there is less light during the fall and winter months. In addition, younger individuals are at higher risk for seasonal depressive episodes than are older persons. Although 60–90% of individuals with a seasonal component to their depressive disorder are women, it is currently unclear whether this reflects a gender factor specifically for SAD or merely reflects the underlying risks associated with recurrent major depressive disorder. Although cases of SAD have been seen in children and adolescents, the disorder usually begins when one is in one's twenties.

Seasonal affective disorder is believed to be relatively common. It is estimated that up to 6% of the population may experience SAD, and up to 20% of Americans may suffer from a mild version of the symptoms (subsyndromal) associated with SAD.

As of 2009, it is not known whether a seasonal pattern is more likely in recurrent major depressive disorder or in bipolar disorders. However, the seasonal pattern is more likely to occur bipolar I disorder (BID) than bipolar II disorder (BIID).

Description

According to the *Diagnostic and Statistic Manual, Fourth Edition*, text revision (*DSM-IV-TR*) of the American Psychiatric Association, a seasonal pattern can exist with major depressive disorder or with major depressive episodes in bipolar I disorder or bipolar II disorder. To be characterized as a seasonal disorder, the onset and remission of the major depressive episodes must occur at characteristic times of year. In most cases of SAD, major depressive episodes occur in the fall and winter months and remit during the spring and summer. Less frequently, some individuals suffer from predictable major depressive episodes during the summer.

Risk factors

Studies have found that women are more likely to be diagnosed with seasonal affective disorder than men, however men with the disorder are more likely to have severe symptoms. SAD is believed to have a hereditary component, so having a close family member who has been diagnosed with SAD is a significant risk factor for the disorder. SAD is more likely to be diagnosed in young adults than in older adults. Individuals who live in areas far from the equator where the duration of sunlight changes substantially during the year are at higher risk for SAD.

Causes and symptoms

It is not known with certainty what causes SAD. Most theories concerning the origins of SAD postulate that it is caused by irregularities in an individual's biological rhythms that are triggered by lengthening or shortening of daylight that occurs with the changing seasons. Among these theories, the phase shift hypothesis (PSH) theorizes that most SAD patients become depressed in the fall and winter because the later dawn at this time of year causes circadian rhythms to become out of synchronization with respect to clock time and the body's sleep-wake cycle. Specifically, the PSH theorizes that SAD is a result of a mismatch between an individual's circadian rhythms related to the sleep-wake

KEY TERMS

Bipolar disorder—Formerly called manic-depressive disorder. A mood disorder characterized alternating periods of overconfidence and activity (manic highs) and depressive lows.

Melatonin—A naturally occurring hormone involved in regulating the body's "internal clock."

Phototherapy—Also called light therapy, the patient is exposed to a bright light to compensate for reduced exposure to sunlight.

Serotonin—5-Hydroxytryptamine; a substance that occurs throughout the body with numerous effects including neurotransmission. Inadequate amounts of serotonin are implicated in some forms of depression and obsessive-compulsive disorder.

cycle and the biological circadian pacemaker in the hypothalamus of the brain.

Another suggested cause of SAD is related to the body's melatonin levels. The body produces more melatonin at night than during the day, and scientists believe it helps people feel sleepy at nighttime. There is also more melatonin in the body during winter, when the days are shorter. Some researchers believe that excessive melatonin release during winter in people with SAD may account for their feelings of drowsiness or depression.

Researchers have also suggested that SAD may be caused at least in part by reduced serotonin levels. Serotonin is a neurotransmitter important for the regulation of mood. Reduced levels of sunlight have been shown to be linked to drops in serotonin levels in the brain, which can cause symptoms of depression.

Common symptoms of SAD include:

- depression and irritability
- lack of energy
- excessive sleepiness during the day or abnormally prolonged sleep at night (hypersomnia)
- tendency to overeat (hyperphagia), including weight gain and/or craving for carbohydrates
- significant impairment of social and occupational functioning (e.g., lack of interest in social interactions, increased sensitivity to negative reactions from others, or lack of interest in normally enjoyable activities)

Diagnosis

Four criteria must be met for a major depressive disorder, BID, or BIID to be characterized as seasonal. First, there must be a regular relationship between the onset of the depressive episodes and the time of year. For most cases of SAD, depressive episodes occur during the fall and winter seasons. Second, full remission of the depressive episodes (or a change from depression to **mania** or hypomania in the case of bipolar disorders) must also occur at predictable times of the year. Third, the seasonal cycle of onset and remission of major depressive episodes must have occurred within the last two years without any nonseasonal depressive episodes during that time. Fourth, seasonal episodes of depression must occur significantly more frequently than non-seasonal depressive episodes over the course of the person' lifetime.

When diagnosing SAD, it is important to distinguish it from depression caused by other factors that may cause depression such as seasonal unemployment or school schedule. In addition, SAD should be distinguished from the "holiday blues." The holiday blues are not related to circadian rhythms but to such psychosocial factors as increased obligations, expectations that one should be joyous, or early childhood memories or unresolved childhood conflicts.

Treatment

The goal of treatment for SAD is to alleviate the symptoms associated with the disorder. In most cases, the prescribed treatment regimen needs to be followed for the months in which the SAD occurs and can be stopped during the remainder of the year. Many people with SAD can be treated effectively with **light therapy**, although some may require the addition of other therapies or medications.

Traditional

The first-line treatment for seasonal affective disorder is **phototherapy**, also called light therapy, which exposes the patient to bright artificial light to compensate for the gloominess of winter. Light therapy uses a device called a light box that contains a set of fluorescent or incandescent lights in front of a reflector. Typically, the patient sits for 30 minutes next to a 10,000-lux box (which is about 50 times brighter than ordinary indoor light). Light therapy appears to be safe for most people. However, it may be harmful for those with eye diseases. The most common side effects are vision problems such as eyestrain, headaches, irritability, and **insomnia**. In addition, hypomania (elevated or expansive mood,

characterized by hyperactivity and inflated self esteem) may occasionally occur.

Drugs

When a major depressive disorder or a bipolar disorder has seasonal characteristics, it may be treated with antidepressant medication. Research has found that fluoxetine (Prozac) is as effective as light therapy in controlled clinical trials. In 2006, the U.S. Food and Drug Administration approved the prescription medication Wellbutrin XL (bupropion HCl extended release tablets) for the prevention of SAD. The effectiveness of Wellbutrin XL has been demonstrated in clinical trials with adults having a history of a major depressive disorder occurring in the fall and winter months. Wellbutrin XL, however, is recommended only for individuals whose SAD symptoms meet the criteria for a major depressive disorder. Other antidepressants may also be prescribed for SAD.

Alternative

The literature also suggests that the over-the-counter compound melatonin may be of help in alleviating SAD symptoms. Melatonin is a hormone produced by the pineal gland that helps regulate the body' seasonal changes. Research funded by the National Institute of Mental Health suggests that a low dose of synthetic or pharmacy-grade melatonin taken in the evening along with exposure to bright light in the morning may be effective in relieving the symptoms of SAD. However, more research needs to be done to determine the safety and effectiveness of such treatment.

Home remedies

Cases of SAD that do not meet all the criteria for formal diagnosis (subsyndromal cases) may also be improved with phototherapy. Activities such as getting outdoors in the sunshine in the morning or rearranging one's home or office to maximize exposure to sunlight during the day may help mild SAD symptoms. Although a trip to the tropics or other sunny place is also of help in overcoming the effects of SAD, the problem returns once the individual is again exposed to shortened daylight hours.

Prognosis

For cases of subsyndromal SAD, the prognosis for control of symptoms through phototherapy treatment is good. For cases in which SAD is a seasonal characteristic of a major depressive disorder or bipolar disorder, the prognosis is generally the same as for the underlying disorder.

Prevention

There is no known way to prevent SAD with certainty. Spending time during waking hours in direct sunlight may help to prevent or reduce symptoms. If an individual has experienced SAD in the past, a doctor or therapist may recommend beginning treatment before the symptoms are expected to occur to help control the symptoms and to minimize the negative effects of the disorder.

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ORGANIZATIONS

- Depression and Bipolar Support Alliance (DBSA), 730 N. Franklin Street, Suite 501, Chicago, IL, 60654-7225, (800) 826-3632, (312) 642-7234, <http://www.dbsalliance.org>.
 Mental Health America, 2000 N. Beauregard St., 6th Floor, Alexandria, VA, 22311, (703) 684-7722, (800) 969-6642, (703) 684-5968, <http://www.nmha.org>.
 National Institute of Mental Health, 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD, 20892-9663, (301) 443-4513. TTY (301) 443-8431, (866) 615-6464 TTY (866) 415-8051, (301) 443-4279, <http://www.nimh.nih.gov>.

Ruth A. Wienclaw, PhD
 Tish Davidson, AM

Seasonal depression see **Seasonal affective disorder**

Seatworm infection see **Enterobiasis**



This young boy is afflicted with seborrheic dermatitis.
(Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

Seborrheic dermatitis appears as red, inflamed skin covered by greasy or dry scales that may be white, yellowish, or gray. It can effect the scalp, eyebrows, forehead, face, folds around the nose and ears, the chest, armpits (axilla), and groin. Dandruff and cradle cap are mild forms of seborrheic dermatitis, and appear as fine white scales without inflammation.

Causes and symptoms

The cause of seborrheic dermatitis is unclear, though it has been linked to genetic or environmental factors. *Pityrosporum ovale*, a species of yeast normally found in hair follicles, has been proposed as one possible causative factor. A high fat diet and alcohol ingestion are thought to play some role. Other possible risk factors include:

- stress and fatigue
- weather extremes (e. g. hot, humid weather or cold, dry weather)
- oily skin
- infrequent shampoos

Seborrheic dermatitis

Definition

Seborrheic **dermatitis** is a common inflammatory disease of the skin characterized by scaly lesions usually on the scalp, hairline, and face.

KEY TERMS

Acne—A chronic inflammation of the sebaceous glands that manifests as blackheads, whiteheads, and/or pustules on the face or trunk.

Psoriasis—A skin disorder of chronic, itchy scaling most commonly at sites of repeated minor trauma (e.g. elbows, knees, and skin folds). It affects up to 2% of the population in Western countries—males and females equally.

Rosacea—A chronic inflammation of the face, with associated scattered round nodules and increased reactivity of the facial capillaries to heat. It is most common in females, aged 30–50 years.

- obesity
- Parkinson's disease
- AIDS
- use of drying lotions that contain alcohol
- other skin disorders (for example acne, rosacea, or psoriasis)

Mild forms of the disorder may be asymptomatic. Symptoms also disappear and reappear, and vary in intensity over time. When scaling is present, it may be accompanied by **itching** that can lead to secondary infection.

Diagnosis

The diagnosis of seborrheic dermatitis is based on assessment of symptoms, accompanied by consideration of medical history.

Treatment

Treatment consists of vigorous shampoos with preparations that assist with softening and removing the scaly accumulations. For mild cases, a non-prescription shampoo with selenium sulfide or zinc pyrithione may be used. For more severe problems, the doctor may prescribe shampoos containing coal tar or scalp creams containing cortisone. The antiseborrheic shampoo should be left on the scalp for approximately five minutes before rinsing out. Hydrocortisone cream may also be ordered for application to the affected areas on the face and body. Application of the hydrocortisone should be discontinued when the condition clears and restarted with recurrence.

Prognosis

This chronic condition may be characterized by long periods of inactivity. Symptoms in the acute phase can be controlled with appropriate treatment.

Prevention

The condition cannot be prevented. The severity and frequency of flare-ups may be minimized with frequent shampoos, thorough drying of skin folds after bathing, and wearing of loose, ventilating clothing. Foods that appear to worsen the condition should be avoided.

Resources

BOOKS

James, William D., et al. *Andrews' Diseases of the Skin: Clinical Dermatology*. 10th ed. Philadelphia: Saunders Elsevier, 2006.

Kathleen D. Wright, RN

Secobarbital see **Barbiturates**

Secondary erythrocytosis see **Secondary polycythemia**

Secondary polycythemia

Definition

Secondary polycythemia is an acquired form of a rare disorder characterized by an abnormal increase in the number of mature red cells in the blood.

Secondary polycythemia is also called secondary erythrocytosis.

Description

Polycythemia means too many red blood cells. The resulting excess of red cells thickens the blood and impedes its passage through small blood vessels.

Secondary polycythemia usually affects people between the ages of 40 and 60.

Types of secondary polycythemia

Known as spurious polycythemia, stress polycythemia, or Gaisböck's syndrome, relative polycythemia is characterized by normal numbers of red blood cells but decreased levels of plasma (the fluid part of the blood). Overweight, middle-aged white men who smoke, have high blood pressure, and are on diuretic medicines to

remove excess water from their bodies may develop Gaisbock's syndrome.

In smoker's polycythemia, the number of red blood cells is elevated. Plasma levels are abnormally low.

Causes and symptoms

Smoking, which impairs red blood cells' ability to deliver oxygen to body tissues, can cause secondary polycythemia. So can the following conditions:

- carbon monoxide poisoning
- chronic heart or lung disease
- hormonal (endocrine) disorders
- exposure to high altitudes
- kidney cysts
- tumors of the brain, liver, or uterus.

Causes of spurious polycythemia include:

- burns
- diarrhea
- hemoconcentration (higher-than-normal concentration of cells and solids in the blood, usually due to becoming dehydrated or taking diuretics)
- stress

Weakness, headaches, and **fatigue** are usually the first symptoms of secondary polycythemia. Patients may feel lightheaded or experience **shortness of breath**.

Visual disturbances associated with this disorder include distorted vision, blind spots, and flashes of light. The gums and small cuts are likely to bleed, and the hands and feet may burn. Extensive **itching** often occurs after taking a bath or shower.

Pain in the chest or leg muscles is common. The face often becomes ruddy, then turns blue after **exercise** or other exertion. Confusion and ringing in the ears (**tinnitus**) may also occur.

Diagnosis

A very important part of diagnosing secondary polycythemia is differentiating it from primary polycythemia (also called polycythemia rubra vera or Vaquez' disease). Unlike secondary polycythemia, primary polycythemia cannot be traced to an underlying condition such as smoking, high altitude, or chronic lung disease.

Doctors diagnose polycythemia by measuring oxygen levels in blood drawn from an artery. A patient whose oxygen level is abnormally low probably has secondary polycythemia. Erythropoietin may also be measured. Levels of this hormone, which stimulates the bone marrow to produce red blood cells, may be normal or elevated in a patient with secondary polycythemia. Red

blood cell mass is also frequently measured in diagnosing the disorder.

Imaging studies are sometimes performed to determine whether the spleen and liver are enlarged and to detect erythropoietin-producing kidney lesions. Other diagnostic procedures include chest x rays and an electrocardiogram (EKG).

Treatment

Secondary polycythemia is treated primarily by treating the underlying condition causing the disorder. For example, patients with Gaisbock's syndrome are often taken off **diuretics** and encouraged to lose weight. Lung disorders, such as **chronic obstructive pulmonary disease** (COPD), may cause secondary polycythemia; treating the lung disorder generally improves the polycythemia.

Some medications may also be taken to treat symptoms caused by polycythemia. For example, **antihistamines** can alleviate itching, and **aspirin** can soothe burning sensations and bone pain.

Until the underlying condition is controlled, doctors use bloodletting (**phlebotomy**) to reduce the number of red blood cells in the patient's body. In most instances, a pint of blood is drained from the patient as needed and tolerated, until the **hematocrit** (the proportion of red cells in the blood) reaches an acceptable level. **Chemotherapy** is not used to treat secondary polycythemia; however, it may be used to treat the primary form.

Prognosis

Curing or removing the underlying cause of this disorder generally eliminates the symptoms.

Resources

OTHER

"Secondary Erythrocytosis." General Practice Notebook. 2010. <http://www.gpnotebook.co.uk/simplepage.cfm?ID=-919273435> (accessed December 16, 2010).

Maureen Haggerty

SED rate see **Erythrocyte sedimentation rate**

Sedation

Definition

Sedation is the act of calming by administration of a sedative. A sedative is a medication that commonly induces the nervous system to calm.

Purpose

The process of sedation has two primary intentions. First, sedation is recommended to allow patients the ability to tolerate unpleasant diagnostic or surgical procedures and to relieve **anxiety** and discomfort. Second, sedation for uncooperative patients may expedite and simplify special procedures that require little or no movement. Additionally, sedation is often desirable to diminish fear associated with operative procedures. Sedation is typically used for common diagnostic tests that require prolonged **immobilization** such as **magnetic resonance imaging** (MRI) and computed axial tomography (CAT) scanning. Some cases that require sedation may also necessitate the use of **analgesics** to decrease **pain** associated with a procedure or test.

Precautions

Benzodiazepines (common sedative medication) have a cumulative effect. This means that if the patient has not had time to metabolize the previous dose and ingests more, then the sedative effect may increase. Because of these additive effects, these medications taken with other sedatives or alcohol (also a sedative hypnotic drug) may increase chances for accidental **death**. In general, most of the medications that induce sedation may alter breathing and cardiac stability. In patients with preexisting lung and/or heart disease, these medications should be monitored closely or not prescribed.

Description

The future of anesthetic care involves the simultaneous administration of several drugs including IV medications and inhaled anesthetics. An extensive survey of death in 100,000 cases published in 1988 revealed that death within seven days was 2.9 times greater when one or two anesthetic drugs were used than when using three or more medications. This study is accepted as standard practice and multiple IV anesthetics is the preferable recommendation for optimal patient care.

The procedure for sedation is usually explained to the patient by an attending clinician. An IV access line is set in place for fluid replacement and injection of medications. A history is usually taken to assess risk and choice of medication. The patient typically signs consent forms and the possible side effects are explained. The day before the test, the patient may be required to maintain specified dietary restriction.

For outpatient surgery there are two types of sedation, conscious and unconscious sedation. Patients receiving conscious sedation are capable of rational responses, and they are able to maintain their airway

for ventilation. The hallmark of conscious sedation is that it does not alter respiratory, cardiac, or reflex functions (nerve reflexes from the brain) to the level that requires external support for these vital functions. Patients receiving conscious sedation are cooperative, have stable vital signs (pulse, respiratory rate, and temperature), shorter recovery room convalescence, and lower risk of developing drug-induced complications. Unconscious sedation is a controlled state of anesthesia, characterized by partial or complete loss of protective nerve reflexes, including the ability to independently breathe and respond to commands. The patient is unable to cooperate, has labile (fluctuating) vital signs, prolonged recovery room convalescence, and higher risk of anesthetic complications.

Preparation

Usually procedures for conscious sedation do not require preoperative or pre-testing orders. Clinical situations for unconscious sedation typically involve eating and drinking protocols starting the day before the procedure.

The age and physical status of the patient is useful in determining sensitivity. A detailed past history, especially prior experiences with sedatives and other anesthetics is an important part of preparatory assessment. It is important to determine if there were any untoward side effects associated with a previous medication. Patient positioning is important to prevent blood pressure changes or nerve damage associated with abnormal position.

Patients are also monitored for pulse rate, respiration, blood pressure, and temperature. Additionally, the heart is monitored using **electrocardiography** (ECG). Ventilation is assessed using a pulse oximeter. This machine is clipped with a special probe on one finger and can measure the levels of oxygen and carbon dioxide, which are reliable indicators of respiratory status.

Aftercare

The major goal for recovery room monitoring is assessment of residual drug effects. Recovery room monitoring primarily focuses on heart stability, respiratory adequacy and return to previous brain functioning.

Risks

The original forms of diazepam (Valium, a very common sedative) caused irritation of veins and phlebitis. Newer forms of diazepam (Dizac) are chemically improved to lower the possibility of vein irritation. Age and physical health are important risk factors. Preexisting medical conditions such as high blood pressure and

KEY TERMS

Baseline—A return to an original state.

Diazepam—One of the most commonly used sedative-hypnotic medications.

heart and lung disease may increase the chance of developing undesirable side effects.

Normal results

Normal or uncomplicated results for sedation include alleviation of anxiety and discomfort. Coupled with analgesic, patients are usually pain-free. The normal progression post procedure or post operatively would be to return to baseline brain functioning, unassisted breathing, and normal heart rate and rhythm.

Abnormal results

Patients may have excessive **nausea and vomiting** associated with narcotic analgesia (if this is indicated). Excessive drowsiness can occur secondary to benzodiazepine-induced sedation. The patient can also develop hypoventilation (a decrease in ventilation), airway obstruction, high or low blood pressure, abnormal heart rhythms, **nausea, vomiting**, and shivering.

Resources

BOOKS

Fleisher, Gary R., et al. *Textbook of Pediatric Emergency Medicine* 6th ed. Lippincott Williams & Wilkins, 2010.
 Miller, Ronald D., et al, eds. *Anesthesia*. 7th ed. Philadelphia: Churchill Livingstone, 2010.

Laith Farid Gulli, M.D.
 Bilal Nasser, M.Sc.

Sedative-hypnotic drugs see **Anti-insomnia drugs**

Sedimentation rate see **Erythrocyte sedimentation rate**



This patient's brain is exposed during surgery in order for surgeons to remove the mass responsible for his epilepsy. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

manifestations. **Epilepsy** is a condition characterized by recurrent seizures that may include repetitive muscle jerking called convulsions.

Description

There are more than 20 different seizure disorders. One in ten Americans will have a seizure at some time during their lifetime. More than 3 million Americans are affected by seizures.

Epilepsy affects 1–2% of the population of the United States. About 2.7 million Americans have active epilepsy, meaning that in the past 5 years they have had a seizure or been on medication for epilepsy.) Epilepsy becomes more prevalent with increased age. About 1% of people under age 20 have epilepsy, and about 3% of people over age 75 have it. About 200,000 new cases of epilepsy are diagnosed each year.

Most seizures are benign, but a seizure that lasts a long time can lead to status epilepticus, a life-threatening condition characterized by continuous seizures, sustained loss of consciousness, and respiratory distress. Non-convulsive epilepsy can impair physical coordination, vision, and other senses. Undiagnosed seizures can lead to conditions that are more serious and more difficult to manage.

Seizure disorder

Definition

A seizure is a sudden disruption of the brain's normal electrical activity accompanied by altered consciousness and/or other neurological and behavioral



This abstract artwork is based on a patient's description of what an epileptic seizure feels like. Epileptic seizures are caused by chaotic electrical activity in the brain. They can be triggered by a variety of factors, such as illness or stress, although the underlying causes are not completely understood. (John Bavosi/Photo Researchers, Inc.)

Types of seizures

Generalized epileptic seizures occur when electrical abnormalities occur throughout the brain. A partial seizure does not involve the entire brain. The area in which the seizure begins is called the epileptic focus. Many seizures stay localized, but some spread to other parts of the brain and cause a generalized seizure. Some people who have epilepsy have more than one type of seizure.

Seizures which involve areas of the brain necessary for motor control can cause parts of the body to jerk repeatedly. This type of seizure can last seconds or minutes, or rarely, more than an hour. Seizures that last more than a few minutes can cause serious long-term disability and even **death**. Sensory seizures begin with **numbness** or **tingling** in one area. The sensation may move along one side of the body or the back before subsiding.

Visual seizures, which affect the area of the brain that controls sight, cause people to see things that are not there. Auditory seizures affect the part of the brain that controls hearing and may cause the individual to imagine voices, music, or other sounds. Other types of seizures can cause confusion, upset stomach, or emotional distress.

GENERALIZED SEIZURES. A generalized tonic-clonic (grand-mal) seizure begins with a loud cry before the person having the seizure loses consciousness and falls to the ground. The muscles become rigid for about 30 seconds during the tonic phase of the seizure and alternately contract and relax during the clonic phase, which lasts 30–60 seconds. The skin sometimes acquires a bluish tint and the person may bite his tongue, lose bowel or bladder control, or have trouble breathing.

A grand mal seizure usually last one to two minutes, and the person may be confused or have trouble talking when consciousness is regained (postictal state). The individual may complain of head or muscle aches, weakness in the arms or legs, or be extremely drowsy or fatigued.

PRIMARY GENERALIZED SEIZURES. A primary generalized seizure occurs when electrical discharges begin in both halves (hemispheres) of the brain at the same time. Primary generalized seizures are more likely to be major motor attacks than to be absence seizures.

ABSENCE SEIZURES. Absence (petit mal) seizures generally begin between the ages of 5 and 15. The seizures usually begin with a brief loss of consciousness and last between 2 and 3 seconds. Sometimes the episodes may last up to 30 seconds. An individual having a petit mal seizure becomes very quiet and may blink or stare blankly and may exhibit facial twitching, eye rolling, or lip movement. When it ends, the individual who had the seizure resumes whatever he or she was doing before the seizure began. The individual will not remember the seizure and may not realize that anything unusual has happened. Untreated, petit mal seizures can recur as many as 100 times a day and may progress to grand mal seizures.

MYOCLONIC SEIZURES. Myoclonic seizures are characterized by brief, involuntary spasms of the tongue or muscles of the face, arms, or legs. Myoclonic seizures are most apt to occur when waking after a night's sleep. These seizures do not generally cause loss of consciousness.

A Jacksonian seizure is a partial seizure characterized by tingling, stiffening, or jerking of an arm or leg. Loss of consciousness is rare. The seizure may progress in characteristic fashion along the limb.

Limp posture and a brief period of unconsciousness are features of atonic seizures, which occur in young children. Atonic seizures, which cause the child to fall, also are called drop attacks.

PARTIAL SEIZURES. Simple partial seizures do not spread from the focal area where they arise. Symptoms are determined by the part of the brain affected. The

individual usually remains conscious during the seizure and may be able to describe it later.

COMPLEX PARTIAL SEIZURES. A distinctive smell, taste, or other unusual sensation (aura) may signal the start of a complex partial seizure. These seizures start as simple partial seizures, but move beyond the focal area and cause loss of consciousness. Complex partial seizures can become major motor seizures. Although a person having a complex partial seizure may not seem to be unconscious, he or she does not know what is happening and may behave strangely or inappropriately. The individual will not remember the seizure, and may seem confused or intoxicated for a few minutes after it ends.

Causes and symptoms

The origin of 50–70% of all cases of epilepsy is unknown. Epilepsy sometimes is the result of trauma at birth. Such causes include insufficient oxygen to the brain; **head injury**; heavy bleeding or incompatibility between a woman's blood and the blood of her newborn baby; and infection immediately before, after, or at the time of birth.

Other causes of epilepsy include:

- head trauma resulting from a car accident, gunshot wound, or other injury.
- alcoholism
- brain abscess or inflammation of membranes covering the brain or spinal cord
- phenylketonuria (PKU, a disease that is present at birth, often is characterized by seizures, and can result in mental retardation) and other inherited disorders
- infectious diseases like measles, mumps, and diphtheria
- degenerative disease
- lead poisoning, mercury poisoning, carbon monoxide poisoning, or ingestion of some other poisonous substance
- genetic factors

Status epilepticus, a condition in which a person suffers from continuous seizures and may have trouble breathing, can be caused by:

- suddenly discontinuing anti-seizure medication
- hypoxic or metabolic encephalopathy (brain disease resulting from lack of oxygen or malfunctioning of other physical or chemical processes)
- acute head injury
- blood infection caused by inflammation of the brain or the membranes that cover it

Diagnosis

Personal and family medical history, description of seizure activity, and physical and neurological examinations help primary care physicians, neurologists, and epileptologists diagnose this disorder. Doctors rule out conditions that cause symptoms that resemble epilepsy, including small strokes (transient ischemic attacks, or TIAs), **fainting**, (syncope), pseudoseizures, and sleep attacks (narcolepsy.) It is often helpful for a family member or other person present during the seizure to describe the event to the physician, as the individual who had the seizure may not remember it.

Neuropsychological testing can uncover any learning or memory problems that may be related to the seizure or seizures. Neuroimaging provides views of brain areas involved in seizure activity.

The electroencephalogram (EEG) is the main test used to diagnose epilepsy. EEGs use electrodes placed on or within the skull to record the brain's electrical activity and pinpoint the exact location of abnormal discharges.

The patient may be asked to remain motionless during a short-term EEG or to go about his normal activities during extended monitoring. Some patients are deprived of sleep or exposed to seizure triggers, such as rapid, deep breathing (hyperventilation) or flashing lights (photic stimulation). In some cases, people may be hospitalized for EEG monitoring that can last as long as two weeks. Video EEGs also document what the patient was doing when the seizure occurred and how the seizure changed his or her behavior.

Other techniques used to diagnose epilepsy include:

- Magnetic resonance imaging (MRI), which provides clear, detailed images of the brain. Functional MRI (fMRI), performed while the patient does various tasks, can measure shifts in electrical intensity and blood flow and indicate which brain region each activity affects.
- Positron emission tomography (PET) and single photon emission tomography (SPECT) monitor blood flow and chemical activity in the brain area being tested. PET and SPECT are very effective in locating the brain region where metabolic changes take place between seizures.

Treatment

The goal of epilepsy treatment is to eliminate seizures or make the symptoms less frequent and less severe. Long-term anticonvulsant drug therapy is the most common form of epilepsy treatment.

Medication

A combination of drugs may be needed to control some symptoms, but most patients who have epilepsy take one of the following medications:

- Dilantin (phenytoin)
- Tegretol (carbamazepine)
- Barbita (phenobarbital)
- Mysoline (primidone)
- Depakene (valproic acid, sodium valproate)
- Klonopin (clonazepam)
- Zarontin (ethosuximide).

Dilantin, Tegretol, Barbita, and Mysoline are frequently used to manage or control generalized tonic-clonic and complex partial seizures. Depakene, Klonopin, and Zarontin are often prescribed for patients who have absence seizures.

Neurontin (gabapentin), Lamictal (lamotrigine), and topiramate (Topamax) are among the medications more recently approved in the United States to treat adults who have partial seizures or partial and grand mal seizures. Another new medication called Levetiracetam (Keppra) has been approved and shows particularly good results in reducing partial seizures among elderly patients with few side effects. This is important, because elderly patients often have other conditions and must take other medications that might interact with seizure medications. Available medications frequently change, and it the physician will determine the best treatment for an individual patient. It is believed that monotherapy, or using just one medication rather than a combination, may work better for most patients. The less complicated the treatment, the more likely the patient will comply and better manage the seizure disorder.

Even an individual whose seizures are well controlled should have regular blood tests to measure levels of anti-seizure medication in his or her system and to check to see if the medication is causing any changes in the blood or liver. A doctor should be notified if any signs of drug toxicity appear, including uncontrolled eye movements; sluggishness, **dizziness**, or hyperactivity; inability to see clearly or speak distinctly; **nausea** or **vomiting**; or sleep problems.

Status epilepticus requires emergency treatment, usually with Valium (Ativan), Dilantin, or Barbita. An intravenous dextrose (sugar) solution is given to patients whose condition is due to low blood sugar, and a vitamin B₁ preparation is administered intravenously when status epilepticus results from chronic alcohol withdrawal. Because dextrose and thiamine are essentially harmless and because delay in treatment can be disastrous, these medications are given routinely, as it is usually difficult to

obtain an adequate history from a patient suffering from status epilepticus.

Intractable seizures are seizures that cannot be controlled with medication or without **sedation** or other unacceptable side effects. Surgery may be used to eliminate or control intractable seizures.

Surgery

Surgery can be used to treat patients whose intractable seizures stem from small focal lesions that can be removed without endangering the patient, changing the patient's personality, dulling the patient's senses, or reducing the patient's ability to function.

Each year, as many as 5,000 new patients may become suitable candidates for surgery, which most often is performed at a comprehensive epilepsy center. Potential surgical candidates include patients with:

- partial seizures and secondarily generalized seizures (attacks that begin in one area and spread to both sides of the brain)
- seizures and childhood paralysis on one side of the body (hemiplegia)
- complex partial seizures originating in the temporal lobe (the part of the brain associated with speech, hearing, and smell) or other focal seizures. (However, the risk of surgery involving the speech centers is that the patient will lose speech function.)
- Generalized myoclonic seizures or generalized seizures featuring temporary paralysis (akinetic) or loss of muscle tone (atonic)

A physical examination is conducted to verify that a patient's seizures are caused by epilepsy. Surgery is not used to treat patients with severe psychiatric disturbances or medical problems that raise risk factors to unacceptable levels.

Surgery is never indicated unless:

- the best available anti-seizure medications have failed to control the patient's symptoms satisfactorily
- the origin of the patient's seizures has been precisely located
- there is good reason to believe that surgery will significantly improve the patient's health and quality of life.

Every patient considering epilepsy surgery is carefully evaluated by one or more neurologists, neurosurgeons, neuropsychologists, and/or social workers. A psychiatrist, chaplain, or other spiritual advisor may help the patient and his family cope with the **stress** that occurs during and after the selection process.

TYPES OF SURGERY. Surgical techniques used to treat intractable epilepsy include:

- Lesionectomy. Removing the lesion (diseased brain tissue) and some surrounding brain tissue is very effective in controlling seizures. Lesionectomy is generally more successful than surgery performed on individuals whose seizures are not caused by clearly defined lesions. Removing only part of the lesion lessens the effectiveness of the procedure.
- Temporal resections. Removing part of the temporal lobe and the part of the brain associated with feelings, memory, and emotions (the hippocampus) provides good or excellent seizure control in 75–80% of properly selected patients with appropriate types of temporal lobe epilepsy. Some patients experience post-operative speech and memory problems.
- Extra-temporal resection. This procedure involves removing some or all of the frontal lobe, the part of the brain directly behind the forehead. The frontal lobe helps regulate movement, planning, judgment, and personality. Special care must be taken to prevent post-operative problems with movement and speech. Extra-temporal resection is most successful in patients whose seizures are not widespread.
- Hemispherectomy. This method of removing brain tissue is restricted to patients with severe epilepsy and abnormal discharges that often extend from one side of the brain to the other. Hemispherectomies most often are performed on infants or young children who have had an extensive brain disease or disorder since birth or from a very young age.
- Corpus callosotomy. This procedure, an alternative to hemispherectomy in patients with congenital hemiplegia, removes some or all of the white matter that separates the two halves of the brain. Corpus callosotomy is performed almost exclusively on children who are frequently injured during falls caused by seizures. If removing two-thirds of the corpus callosum does not produce lasting improvement in the patient's condition, the remaining one-third will be removed during another operation.
- Multiple subpial transection. This procedure is used to control the spread of seizures that originate in or affect the “eloquent” cortex, the area of the brain responsible for complex thought and reasoning.

Other forms of treatment

KETOGENIC DIET. A special high-fat, low-protein, low-carbohydrate diet sometimes is used to treat patients whose severe seizures have not responded to other treatment. Calculated according to age, height, and weight, the ketogenic diet induces mild **starvation** and **dehydration**. This forces the body to create an excessive supply of ketones, natural chemicals with seizure-suppressing properties.

The goal of this controversial approach is to maintain or improve seizure control while reducing medication. The ketogenic diet works best with children between the ages of one and 10. It is introduced over a period of several days, and most children are hospitalized during the early stages of treatment.

If a child following this diet remains seizure-free for at least six months, increased amounts of carbohydrates and protein gradually are added. If the child shows no improvement after three months, the diet is gradually discontinued.

Introduced in the 1920s, the ketogenic diet has had limited, short-term success in controlling seizure activity. Its use exposes patients to such potentially harmful side effects as:

- staphylococcal infections
- stunted or delayed growth
- low blood sugar (hypoglycemia)
- excess fat in the blood (hyperlipidemia)
- disease resulting from calcium deposits in the urinary tract (urolithiasis)
- disease of the optic nerve (optic neuropathy)

VAGUS NERVE STIMULATION. The United States Food and Drug Administration (FDA) has approved the use of vagus nerve stimulation (VNS) in patients over the age of 16 who have intractable partial seizures. This non-surgical procedure uses a pacemaker-like device implanted under the skin in the upper left chest, to provide intermittent stimulation to the vagus nerve. Stretching from the side of the neck into the brain, the vagus nerve affects swallowing, speech, breathing, and many other functions, and VNS may prevent or shorten some seizures. Approximately 80% of patients experience fewer seizures after the procedure. Individuals having undergone this procedure may experience side effects such as dizziness, **memory loss**, weight gain, and slurred speech.

First aid for seizures

A person having a seizure should not be restrained, but sharp or dangerous objects should be moved out of reach. Anyone having a complex partial seizure may be warned away from danger by someone calling his or her name in a clear, calm voice.

A person having a grand mal seizure should be helped to lie down. Tight clothing should be loosened. A soft, flat object like a towel or the palm of a hand should be placed under the person's head. Forcing objects into the mouth of someone having a grand mal seizure could cause injuries or breathing problems, and the individual trying to help may be injured if the jaw

KEY TERMS

Acupressure—Needleless acupuncture.

Acupuncture—An ancient Chinese method of relieving pain or treating illness by piercing specific areas of the body with fine needles.

Biofeedback—A learning technique that helps individuals influence automatic body functions.

Epileptologist—A physician who specializes in the treatment of epilepsy.

clenches shut. The individual having the seizure should be turned on his or her side if consciousness has been lost. This will ensure the individual is able to breathe. After a grand mal seizure has ended, the person who had the seizure should be calmly told what has happened and reminded of where he or she is.

Alternative treatment

Stress increases seizure activity in 30% of people who have epilepsy. Relaxation techniques can provide some sense of control over the disorder, but they should never be used instead of anti-seizure medication or used without the approval of the patient's doctor. **Yoga**, **meditation**, and favorite pastimes help some people relax and manage stress more successfully. **Biofeedback** can teach adults and older adolescents how to recognize an aura and what to do to stop its spread. Children under 14 are not usually able to understand and apply principles of biofeedback. **Acupuncture** treatments (acupuncture needles inserted for a few minutes or left in place for as long as 30 minutes) make some people feel pleasantly relaxed. **Acupressure** can have the same effect on children or on adults who dislike needles.

Aromatherapy involves mixing aromatic plant oils into water or other oils and massaging them into the skin or using a special burner to waft their fragrance throughout the room. Aromatherapy oils affect the body and the brain, and undiluted oils should never be applied directly to the skin. Ylang ylang, chamomile, or lavender can create a soothing mood. People who have epilepsy should not use rosemary, hyssop, sage or sweet fennel, which seem to make the brain more alert.

Dietary changes that emphasize whole foods and eliminate processed foods may be helpful. Homeopathic therapy also can work for people with seizures, especially constitutional homeopathic treatment that acts at the deepest levels to address the needs of the individual person.

Prognosis

People who have epilepsy have a higher-than-average rate of **suicide**; sudden, unexplained death; and drowning and other accidental fatalities.

Benign focal epilepsy of childhood and some absence seizures may disappear in time, but remission is unlikely if seizures occur several times a day, several times in a 48-hour period, or more frequently than in the past.

Seizures that occur repeatedly over time and always involve the same symptoms are called stereotypic seizures. The probability that stereotypic seizures will abate is poor.

About 85% of all seizure disorders can be partially or completely controlled if the patient takes anti-seizure medication according to directions, avoids seizure-inducing sights, sounds, and other triggers, gets enough sleep, and eats regular, balanced meals.

Anyone who has epilepsy should wear a bracelet or necklace identifying the seizure disorder and listing the medication that he or she takes.

Prevention

Eating properly, getting enough sleep, and controlling stress and fevers can help prevent seizures. A person who has epilepsy should be careful not to hyperventilate. A person who experiences an aura should find a safe place to lie down and stay there until the seizure passes. Anticonvulsant medications should not be stopped suddenly and, if other medications are prescribed or discontinued, the doctor treating the seizures should be notified. In some conditions, such as severe head injury, brain surgery, or **subarachnoid hemorrhage**, anticonvulsant medications may be given to the patient to prevent seizures.

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- "New Drug Candidate Shows Promise." *Clinical Trials Week* April 7, 2003: 26.

ORGANIZATIONS

- American Epilepsy Society (AES), 342 North Main Street, West Hartford, CT, 06117-2507, (860) 586-7505, (860) 586-7550, <http://www.aesnet.org/>.
- Epilepsy Foundation of America, 8301 Professional Place, Landover, MD, 20785-7223, (301) 577-2684, (800) 332-1000, info@efa.org, <http://www.efa.org>.
- Epilepsy Information Service, Medical Center Boulevard, Winston-Salem, NC, 27157, (800) 642-0500, pgibson@wfubmc.edu, <http://www.wfubmc.edu/>
- Neurosciences/Comprehensive-Epilepsy-Center/Epilepsy-Information-Service.htm.
- International Bureau for Epilepsy, 100 Priory Hall, Stillorgan, Blackrock, Dublin, Ireland, 3531210-8850, 3531210-8450, ibedublin@eircom.net, <http://www.ib-epilepsy.org>.

Maureen Haggerty
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Selective abortion see **Abortion, selective**

Selective mutism see **Mutism**

Selective serotonin reuptake inhibitors

Definition

Selective serotonin reuptake inhibitors are medicines that relieve symptoms of depression.

Purpose

Selective serotonin reuptake inhibitors are used to treat serious, continuing depression that interferes with a person's ability to function. Like other **antidepressant drugs**, they help reduce the extreme sadness, hopelessness, and lack of interest in life that are typical in people with depression. Selective serotonin reuptake inhibitors also are used to treat **panic disorder**, obsessive compulsive disorder (OCD), and have shown promise for treating a variety of other conditions, such as **premenstrual syndrome**, **eating disorders**, **obesity**, **self-mutilation**, and **migraine headache**.

As of late 2003, SSRIs have been found to have other off-label applications, including treatment of **premature ejaculation** and **diabetic neuropathy**.

Description

Selective serotonin reuptake inhibitors, also known as SSRIs or serotonin boosters, are thought to work by correcting chemical imbalances in the brain. Normally,

chemicals called neurotransmitters carry signals from one nerve cell to another. These chemicals are constantly being released and taken back up at the ends of nerve cells. Selective serotonin reuptake inhibitors act on one particular neurotransmitter, serotonin, reducing its re-entry into nerve cells and thus allowing serotonin to build up. Although scientists are not exactly sure how it works, serotonin is involved in the control of moods, as well as other functions such as sleep, body temperature, and appetite for sweets and other carbohydrates. Somehow, drugs that prevent the uptake of serotonin improve the moods of people with serious depression, OCD, and some types of **anxiety disorders**.

Selective serotonin reuptake inhibitors are available only with a doctor's prescription and are sold in tablet, capsule, and liquid forms. Commonly used selective serotonin reuptake inhibitors are fluoxetine (Prozac), paroxetine (Paxil), sertraline (Zoloft), and fluvoxamine (Luvox).

Recommended dosage

The recommended dosage depends on the type of SSRI and the type and severity of depression for which it is being taken. Dosages may be different for different people. It is important for people taking SSRIs to take the drug exactly as prescribed. Taking larger or more frequent doses or taking the drug for longer than directed, for example, can cause unwanted effects.

SSRIs are about as effective as other antidepressants. About 60–80% of people taking the drugs as directed will find that their conditions improve. However, it may take four weeks or more for the effects of this medicine to be felt. Therefore, when people begin SSRI therapy, it is important to continue taking the medication, even if an improvement in mood doesn't begin immediately.

People who take SSRIs should ask their doctors about how to stop taking the medication. Usually, doctors advise patients to taper down gradually to reduce the chance of withdrawal symptoms or SSRI discontinuation syndrome.

SSRIs may be taken with food to prevent stomach upset.

Precautions

There have been reports that some patients taking SSRIs have an increase in thoughts about **suicide**. It is not clear whether the medicine causes this effect because suicidal thoughts are very often a part of depression itself. While some patients may experience

worsening of such thoughts early in the treatment of their depression, there is no credible evidence that SSRIs alone cause people to become suicidal or violent.

Serious and possibly life-threatening reactions may occur when SSRIs are used in combination with **monoamine oxidase inhibitors** (MAO inhibitors), such as Nardil and Parnate, which also are used to treat depression. These reactions also are possible when a person stops taking an SSRI and immediately begins taking an MAOI. SSRIs and MAO inhibitors should never be taken at the same time. When switching from an SSRI to an MAOI or vice versa, it may be necessary to allow two to five weeks or more between stopping one and starting the other. The physician prescribing the medications should tell the patient exactly how much time to allow before beginning the other medication.

People with a history of manic disorders should use any antidepressant, including an SSRI, with caution.

It is important to see a doctor regularly while taking SSRIs. The doctor will check to make sure the medicine is working as it should and will watch for unwanted side effects. The doctor may also need to adjust the dosage during this period.

Some people feel drowsy, dizzy, or lightheaded when using SSRIs. The drugs may also cause blurred vision in some people. Since SSRIs can sometimes cause drowsiness, driving or operating heavy machinery should be undertaken cautiously, particularly when the person first begins taking the medication.

These medicines make some people feel light-headed, dizzy, or faint when they get up after sitting or lying down, a condition known as **orthostatic hypotension**. People may try to lessen the problem by getting up gradually and holding onto something for support if possible. If the problem is severe or doesn't improve, the patient should discuss it with his or her doctor.

Because SSRIs work on the central nervous system, they may add to the effects of alcohol and other drugs that slow down the central nervous system, such as **antihistamines**, cold medicine, allergy medicine, sleep aids, medicine for seizures, tranquilizers, some **pain** relievers, and **muscle relaxants**. They may also add to the effects of anesthetics, including those used for dental procedures. Anyone taking SSRIs should check with his or her doctor before taking any of the drugs mentioned above.

SSRIs may occasionally cause **dry mouth**, although this side effect is much more common with an older class of antidepressants known as tricyclics. To temporarily relieve the discomfort, doctors sometimes suggest

chewing sugarless gum, sucking on sugarless candy or ice chips, or using saliva substitutes, which come in liquid and tablet forms and are available without a prescription. If the problem continues for more than two weeks, check with a doctor or dentist. Mouth dryness that continues over a long time may contribute to **tooth decay** and other dental problems.

Changes in sexual functioning are among the more common side effects with SSRIs. Depending on the particular SSRI prescribed, 8–15% of patients may report these side effects. The most common problem for men is delayed ejaculation. Women may be unable to have orgasms. A doctor should be contacted if any changes in sexual functioning occur.

Special conditions

People with certain medical conditions or who are taking certain other medicines can have problems if they take SSRIs. Before taking these drugs, a patient should let the doctor know about any of these conditions:

ALLERGIES. Anyone who has had unusual reactions to SSRIs in the past should let his or her doctor know before taking the drugs again. The doctor should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

PREGNANCY. In studies of laboratory animals, some SSRIs have caused **miscarriage** and other problems in pregnant females and their offspring. However, at least two studies in humans (by Pastuszak in 1993 and Kuhlin in 1998) have shown SSRIs to be safe during **pregnancy**, and newer studies done in 2003 have reported that SSRIs do not appear to increase the risks of **birth defects** in the offspring. Still, women who are pregnant or who may become pregnant should check with their doctors before using SSRIs.

BREASTFEEDING. SSRIs pass into breast milk and some may occasionally cause unwanted side effects in nursing babies whose mothers take the drugs. These effects include **vomiting**, watery stools, crying, and sleep problems. Women who are **breastfeeding** should talk to their doctors about the use of SSRIs. They may need to switch to a different medicine while breastfeeding. If SSRIs must be taken, it may be necessary to stop breastfeeding while being treated with these drugs. However, several studies in people (for example, Yoshida in 1998) have indicated that SSRIs in breast milk have no effect on infant development.

DIABETES. SSRIs may affect blood sugar levels. People with diabetes who notice changes in their blood or urine tests while taking this medicine should check with their doctors.

KEY TERMS

Anesthetic—Medicine that causes a loss of feeling, especially of pain. Some anesthetics also cause a loss of consciousness.

Anxiety—Worry or tension in response to real or imagined stress, danger, or dreaded situations. Physical reactions, such as fast pulse, sweating, trembling, fatigue, and weakness may accompany anxiety.

Central nervous system—The brain and spinal cord.

Depression—A mental condition in which people feel extremely sad and lose interest in life. People with depression may also have sleep problems and loss of appetite and may have trouble concentrating and carrying out everyday activities.

Metabolism—All the physical and chemical changes that occur in cells to allow growth and maintain body functions. These include processes that break down

substances to yield energy and processes that build up other substances necessary for life.

Obsessive-compulsive disorder—An anxiety disorder in which people cannot prevent themselves from dwelling on unwanted thoughts, acting on urges, or performing repetitious rituals, such as washing their hands or checking to make sure they turned off the lights.

Off-label application—The use of a prescription medication to treat conditions outside the indications approved by the Food and Drug Administration (FDA).

Premenstrual syndrome—(PMS) A set of symptoms that occur in some women 2–14 days before they begin menstruating each month. Symptoms include headache, fatigue, irritability, depression, abdominal bloating, and breast tenderness.

OTHER MEDICAL CONDITIONS. Before using SSRIs, people with any of these medical problems should make sure their doctors are aware of their conditions: diabetes, **kidney disease**, **liver disease**, seizure disorders, current or past drug **abuse** or dependence, or diseases or conditions that affect the metabolism or blood circulation.

Side effects

The most common side effects are **anxiety** and nervousness (reported by 5–13% of people taking various SSRIs), tremor (5–14%), trouble sleeping (2–8%), tiredness or weakness (4–15%), **nausea** (11–26%), **diarrhea** (11–26%), **constipation** (1–8%), loss of appetite (3–18%), weight loss (1–13%), dry mouth (10–22%), **headache** (1–5%), sweating (5–9%), trouble urinating (1–2%), and decreased sexual ability (8–15%). Many of these problems diminish or disappear as the body adjusts to the drug and do not require medical treatment unless they interfere with normal activities. Persistent problems, such as **sexual dysfunction**, should be discussed with the doctor.

More serious side effects are possible, but extremely rare. People taking SSRIs who notice unusual joint or muscle pain; breathing problems; chills or **fever**; excessive excitement, fast talking, or actions that are out of control; or mood swings should contact their doctors. People who develop skin **rashes** or **hives** after taking an SSRI should stop taking the medication and contact

their doctors as soon as possible. Other rare side effects may occur. Anyone who has unusual symptoms after taking an SSRI should get in touch with his or her doctor.

Side effects may continue for some time after treatment with this medicine ends. How long the effects continue depends on how long the drug was taken and how much of it was used. In most cases, doctors recommend that patients taper off SSRIs rather than abruptly stopping them, because of the risk of developing a condition known as SSRI discontinuation syndrome. This syndrome can mimic serious illness. People who experience agitation, confusion, or restlessness; **dizziness** or lightheadedness; vision problems; tremor; sleep problems; unusual tiredness or weakness; **nausea and vomiting** or diarrhea; headache; excessive sweating; runny nose; or muscle pain for more than a few days after stopping or tapering an SSRI should consult their doctors.

Interactions

SSRIs may interact with other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. Anyone who takes SSRIs should let the doctor know about all other medicines he or she is taking. Among the drugs that may interact with SSRIs are:

- such central nervous system (CNS) depressants as medicine for allergies, colds, hay fever, and asthma;

- sedatives; tranquilizers; prescription pain medicine; muscle relaxants; medicine for seizures; sleep aids; barbiturates; and anesthetics.
- blood thinners (anticoagulants)
- such monoamine oxidase inhibitors (MAOIs) as Nardil or Parnate, used to treat conditions including depression and Parkinson's disease
- the antiseizure drug phenytoin (Dilantin)
- the food supplement (and sleep aid) tryptophan, which has been withdrawn from the United States market, but may be found in some herbal preparations
- digitalis and other heart medicines
- St. John's wort (*Hypericum perforatum*). St. John's wort is a herb used in Europe and the United States to relieve mild-to-moderate symptoms of depression. Research indicates that it acts as an SSRI and not as an MAO inhibitor, as previously believed. People who are using St. John's wort to relieve depression should not take a prescription SSRI at the same time.

The list above does not include every drug that may interact with SSRIs. Patients should be sure to check with a doctor or pharmacist before combining SSRIs with any other prescription or nonprescription (over-the-counter) medicine, including herbal preparations.

Resources

BOOKS

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ORGANIZATIONS

- American Psychiatric Association (APA), 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209, (888) 357-7924, apa@psych.org, <http://www.psych.org>.
- National Center for Complementary and Alternative Medicine (NCCAM), P.O. Box 7923, Gaithersburg, MD, 20898, (866) 464-3616, (888) 644-6226, info@nccam.nih.gov, <http://nccam.nih.gov>.
- National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Bethesda, MD, 20892, (301) 443-4513, (301) 443-4279, (866) 615-6464, nimhinfo@nih.gov, <http://www.nimh.nih.gov>.
- United States Food and Drug Administration (FDA), 10903 New Hampshire Ave, Silver Spring, MD, 202993-0002, (888) 463-6332, <http://www.fda.gov>.

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Self-mutilation

Definition

Self-mutilation is a general term for a variety of forms of intentional self-harm without the wish to die. Cutting one's skin with razors or knives is the most common pattern of self-mutilation. Others include biting, hitting, or bruising oneself; picking or pulling at skin or hair; burning oneself with lighted cigarettes, or amputating parts of the body.

Description

Self-mutilation has become a major public health concern as its incidence appears to have risen since the early 1990s. One source estimates that 0.75% of the general American population practices self-mutilation. The incidence of self-mutilation is highest among teenage females, patients diagnosed with **borderline personality disorder**, and patients diagnosed with one of the **dissociative disorders**. Over half of self-mutilators were sexually abused as children, and many also suffer from **eating disorders**.

Self-mutilation should not be confused with current fads for **tattoos** and **body piercing**. In some cases, however, it may be difficult to distinguish between an interest in these fads and the first indications of a disorder.

The relationship of self-mutilation to **suicide** is still debated even though statistics show that nearly 50% of individuals who injure themselves also attempt suicide at some point in their lives. Many researchers think

that suicide attempts reflect feelings of rejection or hopelessness, while self-mutilation results from feelings of shame or a need to relieve tension.

Causes and symptoms

Several different theories have been proposed to explain self-mutilation:

- self-mutilation is an outlet for strong negative emotions, especially anger or shame, that the person is afraid to express in words or discuss with others.
- self-mutilation represents anger at someone else directed against the self.
- self-mutilation relieves unbearable tension or anxiety. Many self-mutilators do report feeling relief after an episode of self-cutting or other injury.
- self-mutilation is a technique for triggering the body's biochemical responses to pain. Stress and trauma release endorphins, which are the body's natural pain-killing substances
- self-mutilation is a way of stopping a dissociative episode. Dissociation is a process in which the mind splits off, or dissociates, certain memories and thoughts that are too painful to keep in conscious awareness. Some people report that they feel "numb" or "dead" when they dissociate, and self-injury allows them to feel "alive."
- self-mutilation is a symbolic acting-out of the larger culture's mistreatment of women. This theory is sometimes offered to explain why the great majority (about 75%) of self-mutilators are girls and women

The symptoms of self-mutilation typically include wearing long-sleeved or baggy clothing, even in hot weather; and an unusual need for privacy. Self-mutilators are often hesitant to change their clothes or undress around others. In most cases the person has also shown signs of depression.

Diagnosis

Self-mutilation is usually diagnosed by a psychiatrist or psychotherapist. A family practitioner or nurse who notices **scars**, **bruises**, or other physical evidence of self-injury may refer the person to a specialist for evaluation.

Treatment

Persons who mutilate themselves should seek treatment from a therapist with some specialized training and experience with this behavior. Most self-mutilators are treated as outpatients, although there are some inpatient programs, such as S.A.F.E., for adolescent females. A number of different treatment approaches are used with self-mutilators, including psychodynamic

KEY TERMS

Borderline personality disorder (BPD)—A pattern of behavior characterized by impulsive acts, intense but chaotic relationships with others, identity problems, and emotional instability.

Dissociation—The splitting off of certain mental processes from conscious awareness.

Dissociative disorders—A group of mental disorders in which dissociation is a prominent symptom. Patients with dissociative disorders have a high rate of self-mutilation.

Endorphins—Pain-killing substances produced in the human body and released by stress or trauma. Some researchers think that people who mutilate themselves are trying to trigger the release of endorphins.

psychotherapy, **group therapy**, journaling, and behavioral therapy.

Although there are no medications specifically for self-mutilation, antidepressants are often given, particularly if the patient meets the diagnostic criteria for a depressive disorder.

Alternative treatment

Mindfulness training, which is a form of **meditation**, has been used to teach self-mutilators to observe and identify their feelings in order to have some control over them.

Prognosis

The prognosis depends on the presence and severity of other emotional disorders, and a history of **sexual abuse** and/or suicide attempts. In general, teenagers without a history of abuse or other disorders have a good prognosis. Patients diagnosed with borderline personality disorder and/or a history of attempted suicide are considered to have the worst prognosis.

Prevention

Some society-wide factors that influence self-mutilation, such as the high rate of sexual abuse of children and media stereotypes of women, are difficult to change. In general, however, young people who have learned to express themselves in words or through art and other creative activities are less likely to deal with painful feelings by injuring their bodies.

Resources

BOOKS

McPhee, Stephen, and Maxine Papadakis. *Current Medical Diagnosis and Treatment*, 2010, 49th ed. New York: McGraw-Hill Medical, 2009.

ORGANIZATIONS

American Psychiatric Association (APA), 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209, (888) 357-7924, apa@psych.org, <http://www.psych.org>.
Focus Adolescent Services, P. O. Box 4514, Salisbury, MD, 21803, (410) 341-4216, <http://www.focusas.com>.
National Institute of Mental Health (NIMH).

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KEY TERMS

Infertility—The inability of a man and woman to conceive a child after 12 months of unprotected sexual intercourse.

Morphology—The size and shape of sperm.

Motility—The movement of sperm within the semen.

Drops of semen are placed on a microscope slide and examined under the microscope. Motility, or movement, of 100 sperm are observed and graded in categories, such as rapid progressive or immotile.

The structure of sperm (sperm morphology) is assessed by carefully examining sperm for abnormalities in the size and shape in the head, tail, and neck regions. WHO standards define normal as a specimen with less than 30% abnormal forms. An alternative classification system (Kruger's) measures the dimensions of sperm parts. Normal specimens are allowed 14% or less abnormalities.

Sperm are counted by placing semen in a special counting chamber. The sperm within the chamber are counted under a microscope. White blood cells are recorded; these may indicate a reproductive tract infection. Laboratories may test for other biochemicals such as fructose, zinc, and citric acid. These are believed to contribute to sperm health and fertility.

Results of semen analysis for infertility must be confirmed by a second analysis seven days to three months after the first. Sperm counts may vary from day to day.

Semen analysis to confirm success of vasectomy is concerned only with discovering if sperm are still present. Semen is collected six weeks after surgery. If sperm are seen, another specimen is collected 2 to 4 weeks later. The test is repeated until two consecutive specimens are free of sperm.

Description

The semen analysis test is usually done manually, though computerized test systems are available. Many laboratories base their procedures on standards published by the World Health Organization (WHO).

The volume of semen in the entire ejaculate is measured. The appearance, color, thickness, and pH is noted. A pH test looks at the range from a very acid solution to a very alkaline solution. Semen, like many other body fluids, has a standard pH range that would be considered optimal for fertilization of the egg to take place. The thick semen is then allowed to liquify; this usually takes 20-60 minutes.

Preparation

A man should collect an entire ejaculate, by masturbation, into a container provided by his physician. To examine the best quality sperm, the specimen must be collected after two to three days of sexual abstinence, but not more than five to seven days. The specimen must not come into contact with any spermicidal agents used by a female partner for birth control purposes. The man should not have alcohol before the test.

A semen specimen to investigate infertility must be brought to the testing laboratory within one hour of obtaining it. Timing is not as critical for the postvasectomy test but the semen must be kept at body temperature. The most satisfactory sample is one obtained in the lab rather than at home.

Normal results

WHO standards have established these normal values:

- volume less than or equal to 2.0 mL
- sperm count greater than or equal to 20 million per mL
- motility (movement of the sperm) value is greater than or equal to 50% with forward progression, or greater than or equal to 25% with rapid progression within 60 minutes of ejaculation
- morphology greater than or equal to 30% with normal forms
- white blood cell count less than 1 million per mL.

If infertility continues, despite normal semen analysis and female studies, further tests are done to evaluate sperm function.

Abnormal results

Abnormalities of semen volume and liquidity, and sperm number and morphology decrease fertility. These abnormalities may be inherited or caused by a hormone imbalance, medications, or a recent infection. Further tests may be done to determine the cause of abnormalities.

Resources

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Kamada, M., et al. "Semen Analysis and Antisperm Antibody." *Archives of Andrology* (March-April 1998): 117-128.

Nancy J. Nordenson

Senile tremor see **Tremors**



Senior women in a Zumba class. (AP Images.)

Demographics

The proportion of people age 65 years and older in the United States is on the rise and will continue to increase through 2050. As of August 14, 2008 there were 40.2 million Americans age 65 and older, 13% of the total U.S. population, according to the U.S. Census Bureau. This will grow to 88.5 million, or 20% of the total U.S. population by 2050, the Census Bureau estimates. Worldwide, there were nearly 500 million people age 65 and older as of July 1, 2007, according to the United Nations Statistics Division. This represented 7.5% of the world's estimated population of 6.6 billion.

Description

For a senior, the **aging** process and a person's lifestyle will affect health. People who maintain a healthy weight, **exercise** regularly, eat nutritionally, and do not smoke reduce the risk for many health conditions. This wellness allows people to live longer and to remain independent for more years. **Smoking**, **obesity** (excess weight), and lack of exercise shorten life and increase the risk for many health conditions. According to the Centers for Disease Control and Prevention, about 80% of people in the United States age 65 and older have at least one chronic (long-lasting) condition and 50% have two.

Diet and exercise

Proper diet and regular exercise form the foundation of senior health. A nutritional diet and physical activity can help prevent diseases such as **cancer**, **stroke**, heart disease, and diabetes. A healthy diet also can help manage diabetes, high blood pressure, and heart disease.

Seniors' health

Definition

Seniors' health refers to the physical and mental conditions of senior citizens, those who are in their 60s and older.

Leading causes of death in persons 65 and older

Cause of death	Number of deaths	Percentage of all deaths in age group (65+)
Heart disease	496,095	28.3%
Malignant neoplasms (Cancer)	389,730	22.2%
Cerebrovascular diseases	115,961	6.6%
Chronic lower respiratory diseases	109,562	6.2%
Alzheimer's disease	73,797	4.2%
Diabetes mellitus	51,528	2.9%
Influenza and pneumonia	45,941	2.6%
Nephritis	38,484	2.2%
Unintentional injury	38,292	2.2%
Septicemia	26,362	1.5%

SOURCE: Centers for Disease Control and Prevention, National Center for Injury Prevention and Control.

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

As people age, there is more of a need to exercise on a regular basis. According to the American Heart Association, the inactive person loses from 3–5% of muscle fiber each decade after age 30. That loss would total 30% of lost muscle fiber at age 60. Exercise helps to boost muscle strength. It can help improve balance and coordination, and help to prevent falls.

Organizations including the American Heart Association advise that regular physical activity helps prevent bone loss (**osteoporosis**) and the risk of conditions such as heart disease, Type II diabetes, **colon cancer**, **stress**, and depression. In addition, exercise can help extend the lives of people with conditions such as diabetes, high blood pressure, and high cholesterol. Good health later in life helps to prevent serious illness or **death** from common infections as well. If a senior catches the flu, for instance, it can have more detrimental effects than in a healthier, younger person. When the SARS outbreak occurred in 2002 and 2003, clinicians expressed concern about the elderly Americans and again expressed the importance of diet and exercise. As people age, their immune system response weakens. Seniors need to be proactive in keeping their systems strong.

Causes and symptoms

A number of health problems begin to occur as an individual ages. Early recognition of these conditions and proper treatment can improve a senior's health and longevity.

Osteoporosis

Osteoporosis is a condition in which bones become less dense (solid). Bones become brittle, thinner, and

break easily. Although osteoporosis is associated with aging, it is only the risk of osteoporosis that increases as a person ages. It is linked to approximately 70% of bone **fractures** in people age 46 and older. According to the National Institutes of Health (NIH), one out of two women over age 50 and one out of eight men over 50 will experience an osteoporosis-related fracture.

Osteoporosis is associated primarily with the changes that occur to women during **menopause**. During menopause, there is a decrease in the level of estrogen, the hormone that helps maintain bone mass. Other causes of osteoporosis include lack of exercise and a diet deficient in vitamin D.

Osteoporosis is largely preventable. However, there is increasing evidence to suggest that the condition starts as far back as in the womb. If this is true, it still is preventable, but by the behavior of the mother carrying a child. More research needs to be done, but it is clear that childhood growth rates are linked to hip fractures that happen decades later.

Osteoarthritis

Osteoarthritis is a joint disease in which cartilage wears out and bones rub against each other. This condition can occur gradually over time as activities performed throughout the years cause wear on joints. In addition, bones thin as a person ages.

Excess weight and injuries can aggravate this condition. About 16 million Americans experience some form of osteoarthritis. It generally affects the neck, fingers, lower back, knees, and toes. Symptoms include **pain**, stiffness, swelling, and creaking. The pain may disrupt sleep, and joint stiffness may make it difficult for a person to dress.

Falls

More than two million Americans each year fall and experience serious injuries, according to the American Academy of Otolaryngology-Head and Neck Surgery. For seniors, fall-related injuries can reduce mobility and hinder independence.

As people age, their reflexes slow down so it may be more difficult to prevent a fall. Deteriorating vision and hearing can affect balance, which can cause an accidental fall. Furthermore, conditions such as arthritis, **dizziness**, and sleeping disorders can increase the likelihood of a fall. In addition, a person may fall at the start of a condition such as a stroke or **heart attack**.

Falls can result in broken bones or fractures because bones are weakened by osteoporosis. In addition, healing takes longer. Head injuries could affect

sight and hearing. Injuries sustained during falls could reduce an active person's mobility and independence.

Vision

Eyesight changes as people age. Generally, people are in their 40s when they experience **presbyopia**, a form of farsightedness. This is a progressive condition involving a decrease in the eye's ability to focus on close objects (near vision). By age 65, little near focusing ability remains.

Glaucoma is a condition caused by pressure from the build-up of a large amount of fluid in the eye. This progressive condition is often seen in people in their fifties. It starts with the gradual loss of peripheral vision. If not treated, it can lead to some vision loss.

People in their sixties may experience the first signs of age-related **macular degeneration** (AMD). It is a progressive condition affecting the retina. The macula in the retina distinguishes detail. Degeneration in the macula could cause scarring and a gradual reduction in vision. The person experiences a circle of blindness, an area of sightlessness that grows as the condition progresses.

More than half of people age 65 or older will be diagnosed with **cataracts**. Cataract refers to the loss of the transparency in the lens of the eye. As the loss progresses, the person is able to see less detail. This condition generally affects both eyes.

Hearing

Presbycusis, age-related **hearing loss**, is a progressive condition. It usually starts with a difficulty in hearing high-frequency sound such as people talking. A senior has less trouble with low-frequency tones. Background noise makes it even more difficult to hear. Presbycusis affects approximately 25% of people between the ages of 65 and 75 and half of those over 75. Many people diagnosed with this condition say they have lost hearing in both ears. They also report feelings of dizziness and that they experience a ringing in their ears.

Sleep disorders

Sleep patterns change when a person ages. Many people in their sixties and seventies experience less time in the stages of deep sleep known as delta sleep. Despite this change, many healthy older people do not experience **sleep disorders**. Overall health plays a role in whether a senior experiences trouble sleeping.

Obesity is linked to **snoring** and **sleep apnea**. Snoring can turn into apnea. A person with apnea stops breathing for up to one minute until the brain restarts

the breathing process. This action could be repeated several hundred times each night.

Furthermore, a senior's sleep can be disrupted by conditions such as arthritis, osteoporosis, and **Alzheimer's disease**. **Insomnia**, or the inability to stay asleep, is a symptom of conditions including depression, **anxiety**, chronic pain, and **restless legs syndrome** (RLS).

RLS involves movement of legs when a person is at rest. The person moves legs in response to a **tingling** sensation in the upper leg, calf, or foot. In other cases, legs move involuntarily. Sensations that trigger movement can re-occur within seconds.

A person with RLS is likely to have PLMD (periodic limb movement disorder). A sleeping person with this condition kicks their legs or moves their arms repeatedly. These involuntary movements can last from 20 seconds to an hour. Approximately 45% of the elderly have a mild form of PLMD, according to the National Sleep Foundation.

The cause of these disorders is not known. They are thought to be caused by a chemical reaction in the brain. In addition, the conditions may be hereditary.

Mental health

While age has little effect on the mind, social and emotional factors affect an older person's health. After a lifetime of work or raising a family, retirement brings several challenges. A person who has been identified for years by a profession may experience a sense of lost identity.

A senior may find that the thinking process has changed. Learning something new may take longer. However, older people have excellent recall of new information.

Memory loss may be a concern, particularly since this is a symptom of Alzheimer's disease.

Dementia

Alzheimer's disease is a form of **dementia**, a condition in which mental abilities decline. Symptoms of dementia include memory loss that goes beyond forgetting a word or where an item was placed. The person with dementia may never recognize family members or remember how to perform functions such as preparing a meal. Sometimes they experience a change in personality, with some uncharacteristic aggression or **paranoia**.

Alzheimer's disease is the most prevalent form of dementia. Although the cause of this condition is not known, the risk of Alzheimer's increases as a person ages. In 2010, the condition affected one in eight people

over the age of 65. The ratio rises to one in three people age 85 and older.

Alzheimer's is a progressive condition. In most cases, after five to eight years, a patient with this condition is unable to perform basic functions. There is no known cure for Alzheimer's. However, as of 2008, the U.S. Food and Drug Administration (FDA) approved five medications that could help delay the degenerative process.

Precautions

A health condition may result in a doctor recommending against some forms of exercise. Even if a person cannot jog, other forms of exercise include those designed for people in wheelchairs and those who are bedridden.

Treatment

The cost of treatment varies. Cost of medical treatment is determined by the type of procedure and whether a person has medical insurance. Health plan and Medicare coverage and copayments impact an individual's cost for various preventions and treatments.

Osteoporosis

Prevention is the best method of treating osteoporosis. Methods of preventing osteoporosis include regular weight-bearing exercise such as walking, jogging, weight lifting, **yoga**, and stair climbing.

People should not smoke since smoking makes the body produce less estrogen. Care should be taken to avoid falling.

Diet should include 1,000–1,300 mg of **calcium** each day. Sources of calcium include:

- leafy, dark-green vegetables such as spinach, kale, mustard greens, and turnip greens
- low-fat dairy products such as milk, yogurt, and cheeses such as cheddar, Swiss, mozzarella, and parmesan; also foods made with milk such as pudding and soup
- canned fish such as salmon, sardine, and anchovies
- tortillas made from lime-processed corn
- tofu processed with calcium-sulfate
- calcium and vitamin D tablets

An x ray can indicate bone loss when much of the density has decreased. A more effective way of detecting osteoporosis is the DEXA-scan (dual-energy x-ray absorptiometry). This whole-body scan indicates whether a person is at risk for fractures. It could be useful for people at risk for osteoporosis as well as

women near the age of menopause or older. People should ask their doctors about whether this test is needed.

During menopause, a woman loses estrogen. A pill or skin patch containing estrogen and progesterone eases symptoms of menopause has been used to treat osteoporosis. This treatment is known as **hormone replacement therapy** (HRT). In 2002, the Women's Health Initiative found that HRT produced harmful effects in postmenopausal women, including increased incidence of **breast cancer**, heart disease, and dementia. The effects were bad enough to stop the study. Women and physicians are advised to closely weigh the risks and benefits of hormone therapy.

Several drugs are available to help reduce the risk of fractures in seniors with osteoporosis. In 2003, the FDA approved a new treatment option called Teriparatide. Some alternative treatments show promise in studies, including SAMe, (S-adenosylmethionine). Long-term safety and effectiveness of SAMe have yet to be established. Another treatment option for menopause and osteoporosis is Raloxifene, a medication that may cause **blood clots**.

Osteoarthritis

Treatments for osteoarthritis range from preventative measures such as walking to **joint replacement** surgery. Treatment costs vary from no cost for soaking a joint in cold water, the price of over-the-counter remedies, to fees for surgery.

If osteoarthritis is suspected, a doctor's diagnosis will include an assessment of whether joint pain is part of a patient's medical history. The doctor may take an x ray to determine the presence of cartilage loss and how much degeneration occurred.

Over-the-counter (OTC) remedies such as **aspirin** and ibuprofen and salves containing capsaicin can be helpful. A doctor may recommend anti-inflammatory medications.

In cases of severe osteoarthritis, joint replacement surgery or joint **immobilization** may be required. Joints are replaced with metal, plastic, or ceramic material.

The Arthritis Foundation recommends several remedies for easing pain. To treat inflammation, a person should use a cold treatment. Methods include soaking the affected area in cold water or applying an ice pack. To soothe aches and stimulate circulation, a person applies heat to the affected area for 20 minutes. This should be done three times a day.

Acupuncture may be helpful in treating mild osteoarthritis. Generally, a person should have one to two treatments a week for several weeks. Afterward, one treatment is recommended. An assessment of results should be made after 10 treatments.

Preventive and maintenance remedies include low-impact exercise such as swimming and walking, along with maintaining proper posture. Nutritional aids include foods rich in vitamin C such as citrus fruits and broccoli. Daily consumption of 400 IU of Vitamin E is recommended. Cutting back on fats, sugar, salt, cholesterol, and alcohol helps relieve the symptoms of osteoarthritis.

Fall prevention

Fall prevention starts with regular exercise such as walking. This improves balance and muscles. The walk route should be on level ground. Other methods for preventing falls include:

- moving slowly when rising from a chair or bed to avoid dizziness
- quitting smoking
- wearing shoes with low heels and rubber soles
- monitoring medications because of side effects that increase the probability of a fall
- checking vision and hearing periodically
- fall-proofing the home, including the installation of lighting, especially on stairways, clearing clutter and electrical cords that can cause falls, and installing handrails and strips in bathtubs and rails on stairs

After a fall, a senior may need **first aid** treatment for cuts or fractures. The doctor may evaluate whether medications cause balance problems. If indicated, the doctor may examine the patient's central nervous system function, balance, and muscle/joint function. A hearing or vision test may be ordered.

Corrective measures include adjusting prescriptions, vision surgery or having the patient use a cane or walker.

Vision

A person diagnosed with presbyopia may need bifocals or reading glasses to read print that appears too small. These lenses may need to be changed as vision changes over the years. Eventually, a person relies on glasses to focus on items that are near. Other seniors who never needed corrective lenses may need to wear eyeglasses. Publishers aware of this condition produce books with large print.

A senior should schedule periodic vision exams because early treatment helps prevent or lessen a risk of cataracts or glaucoma. Diet also plays a role in vision care. Dark green vegetables like broccoli are said to help prevent cataracts from progressing. Physical exercise is thought to reduce the pressure associated with glaucoma.

Glaucoma can be treated with eye drops. For cataracts, surgery can remove the affected lens and replace it with a permanent synthetic lens called an intraocular lens. Macular degeneration is the leading cause of vision loss and blindness in Americans age 65 and older, affecting 1.75 million Americans. There are two types of macular degeneration: wet and dry. There was no successful treatment for dry macular degeneration as of 2008 but there are three FDA-approved medications for treating wet macular degeneration.

Hearing

An audiologist can administer tests to determine the amount of hearing loss. Although there is no cure for presbycusis, **hearing aids** can help a senior affected by age-related hearing loss. If this treatment is not effective, the person might need to learn to read lips.

Sleep disorders

Losing weight can help with conditions such as snoring and sleep apnea. A doctor may advise the senior to quit smoking, reduce alcohol consumption, or to sleep on his or her side. In some cases, a doctor may refer the senior to a sleep disorder clinic. The senior may be prescribed a continuous positive airway pressure device. Known as a CPAP, the device is placed over the nose and administers a continuous flow of air.

PLMD and restless leg syndrome may be treated with the prescription drugs Dopar, Requip, and Mirapex. These disorders could be signs of kidney or circulation conditions. Treatment of those conditions should end these sleeping disorders.

Insomnia treatments include exercising and treating depression, stress, and other causes of sleeplessness.

Mental health

After retirement, a senior must find activities and interests to provide a sense of fulfillment. Otherwise, feelings of loneliness and isolation can lead to depression and susceptibility to poor health.

Activities that stimulate a person physically and intellectually contribute to good health. A senior can start an exercise program, take up hobbies, take classes, or volunteer. Senior centers offer numerous activities. Lunch programs provide nutritional meals and companionship. This is important because a senior living alone may not feel motivated to prepare healthy meals.

Dementia

Diagnosis of Alzheimer's disease starts with a thorough medical examination. The doctor should administer memory tests. Blood tests may be required, as well as a CT scan or MRI scan of the brain. If Alzheimer's is diagnosed, the doctor may prescribe medication to slow down progression of this form of dementia.

As of 2007, the FDA had approved five prescription medications for treatment of Alzheimer's Disease (AD). Tacrine, donepezil, rivastigmine, and galantamine are cholinesterase inhibitors that enhance memory. Modest improvement was reported in clinical trials on donepezil, rivastigmine, and galantamine. Tacrine's possible side effects include liver damage, so it is seldom prescribed. Namenda (memantine) is approved for moderate to severe AD. It is in a class of drugs called N-methyl D-aspartate (NMDA) antagonists.

Prognosis

Some recovery time may be needed after surgery. However, a healthy person will heal more quickly. A senior needs to maintain a schedule of regular exercise in order to remain mobile. Otherwise, a minor illness could make them dependent on others for daily care, according to the American Heart Association.

If mobility becomes limited due to a condition such as osteoarthritis, equipment like a walker and devices that make it easier to open bottles and grip cutlery can be helpful.

Exercising too long or too strenuously can be physically harmful. The over-exertion could cause the person to lose interest in exercise and put off establishing a regular routine. Experts recommend starting out slowly and building up to more intense or longer sessions. This is particularly important for a sedentary person.

Seniors who stay active and eat healthy are at less risk for conditions such as diabetes. A senior should seek mental stimulation and social interaction, which provide enjoyment, boost self-esteem,

and help reduce feelings of isolation and depression. Although eyesight and hearing will weaken, glasses and hearing aids help seniors keep the senses of sight and hearing.

When surgery is required for osteoarthritis, hip replacement surgery is extremely successful. In about 98% of surgeries, flexibility returns and pain is eased. Knee replacement surgery also is effective.

If a person maintains a healthy lifestyle, the ability to avoid falls and recover from them is increased.

After a fall, seniors need to build up physical strength and confidence so they do not fear falling again. Care should be taken so that seniors do not feel isolated by their injuries. Isolation could lead to decreased mobility and loss of independence.

There is no cure for Alzheimer's disease. However, several medications have proved moderately effective in stopping memory loss. Since Alzheimer's is progressive, a person diagnosed with this condition should make arrangements for the future. Finances should be taken care of and plans should be made for future care. Family should be brought into the discussion.

After diagnosis, a person should stay active for as long as possible. Not only does this help with enjoying this stage of life, activities can help to fight depression. Alzheimer and other support groups can be helpful. In addition, modifications to environment can be effective.

Prevention

Nutrition

Nutrition plays an important role in senior health. Not only does a well-balanced diet keep a person from becoming obese, that same diet is a safeguard against health conditions that seniors face. Proper diet can help prevent a condition like diabetes or keep it from worsening.

The senior diet should consist of foods that are low in fat, particularly saturated fat and cholesterol. A person should choose foods that provide nutrients such as iron and calcium. Other healthy menu choices include:

- fish, skinless poultry, and lean meat
- proteins such as dry beans (red beans, navy beans, and soybeans), lentils, chickpeas, and peanuts
- low-fat dairy products
- vegetables, especially those that are dark green and leafy

- citrus fruits or juices, melons, and berries
- whole grains like wheat, rice, oats, corn, and barley
- whole grain breads and cereals

Exercise

Physical activity should be rhythmic, repetitive, and should challenge the circulatory system. It also should be enjoyable so that a senior gets in the habit of exercising regularly for 30 minutes each day. It may be necessary to check with a doctor to determine the type of exercise that can be done.

Walking is recommended for weight loss, stress release, and many other conditions. Brisk walking is said to produce the same benefits as jogging. Other forms of exercise can include gardening, bicycling, hiking, swimming, dancing, skating, or ice-skating. If weather prohibits outdoor activities, a person can work out indoors with an exercise video.

Exercise also offers a chance to socialize. In some cities, groups of seniors meet for regular walks at shopping malls. Senior centers offer exercise classes ranging from line dancing to belly dancing.

Costs for exercise range from the price of walking shoes to the fees for joining a gym.

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American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Road, Alexandria, VA, 22314-2857, (703) 836-4444, <http://www.entnet.org>.

American Dietetic Association, 120 S. Riverside Plaza, Suite 2000, Chicago, IL, 60606-6995, (312) 899-0040, (800) 877-1600, <http://www.eatright.org>.

American Heart Association, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org, <http://www.americanheart.org>.

Division of Aging and Seniors, Health Canada, Public Health Agency of Canada 1908A1, Ottawa, Canada, Ontario, K1A 0K9, (613) 952-7606, (613) 957-9938, (800) 267-1245, seniors-aines@phac-aspc.gc.ca, <http://www.phac-aspc.gc.ca/seniors-aines>.

Foundation for Health in Aging, 350 Fifth Avenue, Suite 801, New York, NY, 10118, (212) 755-6810, <http://www.healthinaging.org>.

Meals on Wheels Association of America, 203 South Union, Alexandria, VA, 22314, (703) 548-55580, <http://www.mowaa.org>.

National Institute on Aging, Building 31, Room 5C27, 31 Center Dr., Bethesda, MD, 20892, (800) 222-2225, <http://www.nia.nih.gov>.

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Sensory hearing loss see **Hearing loss**

Sensory integration disorder

Definition

Sensory integration disorder or dysfunction (SID) is a neurological disorder that results from the brain's inability to integrate certain information received from the body's five basic sensory systems. These sensory systems are responsible for detecting sights, sounds, smell, tastes, temperatures, **pain**, and the position and movements of the body. The brain then forms a combined picture of this information in order for the body to make sense of its surroundings and react to them appropriately. The ongoing relationship between behavior and brain functioning is called sensory integration (SI), a theory that was first pioneered by A. Jean Ayres, Ph.D., OTR in the 1960s.

Description

Sensory experiences include touch, movement, body awareness, sight, sound, smell, taste, and the pull of gravity. Distinguishing between these is the process of sensory integration (SI). While the process of SI occurs automatically and without effort for most, for some the process is inefficient. Extensive effort and attention are required in these individuals for SI to occur, without a guarantee of it being accomplished. When this happens, goals are not easily completed, resulting in sensory integration disorder (SID).

The normal process of SI begins before birth and continues throughout life, with the majority of SI development occurring before the early teenage years. The ability for SI to become more refined and effective coincides with the **aging** process as it determines how well motor and speech skills, and emotional stability develop. The beginnings of the SI theory by Ayres instigated ongoing research that looks at the crucial foundation it provides for complex learning and behavior throughout life.

Causes and symptoms

The presence of a sensory integration disorder is typically detected in young children. While most children develop SI during the course of ordinary childhood activities, which helps establish such things as the ability for motor planning and adapting to incoming sensations, others' SI ability does not develop as efficiently. When their process is disordered, a variety of problems in learning, development, or behavior become obvious.

Those who have sensory integration dysfunction may be unable to respond to certain sensory information by planning and organizing what needs to be done in an appropriate and automatic manner. This may cause a primitive survival technique called "fright, flight, and fight," or withdrawal response, which originates from the "primitive" brain. This response often appears extreme and inappropriate for the particular situation.

The neurological disorganization resulting in SID occurs in three different ways: the brain does not receive messages due to a disconnection in the neuron cells; sensory messages are received inconsistently; or sensory messages are received consistently, but do not connect properly with other sensory messages. When the brain poorly processes sensory messages, inefficient motor, language, or emotional output is the result.

According to Sensory Integration International (SII), a non-profit corporation concerned with the impact of sensory integrative problems on people's lives, the following are some signs of sensory integration disorder (SID):

- oversensitivity to touch, movement, sights, or sounds
- underreactivity to touch, movement, sights, or sounds
- tendency to be easily distracted
- social and/or emotional problems
- activity level that is unusually high or unusually low
- physical clumsiness or apparent carelessness
- impulsive, lacking in self-control
- difficulty in making transitions from one situation to another
- inability to unwind or calm self
- poor self concept
- delays in speech, language, or motor skills
- delays in academic achievement

While research indicates that sensory integrative problems are found in up to 70% of children who are considered learning disabled by schools, the problems of sensory integration are not confined to children with learning disabilities. SID transfers through all age groups, as well as intellectual levels and socioeconomic groups. Factors that contribute to SID include: premature birth; **autism** and other developmental disorders; learning disabilities; delinquency and **substance abuse** due to learning disabilities; stress-related disorders; and brain injury. Two of the biggest contributing conditions are autism and attention-deficit hyperactivity disorder (**ADHD**).

Diagnosis

In order to determine the presence of SID, an evaluation may be conducted by a qualified occupational or physical therapist. An evaluation normally consists of both standardized testing and structured observations of responses to sensory stimulation, posture, balance, coordination, and eye movements. These test results and assessment data, along with information from other professionals and parents, are carefully analyzed by the therapist who then makes recommendations about appropriate treatment.

Treatment

Occupational therapists play a key role in the conventional treatment of SID. By providing sensory integration therapy, occupational therapists are able to supply the vital sensory input and experiences that children with SID need to grow and learn. Also referred to as a “sensory diet,” this type of therapy involves a planned and scheduled activity program implemented by an occupational therapist, with each “diet” being designed and developed to meet the needs of the child’s nervous system. A sensory diet stimulates the “near” senses (tactile, vestibular, and proprioceptive) with a combination of alerting, organizing, and calming techniques.

Motor skills training methods that normally consist of adaptive physical education, movement education, and gymnastics are often used by occupational and physical therapists. While these are important skills to work on, the sensory integrative approach is vital to treating SID.

The sensory integrative approach is guided by one important aspect—the child’s motivation in selection of the activities. By allowing them to be actively involved, and explore activities that provide sensory experiences most beneficial to them, children become more mature and efficient at organizing sensory information.

Alternative treatment

Sensory integration disorder (SID) is treatable with **occupational therapy**, but some alternative methods are emerging to complement the conventional methods used for SID.

Therapeutic body brushing is often used on children (not infants) who overreact to tactile stimulation. A specific non-scratching surgical brush is used to make firm, brisk movements over most of the body, especially the arms, legs, hands, back and soles of the feet. A technique of deep joint compression follows the brushing. Usually begun by an occupational therapist, the technique is taught to parents who need to complete

KEY TERMS

Axon—A process of a neuron that conducts impulses away from the cell body. Axons are usually long and straight.

Cortical—Regarding the cortex, or the outer layer of the brain, as distinguished from the inner portion.

Neurotransmission—When a neurotransmitter, or chemical agent released by a particular brain cell, travels across the synapse to act on the target cell to either inhibit or excite it.

Proprioceptive—Pertaining to proprioception, or the awareness of posture, movement, and changes in equilibrium and the knowledge of position, weight, and resistance of objects as they relate to the body.

Tactile—The perception of touch.

Vestibular—Pertaining to the vestibule; regarding the vestibular nerve of the ear which is linked to the ability to hear sounds.

the process for three to five minutes, six to eight times a day. The time needed for brushing is reduced as the child begins to respond more normally to touch. In order for this therapy to be effective, the correct brush and technique must be used.

A report in 1998 indicated the use of cerebral electrical stimulation (CES) as being helpful to children with conditions such as moderate to severe autistic spectrum disorders, learning disabilities, and sensory integration dysfunction. CES is a modification of Transcutaneous **Electrical Nerve Stimulation** (TENS) technology that has been used to treat adults with various pain problems, including arthritis and **carpal tunnel syndrome**. TENS therapy uses a low voltage signal applied to the body through the skin with the goal of replacing painful impressions with a massage-like sensation. A much lower signal is used for CES than that used for traditional TENS, and the electrodes are placed on the scalp or ears. Occupational therapists who have studied the use of CES suggest that CES for children with SID can result in improved brain activity. The device is worn by children at home for 10 minutes at a time, twice per day.

Music therapy helps promote active listening. Hypnosis and **biofeedback** are sometimes used, along with **psychotherapy**, to help those with SID, particularly older patients.

Prognosis

By providing treatment at an early age, sensory integration disorder may be managed successfully. The ultimate goal is for the individual to be better able to interact with his or her environment in a more successful and adaptive way.

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Sepsis

Definition

Sepsis refers to a bacterial infection in the bloodstream or body tissues. This very broad term covers the presence of many types of microscopic disease-causing organisms.

Demographics

In the general population, the incidence of sepsis is two people in 10,000. The number of deaths from sepsis each year has almost doubled in the United States since 1980 because more patients are developing the condition. There are three major factors responsible for this increase: a rise in the number of organ transplants and other surgical procedures that require suppressing the patient's immune system; the greater number of elderly people in the population; and the overuse of **antibiotics** to treat infectious illnesses, resulting in the development of drug-resistant bacteria.

Description

Sepsis is also called **bacteremia**. Closely related terms include septicemia and septic syndrome. In sepsis, there is active multiplication of bacteria in the bloodstream which may or may not result in organ dysfunction. If sepsis is not promptly recognized and treated, pulmonary, hepatic, and renal function may be impaired.

KEY TERMS

Bacteremia—The medical term for sepsis.

Prophylactic—Referring to medications or other treatments given to prevent disease.

Causes and symptoms

Sepsis can originate anywhere bacteria can gain entry to the body. Common sites include the genitourinary tract, the liver and its bile ducts, the gastrointestinal tract, and the lungs. Broken or ulcerated skin can also provide access to bacteria commonly present in the environment. Invasive medical procedures, including dental work, can introduce bacteria or permit them to accumulate in the body. Entry points and equipment left in place for any length of time present a particular risk. **Heart valve replacement**, catheters, **ostomy** sites, intravenous (IV) or arterial lines, surgical **wounds**, or surgical drains are examples. IV drug users are at high risk as well.

People with inefficient immune systems, such as those with HIV infection; spinal cord injuries; or blood disorders are at particular risk for sepsis and have a higher **death** rate (up to 60%). In people who have no underlying chronic disease, the death rate is far lower (about five percent). The growing problem of antibiotic resistance has increased the incidence of sepsis, partly because ordinary preventive measures (such as prophylactic antibiotics) are less effective.

Cancer patients are at an increased risk of developing sepsis because **chemotherapy** and other forms of treatment for cancer weaken their immune systems.

The most common symptom of sepsis is **fever**, often accompanied by chills or shaking, or other flu-like symptoms. A history of any recent invasive procedure or dental work should raise the suspicion of sepsis and medical help should be sought promptly.

Diagnosis

An accurate and detailed patient history is helpful in determining the source of the sepsis and in designing an appropriate course of treatment.

The presence of sepsis is indicated by blood tests showing particularly high or low white blood cell counts. The causative agent is determined by **blood culture**.

In some cases the doctor may order imaging studies to rule out **pneumonia**, or to determine whether

the sepsis has developed from a ruptured appendix or other leakage from the digestive tract into the abdomen.

Treatment

Identifying the specific causative agent ultimately determines how sepsis is treated. However, time is of the essence, so a broad-spectrum antibiotic or multiple antibiotics will be administered until blood cultures reveal the culprit and treatment can be designed specific to the organism. Intravenous antibiotic therapy is usually necessary and is administered in a hospital setting.

Prognosis

The prognosis associated with sepsis is dependent on several factors such as the general condition of the patient, including the patient's immune status, and early recognition and initiation of prompt, appropriate treatment of the cause of the sepsis.

In severe cases, the patient's chances of survival are enhanced by rapid admission to an intensive care unit followed by aggressive treatment with antibiotics and by careful monitoring of response to treatment.

Prevention

Prompt recognition and appropriate treatment of bacterial infections can often prevent the progression of bacteremia to sepsis.

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American College of Epidemiology (ACE), 1500 Sunday Dr., Suite 102, Raleigh, NC, 27607, (919) 861-5573, <http://www.acepidemiology.org>.

American Public Health Association (APHA), 800 I St. NW, Washington, DC, 20001-3710, (202) 777-APHA, <http://www.apha.org>.

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Sepsis syndrome see **Septic shock**
Septal deviation see **Deviated septum**
Septic arthritis see **Infectious arthritis**

Septic shock

Definition

Septic shock is a potentially lethal drop in blood pressure due to the presence of bacteria in the blood.

Demographics

The incidence of septic shock in the United States continues to increase. Approximately 200,000 cases of septic shock are documented annually in the United States with 100,000 deaths caused by the condition. In the last ten years, the mortality rate from septic shock has declined. Individuals most susceptible to septic shock are people over the age of 60 years.

Description

Septic shock is a possible consequence of **bacteremia**, or bacteria in the bloodstream. Bacterial toxins, and the immune system response to them, cause a dramatic drop in blood pressure, preventing the delivery of blood to the organs. Septic shock can lead to multiple organ failure including **respiratory failure**, and may cause rapid **death**. **Toxic shock syndrome** is one type of septic shock.

Causes and symptoms

During an infection, certain types of bacteria can produce and release complex molecules, called cytokines (previously referred to as endotoxins), that may provoke a dramatic response by the body's immune system. Released in the bloodstream, cytokines are particularly dangerous, because they become widely dispersed and affect the blood vessels themselves. The most critical

hemodynamic manifestation of septic shock is vasodilation of the arteries. Arteries and the smaller arterioles open wider, increasing the total volume of the circulatory system. At the same time, the walls of the blood vessels become leaky, allowing fluid to seep out into the tissues, lowering the amount of fluid left in circulation. This combination of increased system volume and decreased fluid causes a dramatic decrease in blood pressure and reduces the blood flow to the organs. Other changes brought on by immune response may cause coagulation of the blood in the extremities, which can further decrease circulation through the organs.

Septic shock is seen most often in patients with suppressed immune systems and is usually due to bacteria acquired during treatment at the hospital. The immune system is suppressed by drugs used to treat **cancer**, **auto-immune disorders**, organ transplants, and diseases of immune deficiency such as **AIDS**. **Malnutrition**, chronic **drug abuse**, and long-term illness increase the likelihood of succumbing to bacterial infection. Bacteremia is more likely with preexisting infections such as urinary or gastrointestinal tract infections, or skin ulcers. Bacteria may be introduced to the blood stream by surgical procedures, catheters, or intravenous equipment.

Toxic shock syndrome most often occurs in menstruating women using highly absorbent tampons. Left in place longer than other types, these tampons provide the breeding ground for *Staphylococcus* bacteria, which may then enter the bloodstream through small tears in the vaginal lining. The incidence of toxic shock syndrome has declined markedly since this type of tampon was withdrawn from the market.

Symptoms

Septic shock is usually preceded by bacteremia, which is marked by **fever**, malaise, chills, and **nausea**. The first sign of shock is often confusion and decreased consciousness. In this beginning stage, the extremities are usually warm. Later, they become cool, pale, and bluish. Fever may give way to lower than normal temperatures later on in **sepsis**.

Other symptoms include:

- rapid heartbeat
- shallow, rapid breathing
- decreased urination.
- reddish patches in the skin

Septic shock may progress to cause adult respiratory distress syndrome, in which fluid collects in the lungs, and breathing becomes very shallow and labored. This condition may lead to ventilatory collapse, in which

KEY TERMS

Bacteremia—Invasion of the bloodstream by bacteria.

the patient can no longer breathe adequately without assistance.

Diagnosis

Diagnosis of septic shock is made by measuring blood pressure, heart rate, and respiration rate, as well as by a consideration of possible sources of infection. Central venous pressure and cardiac output may be monitored with a catheter device inserted into the pulmonary artery supplying the lungs (Swan-Ganz catheter). Blood cultures are done to determine the type of bacteria responsible. The levels of oxygen, carbon dioxide, and acidity in the blood are also monitored to assess changes in respiratory function.

Treatment

Septic shock is considered a medical emergency and is treated initially with a combination of **antibiotics** and fluid replacement administered intravenously, often in large amounts. The antibiotic is chosen based on the bacteria present, although two or more types of antibiotics may be used initially until the specific organism is identified. Intravenous fluids, either blood or protein solutions, replace the fluid lost by leakage. Coagulation and hemorrhage may be treated with transfusions of plasma or platelets. Dopamine may be given to increase blood pressure further if necessary.

Respiratory distress is treated with mechanical ventilation and supplemental oxygen, either using a nosepiece or a tube into the trachea inserted through the throat.

Rapid identification and treatment of the primary infection site is critical to prevent ongoing proliferation of bacteria.

Prognosis

Septic shock is most likely to develop in the hospital, since it follows infections which are likely to be the objects of treatment. Because of this fact, careful monitoring and early, aggressive therapy can minimize the likelihood of progression. Nonetheless, the mortality rate from septic shock remains high and death occurs in at least 40–75% of all cases.

The likelihood of recovery from septic shock depends on many factors, including the degree of immunosuppression of the patient, underlying disease, promptness of treatment, and type of bacteria responsible. Mortality is highest in the very young and the elderly, those with persistent or recurrent infection, and those with compromised immune systems.

Prevention

The risk of developing septic shock can be minimized through treatment of underlying bacterial infections, and prompt attention to signs of bacteremia. In the hospital, scrupulous aseptic technique on the part of medical professionals lowers the risk of introducing bacteria into the bloodstream.

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Septoplasty

Definition

Septoplasty is a surgical procedure to correct the shape of the septum of the nose. The nasal septum is the

separation between the two nostrils. In adults, the septum is composed partly of cartilage and partly of bone.

Purpose

Septoplasty is performed to correct a crooked (deviated) or dislocated septum, often as part of **plastic surgery** of the nose (**rhinoplasty**). The nasal septum has three functions: to support the nose, regulate air flow, and support the mucous membranes (mucosa) of the nose. Septoplasty is done to correct the shape of the nose caused by a deformed septum or correct deregulated air flow caused by a **deviated septum**. Septoplasty is often needed when the patient is having an operation to reduce the size of the nose (reductive rhinoplasty), because this operation usually reduces the amount of breathing space in the nose.

Septoplasty may also be done as a follow-up procedure following facial trauma, as the nose is frequently broken or dislocated by blows to the face resulting from automobile accidents, criminal assaults, or **sports injuries**.

Precautions

Septoplasty is ordinarily not performed within six months of a traumatic injury to the nose.

Description

Septoplasties are performed in the hospital with a combination of local and intravenous anesthesia. In some cases, hypnosis has been successfully used as anesthesia. After the patient is anesthetized, the surgeon makes a cut (incision) in the mucous tissue that covers the part of the septum that is made of cartilage. The tissue is lifted, exposing the cartilage and bony part of the septum. Usually, one side of the mucous tissue is left intact to provide support during healing. Cartilage is cut away as needed.

As the surgeon cuts away the cartilage, deformities tend to straighten themselves out, reducing the amount of cartilage that must be cut. Once the cartilage is cut, bony deformities can be corrected. For most patients, this is the extent of the surgery required to improve breathing through the nose and correct deformities. Some patients have bony obstructions at the base of the nasal chamber and require further surgery. These obstructions include bony spurs and ridges that contribute to drying, ulceration, or bleeding of the mucous tissue that covers the inside of the nasal passages. In these cases, the extent of the surgery depends on the nature of the deformities that need correcting.

KEY TERMS

Cartilage—A tough, elastic connective tissue found in the joints, outer ear, nose, larynx, and other parts of the body.

Rhinoplasty—Plastic surgery of the nose.

Septum (plural, septa)—The dividing partition in the nose that separates the two nostrils. It is composed of bone and cartilage.

Splint—A thin piece of rigid material that is sometimes used during nasal surgery to hold certain structures in place until healing is underway.

During surgery, the patient's own cartilage that has been removed can be reused to provide support for the nose if needed. External septum supports are not usually needed. Splints may be needed occasionally to support cartilage when extensive cutting has been done. External splints can be used to support the cartilage for the first few days of healing. Tefla gauze is inserted in the nostril to support the flaps and cartilage and to absorb any bleeding or mucus.

A newer option for closing perforations in the septum is a button made of Silastic, a compound of silicone and rubber.

Preparation

Before performing a septoplasty, the surgeon will evaluate the difference in airflow between the two nostrils. In children, this assessment can be done very simply by asking the child to breathe out slowly on a small mirror held in front of the nose.

As with any other operation under **general anesthesia**, patients are evaluated for any physical conditions that might complicate surgery and for any medications that might affect blood clotting time.

Aftercare

Patients with septoplasties are usually sent home from the hospital later the same day or the morning after the surgery. All **dressings** inside the nose are removed before the patient leaves. Aftercare includes a list of detailed instructions for the patient that focus on preventing trauma to the nose.

Risks

The risks from a septoplasty are similar to those from other operations on the face: postoperative **pain**

with some bleeding, swelling, bruising, or discoloration. A few patients may have allergic reactions to the anesthetics. The operation in itself, however, is relatively low-risk in that it does not involve major blood vessels or vital organs. Infection is unlikely if proper surgical technique is observed.

Results

Normal results include improved breathing and airflow through the nostrils, and an acceptable outward shape of the nose.

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Septum perforation see **Perforated septum**

Serenoa repens see **Saw palmetto**

Serotonin boosters see **Selective serotonin reuptake inhibitors**

Serum albumin test see **Protein components test**

Serum globulin test see **Protein components test**

Serum hepatitis see **Hepatitis B**

Serum protein electrophoresis see **Protein electrophoresis**

Serum sickness

Definition

Serum sickness is a type of delayed allergic response, appearing four to 10 days after exposure to some **antibiotics** or antiserum, the portion of serum that contains antibodies, such as **gamma globulin**, which may be given to provide immunization against some diseases.

Description

Serum sickness is very similar to an allergic reaction. The patient's immune system recognizes the proteins in the drug or antiserum as foreign proteins, and produces its own antibodies to protect against the foreign proteins. The newly formed antibodies bind with the foreign protein to form immune complexes. These immune complexes may enter the walls of blood vessels where they set off an inflammatory reaction.

While other types of allergic reactions may produce a rapid response, the serum sickness reaction is delayed because it takes time for the body to produce antibodies to the new protein.

Causes and symptoms

The usual symptoms are severe skin reactions, often on the palms of the hands and soles of the feet. **Fever**, sometimes as high as 104° F, is always present and usually appears before the skin rash.

Joint pain may be reported in up to 50% of cases. This is usually seen in the larger joints, but occasionally the finger and toe joints may also be involved.

Swelling of lymph nodes, particularly around the site of the injection, is seen in 10–20% of cases. There may also be swelling of the head and neck.

Urine analysis may show traces of blood and protein in the urine.

Other symptoms may involve the heart and central nervous system. These may include changes in vision, and difficulty in movement. Breathing difficulty may occur.

Traditionally, antitoxins were the most common cause of serum sickness, but those reports date from a time when most antitoxins were made from horse serum. As many as 16% of the people who received antirabies serum derived from horses developed serum sickness. The risk of a reaction to antitoxins has dropped dramatically since manufacturers have started

using human serum instead of horse serum to make their products.

Although antitoxins are the most common cause of serum sickness, a number of drugs have been reported to cause a serum sickness reaction. The following list is not complete, but indicates some of the drugs that have been associated with this type of reaction:

- allopurinol (Zyloprim)
- barbiturates
- captopril (Capoten)
- cephalosporin antibiotics
- griseofulvin (Fulvicin, Grifulvin)
- penicillins
- phenytoin (Dilantin)
- procainamide (Procan SR, Procanbid, Pronestyl-SR)
- quinidine (Quinaglute, Quinidex, Quinora)
- streptokinase (Streptase, Kabikinase)
- sulfonamide antibacterial drugs

Of cases of serum sickness reported to the United States Food and Drug Administration, the drugs most commonly associated with the reaction have been the cephalosporin antibiotics, including cefaclor (Ceclor) and cefalexin (Keflex) and the sulfonamide combination trimethoprim-sulfamethoxazole (Bactrim, Septra.) This does not mean that these are high-risk drugs, since these drugs are very widely used, so that there are many people exposed to them.

In addition to these substances, allergenic extracts used for testing and immunization, hormones, and vaccines have been known to cause serum sickness.

Diagnosis

Diagnosis is made by observing the symptoms and reviewing the patient's medical and medication history. Although the symptoms of serum sickness may be similar to other conditions, patients who present with symptoms of serum sickness and who have a recent history of exposure to a drug or other product which may cause this type of reaction should be suspected of having serum sickness.

Treatment

The first step in treatment of serum sickness is always to discontinue the drug or other substance which is suspected of causing the reaction. After that, all treatment is symptomatic. **Antihistamines**, pain relievers, and **corticosteroids** may be given to relieve the symptoms. The choice of treatment depends on the severity of the reaction.

KEY TERMS

Allergy—Altered body reaction, usually hypersensitivity, as a response to exposure to a specific substance.

Antibody—Any of a large number of proteins that are produced after stimulation by an antigen and act specifically against the antigen in an immune response.

Antihistamine—A drug that inhibits the actions of histamine. Histamine causes dilatation of capillaries, contraction of smooth muscle, and stimulation of gastric acid secretion.

Antitoxin—An antibody that is capable of neutralizing the specific toxin (a specific cause of disease) that stimulated its production in the body and is produced in animals for medical purposes by injection of a toxin or toxoid with the resulting serum being used to counteract the toxin in other individuals.

Serum—The clear yellowish fluid that remains from blood plasma after fibrinogen, prothrombin, and other clotting factors have been removed by clot formation—called also blood serum.

Sulfonamide—A sulfa drug, one of a large group of drugs used to treat bacterial infections.

Prognosis

Most serum sickness reactions are mild, and disappear on their own after one or two weeks as long as the cause is removed. Sometimes, symptoms of pain and discomfort may continue for several weeks, even after all the observable reactions such as skin rash and protein in the urine have disappeared. In very rare cases, however, there can be severe reactions and permanent damage. In very rare but extreme cases, serum sickness can lead to **shock**, permanent kidney damage, and even **death**.

Prevention

The most effective method of prevention is simple avoidance of antitoxins that may cause serum sickness. If patients have had a reaction in the past, particularly if the reaction was to a commonly used drug, they should be made aware of it, and be advised to alert physicians and hospitals in the future. Patients who have had particularly severe reactions may be advised to wear identification bracelets, or use other means to alert health care providers.

When it is necessary to administer an antitoxin, skin tests may be used to identify people who are at risk of a reaction. If the situation does not allow enough time for skin testing, the antitoxin should be given along with an intravenous antihistamine. Other drugs, such as epinephrine, which may be needed for an emergency, should be available.

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Serum therapy see **Gammaglobulin**

Severe acute respiratory syndrome (SARS)

Definition

Severe acute respiratory syndrome (SARS) is the first emergent and highly transmissible viral disease to appear during the twenty-first century.

Description

Patients with SARS develop flu-like **fever**, **headache**, malaise, dry **cough** and other breathing difficulties. Many patients develop **pneumonia**, and in 5–10% of cases, the pneumonia and other complications are severe enough to cause **death**. SARS is caused by a virus that is transmitted usually from person to person—predominantly by the aerosolized droplets of virus infected material.

The first known case of SARS was traced to a November 2002 case in Guangdong province, China.

By mid-February 2003, Chinese health officials tracked more than 300 cases, including five deaths in Guangdong province from what was at the time described as an acute respiratory syndrome. Many flu-causing viruses have previously originated from Guangdong province because of cultural and exotic cuisine practices that bring animals, animal parts, and humans into close proximity. In such an environment, pathogens can more easily genetically mutate and make the leap from animal hosts to humans. The first cases of SARS showed high rates among Guangdong food handlers and chefs.

Chinese health officials initially remained silent about the outbreak, and no special precautions were taken to limit travel or prevent the spread of the disease. The world health community, therefore, had no chance to institute testing, **isolation**, and quarantine measures that might have prevented the subsequent global spread of the disease.

On February 21, Liu Jianlun, a 64-year-old Chinese physician from Zhongshan hospital (later determined to have been “super-spreader,” a person capable of infecting unusually high numbers of contacts) traveled to Hong Kong to attend a family wedding despite the fact that he had a fever. Epidemiologists subsequently determined that, Jianlun passed on the SARS virus to other guests at the Metropole Hotel where he stayed—including an American businessman en route to Hanoi, three women from Singapore, two Canadians, and a Hong Kong resident. Jianlun’s travel to Hong Kong and the subsequent travel of those he infected allowed SARS to spread from China to the infected travelers’ destinations.

Johnny Chen, the American businessman, grew ill in Hanoi, Vietnam, and was admitted to a local hospital. Chen infected 20 health care workers at the hospital including noted Italian epidemiologist Carlo Urbani who worked at the Hanoi World Health Organization (WHO) office. Urbani provided medical care for Chen and first formally identified SARS as a unique disease on February 28, 2003. By early March, 22 hospital workers in Hanoi were ill with SARS.

Unaware of the problems in China, Urbani’s report drew increased attention among epidemiologists when coupled with news reports in mid-March that Hong Kong health officials had also discovered an outbreak of an acute respiratory syndrome among health care workers. Unsuspecting hospital workers admitted the Hong Kong man infected by Jianlun to a general ward at the Prince of Wales Hospital because it was assumed he had a typical severe pneumonia—a fairly routine admission. The first notice that clinicians

were dealing with an unusual illness came—not from health notices from China of increasing illnesses and deaths due to SARS—but from the observation that hospital staff, along with those subsequently determined to have been in close proximity to the infected persons, began to show signs of illness. Eventually, 138 people, including 34 nurses, 20 doctors, 16 medical students, and 15 other health care workers, contracted pneumonia.

One of the most intriguing aspects of the early Hong Kong cases was a cluster of more than 250 SARS cases that occurred in a cluster of high-rise apartment buildings—many housing health care workers—that provided evidence of a high rate of secondary transmission. Epidemiologists conducted extensive investigations to rule out the hypothesis that the illnesses were related to some form of local contamination (e.g., sewage, bacteria on the ventilation system, etc.). Rumors began that the illness was due to cockroaches or rodents, but no scientific evidence supported the hypothesis that the disease pathogen was carried by insects or animals.

Hong Kong authorities then decided that those suffering the flu-like symptoms would be given the option of self-isolation, with family members allowed to remain confined at home or in special camps. Compliance checks were conducted by police.

One of the Canadians infected in Hong Kong, Kwan Sui-Chu, return to Toronto, Ontario, and died in a Toronto hospital on March 5. As in Hong Kong, because there were no alert from China about the SARS outbreak, Canadian officials did not initially suspect that Sui-Chu had been infected with a highly contagious virus, until Sui-Chu’s son and five health care workers showed similar symptoms. By mid-April, Canada reported more than 130 SARS cases and 15 fatalities.

Increasingly faced with reports that provided evidence of global dissemination, on March 15, 2003, the World Health Organization (WHO) took the unusual step of issuing a travel warning that described SARS as a “worldwide health threat.” WHO officials announced that SARS cases, and potential cases, had been tracked from China to Singapore, Thailand, Vietnam, Indonesia, Philippines, and Canada. Although the exact cause of the “acute respiratory syndrome” had not, at that time, been determined, WHO officials issuance of the precautionary warning to travelers bound for Southeast Asia about the potential SARS risk served as notice to public health officials about the potential dangers of SARS.

Within days of the first WHO warning, SARS cases were reported in United Kingdom, Spain, Slovenia, Germany, and in the United States.

WHO officials were initially encouraged that isolation procedures and alerts were working to stem the spread of SARS, as some countries reporting small numbers of cases experienced no further dissemination to hospital staff or others in contact with SARS victims. However, in some countries, including Canada, where SARS cases occurred before WHO alerts, SARS continued to spread beyond the bounds of isolated patients.

WHO officials responded by recommending increased screening and quarantine measures that included mandatory screening of persons returning from visits to the most severely affected areas in China, Southeast Asia, and Hong Kong.

In early April 2003, WHO took the controversial additional step of recommending against non-essential travel to Hong Kong and the Guangdong province of China. The recommendation, sought by **infectious disease** specialists, was not controversial within the medical community, but caused immediate concern regarding the potentially widespread economic impacts.

Mounting reports of SARS showed a increasing global dissemination of the virus. By April 9, the first confirmed reports of SARS cases in Africa reached WHO headquarters, and eight days later, a confirmed case was discovered in India.

Causes and symptoms

In mid-April 2003, Canadian scientists at the British Columbia **Cancer** Agency in Vancouver announced that they sequenced the genome of the coronavirus most likely to be the cause of SARS. Within days, scientists at the Centers for Disease Control (CDC) in Atlanta, Georgia, offered a genomic map that confirmed more than 99% of the Canadian findings.

Both genetic maps were generated from studies of viruses isolated from SARS cases. The particular coronavirus mapped had a genomic sequence of 29,727 nucleotides—average for the family of coronavirus that typically contain between 29,000–31,000 nucleotides.

Proof that the coronavirus mapped was the specific virus responsible for SARS would eventually come from animal testing. Rhesus monkeys were exposed to the virus via injection and inhalation and then monitored to determine whether SARS like symptoms developed, and then if sick animals exhibited a histological pathology (i.e., an examination of the tissue and cellular level pathology) similar to findings in human patients. Other tests, including polymerase chain reaction (PCR) testing helped positively match the specific coronavirus present in the lung tissue, blood, and feces of infected animals to the exposure virus.

Identification of a specific pathogen can be a complex process, and positive identification requires thousands of tests. All testing is conducted with regard to testing Koch's postulates—the four conditions that must be met for an organism to be determined to be the cause of a disease. First, the organism must be present in every case of the disease. Second, the organism must be able to be isolated from the host and grown in laboratory conditions. Third, the disease must be reproduced when the isolated organism is introduced into another, healthy host. The fourth postulate stipulates that the same organism must be able to be recovered and purified from the host that was experimentally infected.

Early data indicates that SARS has an incubation period range of two to 10 days, with an average incubation of about four days. Much of the inoculation period allows the virus to be both transported and spread by an asymptomatic carrier. With air travel, asymptomatic carriers can travel to anywhere in the world. The initial symptoms are non-specific and common to the flu. Infected cases then typically spike a high fever 100.4°F (38°C) as they develop a cough, **shortness of breath**, and difficulty breathing. SARS often fulminates (reaches its maximum progression) in a severe pneumonia that can cause **respiratory failure** and death in about 10% of its victims.

Diagnosis

Currently, initial tests include blood cultures, Gram stain, chest radiograph, and tests for other viral respiratory pathogens such as **influenza A** and **B**. Other serologic techniques are used, and if SARS is suspected, samples are forwarded to state/local public health departments and/or the CDC for coronavirus antibody testing.

Treatment

As of May 1, 2003, no therapy was demonstrated to have clinical effectiveness against the virus that causes SARS, and physicians could offer only supportive therapy (e.g. administration of fluids, oxygen, ventilation, etc.).

Prognosis

By late April/early May 2003, WHO officials had confirmed reports of more than 3,000 cases of SARS from 18 different countries with 111 deaths attributed to the disease (about a 5–10% death rate). United States health officials reported 193 cases with no deaths. Significantly, all but 20 of the U.S. cases were linked to travel to infected areas, and the other 20 cases were

accounted for by secondary transmission from infected patients to family members and health care workers.

Information on countries reporting SARS and the cumulative total of cases and deaths is updated each day on the WHO SARS web site at <http://www.who.int/csr/sarscountry/en/>.

Prevention

Until a vaccine is developed, isolation and quarantine remain potent tools in the modern public health arsenal. Both procedures seek to control exposure to infected individuals or materials. Isolation procedures are used with patients with a confirmed illness. Quarantine rules and procedures apply to individuals who are not currently ill, but are known to have been exposed to the illness (e.g., been in the company of an infected person or come in contact with infected materials).

Isolation and quarantine both act to restrict movement and to slow or stop the spread of disease within a community. Depending on the illness, patients placed in isolation may be cared for in hospitals, specialized health care facilities, or in less severe cases, at home. Isolation is a standard procedure for TB patients. In most cases, isolation is voluntary; however, isolation can be compelled by federal, state, and some local law.

States governments within the United States have a general authority to set and enforce quarantine conditions. At the federal level, the Centers for Disease Control and Prevention's (CDC) Division of Global Migration and Quarantine is empowered to detain, examine, or conditionally release (release with restrictions on movement or with a required treatment protocol) individuals suspected of carrying certain listed communicable diseases.

As of April 27, 2003, the CDC in Atlanta recommended SARS patients be voluntarily isolated, but had not recommended enforced isolation or quarantine. Regardless, CDC and other public health officials, including the Surgeon General, sought and secured increased powers to deal with SARS. On April 4, 2003, U.S. President George W. Bush signed Presidential Executive Order 13295 that added SARS to a list of quarantinable communicable diseases. The order provided health officials with the broader powers to seek "...apprehension, detention, or conditional release of individuals to prevent the introduction, transmission, or spread of suspected communicable diseases..."

Travel advisories issued by WHO should be reviewed and people who must travel to areas with SARS outbreaks should follow such preventative measures as frequent hand washing and avoidance of large crowds. Likewise, family members caring for suspected

and/or confirmed SARS patients should wash hands frequently, avoid direct contact with the patient's bodily fluids, and monitor their own possible development of symptoms closely.

Brenda Wilmoth Lerner

Severe combined immunodeficiency

Definition

Severe combined immunodeficiency (SCID) is the most serious human immunodeficiency disorder(s). It is a group of congenital disorders in which both the humoral part of the patient's immune system and the cells involved in immune responses fail to work properly. Children with SCID are vulnerable to recurrent severe infections, retarded growth, and early death.

Description

SCID is thought to affect between one in every 100,000 persons, and one in every 500,000 infants. Several different immune system disorders are currently grouped under SCID:

- Swiss-type agammaglobulinemia. This was the first type of SCID discovered, in Switzerland in the 1950s.
- Adenosine deaminase deficiency (ADA). About 50% of SCID cases are of this type. ADA deficiency leads to low levels of B and T cells in the child's immune system.
- Autosomal recessive. About 40% of SCID cases are inherited from the parents in an autosomal recessive pattern.
- Bare lymphocyte syndrome. In this form of SCID, the white blood cells (lymphocytes) in the baby's blood are missing certain proteins. Without these proteins, the lymphocytes cannot activate the T cells in the immune system.
- SCID with leukopenia. Children with this form of SCID are lacking a type of white blood cell called a granulocyte.

In order to understand why SCID is considered the most severe immunodeficiency disorder, it is helpful to have an outline of the parts of the human immune system. It has three parts: cellular, humoral, and nonspecific. The cellular and humoral parts of the system are both needed to fight infections—they recognize disease agents and attack them. The cellular system is composed

KEY TERMS

Adenosine deaminase (ADA)—An enzyme that is lacking in a specific type of SCID. Children with an ADA deficiency have low levels of both B and T cells.

Antigens—A substance that usually causes the formation of an antibody. A foreign invaders in the body.

Autosomal recessive inheritance—A pattern of inheritance of a recessive gene where, among other things, both parents may not show symptoms.

B cell—A type of lymphocyte or white blood cell that is derived from precursor cells in the bone marrow.

Congenital—Present at the time of birth. Most forms of SCID are hereditary as well as congenital.

Gene therapy—An experimental treatment for SCID that consists of implanting a gene for ADA into an activated virus and merging it with some of the patient's own T cells. The corrected T cells are infused back into the patient every few months.

Humoral—Pertaining to or derived from a body fluid. The humoral part of the immune system includes antibodies and immunoglobulins in blood serum.

Lymphocyte—A type of white blood cell that is important in the formation of antibodies.

Orphan drug—A drug that is known to be useful in treatment but lacks sufficient funding for further research and development.

PEG-ADA—An orphan drug that is useful in treating SCID related to ADA deficiency.

T cells—Lymphocytes that originate in the thymus gland. T cells regulate the immune system's response to infections. The thymus gland is small or underdeveloped in children with SCID.

Thrush—A disease of the mouth caused by a yeast, *Candida albicans*.

of many classes of T-lymphocytes (white blood cells that detect foreign invaders called antigens). The humoral system is made up of B cells, which are the only cells in the body that make antibodies. In SCID, neither the cellular nor the humoral part of the immune system is working properly.

Causes and symptoms

SCID is an inherited disorder. There are two ways in which a developing fetus' immune system can fail to develop normally. In the first type of genetic problem, both B and T cells are defective. In the second type, only the T cells are abnormal, but their defect affects the functioning of the B cells.

For the first few months of life, a child with SCID is protected by antibodies in the mother's blood. As early as three months of age, however, the SCID child begins to suffer from mouth infections (thrush), chronic **diarrhea**, **otitis media** and pulmonary infections, including **pneumocystis pneumonia**. The child loses weight, becomes very weak, and eventually dies from an opportunistic infection.

Diagnosis

SCID is diagnosed by the typing of T and B cells in the child's blood. B cells can be detected by immunofluorescence tests for surface markers (unique proteins) on the

cells. T cells can be identified in tissue sections (samples) using enzyme-labeled antibodies.

Treatment

Patients with SCID can be treated with **antibiotics** and immune serum to protect them from infections, but these treatments cannot cure the disorder. Bone marrow transplants are currently regarded as one of the few effective standard treatments for SCID.

Investigational treatments

In 1990, the Food and Drug Administration (FDA) approved PEG-ADA, an orphan drug (not available in the United States but available elsewhere), for the treatment of SCID. PEG-ADA, which is also called pegademase bovine, works by replacing the ADA deficiency in children with this form of SCID. Children who receive weekly injections of PEG-ADA appear to have normal immune functions restored. Another treatment that is still in the experimental stage is **gene therapy**. In gene therapy, the children receive periodic infusions of their own T cells corrected with a gene for ADA that has been implanted in an activated virus.

Prognosis

Currently, there is no cure for SCID. Most untreated patients die before age two.

Prevention

Genetic counseling is recommended for parents of a child with SCID.

ORGANIZATIONS

Immune Deficiency Foundation, 40 West Chesapeake Avenue, Suite 308, Towson, MD, 21204, (800) 296-4433, <http://www.primaryimmune.org/>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Rebecca J. Frey, PhD

Sex hormones tests

Definition

Sex hormones tests measure levels of the sex hormones, including estrogen, progesterone, and testosterone.

Purpose

The sex hormone tests are ordered to determine if secretion of these hormones is normal. Estrogen fraction test is done to evaluate sexual maturity, menstrual problems, and fertility problems in females. This test may also be used to test for tumors that excrete estrogen. In pregnant women it aids in determining fetal-placental health. Estrogen fraction is also used to evaluate males who have enlargement of one or both breasts (**gynecomastia**), or who have feminization syndromes, where they display female sex characteristics.

Progesterone assay test is ordered to evaluate women who are having difficulty becoming pregnant or maintaining a **pregnancy**, and to monitor high-risk pregnancies.

Testosterone levels are ordered to evaluate:

- ambiguous sex characteristics
- precocious puberty
- virilizing syndromes in the female
- infertility in the male
- rare tumors of the ovary and testicle

Description

The sex hormones control the development of primary and secondary sexual characteristics. They regulate the sex-related functions of the body, such as the menstrual cycle or the production of eggs or sperm. There are three main types of sex hormones:

- the female sex hormones (called the estrogen hormones)
- the progesterone hormones (which help the body prepare for and maintain pregnancy)
- the male sex hormones, or the androgen hormones

Female sex hormones are responsible for normal menstruation and the development of secondary female characteristics. Testosterone is a hormone that induces **puberty** in the male and maintains male secondary sex characteristics. In females, the adrenal glands and the ovaries secrete small amounts of testosterone.

Estrogen

Estrogen is tested to evaluate menstrual status, sexual maturity, and gynecomastia (or feminization syndromes). It is a tumor marker for patients with certain ovarian tumors. E1, a type of estrogen, is the most active estrogen in the nonpregnant female.

E3 (estriol) is the major estrogen in the pregnant female. It is produced in the placenta. Excretion of estriol increases around the eighth week of gestation and continues to rise until shortly before delivery. Serial urine and blood studies of this hormone are used to assess placental function and fetal normality in high-risk pregnancies. Falling values during pregnancy suggest fetal-placental deterioration and require prompt reassessment of the pregnancy, including the possibility of early delivery.

Progesterone

Progesterone is essential for the healthy functioning of the female reproductive system. Produced in the ovaries during the second half of the menstrual cycle, and by the placenta during pregnancy, small amounts of progesterone are also produced in the adrenal glands and testes.

After ovulation, an increase of progesterone causes the uterine lining to thicken in preparation for the implantation of a fertilized egg. If this event does not take place, progesterone and estrogen levels fall, resulting in shedding of the uterine lining.

Progesterone is essential during pregnancy, not only ensuring normal functioning of the placenta, but passing into the developing baby's circulation, where it is converted in the adrenal glands to corticosteroid hormones.

Testosterone

Testosterone is the most important of the male sex hormones. It is responsible for stimulating bone and muscle growth, and sexual development. It is produced

by the testes and in very small amounts by the ovaries. Most testosterone tests measure total testosterone.

Testosterone stimulates sperm production (spermatogenesis), and influences the development of male secondary sex characteristics. Overproduction of testosterone caused by testicular, adrenal, or **pituitary tumors** in the young male may result in **precocious puberty**.

Overproduction of testosterone in females, caused by ovarian and adrenal tumors, can result in masculinization, the symptoms of which include cessation of the menstrual cycle (**amenorrhea**) and excessive growth of body hair (**hirsutism**).

When reduced levels of testosterone in the male indicate underactivity of the testes (**hypogonadism**), testosterone stimulation tests may be ordered.

Preparation

The progesterone and testosterone tests require a blood sample; it is not necessary for the patient to restrict food or fluids before the test. Testosterone specimens should be drawn in the morning, as testosterone levels are highest in the early morning hours. The estrogen fraction test can be performed on blood and/or urine. It is not necessary for the patient to restrict food or fluids for either test. If a 24-hour urine test has been requested, the patient should call the laboratory for instructions.

Risks

Risks for these blood tests are minimal, but may include slight bleeding from the puncture site, **fainting** or feeling lightheaded after having blood drawn, or blood accumulating under the puncture site (hematoma).

Normal results

Estrogen levels vary in women, ranging from 24–149 picograms per mL of blood. In men, the normal range is between 12–34 picograms per mL of blood.

Progesterone levels vary from less than 150 nanograms per deciliter (ng/dL) of blood to 2,000 nanograms in menstruating women. During pregnancy, progesterone levels range from 1,500–20,000 ng/dL of blood.

Testosterone values vary from laboratory to laboratory, but can generally be found within the following levels:

- Men: 300–1,200 ng/dL
- Women: 30–95 ng/dL
- Prepubertal children: Less than 100 ng/dL (boys) less than 40 ng/dL (girls)

Abnormal results

Increased levels of estrogen are seen in feminization syndromes:

- when a male begins to develop female secondary sex characteristics
- during precocious puberty
- when children develop secondary sexual characteristics at an abnormally early age
- because of ovarian, testicular, or adrenal tumor
- During normal pregnancy, cirrhosis, and increased thyroid levels (hyperthyroidism)

Decreased levels of estrogen are found in the following conditions:

- a failing pregnancy
- during menopause
- anorexia nervosa
- primary and secondary hypogonadism
- turner's syndrome, seen in females with one missing X chromosome

Increased levels of progesterone are seen:

- during ovulation and pregnancy
- with certain types of ovarian cysts
- with a tumor of the ovary known as a choriocarcinoma

Decreased levels of progesterone are seen:

- in toxemia of pregnancy
- with a threatened abortion
- during placental failure
- after fetal death
- with amenorrhea
- due to ovarian dysfunction

Increased levels (male) of testosterone are found in:

- sexual precocity
- the viral infection of encephalitis
- tumors involving the adrenal glands
- testicular tumors
- excessive thyroid production (hyperthyroidism)
- testosterone resistance syndromes

Decreased levels (male) of testosterone are seen in:

- Klinefelter syndrome
- a chromosomal deficiency
- primary and secondary hypogonadism
- down syndrome
- surgical removal of the testicles
- cirrhosis

Increased levels (females) of testosterone are found in ovarian and adrenal tumors and in the presence of excessive hair growth of unknown cause (hirsutism).

Resources

BOOKS

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Janis O. Flores

- Newborns with intersex deformities must be assigned to one sex or the other. These deformities represent intermediate stages between the primordial female genitals and the change into male genitals caused by male hormone stimulation.
- Both men and women occasionally believe they are physically a different sex than they are mentally and emotionally. This dissonance is so profound that they are willing to be surgically altered.

In both cases, technical considerations favor successful conversion to a female rather than a male. Newborns with ambiguous organs will almost always be assigned to the female gender unless the penis is at least an inch (2.5 cm) long. Whatever their chromosomes, they are much more likely to be socially well-adjusted as females, even if they cannot have children.

Sex reassignment surgery

Definition

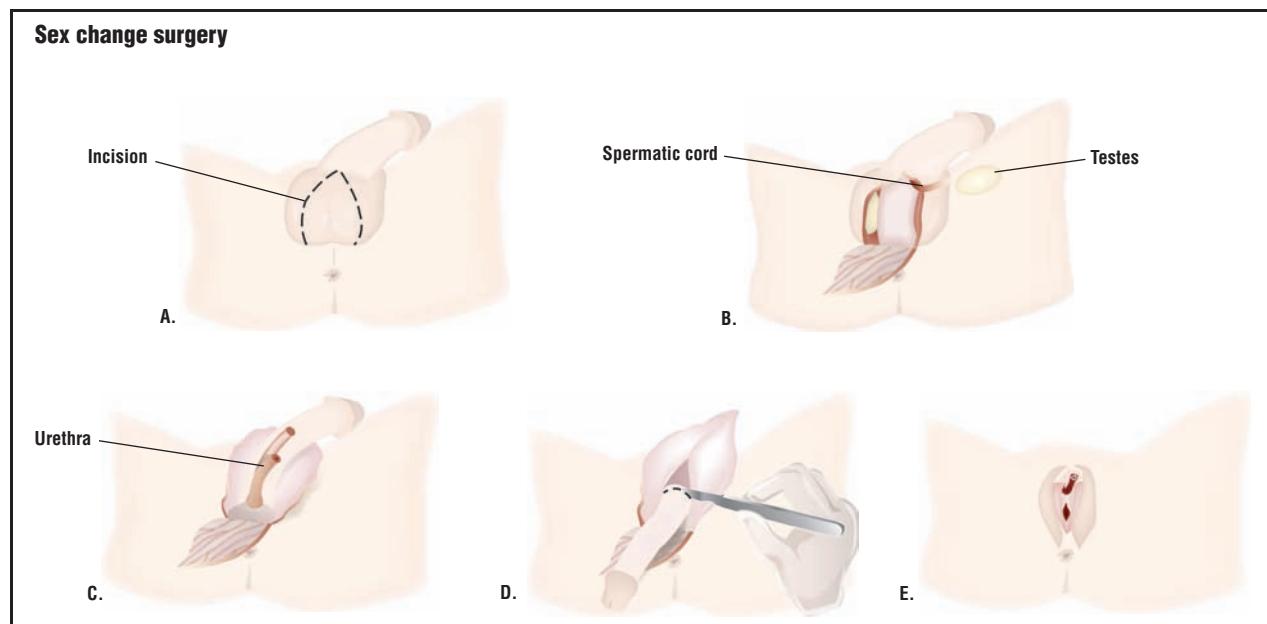
Also known as sex change or **gender reassignment surgery**, sex reassignment surgery is a procedure that changes genital organs from one gender to another.

Purpose

There are two main reasons to alter the genital organs from one sex to another.

Demographics

Reliable statistics are extremely difficult to obtain. Many sexual reassignment procedures are conducted in private facilities that are not subject to reporting requirements. Sexual reassignment surgery is often conducted outside of the United States. The number of gender reassignment procedures conducted in the United States each year is estimated at between 100 and 500. The number worldwide is estimated to be two to five times larger.



To change male genitalia to female genitalia, an incision is made into the scrotum (A). The flap of skin is pulled back, and the testes are removed (B). The skin is stripped from the penis but left attached, and a shorter urethra is cut (C). All but a stump of the penis is removed (D). The excess skin is used to create the labia (external genitalia) and vagina (E). (Illustration by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

KEY TERMS

Androgens—A class of chemical compounds (hormones) that stimulates the development of male secondary sexual characteristics.

Chromosomes—The carriers of genes that determine gender and other characteristics.

Estrogens—A class of chemical compounds (hormones) that stimulates the development of female secondary sexual characteristics.

Hysterectomy—Surgical removal of the uterus.

Oophorectomy—Surgical removal of the ovaries.

Description

Converting male to female anatomy requires removal of the penis, reshaping genital tissue to appear more female, and constructing a vagina. A vagina can be successfully formed from a skin graft or an isolated loop of intestine. Following the surgery, female hormones (estrogen) will reshape the body's contours and stimulate the growth of satisfactory breasts.

Female to male surgery has achieved lesser success due to the difficulty of creating a functioning penis from the much smaller clitoral tissue available in the female genitals. Penis construction is not attempted less than a year after the preliminary surgery to remove the female organs. One study in Singapore found that a third of the persons would not undergo the surgery again. Nevertheless, they were all pleased with the change of sex. Besides the genital organs, the breasts need to be surgically altered for a more male appearance. This can be successfully accomplished.

The capacity to experience an orgasm, or at least "a reasonable degree of erogenous sensitivity," can be expected by almost all persons after gender reassignment surgery.

Diagnosis/Preparation

Gender identity is an extremely important characteristic for human beings. Assigning sex must take place immediately after birth for the mental health of both children and their parents. Changing sexual identity is among the most significant changes that a human can experience. It should therefore be undertaken with extreme care and caution. By the time most adults come to surgery, they have lived for many years with a dissonant identity. The average in one study was 29

years. Nevertheless, even then they may not be fully aware of the implications of becoming a member of the opposite gender.

In-depth psychological counseling should precede and follow any gender reassignment surgical procedure.

Sex reassignment surgery is expensive. The cost for male to female reassignment is \$10,000–\$20,000. The cost for female to male reassignment can exceed \$50,000.

Aftercare

Social support, particularly from one's family, is important for readjustment as a member of the opposite gender. If surgical candidates are socially or emotionally unstable before the operation, over the age of 30, or have an unsuitable body build for the new gender, they tend not to fare well after gender reassignment surgery; however, in no case studied did the gender reassignment procedure diminish the ability to work.

Risks

All surgery carries the risks of infection, bleeding, and a need to return for repairs. Gender reassignment surgery is irreversible, so a candidate must have no doubts about accepting the results and outcome.

Normal results

Persons undergoing gender reassignment surgery can expect to acquire the external genitalia of a member of the opposite gender. Persons having male to female gender reassignment surgery retain a prostate. Individuals undergoing female to male gender reassignment surgery undergo a **hysterectomy** to remove the uterus and **oophorectomy** to remove their ovaries. Developing the habits and mannerisms characteristic of the patient's new gender requires many months or years.

Morbidity and mortality rates

The risks that are associated with any surgical procedure are present in gender reassignment surgery. These include infection, postoperative **pain**, and dissatisfaction with anticipated results. Accurate statistics are extremely difficult to find. Intraoperative **death** has not been reported.

The most common complication of male to female surgery is narrowing of the new vagina. This can be corrected by dilation or using a portion of colon to form a vagina.

A relatively common complication of female to male surgery is dysfunction of the penis. Implanting a penile prosthesis is technically difficult and does not have uniformly acceptable results.

Psychiatric care may be required for many years after sex-reassignment surgery.

The number of deaths in male-to-female transsexuals was five times the number expected, due to increased numbers of **suicide** and death from an unknown cause.

Alternatives

There is no alternative to surgical reassignment to alter one's external genitalia. The majority of persons who experience gender disorder problems never surgically alter their appearance. They dress as members of the desired gender, rather than gender of birth. Many use creams or pills that contain hormones appropriate to the desired gender to alter their bodily appearance. Estrogens (female hormones) will stimulate breast development, widening of the hips, loss of facial hair and a slight increase in voice pitch. Androgens (male hormones) will stimulate the development of facial and chest hair and cause the voice to deepen. Most individuals who undergo gender reassignment surgery lead happy and productive lives.

Resources

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ORGANIZATIONS

American Medical Association, 515 N. State Street, Chicago, IL, 60610, (800) 621-8335, <http://www.ama-assn.org/>.

American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209-3901, (703) 907-7300, apa@psych.org, <http://www.psych.org/>.

American Psychological Association, 750 First Street, NE, Washington, DC, 20002-4242, (202) 336-5500, (800) 374-2721, <http://www.apa.org/>.

American Urological Association, 1000 Corporate Boulevard, Linthicum, MD, 21090, (410) 689-3700, (866) 746-4282, <http://www.auanet.org/>.

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Sex therapy

Definition

Sex therapy is the treatment of **sexual dysfunction**.

Purpose

Sex therapy utilizes various techniques in order to relieve sexual dysfunction commonly caused by **premature ejaculation** or sexual **anxiety** and to improve the sexual health of the patient.

Precautions

Sexual dysfunction conjures up feelings of guilt, anger, insecurity, frustration, and rejection. Therapy is slow and requires open communication and understanding between sexual partners. Therapy may inadvertently address interpersonal communication problems.

Description

Sex therapy is conducted by a trained therapist, doctor, or psychologist. The initial sessions should cover a complete history not only of the sexual problem but of the entire relationship and each individual's background and personality. The sexual relationship should be discussed in the context of the entire relationship. In

fact, sexual counseling may de-emphasize sex until other aspects of the relationship are better understood and communicated.

There are several techniques that combat sexual dysfunction and are used in sex therapy. They include:

- Semans' technique: which is used to help combat premature ejaculation with a "start-stop" approach to penis stimulation. By stimulating the man up to the point of ejaculation and then stopping, the man will become more aware of his response. More awareness leads to greater control, and open stimulation of both partners leads to greater communication and less anxiety. The start-stop technique is conducted four times until the man is allowed to ejaculate.
- Sensate focus therapy, the practice of nongenital and genital touching between partners in order to decrease sexual anxiety and build communication. First, partners explore each other's bodies without touching the genitals or breasts. Once the couple is comfortable with nongenital touching, they can expand to genital stimulation. Intercourse is prohibited in order to allow the partners to expand their intimacy and communication.
- Squeeze technique, which is used to treat premature ejaculation. When the man feels the urge to ejaculate, his partner squeezes his penis just below the head. This stops ejaculation and gives the man more control over his response.

Aftercare

Habits change slowly. All the techniques must be practiced faithfully for long periods of time to relearn behaviors. Communication is imperative.

Resources

BOOKS

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J. Ricker Polsdorfer, MD

Sexual abuse

Definition

Sexual **abuse** is defined as any nonconsensual sexual exposure or activity. Any inappropriate exposure or sexual behavior, contact, or activity between a child

and an older child, teen, or adult is considered to be both sexual abuse and a form of **child abuse**.

Demographics

Of the estimated 3.2 million reports of child abuse in the United States in 2007, affecting about 5.8 million children, 7.6% involved sexual abuse. However the number of sexually abused children is much greater, since many—if not the majority of cases—go unreported. According to the U.S. Department of Justice (DOJ), as many as one out of three girls and one out of five boys under age 18 have been sexually abused. Serious underreporting of abuse occurs because children are too ashamed or afraid to tell anyone or because they think that they will not be believed. Legal procedures for corroborating sexual abuse can also be an impediment. However in recent years increased public awareness of child sexual abuse has resulted in more reporting and more prosecutions.

Although children of any age—from infants to teens—may be victims of sexual abuse, elementary-school-age children and the disabled are at particular risk. The vast majority of child sexual abuse—approximately 90%—is committed by someone the child knows and 68% of cases involve incest—abuse by a family member. The majority of sexual abusers are male and incest most often involves fathers or stepfathers abusing their daughters. However parents, stepparents, and foster parents of either gender abuse their daughters, sons, stepchildren, or foster children. Sexual abuse also occurs between siblings. Sexual abusers include other relatives, neighbors, caregivers, teachers, friends, and strangers. Sexual abuse occurs everywhere, among all racial, ethnic, religious, and socio-economic groups.

Description

The definition of sexual abuse varies among cultures and has changed over time. Although most societies view sex between children and adults as inappropriate, mores concerning the appropriate ages and age differences for sexual partners vary. Feminist movements in Western countries have successfully promoted definitions of sexual abuse that include a wide range of interactions between a child and an older child or adult, including some interactions that do not involve bodily contact.

Sexual abuse includes:

- voyeurism, including pictures on a Web site
- exposing a child to erotic material or pornography

- exposing one's genitals to a child
- sexual comments or gestures directed at a child
- solicitation of a child
- inappropriate touching, including oral, anal, breast, genital, or buttock contact
- forcing, pressuring, tricking, or talking a child into engaging in inappropriate touching, fondling, or other sexual activity
- rape or incest
- using objects for oral, vaginal, or anal fondling, stimulation, or penetration
- using a child for pornography, prostitution, Internet crimes, or other exploitative endeavors
- inadequately or inappropriately supervising a child's voluntary sexual activity

Although child sexual abuse can involve violence, it usually involves coercion. Whereas infants and young children are defenseless, older children may be susceptible to bribes or threats. However most often they acquiesce to the demands of adults who hold inherent power over them. Abusers may manipulate a child through a process called grooming, which can involve gaining the child's trust, confusing the child, preparing the child for sexual abuse, and ensuring that the child will not tell. Grooming can also involve creating a public persona in which the abuser is held in high regard by the child's family and community.

Sexual abuse can overwhelm a child with horror, confusion, disbelief, fear, and shame. Sometimes children become passive in an attempt to dissociate their minds from the physical reality. This passivity can be misconstrued as consent. Some children attempt to repress memories of the abuse or rationalize it into insignificance.

Sexual abuse can leave a child emotionally devastated. Even very young children, who may not understand what has happened, are unable to cope with the over-stimulation. Children aged five and older may feel caught between loyalty or affection toward their abuser and their knowledge that the sexual activity is wrong. If the abuser is a family member, the child may be afraid of breaking up the family. Some abusers threaten children with withdrawal of love. Children often feel that they are to blame for the abuse and this combination of shame and guilt reinforces the abuser's insistence on secrecy.

Risk factors

Risk factors for sexual abuse are similar to those for other types of child abuse, including family stresses, poverty, and alcohol and drug use. Many abusers were themselves physically or sexually abused as children.

Causes and symptoms

Perpetrators of sexual abuse often have multiple victims. They tend to be angry people with a need to control or dominate others. Some abusers suffer from **personality disorders** or are psychotic. Others have a psychiatric disorder known as pedophilia—sexual attraction to prepubescent children. Some sexual abuse is situational, such as parents who abuse their children only when they are under the influence of alcohol or drugs or are under severe **stress**.

Sometimes children report sexual abuse and occasionally there are physical signs, including:

- vaginal or rectal bleeding, pain, itching, redness, swelling, or discharge
- painful urination or bowel movements
- difficulty sitting or walking
- rarely, injury to the buttocks, lower abdomen, or extremities
- pregnancy or a sexually transmitted infection (STI), especially in a child under age 14

Symptoms of sexual abuse in children can be similar to symptoms of **post-traumatic stress disorder** (PTSD):

- stomachaches, headaches, or other vague complaints
- sleep disturbances
- sudden fear of the dark
- nightmares
- bedwetting
- soiling oneself or other bowel disorders
- difficulty eating or swallowing
- loss of appetite or eating disorders, such as anorexia nervosa
- severe nervousness or anxiety
- excessive fears
- poor concentration
- sudden or extreme mood swings, including fear anger or excessive crying
- lethargy
- depression
- withdrawal from family, friends, or usual activities

Other behavioral symptoms of sexual abuse may include:

KEY TERMS

Hymen—A membrane that partially or completely covers the vaginal opening.

Incest—Sexual intercourse between people who are too closely related to legally marry.

Pedophilia—A sexual perversion in which children are the preferred sexual object.

Pornography—Sexually explicit pictures, writings, or other material produced for the purpose of sexual arousal.

Post-traumatic stress disorder (PTSD)—A psychological reaction that continues long after a highly stressful event and is characterized by depression, anxiety, flashbacks, and nightmares.

Sexually transmitted infection (STI)—An infectious disease that is transmitted through sexual activity.

Voyeurism—Sexual stimulation by visual means, usually by observing an unsuspecting individual.

- secretiveness
- talk of a new, older friend
- the sudden appearance of unexplained money or gifts
- a belief that their body is dirty or damaged or that there is something wrong with their genitals
- refusal to change into gym clothes or participate in physical activities
- poor school performance or refusal to attend school
- fear or avoidance of particular people, places, or activities
- excessive obedience
- aggression or other behavioral problems
- regression to earlier developmental stages, including infantile behaviors such as thumb sucking
- running away from home
- avoidance of anything sexual
- sexual interest, language, or knowledge that is inappropriate for the child's age
- unusual or aggressive sexual activities with other children or toys
- excessive or public masturbation

Additional signs of sexual abuse in an adolescent may include:

- high-risk sexual behaviors
- alcohol or drug use
- self-mutilation, such as cutting or burning

Children who are victims of prolonged sexual abuse may develop:

- low self-esteem and feelings of worthlessness
- distrust of adults
- abnormal attitudes toward sex
- difficulties relating to others on nonsexual terms
- multiple personality disorder
- suicidal tendencies

In a process called “secondary victimization,” a non-abusive parent of a sexually abused child may develop many of these same symptoms. Possible signs of a sexually abusive parent or other adult may include:

- overprotection of a child
- severely limiting a child’s interactions with others, especially children of the opposite sex
- secretiveness
- isolation
- jealous or controlling behaviors

Diagnosis

Examination

A child who reports sexual abuse or is suspected of having been abused should have a complete **physical examination** as soon as possible, preferably within 72 hours. An examination is ordered whenever a case of suspected sexual abuse is reported to the police or child protection agency. Medical professionals, teachers, and childcare professionals are required by law to report suspected cases of sexual abuse.

The physician will look for any signs of physical injury or sexual abuse, particularly in the mouth and throat and around the anus and penis or vagina. The hymen—a thin membrane covering the opening to the vagina—may be affected in abused girls. However most children are not physically harmed during sexual abuse; signs of abuse are usually temporary; and the abuse is often not reported or discovered until some time after the last occurrence. Therefore diagnostic findings from a physical exam are rare.

Evaluation of sexually abused children by a trained professional is essential to determine whether treatment is required. Children are often afraid to talk openly about their abuse and therefore must be made to feel very safe. The assessment includes the child’s:

- abuse history and other life stresses, such as frequent moves or personal losses
- current stresses, such as medical problems or learning disabilities

- emotional state
- coping strategies, such as withdrawal or behavioral symptoms
- strengths, such as creativity or athletic ability
- communication skills
- friendships
- attachments to adults

Tests

Blood and/or urine tests may be performed to check for STIs such as **syphilis** and HIV. Adolescent girls may be tested for **pregnancy**.

Procedures

If the sexual abuse included physical harm, injuries may be photographed for use in prosecution of the perpetrator. Serious injuries may require diagnostic imaging procedures.

Treatment

Traditional

The child will be treated for any physical injuries. Police and child protection agencies investigate reports of sexual abuse and are responsible for protecting the child from additional harm. This may involve placing a child with a non-abusive parent or other relative or in a foster home.

Sexually abused children and their families may receive mental health treatment from a counselor, therapist, social worker, psychologist, or psychiatrist:

- Individual therapy is geared to the child's age. It may involve traditional talk therapy or art, play, or music therapy for children who are unable to talk about their experiences.
- Group therapy with other sexually abused children can help a child feel less isolated and learn new skills through games, role playing, and discussions.
- Family therapy can improve parent-child communications and help parents and children learn new coping skills.

Drugs

A sexually abused child may require **antibiotics** or other drugs to prevent or treat STIs. Older girls may be treated with drugs to prevent pregnancy.

Alternative

Support groups for sexually abused children, as well as for their parents or caregivers, can provide an alternative to traditional treatments.

Home remedies

A child's family and home life are critical for recovery from sexual abuse. Important factors include:

- ongoing acceptance of the child's experiences and emotional responses
- respect for the child's level of comfort with physical contact, including touching, hugging, kissing, tickling, and playful wrestling
- encouraging children to respect the comfort levels and privacy preferences of others, including knocking before entering bedrooms and bathrooms and bathing and dressing in private, if possible
- preventing children's exposure to adolescent and adult sexual behaviors
- monitoring children's exposure to television, movies, music and music videos, video games, and magazines with sexual messages, including language and nudity
- monitoring children's Internet usage

Some abused children exhibit sexually aggressive behaviors that require extra safety precautions:

- supervision when playing with friends
- avoidance of sleepovers
- informing the school counselor or other personnel about the possibility of inappropriate behaviors
- extra supervision in other group situations such as daycare, after-school programs, or camp

Prognosis

Various factors influence the effects of sexual abuse on a child, including:

- the type of abuse
- the frequency and duration of the abuse
- the gender of the child and the abuser
- the child's age and emotional development, with younger children being more vulnerable
- the child's relationship to the abuser and the degree to which the abuse is a betrayal of trust
- the form of coercion or seduction
- threats of harm to the child or to the child's family, friends, or pets
- emphasis on secrecy
- the child's ability to cope with his or her physical and emotional responses
- the degree to which the child feels responsible for the abuse

If untreated, the emotional and psychological damage from sexual abuse can be devastating. Children who are coerced into hiding the abuse are most likely to suffer

long-term effects. Fear, anger, guilt, and shame can continue into adulthood, resulting in:

- low self-esteem
- chronic feelings of guilt, helplessness, or hopelessness
- self-destructive behaviors
- anxiety disorders
- sleep disorders
- eating disorders
- depression
- problems with intimacy and trust
- sexual disorders
- unsafe sexual behaviors
- marital and family problems
- PTSD
- psychotic symptoms
- multiple personality disorder from attempts to dissociate from the experience
- suicidal tendencies

There may be other long-term consequences of childhood sexual abuse, including an increased risk for:

- teen pregnancy
- sexually abusing others
- prostitution
- homelessness
- alcohol abuse
- drug addiction

Sexually abused children who receive professional treatment and support from families and friends are much less likely to experience long-term consequences. Many sexually abused children grow up to live happy and productive lives. Most do not become adult abusers.

Prevention

- Parents should be aware that most sexual abuse is perpetrated by someone the child knows.
- Toddlers should be taught the proper names of body parts and that the parts covered by a bathing suit are private.
- Preschoolers should be taught about their private body parts and how to talk to and touch others respectfully.
- Children aged five to eight should be taught to respect the private parts of others and expect the same respect from others. They should also be taught how to recognize potentially dangerous situations, such as being accosted by a stranger.

- Parents should visit their child's caregivers unannounced.
- Children should be taught that if someone tries to touch their body, look at their private parts, show them their parts, or otherwise makes them uncomfortable, they should immediately say "no" and tell a parent or trusted adult.
- Children should not be taught to automatically obey adults.
- Children should be taught not to keep secrets.
- Children should be taught what to do if they are separated from their family in a public place.
- Children should be taught never to go with anyone without their parent's permission.
- Children should be taught to run away and scream if someone tries to take them.
- Children aged eight through 12 should be taught about personal safety when away from home, about sexual abuse, and about peer pressure.
- Parents should talk with their children about sex openly, honestly, and frequently, and give understandable, age-appropriate answers to questions.
- Children should feel that they can talk to their parents about anything that makes them uncomfortable, afraid, or confused.
- Parents should learn what sexual behaviors are normal at each age and be aware of any abnormal or aggressive behaviors.
- Parents should talk to their teenagers about types of sexual abuse, prevention of STIs and pregnancy, and drugs and alcohol.
- Teens should be taught to respect others and expect the same: No one should ever have to say "no" more than once.
- Parents should know the children and adults with whom their children associate.
- Parents should monitor their children's television and movie viewing and Internet use.

Most school systems have programs to teach young children about sexual abuse and its prevention. High schools usually teach students about avoiding **rape** and date rape.

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- American Academy of Child & Adolescent Psychiatry (AACAP), 3615 Wisconsin Ave., NW, Washington, DC, 20016-3007, (202) 966-7300, (202) 966-2891, <http://www.aacap.org>.
- Child Welfare Information Gateway, Children's Bureau/ACYF, 1250 Maryland Ave., SW, Eighth Floor, Washington, DC, 20024, (800) 394-3366, info@childwelfare.gov, <http://www.childwelfare.gov>.

- Childhelp, 15757 N. 78th St., Suite B, Scottsdale, AZ, 85260, (480) 922-8212, (800) 4-A-CHILD, (480) 922-7061, <http://www.childhelpusa.org>.
- National Center for Missing & Exploited Children (NCMEP), Charles B. Wang International Children's Building, 699 Prince St., Alexandria, VA, 22314-3175, (703) 224-2150, (800) THE-LOST (843-5678), (703) 224-2122, <http://www.missingkids.com>.
- RAINN/Rape, Abuse & Incest National Network, 2000 L St., NW, Suite 406, Washington, DC, 20036, (202) 544-1034, (800) 656-HOPE, info@rainn.org, <http://www.rainn.org>.

Margaret Alic, PhD

Sexual addiction

Definition

Psychiatrists do not agree on the exact definition of sexual addiction. The general elements of sexual addiction are a compulsive pattern of sexual behavior that arises from distorted thinking; sexual behavior that interferes with personal relationships, work, or other responsibilities; and often sex with multiple partners who are seen as objects to be used rather than people.

Demographics

Because there is no professional agreement on the definition of sexual addiction, the number of people who are sex addicts is unknown. What is known is that sexual addiction is much more common in men than women. Sex addicts are not necessarily sex offenders, nor are sexual molesters and rapists all sex addicts. Many sex addicts have other mental health and impulse control problems that contribute to their addictive behaviors.

Description

Although a healthy interest in sex is normal, sexual addiction goes well beyond normal healthy interest. For the sexual addict, thinking about sexual activity and having sex dominate thoughts to a degree that it interferes with healthy personal relationships and activities. According to the organization Sex Addicts Anonymous, this compulsive interest in sex covers a wide range of activities including

- compulsive masturbation
- compulsive viewing of pornography

KEY TERMS

Bipolar disorder—Formerly called manic depressive disorder. A mood disorder characterized alternating periods of overconfidence and activity (manic highs) and depressive lows.

Dissociate—To separate or disconnect thoughts and feelings from oneself and one's actions, usually in response to a painful, traumatic or highly stressful situation.

Obsessive-compulsive disorder (OCD)—An anxiety disorder in which a person cannot prevent himself from dwelling on unwanted thoughts, acting on urges, or performing repetitious rituals, such as washing his hands or checking to make sure he turned off the lights.

Paraphilia—Recurring strong sexual arousal to fantasies, objects, situations, or individuals that are not considered normal in the individual's culture.

- mania
- impulse control disorders
- substance abuse
- depression
- post traumatic stress disorder (PTSD)

Specific symptoms of sexual addiction vary with the sex act involved. However, a cycle of events appears to be common to sex addicts. In stage 1, the individual has some ongoing emotional stressor(s) (e.g., fear, **pain**, anger, and loneliness). The individual is unable to develop a healthy way to relieve or cope with the stressor(s). As a result, the individual moves on to stage 2. In stage 2, the individual begins to dissociate and separate thoughts and emotions from contemplated or anticipated actions. It is as if the and contemplated actions belong to someone else and will have no consequences for the individual. In stage 3, the individual moves from thinking about acting on his or her thoughts to actually acting on them. This may mean making obscene phone calls, viewing pornography, or searching for a sexual partner. The action culminates in sexual release, and then there is a period ranging from hours to weeks before the cycle starts again.

- compulsive phone sex
- multiple affairs outside of an established relationship
- frequent sex with anonymous partners
- multiple one-night stands
- prostitution or using prostitutes
- exhibitionism
- voyeurism
- sexual stalking
- sexual molestation or rape

Causes and symptoms

The cause of sexual addiction is not known. Some researchers have suggested that abnormal brain chemistry is responsible. Others suggest that early experiences and childhood **sexual abuse** contribute to the disorder. Nevertheless, there is general agreement among psychiatrists that sex addicts usually have one or more additional mental health disorders. Where experts disagree is over whether these mental health disorders alone are enough to cause the addict's behavior or whether sexual addiction is a separate disorder present in addition to other psychiatric disorders.

Common psychiatric disorders among individuals exhibiting sexually addictive behavior include

- obsessive-compulsive disorder (OCD)
- paraphilia
- bipolar disorder

Diagnosis

Diagnosis of sexual addiction is difficult, as many sex addicts deny that they have a problem. It is also complicated by the fact that sexual addiction often is associated with other mental health disorders. Sexual addiction is not recognized as a specific diagnosis in the *Diagnostic and Statistical Manual of Mental Disorders-IV, Text Revision (DSM-IV-TR)*) used by the American Psychiatric Association (APA) to classify mental disorders. Instead, it is classified as a sexual disorder not otherwise specified (NOS). This designation is intended to be used when other causes for the behavior, such as **impulse control disorders** and **obsessive-compulsive disorder**, have been eliminated. Within the APA, there is substantial controversy over whether sexual addiction should be classified as a separate psychiatric disorder in the upcoming *DSM-V*, which is to be released in 2013. The way this disorder is classified may effect both treatment and insurance coverage.

Treatment

There are two approaches to treating sexual addiction. Psychiatrists who see the disorder as mainly caused by compulsive disorder or as a variation on an impulse control disorder may treat the disorder with drugs such as fluoxetine (Prozac) or clomipramine (Anafranil) along with **psychotherapy**.

Psychiatrists who see sexual addiction as its own disorder are more likely to use psychotherapy to help the individual control addictive behavior. However, unlike addiction to drugs or alcohol, the goal of treating sexual addiction is not complete abstinence from sex, but rather to develop a normal, healthy approach to sex. Psychotherapy for sexual addiction may involve treatment at a residential center or intensive outpatient therapy. Regardless of where the therapy takes place, the individual should be treated by professionals experienced in dealing with sexual compulsions. Some therapy may involve couples. Often, therapy is supplemented by a 12-step recovery program such as the one designed by Sex Addicts Anonymous. Since many sex addicts have other mental health disorders such as **substance abuse** or depression, these also are treated with drugs and psychotherapy.

Prognosis

Recovery from sexual addiction is difficult. The earlier the addiction is treated (i.e., stage 1 or 2 before thoughts have been translated into actions), the greater the chance for recovery. Most sex addicts, however, do not recognize their disorder in its early stages and thus do not receive early treatment. Often it takes a major life-altering event to propel the sex addicted individual into treatment. Even then, relapses are common. Prognosis is also affected by the success or failure of treatment for other disorders such as substance **abuse**.

Prevention

Since the causes of sexual addiction are not clear, there is no definitive form of prevention. Recognizing the problem and getting early treatment for stressors can help prevent behaviors from becoming full-blown sexual addiction.

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American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209-3901, (703)907-7300, (888) 35-PSYCH [(888) 357-7924], apa@psych.org, <http://www.psych.org>.

Sex Addicts Anonymous, P. O. Box 70949, Houston, TX, 77270, (800) 477-8191 [US and Canada] +1 713-869-4902 [other countries], info@saa-recovery.org, <http://saa-recovery.org>.

Tish Davidson, AM

Sexual arousal disorders see **Sexual dysfunction**

Sexual desire disorders see **Sexual dysfunction**

Sexual dysfunction

Definition

Sexual dysfunction is broadly defined as the inability to fully enjoy sexual intercourse. Specifically, sexual dysfunctions are disorders that interfere with a full sexual response cycle. These disorders make it difficult for a person to enjoy or to have sexual intercourse. While sexual dysfunction rarely threatens physical health, it can take a heavy psychological toll, bringing depression, **anxiety**, and debilitating feelings of inadequacy.

Description

Sexual dysfunction takes different forms in men and women. A dysfunction can be life-long and always present; acquired; situational; or generalized, occurring despite the situation. A man may have a sexual problem if he:

- ejaculates before he or his partner desires
- does not ejaculate, or experiences delayed ejaculation
- is unable to have an erection sufficient for pleasurable intercourse

- feels pain during intercourse
- lacks or loses sexual desire

A woman may have a sexual problem if she:

- lacks or loses sexual desire
- has difficulty achieving orgasm
- feels anxiety during intercourse
- feels pain during intercourse
- feels vaginal or other muscles contract involuntarily before or during sex
- has inadequate lubrication

The most common sexual dysfunctions in men include:

- Erectile dysfunction: an impairment of the erectile reflex. The man is unable to have or maintain an erection that is firm enough for coitus or intercourse.
- Premature ejaculation, or rapid ejaculation with minimal sexual stimulation before, on, or shortly after penetration and before the person wishes it.
- Ejaculatory incompetence: the inability to ejaculate within the vagina despite a firm erection and relatively high levels of sexual arousal.
- Retrograde ejaculation: a condition in which the bladder neck does not close off properly during orgasm so that the semen spurts backward into the bladder.

Until recently, it was presumed that women were less sexual than men. In the past two decades, traditional views of female sexuality were all but demolished, and women's sexual needs became accepted as legitimate in their own right.

Female sexual dysfunctions include:

- Sexual arousal disorder: the inhibition of the general arousal aspect of sexual response. A woman with this disorder does not lubricate, her vagina does not swell, and the muscle that surrounds the outer third of the vagina does not tighten—a series of changes that normally prepare the body for orgasm (“the orgasmic platform”). Also, in this disorder, the woman typically does not feel erotic sensations.
- Orgasmic disorder: the impairment of the orgasmic component of the female sexual response. The woman may be sexually aroused but never reach orgasm. Orgasmic capacity is less than would be reasonable for her age, sexual experience, and the adequacy of sexual stimulation she receives.
- Vaginismus: a condition in which the muscles around the outer third of the vagina have involuntary spasms in response to attempts at vaginal penetration.

- Painful intercourse: a condition that can occur at any age. Pain can appear at the start of intercourse, midway through coital activities, at the time of orgasm, or after intercourse is completed. The pain can be felt as burning, sharp searing, or cramping; it can be external, within the vagina, or deep in the pelvic region or abdomen.

Causes and symptoms

Many factors, of both physical and psychological natures, can affect sexual response and performance. Injuries, ailments, and drugs are among the physical influences; in addition, there is increasing evidence that chemicals and other environmental pollutants depress sexual function. As for psychological factors, sexual dysfunction may have roots in traumatic events such as **rape** or incest, feelings of guilt, a poor self-image, depression, chronic **fatigue**, certain religious beliefs, or marital problems. Dysfunction is often associated with anxiety. If a man operates under the misconception that all sexual activity must lead to intercourse and to orgasm by his partner, and if the expectation is not met, he may consider the act a failure.

Men

With **premature ejaculation**, physical causes are rare, although the problem is sometimes linked to a neurological disorder, prostate infection, or **urethritis**. Possible psychological causes include anxiety (mainly performance anxiety), guilty feelings about sex, and ambivalence toward women. However, research has failed to show a direct link between premature ejaculation and anxiety. Rather, premature ejaculation seems more related to sexual inexperience in learning to modulate arousal.

When men experience painful intercourse, the cause is usually physical; an infection of the prostate, urethra, or testes, or an allergic reaction to spermicide or **condoms**. Painful erections may be caused by **Peyronie's disease**, fibrous plaques on the upper side of the penis that often produce a bend during erection. **Cancer** of the penis or testis and arthritis of the lower back can also cause **pain**.

Retrograde ejaculation occurs in men who have had prostate or urethral surgery, take medication that keeps the bladder open, or suffer from diabetes, a disease that can injure the nerves that normally close the bladder during ejaculation.

Erectile dysfunction is more likely than other dysfunctions to have a physical cause. Drugs, diabetes (the most common physical cause), Parkinson's

disease, **multiple sclerosis**, and spinal cord lesions can all be causes of erectile dysfunction. When physical causes are ruled out, anxiety is the most likely psychological cause of erectile dysfunction.

Female

Dysfunctions of arousal and orgasm in women also may be physical or psychological in origin. Among the most common causes are day-to-day discord with one's partner and inadequate stimulation by the partner. Finally, sexual desire can wane as one ages, although this varies greatly from person to person.

Pain during intercourse can occur for any number of reasons, and location is sometimes a clue to the cause. Pain in the vaginal area may be due to infection, such as urethritis; also, vaginal tissues may become thinner and more sensitive during **breastfeeding** and after **menses**. Deeper pain may have a pelvic source, such as **endometriosis**, pelvic **adhesions**, or uterine abnormalities. Pain can also have a psychological cause, such as fear of injury, guilt feelings about sex, fear of **pregnancy** or injury to the fetus during pregnancy, or recollection of a previous painful experience.

Vaginismus may be provoked by these psychological causes as well, or it may begin as a response to pain, and continue after the pain is gone. Both partners should understand that the vaginal contraction is an involuntary response, outside the woman's control.

Similarly, insufficient lubrication is involuntary, and may be part of a complex cycle. Low sexual response may lead to inadequate lubrication, which may lead to discomfort, and so on.

Diagnosis

In deciding when a sexual dysfunction is present, it is necessary to remember that while some people may be interested in sex at almost any time, others have low or seemingly nonexistent levels of sexual interest. Only when it is a source of personal or relationship distress, instead of voluntary choice, is it classified as a sexual dysfunction.

The first step in diagnosing a sexual dysfunction is usually discussing the problem with a health care professional, who will need to ask further questions in an attempt to differentiate among the types of sexual dysfunction. A physical exam of the genitals may be performed, and further medical tests may be ordered, including measurement of hormone levels in the blood. Men may be referred to a specialist in diseases of the urinary and genital organs (urologist), and primary care physicians may refer women to a gynecologist.

In general, causes of sexual dysfunction are either physical or psychological. Physical causes often have an underlying condition that effect sexual function including:

- diabetes
- heart disease
- neurological disorders
- pelvic surgery or trauma
- alcoholism and drug abuse
- chronic disease such as kidney or liver failure
- side effects of medicines
- hormone imbalance
- heavy smoking

Psychological factors including the following:

- stress or anxiety
- insecurity about sexual performance
- relationship discord
- confusion regarding sexual orientation
- depression
- trauma in previous sexual experiences

The following agents have been associated with sexual dysfunction, so patients should speak to their doctors if they have concerns regarding: Tamoxifen, Luminal, Dilantin, Mysloine, Tegretol, Tricyclic, Anafranil, Prozac, Paxil, Inderal, Lopressor, Corgard, Blocadren, Tenormin, Cimetidine, Tagament, Thorazine, Haldol, Zyprexa, Xanax, Valium, and some progestin-dominant birth control pills. It is important to note that there may be alternate medications available that do not affect sexual function. Other agents may also be available to counteract any sexual dysfunctions experienced with these medications. Prescribed medication should not be discontinued without first speaking with a physician.

Treatment

Treatments break down into two main kinds, physical and behavioral **psychotherapy**.

In many cases, doctors or advance practice nurses may prescribe medications to treat an underlying physical cause or sexual dysfunction. Possible medical treatments include:

- Viagra (Sildenafil) is a treatment for erectile dysfunction in men.
- Papaverine and prostaglandin are used for erectile difficulties.
- MUSE (Medical Urethral System for Erection), a prostaglandin E-1 pellet which can be inserted into the urethra. In addition, Caverject and Edex are

- prostaglandin E-1 injection medications for erectile dysfunction.
- Surgically implanted inflatable penile prosthesis for erectile dysfunction.
- Androgel, a topical gel for testosterone/androgen replacement in men. Testosterone injections and patches may also be used in men and women to stimulate sexual desire.
- Clomipramine, fluoxetine, as well as serotonin re-uptake inhibitors such as Prozac, Zoloft, and Anafranil for premature ejaculation.
- Hormone replacement therapy for female dysfunctions.
- EROS-CTD, a clitoral therapy device approved by the FDA in May 2000 is designed to enhance lubrication and sensation in women who have arousal disorders. With a gentle suction, it increases blood flow to the clitoris and surrounding area.

Other agents include:

- ICOS is an agent for treatment of erectile dysfunction.
- Uprima (apomorphine) claims to induce erection in men and arousal in women.
- Vasomax, an oral tablet, is said to facilitate an erection within 10–15 minutes. It is anticipated that Vasomax may aid women as well as men.
- Viagra for women.
- SS Cream is a topical agent with natural plant extracts which appears to desensitize the penis and is used to treat premature ejaculation.

In some cases, a specific technique may be used during intercourse to correct a dysfunction. One of the most common is the “squeeze technique” to prevent premature ejaculation. When a man feels that an orgasm is imminent, he withdraws from his partner. Then, the man or his partner gently squeezes the head of the penis to halt the orgasm. After 20–30 seconds, the couple may resume intercourse. The couple may do this several times before the man proceeds to ejaculation.

In cases where significant sexual dysfunction is linked to a broader emotional problem, such as depression or **substance abuse**, intensive psychotherapy and/or pharmaceutical intervention may be appropriate.

A variety of alternative therapies can be useful in the treatment of sexual dysfunction. Counseling or psychotherapy is highly recommended to address any emotional or mental components of the disorder. Botanical medicine, either western, Chinese, or ayurvedic, as well as nutritional supplementation, can help resolve biochemical causes of sexual dysfunction. **Acupuncture** and homeopathic treatment can be helpful by focusing on the energetic aspects of the disorder.

KEY TERMS

Ejaculatory incompetence—The inability to ejaculate within the vagina.

Erectile dysfunction—Difficulty achieving or maintaining an erect penis.

Impotence—The inability to achieve and sustain an erection suitable for intercourse.

Orgasmic disorder—The impairment of the ability to reach sexual climax.

Painful intercourse (dyspareunia)—Generally thought of as a female dysfunction but it also affects males. Pain can occur anywhere.

Premature ejaculation—Rapid ejaculation before the person wishes it, usually in less than one to two minutes after beginning intercourse.

Retrograde ejaculation—A condition in which the semen spurts backward into the bladder.

Sexual arousal disorder—The inhibition of the general arousal aspect of sexual response.

Vaginismus—Muscles around the outer third of the vagina have involuntary spasms in response to attempts at vaginal penetration, not allowing for penetration.

Some problems with sexual function are normal. For example, women starting a new or first relationship may feel sore or bruised after intercourse and find that an over-the-counter lubricant makes sex more pleasurable. Simple techniques, such as soaking in a warm bath, may relax a person before intercourse and improve the experience. **Yoga** and **meditation** provide needed mental and physical relaxation for several conditions, such as vaginismus. Relaxation therapy eases and relieves anxiety about dysfunction. Massage is extremely effective at reducing **stress**, especially if performed by the partner.

Prognosis

There is no single cure for sexual dysfunction, but almost all can be controlled. Most people who have a level of sexual dysfunction fare well once they get into a treatment program. For example, a high percentage of men with premature ejaculation can be successfully treated in two to three months. Furthermore, the gains made in **sex therapy** tend to be long-lasting rather than short-lived. Viagra produces an erection in 75% of men with erectile dysfunction. For men who are not responsive to drug treatment, studies with surgically

implanted inflatable penile prosthesis claim a success rate at approximately 98%.

Health care team roles

Nursing and allied health professionals play a critical part in the diagnosis and treatment of sexual dysfunction. Sex therapy, which is ideally provided by a member of the American Association of Sexual Educators, Counselors, and Therapists (AASECT), universally emphasizes correcting sexual misinformation, the importance of improved partner communication and honesty, anxiety reduction, sensual experience and pleasure, and interpersonal tolerance and acceptance. Sex therapists believe that many sexual disorders are rooted in learned patterns and values. These are termed psychogenic. An underlying assumption of sex therapy is that relatively short-term outpatient therapy can alleviate learned patterns, restrict symptoms, and allow a greater satisfaction with sexual experiences.

Registered dietitians and nutritionists can be instrumental in giving dietary guidance and **nutrition** supplementation that may improve overall health and energy levels. Health improvements may impact general well-being and sexual function.

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ORGANIZATIONS

American Academy of Clinical Sexologists, 1929 18th Street NW, Suite 1166, Washington, DC, 20009, (202) 462-2122.

American Association for Marriage and Family Therapy, 1100 17th Street NW, 10th Floor, Washington, DC, 20036-4601, (202) 452-0109.

American Association of Sex Educators, Counselors & Therapists, P. O. Box 238, Mt. Vernon, IA, 52314.

American Foundation for Urologic Disease, Sexual Function Health Council, 1126 N. Charles Street, Baltimore, MD, 21201, (410) 468-1800, <http://www.impotence.org>.

National Kidney and Urologic Diseases, Information Clearinghouse, 3 Information Way, Bethesda, MD, 20892-3580, <http://www.niddk.nih.gov>.

Network for Excellence in Women's Sexual Health (NEWSHE), Female Sexual Medicine Center, UCLA Medical Center, 924 Westwood Blvd., Suite 520, Los Angeles, CA, 90095, (310) 825-0025, (310) 794-0211, <http://www.newshe.com>.

Crystal Heather Kaczkowski, MSc.

Sexual perversions

Definition

Sexual perversions are conditions in which sexual excitement or orgasm is associated with acts or imagery that are considered unusual within the culture. To avoid problems associated with the stigmatization of labels, the neutral term paraphilia, derived from Greek roots meaning "alongside of" and "love," is used to describe what used to be called sexual perversions. A paraphilia is a condition in which a person's sexual

arousal and gratification depend on a fantasy theme of an unusual situation or object that becomes the principal focus of sexual behavior.

Description

Paraphilic behaviors can revolve around a particular sexual object or a particular act. They are defined by *DSM-IV* as "sexual impulse disorders characterized by intensely arousing, recurrent sexual fantasies, urges and behaviors considered deviant with respect to cultural norms and that produce clinically significant distress or impairment in social, occupational or other important areas of psychosocial functioning." The nature of a paraphilia is generally specific and unchanging, and most of the paraphilic behaviors are far more common in men than in women.

Paraphilic behaviors differ from what some people might consider "normal" sexual activity in that these behaviors cause significant distress or impairment in areas of life functioning. They do not refer to the normal use of sexual fantasy, activity, or objects to heighten sexual excitement where there is no distress or impairment. The most common signs of sexual activity that can be classified as paraphilia include: the inability to resist an impulse for the sexual act, the requirement of participation by non-consenting or under-aged individuals, legal consequences, resulting **sexual dysfunction**, and interference with normal social relationships.

Paraphilic behaviors include fantasies, behaviors, and/or urges which:

- involve nonhuman sexual objects, such as shoes or undergarments
- require the suffering or humiliation of oneself or partner
- involve children or other non-consenting partners

The most common paraphilic behaviors are:

- exhibitionism, or exposure of the genitals
- fetishism, or the use of nonliving objects
- frotteurism, or touching and rubbing against a non-consenting person
- pedophilia, or the focus on prepubescent children
- sexual masochism, or the receiving of humiliation or suffering
- sexual sadism, or the inflicting of humiliation or suffering
- transvestic fetishism, or cross-dressing
- voyeurism, or watching others engage in undressing or sexual activity

A paraphiliac often has more than one paraphilia. Paraphilic behaviors often result in a variety of associated problems, such as guilt, depression, shame, isolation, and impairment in the capacity for normal social and sexual relationships. A paraphilia can, and often does, become highly idiosyncratic and ritualized.

Causes and symptoms

There is very little certainty about what causes a paraphilia. Psychoanalysts generally theorize that these conditions represent a regression to or a fixation at an earlier level of psychosexual development resulting in a repetitive pattern of sexual behavior that is not mature in its application and expression. In other words, an individual repeats or reverts to a sexual habit arising early in life. Another psychoanalytic theory holds that these conditions are all expressions of hostility in which sexual fantasies or unusual sexual acts become a means of obtaining revenge for a childhood trauma. The persistent, repetitive nature of the paraphilia is caused by an inability to erase the underlying trauma completely. Indeed, a history of childhood **sexual abuse** is sometimes seen in individuals with paraphilic behaviors.

However, behaviorists suggest, instead, that the paraphilia begins via a process of conditioning. Non-sexual objects can become sexually arousing if they are frequently and repeatedly associated with a pleasurable sexual activity. The development of a paraphilia is not usually a matter of conditioning alone; there must usually be some predisposing factor, such as difficulty forming person-to-person sexual relationships or poor self-esteem.

The following are situations or causes that might lead someone in a paraphiliac direction:

- parents who humiliate and punish a small boy for strutting around with an erect penis
- a young boy who is sexually abused
- an individual who is dressed in a woman's clothes as a form of parental punishment
- fear of sexual performance or intimacy
- inadequate counseling
- excessive alcohol intake
- physiological problems
- sociocultural factors
- psychosexual trauma

Diagnosis

Whatever the cause, paraphiliacs apparently rarely seek treatment unless they are induced into it by an

arrest or discovery by a family member. This makes diagnosis before a confrontation very difficult.

Paraphiliacs may select an occupation, or develop a hobby or volunteer work, that puts them in contact with the desired erotic stimuli, for example, selling women's shoes or lingerie in fetishism, or working with children in pedophilia. Other coexistent problems may be alcohol or drug **abuse**, intimacy problems, and personality disturbances, especially emotional immaturity. Additionally, there may be sexual dysfunctions. **Erectile dysfunction** and an inability to ejaculate may be common in attempts at sexual activity without the paraphiliac theme.

Paraphilic urges may be mild, moderate, or severe. An individual with mild paraphilia is markedly distressed by the recurrent paraphiliac urges but has never acted on them. The moderate has occasionally acted on the paraphilic urge. A severe paraphiliac has repeatedly acted on the urge.

Treatment

The literature describing treatment is fragmentary and incomplete. Traditional **psychoanalysis** has not been particularly effective with paraphilia and generally requires several years of treatment. Therapy with hypnosis has also had poor results. Current interests focus primarily on several behavioral techniques that include the following:

- Aversion imagery involves the pairing of a sexually arousing paraphilic stimulus with an unpleasant image, such as being arrested or having one's name appear in the newspaper.
- Desensitization procedures neutralize the anxiety-provoking aspects of nonparaphilic sexual situations and behavior by a process of gradual exposure. For example, a man afraid of having sexual contact with women his own age might be led through a series of relaxation procedures aimed at reducing his anxiety.
- Social skills training is used with either of the other approaches and is aimed at improving a person's ability to form interpersonal relationships.
- Orgasmic reconditioning may instruct a person to masturbate using his paraphilia fantasy and to switch to a more appropriate fantasy just at the moment of orgasm.

In addition to these therapies, drugs are sometimes prescribed to treat paraphilic behaviors. Drugs that drastically lower testosterone temporarily (antiandrogens) have been used for the control of repetitive deviant sexual behaviors and have been prescribed for paraphilia-

KEY TERMS

Exhibitionism—Obtaining sexual arousal by exposing genitals to an unsuspecting stranger.

Fetishism—Obtaining sexual arousal using or thinking about an inanimate object or part of the body.

Frotteurism—Obtaining sexual arousal and gratification by rubbing one's genitals against others in public places.

Masochism—Sexual arousal by having pain and/or humiliation inflicted upon oneself.

Pedophilia—Sex or sexual activity with children who have not reached puberty.

Sadism—Sexual arousal through inflicting pain on another person.

Transvestitism—Sexual arousal from dressing in the clothes of the opposite sex.

Voyeurism—Sexual arousal by observing nude individuals without their knowledge.

related disorders as well. Cyproterone acetate inhibits testosterone directly at androgen receptor sites. In its oral form, the usual prescribed dosage range is 50–200 mg per day.

Serotonergics (drugs that boost levels of the brain chemical serotonin) are prescribed for anxious and depressive symptoms. Of the serotonergic agents reported, fluoxetine has received the most attention, although lithium, clomipramine, buspirone, and sertraline are reported as effective in case reports and open clinical trials with outpatients. Other alternative augmentation strategies that may be effective include adding a low dose of a secondary amine tricyclic antidepressant to the primary serotonergics, but these reports are only anecdotal.

Prognosis

Despite more than a decade of experience with psychotherapeutic treatment programs, most workers in the field are not convinced that they have a high degree of success. Furthermore, because some cases involve severe abuse, many in the general public would prefer to "lock up" the sex offender than to have him out in the community in a treatment program or on parole after the treatment program has been completed.

Paraphilia and paraphilia-related disorders are more prevalent than most clinicians suspect. Since these

disorders are cloaked in shame and guilt, the presence of these conditions may not be adequately revealed until a therapeutic alliance is firmly established. Once a diagnosis is established, appropriate education about possible behavioral therapies and appropriate use of psychopharmacological agents can improve the prognosis for these conditions.

ORGANIZATIONS

American Academy of Clinical Sexologists, Inc., 3203 Lawton Road, Suite 170, Orlando, FL, 32803, (407) 645-1641, <http://www.esextherapy.com>.

American Association for Marriage and Family Therapy, 112 South Alfred Street, Alexandria, VA, 22314-3061, (703) 838-9808, (703) 838-9805, <http://www.aamft.org>.

David James Doermann

body areas of people with an STD. Sexually transmitted diseases are also called venereal diseases.

Demographics

The Centers for Disease Control and Prevention (CDC) has reported that 85% of the most prevalent infectious diseases in the United States are sexually transmitted. The rate of STDs in this country is 50 to 100 times higher than that of any other industrialized nation. One in four sexually active Americans will be affected by an STD at some time in his or her life.

The CDC estimates that about 19 million new STD infections occur in the United States each year. Almost half of these infections occurs in someone between the ages of 15 and 24. It is estimated that STDs have an economic cost of as much as \$15.9 billion dollars each year in the United States alone.

The two most commonly reported STDs are Chlamydia and **gonorrhea**, with more than 1.5 million new cases being reported annually. The most frequently affected group is girls between 15 and 19 years of age, and women between 20 and 24 years of age. Other STDs may occur more frequently than Chlamydia and gonorrhea, but because some STDs such as human papillomavirus (HPV) and **genital herpes** do not get reported to the CDC they tend to be undercounted.

Sexually transmitted diseases

Definition

Sexually transmitted disease (STD) is a term used to describe more than 20 different infections that are transmitted through exchange of semen, blood, and other body fluids, or by direct contact with the affected

Description

Types of STDs

Some of the most common and potentially serious STDs in the United States include:

- Chlamydia. This STD is caused by the bacterium *Chlamydia trachomatis*, a microscopic organism that lives as a parasite inside human cells. In 2008, there were 1,210,523 reported cases of Chlamydia. That means that Chlamydia affects more about 40 out of every 1000 people. Chlamydia has been increasing in frequency in the United States in recent years, with a 9.2% increase in reported cases from 2007 to 2008. Approximately 40% of women will develop pelvic inflammatory disease (PID) as a result of Chlamydia infection, a leading cause of infertility.
- Human papillomavirus (HPV). HPV causes genital warts and is the single most important risk factor for cervical cancer in women. Over 100 types of HPV exist, but only about 30 of them can cause genital warts and are spread through sexual contact. In some instances, warts are passed from mother to child during childbirth, leading to a potentially life-

Antibiotics used to treat STDs

Brand name (generic name)	Possible side effects
Ceftin (cefuroxime axetil)	Diarrhea, nausea and vomiting, skin irritation
Cipro (ciprofloxacin)	Headache, heartburn, nausea and vomiting, stomach pain
Doryx (doxycycline hyclate)	Diarrhea, itching (genital and/or rectal), loss of appetite, nausea and vomiting, swelling
Flagyl (metronidazole)	Numbness or tingling sensation in extremities, seizures
Floxin (ofloxacin)	Diarrhea, dizziness, genital itching, headache, nausea and vomiting
Minocin (minocycline hydrochloride)	Anemia, blurred vision, hives, rash, throat irritation
Noroxin (norfloxacin)	Dizziness, headache, nausea
Zithromax (azithromycin)	Abdominal pain, diarrhea, nausea and vomiting

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

- threatening condition for newborns in which warts develop in the throat (laryngeal papillomatosis).
- Genital herpes. Herpes is an incurable viral infection thought to be one of the most common STDs in the United States. It is caused by one of two types of herpes simplex viruses: HSV-1 (commonly causing oral herpes) or HSV-2 (usually causing genital herpes). The CDC estimates that there were 292,000 new cases of genital herpes in 2008. It is believed to affect more than 45 million Americans (one out of every five individuals 12 years of age or older) are infected with HSV-2; this number has increased 30% since the 1970s. HSV-2 infection is more common in women (one out of every four women) than men (one out of every five men) and in African Americans than Caucasians.
 - Gonorrhea. The bacterium *Neisseria gonorrhoeae* is the causative agent of gonorrhea and can be spread by vaginal, oral, or anal contact. The CDC reports that 336,742 new cases of gonorrhea were reported in 2008. This is about 111.6 cases per 100,000 people. This was a decrease of 5.4% from 2007. Since 1975 reported cases of gonorrhea have declined more than 70%.
 - Syphilis. Syphilis is a potentially life-threatening infection that increases the likelihood of acquiring or transmitting HIV. In 2008, the CDC reported approximately 13,500 new cases of syphilis in the United States. This was the highest number of new cases reported since 1995. The rate of primary and secondary syphilis is about 4.5 cases per 100,000 people, and the rate of congenital syphilis is about 10.1 per 100,000 live births. Congenital syphilis causes irreversible health problems or death in as many as 40% of all live babies born to women with untreated syphilis.
 - Human immunodeficiencyvirus (HIV) infection. The CDC estimates that there are approximately 1,106,400 people in the United States living with HIV/AIDS, and that about one fifth of them were not aware of the HIV infection. In 2007 there were 35,962 diagnosed cases of AIDS in the United States, with 28 of them occurring in children under the age of 13. As of 2010 The World Health Organization estimated that there were 33.4 million people living with HIV worldwide. There is no cure for this STD.

STDs can have very painful long-term consequences as well as immediate health problems. They can cause:

- birth defects
- blindness
- bone deformities
- brain damage
- cancer

- heart disease
- infertility and other abnormalities of the reproductive system
- mental retardation
- death

Social groups and STDs

STDs affect certain population groups more severely than others. Women, young people, and members of minority groups are particularly affected. Women in any age bracket are more likely than men to develop medical complications related to STDs. Ethnic minorities are more likely to be affected by STDs than Caucasians, with African Americans especially at risk, although this may be changing. For example in 2008 the incidence of **syphilis** among white women was 0.5 cases per 100,000, while the incidence among African American women was 7.6 cases per 100,000.

Causes and symptoms

The symptoms of STDs vary according to the disease agent (virus or bacterium), the sex of the patient, and the body systems affected. The symptoms of some STDs are easy to identify, others produce infections that may either go unnoticed for some time or are easy to confuse with other diseases. Syphilis in particular can be confused with disorders ranging from **infectious mononucleosis** to allergic reactions to prescription medications. In addition, the incubation periods of STDs varies. Some produce symptoms close enough to the time of sexual contact—often less than 48 hours later—for the individual to recognize the connection between the behavior and the symptoms. Others have a longer incubation period, so that the individual may not recognize the early symptoms as those of a sexually transmitted infection.

Some symptoms of STDs affect the genitals and reproductive organs:

- A woman who has an STD may bleed when she is not menstruating or have abnormal vaginal discharge. Vaginal burning, itching, and odor are common, and she may experience pain in her pelvic area while having sex.
- A discharge from the tip of the penis may be a sign that a man has an STD. Males may also have painful or burning sensations when they urinate.
- There may be swelling of the lymph nodes near the groin area.
- Both men and women may develop skin rashes, sores, bumps, or blisters near the mouth or genitals.

KEY TERMS

Chlamydia—A microorganism that resembles certain types of bacteria and causes several sexually transmitted diseases in humans.

Condom—A thin sheath worn over the penis during sexual intercourse to prevent pregnancy or the transmission of STDs. There are also female condoms.

Diaphragm—A dome-shaped device used to cover the back of a woman's vagina during intercourse in order to prevent pregnancy.

Pelvic inflammatory disease (PID)—An inflammation of the tubes leading from a woman's ovaries to the uterus (the Fallopian tubes), caused by a bacterial infection. PID is a leading cause of fertility problems in women.

Venereal disease—Another term for sexually transmitted disease.

Homosexual men frequently develop these symptoms in the area around the anus.

Other symptoms of STDs are systemic, which means that they affect the body as a whole. These symptoms may include:

- fever, chills, and similar flu-like symptoms
- skin rashes over large parts of the body
- arthritis-like pains or aching in the joints
- throat swelling and redness that lasts for three weeks or longer

Diagnosis

A sexually active person who has symptoms of an STD should be examined without delay by one of the following health care professionals:

- a specialist in women's health (gynecologist)
- a specialist in disorders of the urinary tract and the male sexual organs (urologist)
- a family physician
- a nurse practitioner
- a specialist in skin disorders (dermatologist).

The diagnostic process begins with a thorough **physical examination** and a detailed medical history that documents the patient's sexual history and assesses the risk of infection.

The doctor or other healthcare professional will:

- Describe the testing process. This includes all blood tests and other tests that may be relevant to the specific infection.
- Explain the meaning of the test results.
- Provide the patient with information regarding high-risk behaviors and any necessary treatments or procedures.

The doctor may suggest that a patient diagnosed with one STD be tested for others, as it is possible to have more than one STD at a time. One infection may hide the symptoms of another or create a climate that fosters its growth. At present, it is particularly important that people who are HIV-positive be tested for syphilis as well.

Notification

The law in some parts of the United States requires public health officials to trace and contact the partners of people with some STDs. Minors, however, can get treatment without their parents' permission. Public health departments in most states can provide information about STD clinic locations, and Planned Parenthood facilities are available to provide testing and counseling. These agencies can also help with or assume the responsibility of notifying sexual partners who should be tested and may require treatment.

Treatment

Although self-care can relieve some of the **pain** of genital herpes or **genital warts** that has recurred after being diagnosed and treated by a physician, other STD symptoms require immediate medical attention.

Antibiotics are prescribed to treat gonorrhea, Chlamydia, syphilis, and other STDs caused by bacteria. Although prompt diagnosis and early treatment can almost always cure these STDs, new infections can develop if exposure continues or is renewed. Viral infections can be treated symptomatically and possibly with antiviral medications.

Prognosis

The prognosis for recovery from STDs varies among the different diseases. The prognosis for recovery from gonorrhea, syphilis, and other STDs caused by bacteria is generally good, provided that the disease is diagnosed early and treated promptly. Untreated syphilis in particular can lead to long-term complications and disability. Viral STDs (genital herpes, genital **warts**, HIV) cannot be cured but must be treated on a long-term basis to relieve symptoms and prevent life-threatening complications.

Prevention

Vaccines

Vaccines for the prevention of **hepatitis A** and **hepatitis B** are currently available, and are recommended, especially for gay and bisexual men, users of illegal drugs, health care workers, and others at risk of contracting these diseases. Vaccine for HPV also is available and is recommended for young women. Vaccines to prevent other STDs are being actively researched and tested.

Research into vaccinations to prevent HIV infection are underway. Although some have undergone clinical trials as of 2010 there are no vaccines approved by the United States Food and Drug Administration (FDA) to prevent the disease.

Lifestyle choices

The risk of becoming infected with an STD can be reduced or eliminated by making certain choices. Abstaining from sexual contact, maintaining a mutually monogamous relationship, or being informed about a partner's medical status can all reduce the risk. The risk of contracting an STD can also be reduced by avoiding sexual contact with partners who are known to be infected with an STD, whose health status is unknown, who **abuse** drugs, or who are involved in the sex trade.

Use of condoms and other contraceptives

Condoms are the only known contraceptive method to reduce the risk of STD transmission. It is important to make sure a new condom is used every time there is genital, oral, or anal contact. Used correctly and consistently, male condoms provide good protection against HIV and other STDs such as gonorrhea, Chlamydia, and syphilis. Female condoms (lubricated sheaths inserted into the vagina) have also been shown to be effective in preventing HIV and other STDs. Condoms also provide a measure of protection against genital herpes, genital warts, and hepatitis B.

There is some evidence that spermicides and diaphragms may provide a small amount of protection from some STDs, but that claim remains extremely controversial, and it is recommended that people do not use these instead of other methods of STD protection. They do not protect women from contracting HIV. Birth-control pills, patches, or injections do not prevent STDs. Neither do surgical sterilization or **hysterectomy**.

Hygienic measures

Urinating and washing the genital area with soap and water immediately after having sex may eliminate

some germs before they cause infection. Douching, however, can spread infection deeper. It may also increase a woman's risk of developing **pelvic inflammatory disease** (PID).

Resources

BOOKS

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Detroit, MI: Greenhaven Press, 2007.

Grimes, Jill. *Seductive Delusions: How Everyday People Catch STDs*. Baltimore : Johns Hopkins University Press, 2008.

Marr, Lisa. *Sexually Transmitted Diseases: A Physician Tells You What You Need to Know* 2nd ed. Baltimore : The Johns Hopkins University Press, 2007.

Nack, Adina. *Damaged goods?: women living with incurable sexually transmitted diseases*. Philadelphia: Temple University Press, 2008.

OTHER

Medline Plus. Sexually Transmitted Diseases. February 3 2010. <http://www.nlm.nih.gov/medlineplus/sexuallytransmitteddiseases.html>

National STD and AIDS Hotline. (800)227-8922.

ORGANIZATIONS

AIDS Education and Training Centers (AETC) National Resource Center, 65 Bergen Street, 8th floor, Newark, NJ, 07101, info@aidsetc.org, <http://www.aidsetc.org>.

CDC National Prevention Information Network, P.O. Box 6003, Rockville, MD, 20849-6003, (404) 639-3113, (888)CDC-INFO (888) 232-4636, a 24-hour information number., cdcinfo@cdc.gov, <http://www.cdc.gov>.

Planned Parenthood Federation of America, 434 West 33rd St., New York, NY, 10001, (212) 541-7800, (800) 230-PLAN, (212) 245-1845, <http://www.plannedparenthood.org>.

United States Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (404) 639-3534, 800-CDC-INFO (800-232-4636). TTY: (888) 232-6348, inquiry@cdc.gov, <http://www.cdc.gov>.

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Sexually transmitted diseases cultures

Definition

Sexually transmitted diseases are infections spread from person to person through sexual contact. A culture is a test in which a laboratory attempts to grow and identify the microorganism causing an infection.



Cultures on agar plates. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Purpose

Sexually transmitted diseases (STDs) produce symptoms such as genital discharge, **pain** during urination, bleeding, pelvic pain, skin ulcers, or **urethritis**. Often, however, they produce no immediate symptoms. Therefore, the decision to test for these diseases must be based not only the presence of symptoms, but on whether or not a person is at risk of having one or more of the diseases. Activities, such as drug use and sex with more than one partner, put a person at high risk for these diseases.

STD cultures are necessary to diagnose certain types of STDs. Only after the infection is diagnosed can it be treated and further spread of the infection prevented. Left untreated, consequences of these diseases range from discomfort to **infertility** to **death**. In addition, these diseases, if present in a pregnant woman, can be passed from mother to fetus.

Description

Gonorrhea, **syphilis**, chlamydia, **chancroid**, herpes, human papillomavirus, human **immunodeficiency** virus (HIV), and mycoplasma are common sexually transmitted diseases. Not all are diagnosed with a culture. For those that are, a sample of material is taken from the infection site, placed in a sterile container, and sent to the laboratory.

Bacterial cultures

In the laboratory, a portion of material from the infection site is spread over the surface of several different types of culture plates and placed in an incubator at body temperature for one to two days. Bacteria present in the sample will multiply and appear on the plates as visible colonies. They are identified by the appearance of

their colonies and by the results of biochemical tests and a gram stain. The Gram stain is done by smearing part of a colony onto a microscope slide. After it dries, the slide is stained with purple and red stains, then examined under a microscope. The color of stain picked up by the bacteria (purple or red), the shape (such as round or rectangle), and the size provide valuable clues as to the identity and which **antibiotics** might work best. Bacteria that stain purple are called Gram-positive; those that stain red are called gram-negative.

The result of the gram stain is available the same day or in less than an hour if requested by the physician. An early report, known as a preliminary report, is usually available after one day. This report will tell if any microorganisms have been found yet, and if so, their Gram stain appearance—for example, a Gram-negative rod or a gram-positive cocci. The final report, usually available in one to seven days, includes complete identification and an estimate of the quantity of the microorganisms isolated.

A sensitivity test, also called antibiotic susceptibility test, commonly done on bacteria isolated from an infection site, is not always done on bacteria isolated from a sexually transmitted disease. These bacteria often are treated using antibiotics that are part of a standard treatment protocol.

GONORRHEA. *Neisseria gonorrhoeae*, also called gonococcus or GC, causes gonorrhea. It infects the surfaces of the genitourinary tract, primarily the urethra in males and the cervix in females. On a gram stain done on material taken from an infection site, the bacteria appear as small gram-negative diplococci (pairs of round bacteria) inside white blood cells. *Neisseria gonorrhoeae* grows on a special culture plate called Thayer-Martin (TM) media in an environment with low levels of oxygen and high levels of carbon dioxide.

The best specimen from which to culture *Neisseria gonorrhoeae* is a swab of the urethra in a male or the cervix in a female. Other possible specimens include vagina, body fluid discharge, swab of genital lesion, or the first urine of the day. Final results usually are available after two days. Rapid nonculture tests are available to test for GC and provide results on the same or following day.

CHANCROID. Chancroid is caused by *Haemophilus ducreyi*. It is characterized by genital ulcers with nearby swollen lymph nodes. The specimen is collected by swabbing one of these pus-filled ulcers. The gram stain may not be helpful as this bacteria looks just like other *Haemophilus* bacteria. This bacteria only grows on special culture plates, so the physician

must request a specific culture for a person who has symptoms of chancroid. Even using special culture plates, *Haemophilus ducreyi* is isolated from less than 80% of the ulcers it infects. If a culture is negative, the physician must diagnose chancroid based on the person's symptoms and by ruling out other possible causes of these symptoms, such as syphilis.

MYCOPLASMA. Three types of mycoplasma organisms cause sexually transmitted urethritis in males and **pelvic inflammatory disease** and **cervicitis** in females: *Mycoplasma hominis*, *Mycoplasma genitalium*, and *Ureaplasma urealyticum*. These organisms require special culture plates and may take up to six days to grow. Samples are collected from the cervix in a female, the urethra or semen in a male, or urine.

SYPHILIS. Syphilis is caused by *Treponema pallidum*, one in a group of bacteria called spirochetes. It causes ulcers or chancres at the site of infection. The organism does not grow in culture. Using special techniques and stains, it is identified by looking at a sample of the ulcer or chancre under the microscope. Various blood tests also may be done to detect the treponema organism.

CHLAMYDIA. Chlamydia is caused by the gram-negative bacterium *Chlamydia trachomatis*. It is one of the most common STDs in the United States and generally appears in sexually active adolescents and young adults. While chlamydia often does not have any initial symptoms, it can, if left untreated, lead to pelvic inflammatory disease and sterility. Samples are collected from one or more of these infection sites: cervix in a female, urethra in a male, or the rectum. A portion of specimen is combined with a specific type of cell and allowed to incubate. Special stains are performed on the cultured cells, looking for evidence of the chlamydia organism within the cells. A swab can also be taken from the woman's vulva. Men and women can now be screened for Chlamydia with a urine sample. Urine-based screening has increased screening significantly, especially among men.

Viral cultures

To culture or grow a virus in the laboratory, a portion of specimen is mixed with commercially prepared animal cells in a test tube. Characteristic changes to the cells caused by the growing virus help identify the virus. The time to complete a viral culture varies with the type of virus. It may take several days or up to several weeks.

HERPES VIRUS. Herpes simplex virus type 2 is the cause of **genital herpes**. Diagnosis is usually made based on the person's symptoms. If a diagnosis needs

confirmation, a viral culture is performed using material taken from an ulcer. A Tzanck smear is a microscope test that can rapidly detect signs of herpes infection in cells taken from an ulcer. The culture takes up to 14 days. In 2004, the FDA approved a blood test to detect the antibodies to herpes virus.

HUMAN PAPILLOMAVIRUS. Human papillomavirus causes **genital warts**. This virus will not grow in culture; the diagnosis is based on the appearance of the **warts** and the person's symptoms. In late 2003, the U.S. Food and Drug Administration (FDA) approved a human papillomavirus (HPV) DNA test with a Pap smear for screening women age 30 and older. The combined test would help physicians determine which women were at extremely low risk for **cervical cancer** and which should be more closely monitored.

HIV. Human immunodeficiency virus (HIV) is usually diagnosed with a blood test. Cultures for HIV are possible, but rarely needed for diagnosis. However, newer rapid tests were developed in 2003 and approved by the FDA in 2004. These tests are cheaper and can deliver results in as little as three minutes. The FDA also approved an HIV test in 2004 that can detect HIV in saliva.

Preparation

Generally, the type of specimen depends on the type of infection. Cultures always should be collected before the person begins taking antibiotics. After collection of these specimens, each is placed into a sterile tube containing a liquid in which the organism can survive while in route to the laboratory. The new rapid HIV tests rely on blood samples collected from a finger stick or vein or on saliva collected from the mouth. Initial results are not sent to a lab but are processed onsite.

Urethral specimen

Men should not urinate one hour before collection of a urethral specimen. The physician inserts a sterile, cotton-tipped swab into the urethra.

Cervical specimen

Women should not douche or take a bath within 24 hours of collection of a cervical or vaginal culture. The physician inserts a moistened, nonlubricated vaginal speculum. After the cervix is exposed, the physician removes the cervical mucus using a cotton ball. Next, he or she inserts a sterile cotton-tipped swab into the endocervical canal and rotates the swab with firm pressure for about 30 seconds.

KEY TERMS

Culture—A laboratory test done to grow and identify microorganisms causing infection.

Gram stain—Microscopic examination of a portion of a bacterial colony or sample from an infection site after it has been stained by special stains. Certain bacteria pick up the purple stain; these bacteria are called gram positive. Other bacteria pick up the red stain; these bacteria are called gram negative. The color of the bacteria, in addition to their size and shape, provide clues as to the identity of the bacteria.

Sensitivity test—A test that determines which antibiotics will kill the bacteria isolated from a culture.

Vulva—The external part of the woman's genital organs, including the vaginal vestibule.

Vaginal specimen

Women should not douche or take a bath within 24 hours of collection of a cervical or vaginal culture. The physician inserts a sterile, cotton-tipped swab into the vagina.

Anal specimen

The physician inserts a sterile, cotton-tipped swab about 1 inch into the anus and rotates the swab for 30 seconds. Stool must not contaminate the swab.

Oropharynx (throat) specimen

The person's tongue is held down with a tongue depressor, as a healthcare worker moves a sterile, cotton-tipped swab across the back of the throat and tonsil region.

Urine specimen

To collect a "clean-catch" urine, the person first washes the perineum, and the penis or labia and vulva. He or she begins urinating, letting the first portion pass into the toilet, then collecting the remainder into a sterile container.

Normal results

These microorganisms are not found in a normal culture. Many types of microorganisms, normally found on a person's skin and in the genitourinary tract, may contaminate the culture. If a mixture of

these microorganisms grow in the culture, they are reported as normal flora.

Abnormal results

If a person has a positive culture for one or more of these microorganisms, treatment is started and his or her sexual partners should be notified and tested. Certain laws govern reporting and partner notification of various STDs. After treatment is completed, the person's physician may want a follow-up culture to confirm the infection is gone.

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- American Social Health Association, P.O. Box 13827, Research Triangle Park, NC, 27709, (919) 361-8400, (919) 361-8425, <http://www.ashastd.org/>.
- Centers for Disease Control and Prevention. National Center for HIV, STD, and TB Prevention, 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdclinfo@cdc.gov, <http://www.cdc.gov/nchhstp/>.

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SGOT see **Aspartate aminotransferase test**

Shaken baby syndrome

Definition

Shaken baby syndrome is a severe form of closed-head injury caused by the forcible shaking of a child. The force is sufficient to cause the brain to bounce against the baby's skull, causing injury or damage to the brain. It is also known as shaken infant syndrome, SBS, abusive head trauma, shaken brain trauma, pediatric traumatic brain injury, and whiplash shaken infant syndrome. The syndrome was first identified by Dr. John Caffey (1895–1978), a pediatric radiologist who published a landmark paper on the subject in 1972.

Demographics

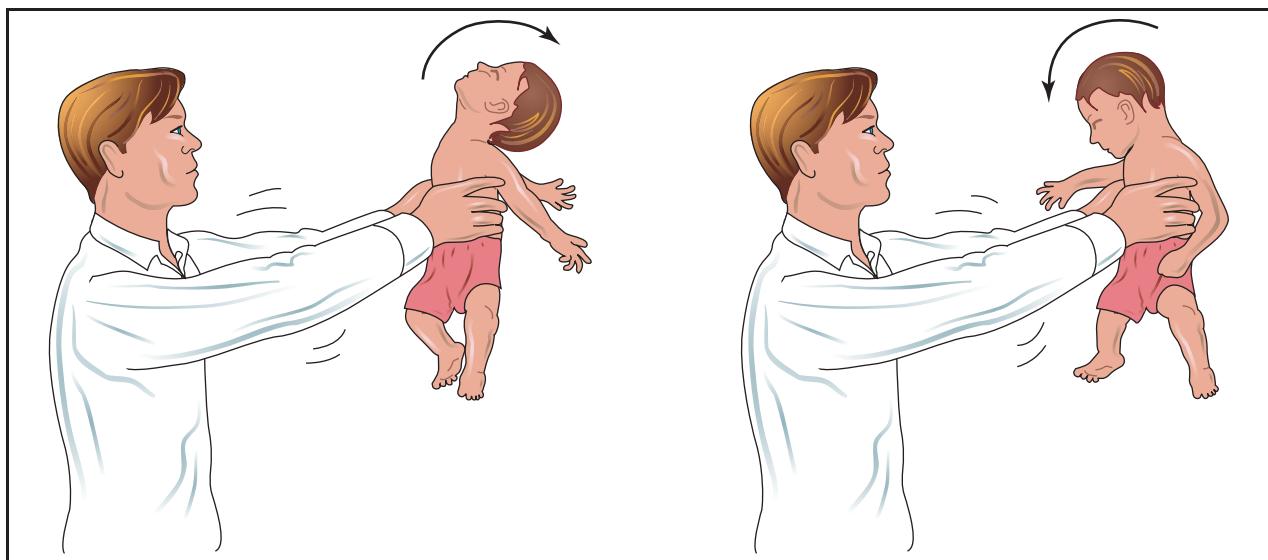
The American Academy of Pediatrics (AAP) estimates that there are between 600 and 1400 cases of SBS in the United States each year, though it is possible that the true number is higher because some cases are misdiagnosed as the result of accidental falls or auto accidents. The National Center on Shaken Baby Syndrome gives a figure of 1200 to 1400 cases annually in the United States. What is known is that shaken baby syndrome is the most common cause of mortality and long-term disability in infants and young children due to physical **abuse**. The syndrome has been reported in infants as young as 5 days and children as old as 5 years, but most victims are 2 years or younger.

SBS occurs in all racial groups in the United States but is more likely to be caused by males than by females. Adult males in their early 20s are the perpetrators in 65 to 90 % of cases; most often they are the baby's father or the mother's boyfriend. Female perpetrators are more likely to be a teenage babysitter or nanny than the baby's mother. The usual trigger for the abuse is crying lasting for several hours or repeated diaper soiling. In some cases involving men, the abuser is angry because he is jealous of the attention the baby receives from its mother.

Description

Shaking an infant forcibly transfers a great deal of energy to the infant. When the shaking occurs as the infant is being held, much of the force is transferred to the neck and the head. The force can be so great that the brain can move within the skull, rebounding back and forth from one side of the skull to the other. The bashing can be very destructive to the brain and neck because it can cause bruising, swelling, or bleeding. Bleeding of the brain is also called intracerebral hemorrhage.

As its name implies, shaken baby syndrome can often be a result of deliberate abuse. The brain damage can also be the result of an accident. The force and length of the force necessary to cause shaken baby syndrome is debatable. What is clear is that not much time is needed, since most shaking events likely tend to last only 20



Shaken baby syndrome is a collective term for the internal head injuries a baby or young child sustains from being violently shaken. Because of the fragile state of an infant's brain tissue and blood vessels, when a baby is vigorously shaken by the chest, as shown in the illustration above, the whiplash motion repeatedly jars the baby's brain with extreme force, causing serious internal damage and bleeding. Nearly 2,000 American children die annually from this condition. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

seconds or less. It is the explosive violence of the shaking that exacts the damage.

Risk factors

Risk factors for shaken baby syndrome include:

- Male sex. Most perpetrators of SBS are male.
- Substance abuse. While alcohol and drug abuse do not cause violence in the strict sense of cause, they lower a person's internal inhibitions against violence.
- Lack of information about normal child development.
- Psychological role reversal in which the caregiver expects the baby to fulfill his or her emotional needs.
- Divorce, separation, or family breakup.
- Financial stress.
- Social isolation. Caregivers who do not have friends or neighbors they can talk to (geographical distance, language barriers, recent move) may take out their loneliness as well as other stressors on the baby.
- Caregiver has personal history of having been abused as a child.

Causes and symptoms

The cause of the brain, neck, and spine damage that can result from shaken baby syndrome is brute force. Brain damage results from bleeding beneath the skull and bruising of brain tissue due to the brain's moving up against the inside of the skull during shaking. In some cases the brain is also damaged by loss of its oxygen supply.

The violent shaking of a baby by a much stronger adult conveys a tremendous amount of energy to the infant. Part of the reason for the damage is because an infant's head is much larger than the rest of the body, in relation to an older child or an adult. This, combined with neck muscles that are still developing and are incapable of adequately supporting the head, can make shaking an explosively destructive event. The amount of brain damage depends on how hard the shaking is and how long an infant is shaken. If accidental, the force and length of the head trauma similarly determines the extent of injury. The normal tossing and light horse play that can occur between an adult and an infant is not sufficient to cause shaken baby syndrome.

The damage to the brain can have dire consequences that include permanent and severe brain damage or **death**. Other symptoms that can develop include behavioral changes, lack of energy or motivation, irritable behavior, loss of consciousness, paling of the skin color or development of a bluish tinge to the skin, **vomiting**, and convulsions. These symptoms are the result of the destruction of brain cells, which is primarily due to the

KEY TERMS

Closed-head injury—An injury to the head in which the skull is not broken or penetrated.

Increased intracranial pressure—Increased overall pressure inside the skull.

Meningitis—Inflammation of the protective membranes that cover the brain and spinal cord.

Perpetrator—The legal term for a person who commits a crime.

Radiologist—A doctor who specializes in medical imaging techniques to diagnose or treat disease.

Retina—The layer of light-sensitive tissue at the back of the eyeball.

Subdural hematoma—A collection of blood or a clot trapped under the dura mater, the outermost membrane surrounding the brain and spinal cord, often causing neurological damage due to pressure on the brain.

trauma of the blow against the skull, and secondarily as a result of oxygen deprivation and swelling of the brain. The banging of the brain against the sides of the skull causes the inflammation and swelling as well as internal bleeding. Increased intracranial pressure can be damaging to the structure and function of the brain.

Additionally, because the neck and head can absorb a tremendous amount of energy due to the shaking force of the adult, bones in the neck and spine can be broken and muscles can be torn or pulled. The eyes can also be damaged by the explosive energy of shaking. Retinal damage occurs in 50–80% of cases. The damage can be so severe that it can permanently blind an infant.

Babies who are less severely injured when shaken may have symptoms that are easy to confuse with the symptoms of flu:

- Vomiting or other flu-like symptoms *without* fever or diarrhea.
- Crankiness and irritability over a period of time.
- Poor feeding, loss of appetite.
- Breathing problems.
- Unusual drowsiness.

Diagnosis

The doctor's greatest help in making a correct diagnosis of shaken baby syndrome is a description of what happened by the perpetrator or a witness. In

many cases an abuser will tell the doctor that the child fell or was in a car accident, or that the abuser shook the baby trying to revive it. One important clue is that the injuries caused by SBS are usually much more severe than would be caused by a fall or other accidental **head injury**.

Examination

Diagnosis depends on the detection of a blood clot below the inner layer of the dura mater (a membrane that surrounds the brain), but external to the brain. The clot is also known as a **subdural hematoma**. Two other critical features of shaken baby syndrome that are used in diagnosis are brain swelling and hemorrhaging in the eyes.

An infant may also have external bruising on parts of the body that were used to grip him or her during shaking. Bone or rib **fractures** can also be apparent. However, these external features may not always be present and the abuse may not be detected during a routine office visit.

Tests

Diagnosis of SBS can also involve the nondestructive imaging of the brain using the techniques of computed tomography (CT), skull x ray, or **magnetic resonance imaging** (MRI). Typically, these procedures are done after an infant has been stabilized and survival is assured. X-rays of the ribs and long bones of the body may also be ordered if the doctor suspects the child was struck with an object, thrown against a wall, or shaken.

In some cases the doctor may order laboratory tests of blood or cerebrospinal fluid to rule out **meningitis** and other infectious diseases that can affect the brain and cause seizures or **coma**.

Treatment team

Treatment in an emergency setting typically involves nurses and emergency room physicians. A neurosurgeon is usually consulted when shaken baby syndrome is suspected. Depending on the extent of injury, neurosurgeons can become involved if surgery for brain repair is needed. An ophthalmologist may be consulted to examine the baby's eyes for evidence of bleeding into the retina.

Police officers and social workers also become involved in cases of shaken baby syndrome in order to ensure that the child is placed in a safe environment.

Treatment

Traditional

Children with severe injuries from shaken baby syndrome require emergency treatment, usually brain surgery to relieve pressure on the brain and respiratory support to help them breathe. Treatment of the blindness, **learning disorders**, **mental retardation**, and other long-term consequences of SBS may last for the rest of the child's life. These children often need special education services, **physical therapy**, **speech therapy**, eye treatment, **psychotherapy**, and **occupational therapy**. Medical costs associated with initial and long-term care for these children can range from \$300,000 to more than \$1,000,000.

Clinical trials

As of May 2010, there are three clinical trials on shaken baby syndrome underway. Two trials concern educational strategies to prevent SBS by teaching parents about the syndrome and how to cope with **stress**; the third is a trial of preventing **colic** in children, as colic is a common cause of the crying and fussiness that may lead to parents' losing self-control. Such other agencies as the National Institute of Neurological Disorders and Stroke (NINDS) also fund studies that seek to better understand the basis of the damage. Other agencies attempt to lessen the occurrence of the syndrome through counseling, anger management, and interventions in abusive situations.

Prognosis

SBS has a high mortality rate. It is estimated that a third of the babies who are abused in this way will die; twenty percent of cases result in death within the first few days after injury. Another third will suffer severe permanent injuries, and the remaining third will recover.

Prevention

Dr. Caffey believed in the value of education to prevent at least some instances of SBS. While some abusers are people with a history of **substance abuse** or poor impulse control, others do not understand how much an angry adolescent or adult can harm a baby by shaking it. Various prevention strategies that are used as of 2010 include showing videos about SBS to new parents; encouraging pediatricians to discuss the stresses of childrearing with parents and teach them some ways to soothe a crying child; asking social workers to help identify families at risk of **child abuse**;

instructing workers in day care centers and others who work with small children about the syndrome; and advising parents to screen babysitters or nannies very carefully before hiring them for child care responsibilities.

The Arc, a community-based organization of and for people with developmental disabilities, offers three key words for caregivers to remember when dealing with a crying or fussy baby: Stop, Calm down, and Try again:

- Stop: Do not handle the baby if you are upset or angry. Place the child in a safe place like a crib or playpen.
- Calm down: Leave the room but stay close enough to hear the baby. Listen to calming music for a short time; then call a friend or a hotline for support or advice. Another approach is to run the vacuum cleaner; the noise will drown out the sound of the crying, and it also calms some babies. Keep in mind that the baby may be crying from an earache, teething, or other illness as well as hunger or a wet diaper. If the baby cannot be soothed and keeps crying for a long time, it is best to call the doctor.
- Try again: After calming down, try again to help the baby.

Keep the number of the Childhelp National Child Abuse Hotline on the refrigerator or near the telephone: 1-800-4-A-CHILD (1-800-422-4453). The hotline is staffed 24 hours a day, 365 days a year.

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National Center on Shaken Baby Syndrome. *All about Shaken Baby Syndrome/Abusive Head Trauma*. <http://www.dontshake.org/sbs.php?topNavID=3&subNavID=317>.

National Institute of Neurological Disorders and Stroke (NINDS). *Shaken Baby Syndrome Information Page*. <http://www.ninds.nih.gov/disorders/shakenbaby/shakenbaby.htm>.

ORGANIZATIONS

American Academy of Neurology (AAN), 1080 Montreal Avenue, Saint Paul, MN, 55116, (651) 695-2717, (800) 879-1960, (651) 695-2791, <http://www.aan.com/>.

American Academy of Pediatrics (AAP), 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007, (847) 434-4000, (847) 434-8000, <http://www.aap.org/>.

The Arc of the United States, 1660 L Street, NW, Suite 301, Washington, DC, 20036, (202) 534-3700, (800) 433-5255, (202) 534-3731, <http://www.thearc.org/NetCommunity/Page.aspx?pid=1386>.

Brain Trauma Foundation (BTF), 415 Madison Avenue, 14th Floor, New York, NY, 10017, (212) 772-0608, http://www.braintrauma.org/site/PageServer?page_name=homepage.

National Center on Shaken Baby Syndrome, 2955 Harrison Blvd #102, Ogden, UT, 84403, (801) 627-3399, (888) 273-0071, (801) 627-3321, mail@dontshake.org, <http://www.dontshake.org/index.php>.

National Institute of Neurological Disorders and Stroke (NINDS), P.O. Box 5801, Bethesda, MD, 20824, (800)352-9424, (301) 496-5751, <http://www.ninds.nih.gov/index.htm>.

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Shiatsu

Definition

Shiatsu is a manipulative therapy developed in Japan that incorporates techniques of *anma* (Japanese traditional massage), **acupressure**, stretching, and Western massage. Shiatsu involves applying pressure to special points or areas on the body in order to maintain physical and mental well being, treat disease, or alleviate discomfort. This therapy is considered holistic because it attempts to treat the whole person instead of a specific medical complaint. All types of acupressure generally focus on the same pressure points and so-called energy pathways but may differ in terms of massage technique. Shiatsu, which can be translated as finger pressure, has been described as needle-free **acupuncture**.

Purpose

Shiatsu has a strong reputation for reducing **stress** and relieving **nausea and vomiting**. Shiatsu is also believed to improve circulation and boost the immune system. Some people use it to treat **diarrhea**, **indigestion**, **constipation**, and other disorders of the gastrointestinal tract; menstrual and menopausal problems; chronic **pain**; migraine; arthritis; **toothache**; **anxiety**; and depression. Shiatsu can be used to relieve muscular pain or tension, especially neck and back pain. It also appears to have sedative effects and may alleviate **insomnia**. In a broader sense, shiatsu is believed to enhance physical vitality and emotional well being.

Description

Origins

Shiatsu is an offshoot of *anma* that developed during the period after the Meiji Restoration in 1868. Traditional massage (*anma*) used during the age of shoguns was being criticized, and practitioners of *koho anma* (ancient way) displeased with it introduced new practices and new names for their therapies.

During the twentieth century, shiatsu distinguished itself from *anma* through the merging of Western knowledge of anatomy, *koho anma*, *ampuku* (abdominal massage), acupressure, *Do-In* (breathing practices), and Buddhism. Based on the work of Tamai Tempaku, shiatsu established itself in Japan and worldwide. The Shiatsu Therapists Association was founded in 1925 and clinics and schools followed. Students of Tempaku began teaching their own brand of shiatsu, creating branch disciplines. By 1955, the Japanese Ministry of Health and Welfare acknowledged shiatsu as a

beneficial treatment, and licensing was established for practitioners.

Shiatsu and other forms of Japanese acupressure are based on the concept of *ki*, the Japanese term for the all-pervading energy that flows through everything in the universe. (This notion is borrowed from the Chinese, who refer to the omnipresent energy as *qi* or *chi*.) *Ki* tends to flow through the body along special energy pathways called meridians, each of which is associated with a vital organ. In Asian systems of traditional medicine, diseases are often believed to occur due to disruptions in the flow of this energy through the body. These disruptions may stem from emotional factors, climate, or a host of other causes including stress, the presence of impurities in the body, and physical trauma.

The aim of shiatsu is to restore the proper flow of bodily energy by massaging the surface of the skin along the meridian lines. Pressure may also be applied to any of the 600 or so acupoints. Acupoints, which are supposedly located just under the skin along the meridians, are tiny energy structures that affect the flow of *ki* through the body. When *ki* either stagnates and becomes deflected or accumulates in excess along one of these channels, stimulation to the acupoints, which are sensitive to pressure, can unblock and regulate the *ki* flow through toning or sedating treatment.

Western medicine has not proven the existence of meridians and acupoints. However, in one study, two French medical doctors conducted an experiment at Necher Hospital in Paris to test validity of the theory that energy is being transported along acupuncture meridians. They injected and traced isotopes with gamma-camera imaging. The meridians may actually correspond to nerve transmission lines. In this view, shiatsu and other forms of healing massage may trigger the emission of naturally occurring chemicals called neurotransmitters. Release of these chemical messengers may be responsible for some of the therapeutic effects associated with shiatsu, such as pain relief.

Preparations

People usually receive shiatsu therapy while lying on a floor mat or massage table or sitting up. The massage is performed through the clothing—preferably a thin garment made from natural fibers—and disrobing is not required. Pressure is often applied using the thumbs, though various other parts of the body may be employed, including fingertips, palms, knuckles, elbows, and knees—some therapists even use their feet. Shiatsu typically consists of sustained pressure (lasting up to 10 seconds at a time), squeezing, and stretching exercises. It

KEY TERMS

Acupressure—An ancient form of Asian healing massage that involves applying pressure to special points or areas on the body in order to maintain good health, cure disease, and restore vitality.

Analgesic—Pain reliever.

Osteoporosis—A disease of the bones due to deficiency of bone matrix, occurring most frequently in postmenopausal women.

Palpate—Feel.

may also involve gentle holding as well as rocking motions. A treatment session lasts anywhere from 30 to 90 minutes.

Before shiatsu treatment begins, the therapist usually performs a general health assessment. This involves taking a family medical history and discussing the physical and emotional health of the person seeking therapy. Typically, the practitioner also conducts a diagnostic examination by palpating the abdomen or back for any energy imbalances present in other parts of the body.

Precautions

While shiatsu is generally considered safe, there are a few precautions to consider. Because it may increase blood flow, this type of therapy is not recommended in people with bleeding problems, heart disease, or **cancer**.

Massage therapy should always be used with caution in those with **osteoporosis**, fresh **wounds** or scar tissue, bone **fractures**, or inflammation.

Applying pressure to areas of the head is not recommended in people with **epilepsy** or high blood pressure, according to some practitioners of shiatsu.

Shiatsu is not considered effective in the treatment of **fever**, **burns**, and infectious diseases.

Shiatsu should not be performed right after a meal.

Side effects

When performed properly, shiatsu is not associated with any significant side effects. Some people may experience mild discomfort, which usually disappears during the course of the treatment session.

Research and general acceptance

Like many forms of massage, shiatsu is widely believed to have a relaxing effect on the body. There is

also a significant amount of research suggesting that acupressure techniques can relieve **nausea** and **vomiting** associated with a variety of causes, including **pregnancy**, anesthetics, and other drugs. In one study, acupressure was shown to significantly reduce the effects of nausea in 12 of 16 women suffering from morning sickness. Five days of this therapy also appeared to reduce anxiety and improve mood. Another investigation, published in 1999, studied the effects of acupressure on nausea resulting from the use of anesthetics. Pressure applied to an acupoint on the inside of the wrist appeared to alleviate nausea in patients who received anesthetics during the course of laparoscopic surgery.

Shiatsu may also produce sedative and analgesic effects. The sedative powers of acupressure were investigated in a study published in the *Journals of Gerontology* 1999, which involved over 80 elderly people who suffered from sleeping difficulties. Compared to the people in the control groups, the 28 participants who received acupressure were able to sleep better. They slept for longer periods of time and were less likely to wake up during the night. The researchers concluded that acupressure may improve the quality of sleep in older adults. The use of acupressure in postoperative pain was investigated in a study published in 1996. In this study, which involved 40 knee surgery patients, one group received acupressure (15 acupoints were stimulated) while the control group received sham acupressure. Within an hour of treatment, members of the acupressure group reported less pain than those in the control group. The pain-relieving effects associated with acupressure lasted for 24 hours.

Shiatsu may benefit **stroke** victims. The results of at least one study (which did not include a control group) suggest that shiatsu may be useful during stroke **rehabilitation** when combined with other treatments.

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ORGANIZATIONS

Acupressure Institute, 1533 Shattuck Avenue, Berkeley, CA, 94709, (510) 845-1059, (800) 442-2232, info@acupressureinstitute.com, <http://www.acupressureinstitute.com>.

American Massage Therapy Association, 500 Davis Street, Suite 900, Evanston, IL, 60201-4695, (847) 864-0123, (847) 864-5196, (877) 905-2700, info@amtamassage.org, <http://www.amtamassage.org/>.

American Organization for Bodywork Therapies of Asia, 1010 Haddonfield-Berlin Road, Suite 408, Voorhees, NJ, 08043-3514, (856) 782-1616, (856) 782-1653, office@aopta.org, http://www.aopta.org/.

International School of Shiatsu, 10 South Clinton Street, Doylestown, PA, 18901, (215) 340-9918, (215) 340-9181, http://www.shiatsubo.com.

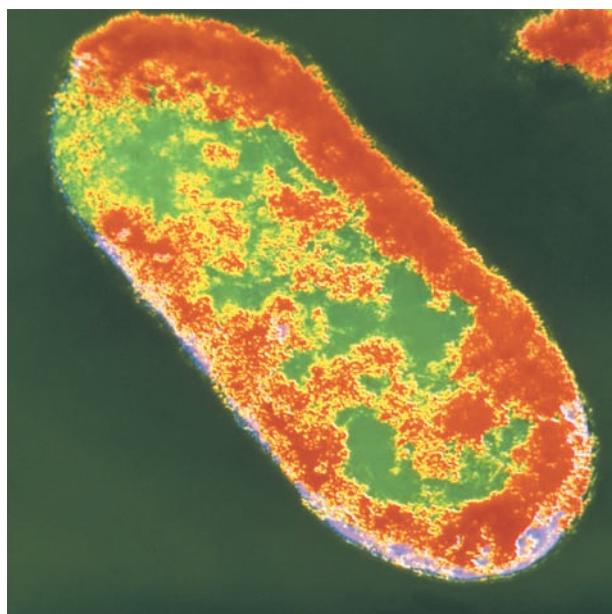
National Certification Board for Therapeutic Massage and Bodywork, 1901 South Meyers Road, Suite 240, Oak Brook, IL, 60181, (630) 627-8000, info@ncbtmb.org, http://www.ncbtmb.org.

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Shigellosis

Definition

Shigellosis is an infection of the intestinal tract by a group of bacteria called *Shigella*. The bacteria is named in honor of Shiga, a Japanese researcher, who discovered the organism in 1897. The major symptoms are **diarrhea**, abdominal cramps, **fever**, and severe fluid loss (**dehydration**). Four different groups of *Shigella* can affect humans; of these, *S. dysenteriae* generally produces the most severe attacks, and *S. sonnei* the mildest.



A transmission electron microscopy (TEM) scan of *Shigella*, a genus of aerobic bacteria that causes dysentery in humans and animals. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

Shigellosis is a well-known cause of **traveler's diarrhea** and illness throughout the world. *Shigella* are extremely infectious bacteria and ingestion of just 10 organisms is enough to cause severe diarrhea and dehydration. *Shigella* accounts for 10-20% of all cases of diarrhea worldwide, and in any given year infects over 140 million persons and kills 600,000, mostly children and the elderly. The most serious form of the disease is called **dysentery**, which is characterized by severe watery (and often blood- and mucous-streaked) diarrhea, abdominal cramping, rectal **pain**, and fever. *Shigella* is only one of several organisms that can cause dysentery, but the term bacillary dysentery is usually another name for shigellosis.

Most deaths are in less-developed or developing countries, but even in the United States, shigellosis can be a dangerous and potentially deadly disease. Poor hygiene, overcrowding, and improper storage of food are leading causes of infection. The following statistics show the marked difference in the frequency of cases between developed and less-developed countries; in the United States, about 30,000 individuals are hit by the disease each year or about 10 cases/100,000 population. On the other hand, infection in some areas of South America is 1,000 times more frequent. Shigellosis is most common in children below age 5 and occurs less often in adults over 20.

Causes and symptoms

Shigella share several of the characteristics of a group of bacteria that inhabit the intestinal tract. *E. coli*, another cause of food-borne illness, can be mistaken for *Shigella* both by physicians and the laboratory. Careful testing is needed to assure proper diagnosis and treatment.

Shigella are very resistant to the acid produced by the stomach, and this allows them to easily pass through the gastrointestinal tract and infect the colon (large intestine). The result is a **colitis** that produces multiple ulcers, which can bleed. *Shigella* also produce a number of toxins (Shiga toxin and others) that increase the amount of fluid secretion by the intestinal tract. This fluid secretion is a major cause of diarrhea symptoms.

Shigella infection spreads through food or water contaminated by human waste. Sources of transmission are:

- contaminated milk, ice cream, vegetables and other foods, which often cause epidemics
- household contacts (40% of adults and 20% of children will develop infection from such a source)
- poor hygiene and overcrowded living conditions
- day care centers

- sexual practices which lead to oral-anal contact, directly or indirectly

Symptoms can be limited to only mild diarrhea or progress to full-blown dysentery. Dehydration results from the large fluid losses due to diarrhea, **vomiting**, and fever. Inability to eat or drink worsens the situation.

In developed countries, most infections are of the less severe type, and are often due to *S. sonnei*. The period between infection and symptoms (incubation period) varies from one to seven days. Shigellosis can last from a few days to several weeks, with an average of seven days.

Complications

Areas outside the intestine can be involved, including:

- nervous system (irritation of the meninges or meningitis, encephalitis, and seizures)
- kidneys (producing **hemolytic-uremic syndrome** or HUS, which leads to kidney failure)
- joints (leading to an unusual form of arthritis called **Reiter's syndrome**)
- skin (rash)

One of the most serious complications of this disease is HUS, which involves the kidney. The main findings are kidney failure and damage to red blood cells. As many as 15% of patients die from this complication, and half of the survivors develop **chronic kidney failure**, which requires dialysis.

Another life-threatening condition is toxic megacolon. Severe inflammation causes the colon to dilate or stretch, and the thin colon wall may eventually tear. Certain medications (particularly those that diminish intestinal contractions) may increase this risk but this interaction is unclear. Clues to this diagnosis include sudden decrease in diarrhea, swelling of the abdomen, and worsening abdominal pain.

Diagnosis

Shigellosis is one of the many causes of acute diarrhea. Culture (growing the bacteria in the laboratory) of freshly obtained diarrhea fluid is the only way to be certain of the diagnosis. But even this is not always positive, especially if the patient is already on **antibiotics**. *Shigella* are identified by a combination of their appearance under the microscope and various chemical tests. These studies take several days but quicker means to recognize the bacteria and its toxins are being developed.

Treatment

The first aim of treatment is to keep up **nutrition** and avoid dehydration. Ideally, a physician should be

consulted before starting any treatment. Antibiotics may not be necessary, except for the more severe infections. Many cases resolve before the diagnosis is established by culture. Medications that control diarrhea by slowing intestinal contractions can cause problems and should be avoided by patients with bloody diarrhea or fever, especially if antibiotics have not been started.

Rehydration

The World Health Organization (WHO) has developed guidelines for a standard solution taken by mouth and prepared from ingredients readily available at home. This Oral Rehydration Solution (ORS) includes salt, baking powder, sugar, orange juice, and water. Commercial preparations, such as Pedialyte, are also available. In many patients with mild symptoms, this is the only treatment needed. Severe dehydration usually requires intravenous fluid replacement.

Antibiotics

In the early and mid-1990s, researchers began to realize that not all cases of bacterial dysentery needed antibiotic treatment. Therefore these drugs are indicated only for treatment of moderate or severe disease, as found in the tropics. Choice of antibiotic is based on the type of bacteria found in the geographical area and on laboratory results. Recommendations include ampicillin, sulfa derivatives such as Trimethoprim-Sulfamethoxazole (TMP-SMX) sold as Bactrim, or **fluoroquinolones** (such as Ciprofloxacin which is not FDA approved for use in children).

Prognosis

Many patients with mild infections need no specific treatment and recover completely. In those with severe infections, antibiotics will decrease the length of symptoms and the number of days bacteria appear in the feces. In rare cases, an individual may fail to clear the bacteria from the intestinal tract; the result is a persistent carrier state. This may be more frequent in **AIDS** (Acquired Immune Deficiency Syndrome) patients. Antibiotics are about 90% effective in eliminating these chronic infections.

In patients who have suffered particularly severe attacks, some degree of cramping and diarrhea can last for several weeks. This is usually due to damage to the intestinal tract, which requires some time to heal. Since antibiotics can also produce a form of colitis, this must be considered as a possible cause of persistent or recurrent symptoms.

KEY TERMS

Antibiotic—A medication that is designed to kill or weaken bacteria.

Anti-motility medications—Medications such as loperamide (Imodium), diphenoxyate (Lomotil), or medications containing codeine or narcotics which decrease the ability of the intestine to contract. These may worsen the condition of a patient with dysentery or colitis.

Carrier state—The continued presence of an organism (bacteria, virus, or parasite) in the body that does not cause symptoms but is able to be transmitted and infect other persons.

Colitis—Inflammation of the colon or large bowel, which has several causes. The lining of the colon becomes swollen and ulcers often develop. The ability of the colon to absorb fluids is also affected and diarrhea often results.

Dialysis—A form of treatment for patients with kidneys that do not function properly. The treatment removes toxic wastes from the body that are normally removed by the kidneys.

Dysentery—A disease marked by frequent watery bowel movements, often with blood and mucus, and characterized by pain, urgency to have a bowel movement, fever, and dehydration.

Fluoroquinolones—A relatively new group of antibiotics that have had good success in treating infections with many gram-negative bacteria, such as *Shigella*. One drawback is that they should not be used in children under 17 years of age because of possible effects on bone and cartilage growth.

Food-borne illness—A disease that is transmitted by eating or handling contaminated food.

Meninges—Outer covering of the spinal cord and brain. Infection is called meningitis, which can lead to damage to the brain or spinal cord and lead to death.

Oral Rehydration Solution (ORS)—A liquid preparation developed by the World Health Organization that can decrease fluid loss in persons with diarrhea. Originally developed to be prepared with materials available in the home, commercial preparations have recently come into use.

Stool—Passage of fecal material; a bowel movement.

Traveler's diarrhea—An illness due to infection from a bacteria or parasite that occurs in persons traveling to areas where there is a high frequency of the illness. The disease is usually spread by contaminated food or water.

Prevention

Shigellosis is an extremely contagious disease; good hand washing techniques and proper precautions in food handling will help in avoiding the spread of infection. Children in day care centers need to be reminded about hand washing during an outbreak to minimize spread. *Shigellosis* in schools or day care settings almost always disappears when holiday breaks occur, which sever the chain of transmission.

Traveler's diarrhea (TD)

Shigella accounts for about 10% of diarrhea illness in travelers to Mexico, South America, and the tropics. Most cases of TD are more of a nuisance than a life-threatening disease. However, bloody diarrhea is an indication that *Shigella* may be responsible.

In some cases though, aside from ruining a well deserved vacation, these infections can interrupt business conference schedules and, in the worst instances, lead to a life-threatening illness. Therefore, researchers have tried to find a safe and effective way of preventing

TD. One of the best means of prevention is to follow closely the rules outlined by the WHO and other groups regarding eating fresh fruits, vegetables, and other foods.

One safe and effective method of preventing TD is the use of large doses of Pepto Bismol. Tablets are now available which are easier for travel; usage must start a few days before departure. Patients should be aware that Bismuth will turn bowel movements black.

Antibiotics have also proven to be highly effective in preventing TD. They can also produce significant side effects, therefore a physician should be consulted before use. Like Pepto Bismol, antibiotics need to be started before beginning travel.

ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

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Shin splints

Definition

Shin splints refer to the sharp pains that occur down the front of the lower leg. They are a common complaint among runners and other athletes.

Description

Shin splints may refer to a number of lower leg complaints and injuries. In most cases, shin splints refer to the **pain** that results from overload on the tissues that connect muscles to the shin bone (tibia). They may come from the small bone of the lower leg and ankle, called the fibula. The medical term for shin splints is medial tibial stress syndrome.

Next to ankle sprains, shin splints are probably the most common complaint of injury to the lower body. Most shin splints occur in the front (anterior) portion of the tibia; some also occur in the inside of the leg, along the tibia. Runners probably suffer shin splints more than other people but they also occur in people who play basketball and tennis and those who walk long distances, particularly on treadmills.

Causes and symptoms

The most common cause of shin splints is overdoing activities that constantly pound on the legs and feet. This may include sports with many stops and starts, running down hills or other tilted surfaces, or repeated walking. Simply training too long or too hard, especially without proper stretching and warm-up, can cause shin splints. People with flat feet, high arches, or feet that turn outward may be more prone to shin splints. Shoes that are worn out or that do not provide proper foot support also add to the problem.

Diagnosis

The physician will check the leg for tenderness. If the pain is in a single area of the tibia and hurts to the touch, the cause may be a stress fracture. The physician may order an x ray to rule out a stress fracture, but shin splints can be diagnosed without x rays.

Treatment

Physicians usually recommend a period of rest for people with shin splints to let the area heal. Usually, about three to four weeks is recommended but the time varies depending on the patient and injury severity. Shin splints may be treated in phases,

KEY TERMS

Podiatrist—A physician who specializes in the medical care and treatment of the human foot.

Stress fracture—A hairline fracture (narrow crack along the surface of a bone) that is caused by repeated stress to the bone, such as from jogging, rather than from a single heavy blow.

beginning with absolute rest and gradual return to activity. Ice and elevation of the foot may be used to help relieve pain and swelling in the first phase. If the person needs to keep in shape, stretching and water exercises that keep the foot from bearing weight may be allowed after initial treatment. As the patient returns to normal function, orthotic footwear and braces may be added to prevent re-injury.

Alternative treatment

Various massage techniques may help speed up recovery. Homeopathic physicians may recommend Rhus tox. Those using alternative remedies should ensure they are certified practitioners and should coordinate care with allopathic providers.

Prognosis

With proper rest, management, and prevention, people with shin splints can return to normal activity in a few weeks or more. However, continuing to perform the activity that caused the shin splints can lead to stress **fractures** of the tibia.

Prevention

Re-injury is most common in the first month after return to normal activity and patients who have had shin splints should return to previous activities cautiously. The following can help prevent shin splints from occurring in people who run and perform stop and start physical activities:

- Warming up and stretching calf muscles before running or jogging. A podiatrist specializing in sports medicine or other sports medicine specialist may recommend specific stretching exercises.
- Strengthening muscles in the front lower leg (anterior tibialis) with resistance exercises or by walking on the heels three times daily for about 30 yards.
- Wearing quality shoes with arch supports. Runners should purchase new shoes about every 400 miles. A

podiatrist can design special arch supports or orthotics for people with flat feet.

- Runs should be started at a slow pace and gradually increased.
- Athletes can cross-train in a sport that does not impact the feet and lower legs as much, such as swimming or riding a bicycle.

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American Academy of Podiatric Sports Medicine, (301) 845-9887, info@aapsm.org, <http://www.aapsm.org>.

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Shingles, or herpes zoster, on patient's buttocks and thigh.
(Custom Medical Stock Photo, Inc. Reproduced by permission.)

there are at least one million cases in the United States each year.

Although shingles can occur at any age, even in children, the incidence increases steadily with age. About half of all cases occur in people aged 60 or older. About 20% of people with shingles develop PHN. It is more common in women than in men. In the United States between 120,000 and 200,000 people suffer from PHN each year. It occurs more frequently among the elderly and is one of the most common causes of pain-related **suicide** in older adults. The incidence of PHN increases with age and tends to last longer in older patients:

- PHN is rare in those under age 30.
- By age 40 the risk of PHN lasting longer than one month is 33%.
- By age 70 the risk increases to 74%.

Some scientists believe that the incidence of shingles is likely to increase over the next 40–50 years due to the introduction of a childhood vaccine against chickenpox in 1995. With far fewer children contracting chickenpox, adults have far less exposure to the virus, which would otherwise boost the immunity they acquired during childhood and help prevent reactivation of latent virus in their bodies.

Shingles

Definition

Shingles, or herpes zoster, is a condition caused by the reactivation of the varicella zoster virus (VZV) that causes **chickenpox** (varicella). After a bout of chickenpox, the virus remains dormant in the sensory nerve ganglia that are adjacent to the spinal cord and brain. Years later the virus reemerges, traveling along the nerves to the skin where it causes red **rashes** that develop into blisters. In the process the virus can damage nerves, leading to a very painful inflammation called post-herpetic **neuralgia** (PHN), which can persist long after the rash disappears.

Demographics

Anyone who has had chickenpox or been vaccinated against varicella can develop shingles. Virtually all American adults have had chickenpox, even if the disease was so mild as to pass unnoticed. Nearly one in three Americans eventually develops shingles and

Description

Varicella zoster virus is a member of the herpes virus family. It causes chickenpox or varicella, which is

highly contagious and spreads through the air. Following this initial or primary VZV infection, which usually occurs in childhood, the virus remains in an inactive or latent state in nerve tissue. Years later—usually after age 50—VZV can be reactivated to cause herpes zoster or shingles. The name “varicella” is derived from “variola,” the Latin name for **smallpox**, a now-eradicated deadly disease, which can resemble chickenpox. “Zoster” is the Greek word for girdle and “shingles” derives from “cingulum,” the Latin word for belt or girdle, which refer to the shingles lesions or blisters that form on one side of the waist. Scientists suspected as early as 1909 that chickenpox and shingles were caused by the same virus; this was confirmed in 1958.

Shingles is an infection of the central nervous system, particularly the dorsal root ganglia of the spine. From there the virus migrates through sensory nerve fibers to the skin, usually on the trunk, where it causes painful, fluid-filled eruptions or vesicles. Because the sensory nerves serve sharply bounded, non-overlapping areas of the skin called dermatomes, the shingles lesions appear within these dermatomes and do not cross the midline of the body.

Unlike chickenpox, shingles is not contagious because the virus is not usually in the lungs from which it could spread through the air. However the fluid-filled eruptions on the skin contain large amounts of virus, which can be transmitted through direct contact and infect a person, usually a child, who has not previously been exposed to VZV. The infected person will develop a case of primary chickenpox. A vaccine that prevents or ameliorates the symptoms of shingles became available in 2006. Immunization against chickenpox does not prevent shingles although it may reduce its incidence.

Risk factors

Anyone who has ever had chickenpox or been vaccinated against it is at risk for shingles. Overall approximately 20% of those who had chickenpox as children eventually develop shingles. Susceptibility to shingles appears to be genetically determined and the condition runs in families. The risk of shingles increases with age and with any condition that weakens the immune system. Those at particular risk for shingles include:

- children who had chickenpox in infancy or whose mothers had chickenpox late in pregnancy
- bone marrow and other transplant recipients
- those with compromised immune systems from diseases such as HIV/AIDS
- those with suppressed immune systems from chemotherapy drugs or other medications

Causes and symptoms

It is not clear why VZV reactivates to cause shingles, but it appears to be related to a decreased immune response due to advancing age, emotional or physical **stress**, **fatigue**, certain medications, **chemotherapy**, or diseases such as **cancer** or **HIV/AIDS**. Shingles is sometimes an early sign of **immunodeficiency** in people infected with HIV. In some cases the virus appears to be reactivated by mechanical irritation or minor surgical procedures.

Mild cases of shingles often go unnoticed. The earliest signs may be vague and can easily be mistaken for other illnesses. The condition may begin with **fever**, chills, gastrointestinal discomfort, and malaise (a vague feeling of weakness or discomfort). Lymph nodes may swell. Within two to four days, localized areas of intense **pain**, **itching**, and numbness/tingling (paresthesia) or extreme sensitivity to touch (hyperesthesia) can develop, usually on the trunk. The second most common place is on one side of the face around the eye (ophthalmic shingles) or on the forehead. However shingles can occur on the arms, legs, or elsewhere on the body. The pain may be continuous or intermittent, usually lasting from one to four weeks. The pain may accompany skin eruptions or precede the eruptions by days.

The red rash or oozing blisters appear along the course of the affected nerve. There is usually a vague streak or band from the spine along the path of the nerve on one side of the body. About five days after they appear, the vesicles begin to crust or scab and the disease resolves within the next two to three weeks. There may be no visible after effects or a slight scarring from the vesicles.

Shingles can be more debilitating in the elderly or those in poor health. The eruptions may be more extensive and inflammatory; they may also include bleeding blisters, areas of skin **death**, secondary bacterial infection, or extensive and permanent scarring. Ophthalmic shingles can cause painful eye infections and vision loss. Shingles infections within or near the ear can cause hearing or balance problems. Sometimes shingles can cause temporary or permanent **tremors** or **paralysis**; rarely, the condition spreads to the brain or spinal cord and causes **stroke** or **meningitis**.

Shingles pain usually subsides when the rash disappears, but it may last much longer, especially in the elderly. PHN can persist for months or years. It is caused by damage to the dorsal root ganglia, with the nerves becoming either spontaneously active—which is perceived as chronic pain—or hypersensitive to slight stimuli such as light touch. In the most severe cases, PHN can cause **insomnia**, weight loss, depression, and disability.

KEY TERMS

Acyclovir—An antiviral drug that is available in oral, intravenous, and topical forms and that blocks replication of the varicella zoster virus.

Antibody—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

Capsaicin—An active ingredient from hot chili peppers that is used in topical ointments to relieve pain. It appears to work by reducing the levels of a chemical substance involved in transmitting pain signals from nerve endings to the brain.

Corticosteroids—A group of hormones produced by the adrenal glands or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Famciclovir—An oral antiviral drug that blocks the replication of the varicella zoster virus.

Ganglion—A mass of nerve tissue outside of the central nervous system.

Immunocompromised—A weakened or poorly functioning immune system due to disease.

Immunosuppressed—Suppression of the immune system by medications during the treatment of diseases such as cancer or following an organ transplantation.

Post-herpetic neuralgia (PHN)—Long-lasting nerve pain caused by herpes zoster.

Tzanck preparation—A procedure in which skin cells from a blister are stained and examined under the microscope.

Valacyclovir—An oral antiviral drug that blocks the replication of the varicella zoster virus.

Vesicle—A small, raised lesion filled with clear fluid.

Diagnosis

Examination

Diagnosis of shingles is based on a medical history and **physical examination**. A definite diagnosis is difficult before eruption of the characteristic vesicles or bumps on the skin. The vesicles have a clear dermatome-bounded distribution usually on the midsection of the body.

Tests

Tests for shingles are rarely necessary but may include:

- polymerase chain reaction (PCR) testing for viral DNA
- viral culture of skin lesions
- a Tzanck preparation—stained cells from a blister, which will appear under the microscope to have many very large dark nuclei if infected with VZV
- a complete blood count (CBC) to test for elevated white blood cells that are indicative of infection
- blood serum levels of antibodies against VZV

Treatment

Traditional

Shingles almost always resolves spontaneously within a few weeks. Unless complicated by conditions such as HIV/AIDS or cancer, a primary care physician can provide treatment for easing painful symptoms. Rarely, transcutaneous **electrical nerve stimulation** (TENS) or a permanent nerve block is used to relieve the pain of PHN.

Drugs

The **antiviral drugs** acyclovir, valacyclovir, and famciclovir are used to treat shingles. These drugs can shorten the course of the illness. If started within 72 hours of the onset of the rash, antiviral therapy can heal the blisters more rapidly and sometimes even halt the disease. If taken after the disease has progressed, these drugs are less effective but may still lessen the pain. Antiviral drug treatment reduces the incidence of PHN by about one half and may also shorten its duration. Severely immunocompromised individuals, such as those with HIV/AIDS, may require intravenous administration of antiviral drugs or taking the drugs on an ongoing basis.

Various other drugs may be prescribed for shingles and PHN:

- corticosteroids, such as prednisone, to reduce inflammation from shingles, especially if the eye or other facial nerves are involved, and to reduce severe pain
- anticonvulsants such as pregabalin (Lyrica) or gabapentin to relieve pain
- the tricyclic antidepressants (TCAs) desipramine and nortriptyline
- opioid painkillers such as oxycodone, morphine, tramadol, or methadone
- tranquilizers or sedatives
- topical local anesthetics for application to the painful skin area and for post-herpetic itch; especially lidocaine, available as a cream, gel, spray, or patch

- capsaicin cream, which is available without a prescription but usually causes burning pain during application

Alternative

Alternative remedies and therapies will not cure shingles but they may relieve pain, reduce inflammation, and speed recovery:

- The amino acid lysine has also been reported to ease the symptoms of shingles and other herpes infections. Foods that are high in lysine include soybeans, black bean sprouts, lentils, parsley, and peas.
- Vitamin B12 supplementation during the first two days of the illness and ongoing vitamin B complex, vitamin C with bioflavonoids, and calcium supplements may boost the immune system.
- Echinacea can boost the immune system and help fight viral infections.
- Red pepper (*capsicum* or *cayenne*) is an ingredient in commercial ointments including *Zostrix* and *Capzasin-P*. It should be applied only to healed blisters and is useful for treating painful PHN. Seasoning food with red pepper may also provide relief.
- Calendula or licorice (*Glycyrrhiza glabra*) ointment or lotion may help treat shingles.
- Topical applications of lemon balm (*Melissa officinalis*), licorice, or peppermint (*Mentha piperita*) may reduce pain and blistering. These can also be consumed as teas.
- Sedative herbs such as passionflower can be brewed for a tea to treat PHN.
- Vervain helps relieve pain and inflammation.
- St. John's wort, lavender, chamomile, and marjoram help relieve inflammation.
- Homeopathic remedies include *Rhus toxicodendron* for blisters, *Mezereum* and *Arsenicum album* for pain, and *Ranunculus* for itching.
- Several drops of "Rescue Remedy" placed under the tongue or taken in water throughout the day are prescribed for relieving stress.
- Ayurvedic treatments for shingles include the application of turmeric paste.
- Acupuncture and acupressure can alleviate pain and PHN.
- Biofeedback or spinal cord stimulators may help relieve PHN.
- Relaxation techniques such as hypnotherapy and yoga may help relieve pain.
- Reflexology may help balance the body.

Practitioners of **traditional Chinese medicine** (TCM) may recommend herbal remedies:

- Chinese gentian root is used to treat the liver.
- Skullcap root in water is a Chinese folk remedy for shingles.
- Long Dan Xie Gan Tang is used to quell the accumulation of damp, toxic heat in the liver.
- For damp, infected, painful eruptions on the torso, Huang Qin Gao can be applied to the surrounding area.

Home remedies

Home remedies for shingles include plenty of rest, a healthy diet, regular **exercise**, and minimizing stress. The skin should be kept clean and contaminated items should not be reused. Cool compresses may help reduce pain from blisters. Blisters or crusting can be treated with compresses made with one-quarter cup (60 mL) of white vinegar in two quarts (1.9 L) of lukewarm water and applied twice daily for 10 minutes. The compresses should be discontinued when the blisters have dried up. Soothing baths and lotions with colloidal oatmeal, starch, or calamine may help to relieve itching and discomfort. If the skin becomes dry, tight, and cracked as the crusts and scabs separate, a small amount of plain petroleum jelly can be applied three or four times daily. The pain of PHN may be relieved with hot and cold compresses.

Prognosis

Shingles is almost never life-threatening in otherwise healthy patients and usually resolves without treatment in a few weeks. Because shingles boosts the immune response to VZV, repeat episodes are rare, occurring in less than 4% of patients. Although PHN usually diminishes over time, it can be disabling and difficult to treat.

Shingles can be much more severe in immunocompromised patients. The condition can last for months, recur frequently, and spread to the lungs, liver, gastrointestinal tract, brain, or other vital organs. Complications of shingles in immunocompromised or immunosuppressed patients may resemble those of primary varicella infection in adults, including viral **pneumonia**, male sterility, acute liver failure, and **birth defects** in children born to infected mothers. Depletion of CD4+ T lymphocytes in HIV/AIDS patients is associated with severe and chronic or recurrent VZV infection.

Prevention

A lifestyle that promotes immune system function and overall health may help prevent shingles. Factors include a well-balanced diet rich in essential **vitamins** and **minerals**, adequate sleep, regular exercise, and reduced stress. Patients with shingles should avoid contact with anyone who has not had chickenpox or been vaccinated against the disease, particularly pregnant women, newborns, and those with weakened immune systems.

In the United States it is now recommended that all children between 18 months and adolescence be immunized against chickenpox. Because a weakened (attenuated) form of the virus is used in this vaccine, it is thought that **vaccination** will reduce the likelihood of shingles later on in life. A single-dose vaccine against shingles (Zostavax) became available in 2006 and is recommended for most people aged 60 and older who have previously had chickenpox. It appears to prevent shingles in about 50% of vaccinated people and reduces the pain associated with shingles in others. It also can help prevent post-herpetic neuralgia.

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American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168, (847) 240-1280, (866) 503-SKIN (7546), (847) 240-1859, <http://www.aad.org>.

American Botanical Council, 6200 Manor Rd., Austin, TX, 78723, (512) 926-4900, (512) 926-2345, abc@herbalgram.org, <http://cms.herbgram.org>.

National Institute of Allergy and Infectious Diseases (NIAID), Office of Communications and Public Liaison, 6610 Rockledge Drive, Bethesda, MD, 20892-66123, (866) 284-4107, <http://www3.niaid.nih.gov>.

National Institute of Neurological Disorders and Stroke (NINDS), NIH Neurological Institute, PO Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov>.

National Shingles Foundation, 590 Madison Ave., 21st Floor, New York, NY, 10022, (212) 222-3390, (212) 222-8627, <http://www.vzvfoundation.org>.

U.S. Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800)-CDC-INFO (232-4636), cdcinfo@cdc.gov, <http://www.cdc.gov>.

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Shock

Definition

Shock is a medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This condition deprives organs and tissues of oxygen (carried in the blood) and allows the buildup of waste products. Shock can result in serious damage to the body, or even **death**.

Demographics

There are four grades of shock: Grade 1 causes up to 15% loss of effective blood volume or about 750 mL in the average adult; Grade 2 causes between 15–30% loss of blood volume or 750–1500 mL, provokes moderate tachycardia, and begins to narrow pulse pressure; Grade 3 causes about 30–40% loss of effective blood volume or 1500–2000 mL, compensatory mechanisms to fail, and **hypotension**, tachycardia, and low urine output to occur. Finally, Grade 4 causes about 40–50% loss of blood volume or 2000–2500 mL, and profound hypotension will develop and, if prolonged, will cause severe organ damage and death.

Description

There are various stages of shock: Stage I (also called compensated, or nonprogressive), Stage II (also called decompensated or progressive), and Stage III (also called irreversible).

In Stage I of shock, when low blood flow (perfusion) is first detected, a number of systems are activated in order to maintain and/or restore perfusion. The result is that the heart beats faster, blood vessels throughout the body become slightly smaller in diameter, and the kidney works to retain fluid in the circulatory system. All these changes serve to maximize blood flow to the most important organs and systems in the body. The patient in this stage of shock has very few symptoms, and treatment can completely halt any progression.

In Stage II of shock, these methods of compensation begin to fail. The systems of the body are unable to improve perfusion any longer, and the patient's symptoms reflect that fact. Oxygen deprivation in the brain causes the patient to become confused and disoriented, while oxygen deprivation in the heart may cause chest **pain**. With quick and appropriate treatment, this stage of shock can be reversed.

In Stage III of shock, the length of time that poor perfusion has existed begins to take a permanent toll on the body's organs and tissues. The heart's functioning continues to spiral downward and the kidneys usually shut down completely. Cells in organs and tissues throughout the body are injured and dying. The endpoint of Stage III shock is the patient's death.

Causes and symptoms

Shock is caused by three major categories of problems: cardiogenic (meaning problems associated with the heart's functioning); hypovolemic (meaning that the total volume of blood available to circulate is low); and **septic shock** (caused by overwhelming infection, usually by bacteria).

Cardiogenic shock can be caused by any disease or event that prevents the heart muscle from pumping strongly and consistently enough to circulate the blood normally. **Heart attack**, conditions that cause inflammation of the heart muscle (**myocarditis**), disturbances of the electrical rhythm of the heart, and any kind of mass or fluid accumulation and/or blood clot that interferes with flow out of the heart can all significantly affect the heart's ability to adequately pump a normal quantity of blood.

Hypovolemic shock occurs when the total volume of blood in the body falls well below normal. This condition can occur when there is excess fluid loss, as

in **dehydration** due to severe **vomiting** or **diarrhea**, diseases which cause excess urination (**diabetes insipidus**, **diabetes mellitus**, and kidney failure), extensive **burns**, blockage in the intestine, inflammation of the pancreas (**pancreatitis**), or severe bleeding of any kind.

Septic shock can occur when an untreated or inadequately treated infection (usually bacterial) is allowed to progress. Bacteria often produce poisonous chemicals (toxins), which can cause injury throughout the body. When large quantities of these bacteria, and their toxins, begin circulating in the bloodstream, every organ and tissue in the body is at risk of their damaging effects. The most serious consequences of these bacteria and toxins include poor functioning of the heart muscle; widening of the diameter of the blood vessels; a drop in blood pressure; activation of the blood clotting system, causing **blood clots**, followed by a risk of uncontrollable bleeding; damage to the lungs, causing acute **respiratory distress syndrome**; liver failure; kidney failure; and **coma**.

Initial symptoms of shock include cold, clammy hands and feet; pale or blue-tinged skin tone; weak, fast pulse rate; fast rate of breathing; low blood pressure. A variety of other symptoms may be present but they are dependent on the underlying cause of shock.

Diagnosis

Diagnosis of shock is based on the patient's symptoms as well as criteria including a significant drop in blood pressure, extremely low urine output, and blood tests that reveal overly acidic blood with a low circulating concentration of carbon dioxide. Other tests are performed, when appropriate, to try to determine the underlying condition responsible for the patient's state of shock.

Treatment

The most important goals in the treatment of shock include: quickly diagnosing the patient's state of shock; quickly intervening to halt the underlying condition (stopping bleeding, re-starting the heart, giving **antibiotics** to combat an infection, etc.); treating the effects of shock (low oxygen, increased acid in the blood, activation of the blood clotting system); and supporting vital functions (blood pressure, urine flow, heart function).

Treatment includes keeping the patient warm with legs raised and head down to improve blood flow to the brain; putting a needle in a vein in order to give fluids or blood transfusions, as necessary; giving the patient extra oxygen to breathe and medications to improve

KEY TERMS

- Cardiogenic**—Originating with the heart.
- Deprivation**—A condition of having too little of something.
- Hypovolemic**—Having a low volume.
- Perfusion**—Blood flow through an organ or tissue.
- Sepsis**—An overwhelming infection throughout the body, usually caused by bacteria in the bloodstream.

the heart's functioning; and treating the underlying condition that led to shock.

Prognosis

The prognosis of an individual patient in shock depends on the stage of shock when treatment was initiated, the underlying condition causing shock, and the general medical state of the patient.

Prevention

The most preventable type of shock is caused by dehydration during illnesses with severe **vomiting** or diarrhea. Shock can be avoided by recognizing that a patient who is unable to drink in order to replace lost fluids needs to be given fluids intravenously (through a needle in a vein). Other types of shock are only preventable insofar as one can prevent his or her underlying conditions, or can monitor and manage those conditions well enough so that they never progress to the point of shock.

Resources

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- Rapp J. et al., "Blood Vessel and Lymphatic Disorders." In: McPhee S., et al. *Current Medical Diagnosis and Treatment*, Los Altos, CA: McGraw-Hill, 2010.

Rosalyn Carson-DeWitt, MD
Karl Finley

Shock I see **Adult respiratory distress syndrome**

Shock therapy see **Electroconvulsive therapy**

Shortness of breath

Definition

Shortness of breath, or dyspnea, is a feeling of difficult or labored breathing that is out of proportion to the patient's level of physical activity. It is a symptom of a variety of different diseases or disorders and may be either acute or chronic.

Description

The experience of dyspnea depends on its severity and underlying causes. The feeling itself results from a combination of impulses relayed to the brain from nerve endings in the lungs, rib cage, chest muscles, or diaphragm, combined with the patient's perception and interpretation of the sensation. In some cases, the patient's sensation of breathlessness is intensified by **anxiety** about its cause. Patients describe dyspnea variously as unpleasant shortness of breath, a feeling of increased effort or tiredness in moving the chest muscles, a panicky feeling of being smothered, or a sense of tightness or cramping in the chest wall.

Causes and symptoms

ACUTE DYSPNEA. Acute dyspnea with sudden onset is a frequent cause of emergency room visits. Most cases of acute dyspnea involve pulmonary (lung and breathing) disorders, cardiovascular disease, or chest trauma.

PULMONARY DISORDERS. Pulmonary disorders that can cause dyspnea include airway obstruction by a foreign object, swelling due to infection, or anaphylactic shock; acute **pneumonia**; hemorrhage from the lungs; or severe bronchospasms associated with **asthma**.

CARDIOVASCULAR DISEASE. Acute dyspnea can be caused by disturbances of the heart rhythm, failure of the left ventricle, mitral valve (a heart valve) dysfunction, or an embolus (a clump of tissue, fat, or gas) that is blocking the pulmonary circulation. Most pulmonary emboli (**blood clots**) originate in the deep veins of the lower legs and eventually migrate to the pulmonary artery.

TRAUMA. Chest injuries, both closed injuries and penetrating **wounds**, can cause **pneumothorax** (the presence of air inside the chest cavity), **bruises**, or fractured ribs. **Pain** from these injuries results in dyspnea. The impact of the driver's chest against the steering wheel in auto accidents is a frequent cause of closed chest injuries.

OTHER CAUSES. Anxiety attacks sometimes cause acute dyspnea although they may or may not be associated

with chest pain. Anxiety attacks are often accompanied by hyperventilation, which is a breathing pattern characterized by abnormally rapid and deep breaths. Hyperventilation raises the oxygen level in the blood, causing chest pain and **dizziness**.

Chronic dyspnea

PULMONARY DISORDERS. Chronic dyspnea can be caused by asthma, **chronic obstructive pulmonary disease** (COPD), **bronchitis**, **emphysema**, inflammation of the lungs, **pulmonary hypertension**, tumors, or disorders of the vocal cords.

HEART DISEASE. Disorders of the left side of the heart or inadequate supply of blood to the heart muscle can cause dyspnea. In some cases a tumor in the heart or inflammation of the membrane surrounding the heart may cause dyspnea.

NEUROMUSCULAR DISORDERS. Neuromuscular disorders cause dyspnea from progressive deterioration of the patient's chest muscles. They include **muscular dystrophy**, **myasthenia gravis**, and **amyotrophic lateral sclerosis**.

OTHER CAUSES. Patients who are severely anemic may develop dyspnea if they **exercise** vigorously. **Hyperthyroidism** or **hypothyroidism** may cause shortness of breath, and so may **gastroesophageal reflux disease** (GERD). Chronic **anxiety disorders** or physical fitness can also cause episodes of dyspnea. Deformities of the chest or **obesity** can cause dyspnea by limiting the movement of the chest wall and the ability to fill the lungs completely.

Diagnosis

Patient history

The patient's history provides the doctor with such necessary information as a history of gastroesophageal reflux disease (GERD), asthma, or other allergic conditions; the presence of chest pain as well as difficulty breathing; recent accidents or recent surgery; information about **smoking** habits; the patient's baseline level of physical activity and exercise habits; and a psychiatric history of panic attacks or anxiety disorders.

ASSESSMENT OF BODY POSITION. How a person's body position affects his/her dyspnea symptoms sometimes gives hints as to the underlying cause of the disorder. Dyspnea that is worse when the patient is sitting up is called **platypnea** and indicates the possibility of **liver disease**. Dyspnea that is worse when the patient is lying down is called **orthopnea**, and is associated with heart disease or **paralysis** of the diaphragm. **Paroxysmal nocturnal dyspnea** (PND)

refers to dyspnea that occurs during sleep and forces the patient to awake gasping for breath. It is usually relieved if the patient sits up or stands. PND may point to dysfunction of the left ventricle of the heart, **hypertension**, or narrowing of the mitral valve.

Physical examination

The doctor will examine the patient's chest in order to determine the rate and depth of breathing, the effort required, the condition of the patient's breathing muscles, and any evidence of chest deformities or trauma. He or she will listen for **wheezing**, **stridor**, or signs of fluid in the lungs. If the patient has a **fever**, the doctor will look for other signs of pneumonia. The doctor will check the patient's heart functions, including blood pressure, pulse rate, and the presence of **heart murmurs** or other abnormal heart sounds. If the doctor suspects a blood clot in one of the large veins leading to the heart, he or she will examine the patient's legs for signs of swelling.

Diagnostic tests

BASIC DIAGNOSTIC TESTS. Patients who are seen in emergency rooms are given a **chest x ray** and electrocardiogram (ECG) to assist the doctor in evaluating abnormalities of the chest wall, also to determine the position of the diaphragm, possible rib **fractures** or pneumothorax, irregular heartbeat, or the adequacy of the supply of blood to the heart muscle. Also, the patient may be given a breathing test on an instrument called a spirometer to screen for airway disorders.

The doctor may order blood tests and arterial blood gas tests to rule out anemia, hyperventilation from an anxiety attack, or thyroid dysfunction. A **sputum culture** can be used to test for pneumonia.

SPECIALIZED TESTS. Specialized tests may be ordered for patients with normal results from basic diagnostic tests for dyspnea. High-resolution CT scans can be used for suspected airway obstruction or mild **emphysema**. Tissue biopsy performed with a bronchoscope can be used for patients with suspected lung disease.

If the doctor suspects a **pulmonary embolism**, he or she may order ventilation-perfusion scanning to inspect lung function, an angiogram of blood vessels, or ultrasound studies of the leg veins. **Echocardiography** can be used to test for pulmonary hypertension and heart disease.

Pulmonary function studies or **electromyography** (EMG) are used to assess neuromuscular diseases. Exercise testing is used to assess dyspnea related to COPD, anxiety attacks, poor physical fitness, and the severity of lung or heart disease. The level of acidity in

the patient's esophagus may be monitored to rule out GERD.

Treatment

Treatment of dyspnea depends on its underlying cause.

Acute dyspnea

Patients with acute dyspnea are given oxygen in the emergency room with the following treatments for specific conditions:

- Asthma. Treatment with Alupent, epinephrine, or aminophylline.
- Anaphylactic shock. Treatment with Benadryl, steroids, or aminophylline, with hydrocortisone if necessary.
- Congestive heart failure. Treatment with oxygen, diuretics, and placing patient in upright position.
- Pneumonia. Treatment with antibiotics and removal of lung secretions.
- Anxiety attacks. Immediate treatment includes anti-depressant medications. If the patient is hyperventilating, he or she may be asked to breathe into a paper bag to normalize breathing rhythm and the oxygen level of the blood.
- Pneumothorax. Surgical placement of a chest tube.

Chronic dyspnea

The treatment of chronic dyspnea depends on the underlying disorder. Asthma can often be managed with a combination of medications to reduce airway spasms and removal of allergens from the patient's environment. COPD requires both medication, lifestyle changes, and long-term physical **rehabilitation**. Anxiety disorders are usually treated with a combination of medication and **psychotherapy**. GERD can usually be managed with **antacids**, other medications, and dietary changes. There are no permanent cures for myasthenia gravis or muscular dystrophy.

Tumors and certain types of chest deformities can be treated surgically.

Alternative treatment

The appropriate alternative therapy for shortness of breath depends on the underlying cause of the condition. When dyspnea is acute and severe, **oxygen therapy** is used either in the doctor's office or in the emergency room. For shortness of breath with an underlying physical cause like asthma, anaphylactic shock, or pneumonia, the physical condition should be treated. Botanical and homeopathic remedies can be

KEY TERMS

Anaphylactic shock—A severe systemic reaction to an allergen that occurs in hypersensitive individuals. It can cause spasms of the larynx that block the patient's airway and cause dyspnea.

Dyspnea—A sensation of difficult or labored breathing.

Electromyography—A technique for recording electric currents in an active muscle in order to measure its level of function.

Orthopnea—Difficulty in breathing that occurs while the patient is lying down.

Paroxysmal nocturnal dyspnea (PND)—A form of dyspnea characterized by the patient's waking from sleep unable to breathe.

Platypnea—Dyspnea that occurs when the patient is sitting up.

Pneumothorax—The presence of air or gas inside the chest cavity.

Spirometer—An instrument that is used to test lung capacity. It is used to screen patients with dyspnea.

Stridor—A harsh or crowing breath sound caused by partial blockage of the patient's upper airway.

Wheezing—A whistling or musical sound caused by tightening of the air passages inside the patient's chest. Wheezing is most commonly associated with asthma.

used for acute dyspnea, if the proper remedies and formulas are prescribed. If the dyspnea has a psychological basis (especially if it is caused by anxiety), **acupuncture**, botanical medicine, and homeopathy can help the patient heal at a deep level.

Prognosis

The prognosis for recovery depends on the underlying cause of the dyspnea, its severity, and the type of treatment required.

Prevention

Dyspnea caused by asthma can be minimized or prevented by removing dust and other triggers from the patient's environment. Long-term prevention of chronic dyspnea includes such lifestyle choices as regular aerobic exercise and avoidance of smoking.

Resources

BOOKS

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Rebecca J. Frey, PhD

Shy-Drager syndrome

Definition

Shy-Drager syndrome (SDS) is a rare condition that causes progressive damage to the autonomic nervous system. The autonomic nervous system controls vital involuntary body functions such as heart rate, breathing, and intestinal, urinary, and sexual functions. The autonomic nervous system also controls skin and body temperature, and how the body responds to **stress**. Shy-Drager syndrome leads to **dizziness** or **fainting** when standing up, **urinary incontinence**, **impotence**, and muscle **tremors**.

Description

SDS was named for neurologists Milton Shy, M.D., from the National Institutes of Health, and Glenn Drager, M.D., from the Baylor College of Medicine, who first described the condition in 1960. It typically affects those between ages 50–70. It affects more men than women. In severe cases, the person cannot even stand up. Symptoms can be mild as well. Sometimes, people with mild cases are misdiagnosed as having **anxiety** or **hypertension**.

Many nonprescription drugs, such as cold medicines and diet capsules, can trigger extremely high blood pressure spikes in patients with SDS, even in very low doses. Therefore, these patients are at risk for strokes and excessive bleeding (hemorrhage) if they take even the recommended dosage of these drugs.

Causes and symptoms

The cause of SDS is unknown. Symptoms develop because of degeneration of certain groups of nerve cells in the spinal cord.

Patients with SDS usually have problems with the function of the autonomic nervous system. Progressive degeneration may occur in other areas of the nervous system as well. The hallmark of the syndrome is dizziness and fainting when arising or after standing still for a long time (postural **hypotension**). This is

caused by low blood pressure and inadequate blood flow to the brain. When this problem becomes severe (for example, a blood pressure below 70/40 mmHg), it can lead to a momentary loss of consciousness. After the person faints, the blood pressure returns to normal and the person wakes up.

Many patients also notice impotence, urinary incontinence, **dry mouth** and skin, and trouble regulating body temperature because of abnormal sweating. Since the autonomic nervous system also controls the narrowing and widening of the iris, some patients with SDS have vision problems, such as trouble focusing.

In later stages, problems in the autonomic nervous system lead to breathing difficulties such as **sleep apnea**, loud breathing, and **snoring**. In advanced stages of the disease, patients can die from irregular heartbeat.

Other symptoms of SDS do not involve the autonomic nervous system. These include parkinsonism (muscle tremor, rigidity, and slow movements), double vision, problems controlling emotions, and wasting of muscles in the hands and feet. Eventually, patients may have problems chewing, swallowing, speaking, and breathing. There may be a loss of color pigment in the iris.

Diagnosis

While no blood test can reveal the disorder, a careful assessment of symptoms should alert a neurologist to suspect SDS. A combination of parkinsonism and certain autonomic problems (especially impotence, incontinence, and postural hypotension) are clear indications of the syndrome.

Tests of the autonomic nervous system may help diagnose the condition. In normal patients, blood levels of norepinephrine rise when they stand up. This doesn't happen in people with SDS. Norepinephrine is a hormone that helps maintain blood pressure by triggering certain blood vessels to constrict when blood pressure falls below normal. Another test for the condition is the **Valsalva maneuver**. In this test, the patient holds his or her breath and strains down as if having a bowel movement while the doctor monitors blood pressure and heart rate for 10 seconds. Patients with SDS will not have the normal increase in blood pressure and heart rate.

A variety of other tests can identify a broad range of autonomic problems in patients with SDS. Brain scans, however, do not usually reveal any problems.

Treatment

Medication can relieve many of the symptoms, especially the parkinsonism and low blood pressure.

KEY TERMS

Autonomic nervous system—The part of the nervous system that controls the involuntary (apparently automatic) activities of organs, blood vessels, glands, and many other body tissues.

Degenerative—Degenerative disorders involve progressive impairment of both the structure and function of part of the body.

Gastrostomy—An artificial opening into the stomach through the abdomen to enable a patient to be fed via a feeding tube. The procedure is given to patients with SDS who are unable to chew or swallow.

Norepinephrine—A hormone that helps maintain blood pressure by triggering certain blood vessels to constrict when blood pressure falls below normal.

Sleep apnea—A sleep disorder characterized by periods of breathing cessation lasting for 10 seconds or more.

Tracheostomy—An opening through the neck into the trachea through which a tube may be inserted to maintain an effective airway and help a patient breathe.

However, typical antiparkinsonism drugs such as carbidopa-levodopa (Sinemet) should be used with caution, since they often worsen the postural low blood pressure and may cause fainting.

Because postural hypotension is the most troublesome of the symptoms in the early years, treatments center on relieving this problem. Patients are encouraged to eat a liberal salt diet and drink plenty of fluids. They are advised to wear waist-high elastic hosiery and to sleep with the head elevated at least 5 inches (13 centimeters). Other drug treatment includes fludrocortisone, indomethacin, **nonsteroidal anti-inflammatory drugs, beta blockers**, central stimulants, and other medications.

Occasionally, a pacemaker, **gastrostomy**, or tracheostomy may be needed. A pacemaker is a device that delivers electrical impulses to the heart to keep it beating regularly. A gastrostomy creates an opening in the stomach to connect a feeding tube from outside the body. In a tracheostomy an opening is made in the windpipe and a tube is inserted to maintain breathing.

Prognosis

While the course of the disease varies, and some patients live for up to 20 years after the symptoms first appear, most patients become severely disabled within 7 or 8 years. It is unusual for someone to survive more than 15 years after diagnosis.

Symptoms (especially tremor) often get worse if the patient smokes because of the nicotine.

Many patients develop swallowing problems which may lead to recurrent episodes of **pneumonia**, a frequent cause of **death**. Others experience Cheyne-Stokes (periodic breathing). One of the most common causes of death is pulmonary embolus. This is caused by a blood clot in the main artery in the lung.

Prevention

Since scientists do not know the cause of Shy-Drager syndrome, there is no way to prevent the condition.

ORGANIZATIONS

American Academy of Neurology, 1080 Montreal Ave., St. Paul, MN, 55116, (651) 695-2717, (651) 695-2791, (800) 879-1960, memberservices@aan.com, <http://www.aan.com/>.

Association for Neuro-Metabolic Disorders, 5223 Brookfield Lane, Sylvania, OH, 43560-1809, (419) 885-1809.

National Institute of Neurological Disorders and Stroke (NINDS), NIH Neurological Institute, P. O. Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov/>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

SDS/MSA Support Group, 8311 Brier Creek Parkway, Suite 105-434, Raleigh, NC, 27617, (866) 737-5999, vjames@shy-drager.org, <http://www.shy-drager.org>.

Carol A. Turkington

Shyness

Definition

Shyness is a personality trait that produces behaviors ranging from feeling uncomfortable at a party to an extreme fear of being watched by others while talking on the telephone.

Description

Shyness affects people of all ages. A toddler might run from strangers and cling to his or her parents. While kindergarten is frightening for many children, some students are anxious about the first day of school until they graduate from college. Job interviews are stressful for people uncomfortable talking about themselves. For some people, feelings of self-worth are related to their careers. Retirement may bring feelings of lower self-esteem.

Shyness is linked to brain activity, how a person was raised and other experiences, and the person's reaction to those experiences.

Social phobia

Extreme shyness is sometimes referred to as a social phobia. Also known as social **anxiety** disorder, a social phobia is a psychiatric condition defined as a "marked and persistent fear" of some situations. Social phobia may cause a person to remain unemployed, according to the National Mental Health Association (NMHA). True social phobia affects about 3% of people.

Introversion

The introvert enjoys being alone and intentionally avoids situations like parties. The shy person wants to be around people. However, shyness is stronger than the desire to be sociable. The shy person is afraid to go to the party and stays home alone.

Causes and symptoms

Temperament is related to the amygdala, the part of the brain related to emotions and new situations. The amygdala evaluates new situations based on memories of past experiences. If the new situation appears threatening, the amygdala sends a warning signal. The amygdala in a shy person is extremely sensitive and much more active than that of an outgoing person. The increased activity causes the person to withdraw either physically or emotionally. This withdrawal is known as inhibition.

The baby runs from strangers; the job applicant laughs nervously when talking about his accomplishments. Brain activity is one component of shyness. Environment also plays a role. If the inhibited child has outgoing, nurturing parents, she will probably imitate their behavior. If parents and teachers are mocking and critical, a child may have a lifelong fear of the first day of school. A person with that background may compare him or herself with others and feel they are

more capable than he or she is. The person embarrassed in a job interview could become anxious in future interviews.

At the root of shyness is a feeling of self-consciousness. This may cause the person to blush, tense up, or start sweating. Those are some reactions caused when the brain signals its warning. The person may avoid eye contact, look down, become very quiet, or fumble over words.

Symptoms vary because there are degrees of shyness. A person might be very quiet when meeting new people, but then become talkative when she feels comfortable with them. The jobseeker may not be afraid of social gatherings.

Social Phobia

Social phobia causes an extreme fear of being humiliated or embarrassed in front of people, according to the NMHA. It may be connected to low self-esteem or feelings of inferiority. The phobic is not fearful in all situations and may feel comfortable around people in most of the time.

However, social **phobias** have caused people to drop out of school, avoid making friends, and keep away from other fear-provoking situations. Phobic fears range from speaking in public and dating to using public restrooms or writing when other people are present.

According to the NMHA, a phobic may feel that everyone is looking at them. A trivial mistake is regarded as much more serious and blushing is painfully embarrassing. Social phobia is frequently accompanied by depression or **substance abuse**.

Diagnosis

In many cases, adults realize they are shy. In a sense they have diagnosed themselves, and may take steps to overcome their shyness. Teenagers may also try to remedy their situations.

Adults and youths may buy self-help books or take classes on subjects like overcoming shyness and assertiveness training. These classes may be taught by counselors, psychologists, or people with experience conquering shyness. Health-care providers often schedule these classes. They are also taught in settings ranging from adult schools to social service agencies. Costs will vary at these classes.

Children may not know there are treatment solutions for their shyness. Parents and educators should be alert for symptoms of shyness in younger children. Schools and family resource centers can provide referrals if it appears counseling will help get their child diagnosed.

Medical diagnosis

Based on the child's circumstances, parents may take the child to their health care provider. Some insurance plans require an appointment with a doctor before a referral to a counselor or a psychologist. The health professional conducts an assessment and then recommends treatment.

Children and adults may need medical treatment for social phobia. The adult's diagnosis also starts with a medical exam to determine if there is a physical cause for symptoms. If that has been ruled out, the patient undergoes a psychiatric evaluation.

Diagnostic fees and the time allocated for evaluation vary for both shy and phobic people. Diagnosis could span several hour-long sessions that cover an initial evaluation, personality tests, and a meeting to set therapy goals. Each session could cost around \$90. Insurance may cover part of the costs.

Treatment

Shyness treatment concentrates on changing behavior so the person feels more at ease in shyness-provoking situations. The person may be guided by a self-help book or participate in individual or **group therapy**.

Books and therapy generally focus on behavioral therapy and **cognitive-behavioral therapy**. One method of behavioral therapy is to expose the person to the situation that triggers fear. This could start with rehearsing a job interview with a friend or making eye contact with a store clerk. Over time, the person goes on interviews to get experience rather than to be hired. Another person might move from eye contact to attending an enjoyable event like a concert to become more at ease around strangers.

Therapy also focuses on developing skills to cope in new situations. These include taking deep breaths to relax and practicing small talk. Cognitive-therapy helps the person learn how thinking patterns contribute to symptoms, according to NMHA. The person is taught techniques to change those thoughts or stop symptoms. This association maintains this therapy is very effective for people with social phobias.

Treatment costs vary from the price of a self-help book to the fees for therapy. Therapy sessions may be led by a licensed marriage and family counselor, a psychologist or psychiatrist. The cost of group therapy is generally an hourly fee with therapy planned for a set time. The therapist might charge \$80 an hour for a social phobia group that meets 3 hours a week for 16 weeks.

Treatment may include medication. Prescription drugs like Paxil (paroxetine) are generally only prescribed to people with social **anxiety disorders**. Paxil is prescribed for depression and other **mood disorders**. The patient takes one tablet daily. Costs will vary, and a 30-day order could be priced at \$74 to \$84.

Insurance may cover part of the costs of therapy and medicine.

Alternative treatment

Alternative treatments for shyness focus on symptoms like tension and **stress**. Relaxation tapes and CDs guide the listener through a series of actions to relieve tension. The activity starts with deep breathing and then the person progressively focuses on the head and different parts of the body. The exercise may start with the head, neck, shoulders, moving down to the one foot and then the other. Some techniques involve tightly tensing and then releasing each part. Another method is to concentrate on relaxing each part or imagine that it becomes warm.

Another self-treatment is **aromatherapy**. Lavender is a relaxing scent and is available in liquid form as an essential oil. Stress can be relieved by adding oil to a bath. Some people carry the oil with them. If they become anxious, the people can dab the oil on a cotton pad. They breathe in the lavender and feel calmer.

Prognosis

Shyness may not be a permanent. Children often outgrow shyness. Behavioral changes and therapy can help people feel more at ease. Furthermore, some aspects of shyness are positive. Shy people are frequently good listeners and are empathetic, aware of others' feelings.

Prevention

Shyness is a personality trait related to a person's biology and experiences. The part of shyness related to the brain cannot be changed. However, parents can provide a nurturing environment that helps prevent shyness. This will provide the child with a healthy mental attitude that helps prevent shyness. When faced with situations that could cause self-defeating shyness, children will have coping skills.

According to the National Mental Health Association, the basics of good mental health for children include:

- A family that provides unconditional love not related on accomplishments.
- Nurturing self-confidence and high self-esteem by praising children. Methods include encouraging a child to

learn a new game. The parents should set realistic goals, assure children, and smile frequently. Parents should avoid sarcastic remarks, set realistic goals and let children know that all people make mistakes.

- Playing with other children helps the young learn how to develop friendships and problem-solving skills.
- Emphasizing that school is fun. Parents can play school with their child to demonstrate that learning is enjoyable. Enrolling children in preschool or children's programs allows them to learn, be creative, and develop social skills.
- When disciplining, parents should criticize the behavior, rather than berating the child.

Shyness prevention and adults

For adults prone to shyness, the issue is related more to treatment than prevention. Shyness for these people has probably been an issue, one that surfaces at various times in their lives. A move, a **death** in the family, job loss, and other unsettling changes could cause emotions that include the fear associated with shyness.

In some circumstances, the person must go through the grieving process. In other situations, the person needs to do things that build self-confidence. Like the child, the adult needs a support system. A network of friends helps with encouragement and listens to the person's concerns.

To combat the avoidance symptom caused by shyness, the person should look into enjoyable pursuits. Recreational activities like walking groups combine physical exercise with the opportunity to socialize. Enrolling in a class at an adult school or community college provides the opportunity to learn and make new friends. Class topics range from upholstery to mystery book discussions. Classes like these can boost confidence as a person learns a hands-on skill or discovers that other mystery readers value her or his opinion.

Resources

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ORGANIZATIONS

- American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (202) 336-5500, (800) 374-2721, <http://www.apa.org/>.
- Mental Health America, 2000 N. Beauregard Street, 6th Floor, Alexandria, VA, 22311, (703) 684-7722, (703) 684-5968, (800) 969-6642, infoctr@mentalhealthamerica.net, <http://www.mentalhealthamerica.net>.
- Shyness Research Institute, 4201 Grant Line Rd., New Albany, IN, 47150, (812) 941-2295, (812) 941-2591, bearducc@ius.edu, <http://homepages.ius.edu>.

Liz Swain

Sick sinus syndrome

Definition

Sick sinus syndrome is a disorder of the sinus node of the heart, which regulates heartbeat. With sick sinus syndrome, the sinus node fails to signal properly, resulting in changes in the heart rate.

Description

The sinus node in the heart functions as the heart's pacemaker, or beat regulator. In sick sinus syndrome, patients normally will experience bradycardia, or slowed heart rate. Also, it is not uncommon to see fluctuations between slow and rapid heart rate (tachycardia). This makes the diagnosis and treatment of sick sinus syndrome more complicated than most other cardiac **arrhythmias** (irregular heart beats). A sick sinus node may be responsible for starting beats too slowly, pausing too long between initiation of heartbeats, or not producing heartbeats at all.

Causes and symptoms

Sick sinus syndrome may be brought on by the use of certain drugs, but is most common in elderly patients. Cardiac **amyloidosis**, a condition in which amyloid, a kind of protein, builds up in heart tissue, may affect the sinus node. Other conditions, such as **sarcoidosis** (round bumps in the tissue surrounding the heart and other organs), **Chagas' disease** (resulting from the bite of a bloodsucking insect), or certain cardiac **myopathies** can cause fiber-like tissue to grow around the normal sinus node causing the node to malfunction.

A patient may not show any symptoms of sick sinus syndrome. In general the common symptoms are those associated with slow heart rate, such as light-headedness, or **dizziness**, **fatigue** and **fainting**. Patients may

also experience confusion, heart **palpitations, angina**, or **heart failure**.

Diagnosis

A slow pulse, especially one that is irregular, may be the first indication of sick sinus syndrome. **Electrocardiography (ECGs)** is a commonly used method of detecting sick sinus syndrome. ECG monitoring for 24 hours is most useful, since with this syndrome, the heart rate may alternate between slow and fast and the determination of this fact can help differentiate sick sinus syndrome from other arrhythmias.

Treatment

If drugs are causing the problem, their withdrawal may effectively eliminate the disorder. However, the treatment of sick sinus syndrome is normally delayed until a patient shows symptoms. Once treatment is indicated, most patients will receive a pacemaker. This is a permanent treatment involving implantation of a small device under the skin below the collarbone. Small electrodes run from the device to the heart; they deliver and regulate the electrical signals that cause the heart to beat. Patients with sick sinus syndrome should generally receive dual chamber pacing systems to prevent **atrial fibrillation** (involuntary contraction of the muscles of the atria). Some drugs are used to treat sick sinus syndrome, but digitalis should be used with caution. Often the use of drugs to regulate the heartbeat should be implemented only after the pacemaker has been placed, since these drugs may further worsen the slow heart rate.

Alternative treatment

The reduction or elimination of certain foods and substances, such as alcohol or **caffeine**, may be advised to control heart rate. **Stress reduction** may also assist with changes in rate. Homeopathic treatment can work on a deep healing level, while **acupuncture** and botanical medicine can offer supportive treatment of symptoms.

Prognosis

Patients with sick sinus syndrome lead relatively normal lives if the disorder is controlled by a pacemaker. However, in some patients, the pacemaker does not adequately control the fluctuations in heart rate. If left untreated, or in severe cases, the heart could stop beating.

Prevention

Elimination of a drug therapy which aggravates sick sinus syndrome is the first line of treatment for

KEY TERMS

Arrhythmia—Irregular heart beat.

Atria—Plural for atrium. The atria are the upper chambers of the heart.

Bradycardia—A heart rate slower than normal.

Electrocardiograph (ECG)—A test of a patient's heartbeat that involves placing leads, or detectors, on the patient's chest to record electrical impulses in the heart. This test will produce a strip, or picture record of the heart's electrical functioning.

Myopathy—Weakness of muscle.

Pacemaker—A device implanted under the skin, below the collarbone, to regulate heartbeat. Leads from the device to the heart stimulate the electrical functions of the heart. Pacemakers are often used to control bradycardia and are usually smaller than a silver dollar.

some patients. Other causes of the syndrome are not preventable. However, proper treatment of those underlying conditions which affect the tissues of the heart may intervene to prevent sick sinus syndrome from becoming a significant problem.

Resources

OTHER

"Sick Sinus Syndrome." Heart Rhythm Society. [Accessed December 3, 2010] http://www.hrspatients.org/patients/heart_disorders/sick_sinus/default.asp.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review. personal.info@heart.org.

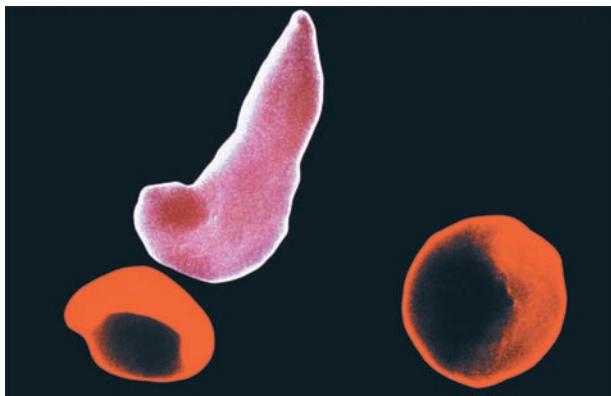
National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

Teresa Odle

Sickle cell disease

Definition

Sickle cell disease describes a group of inherited blood disorders characterized by chronic anemia, painful events, and various complications due to associated tissue and organ damage.



A scanning electron microscopy (SEM) scan of red blood cells taken from a person with sickle cell anemia. The blood cells at the bottom are normal; the diseased, sickle-shaped cells appear at the top. (Dr. Gopal Murti/Photo Researchers, Inc.)

Demographics

As of 2009, the National Heart, Lung and Blood Institute (NHLBI) estimates that sickle cell disease affects about 70,000 people in the United States. It mainly affects African Americans. The disease occurs in about 1 out of every 500 African American births. Sickle cell disease also affects Hispanic Americans, occurring in 1 out of every 36,000 births. Approximately 2 million Americans have the sickle cell trait. The condition occurs in about 1 in 12 African Americans. Sickle cell disease affects millions of people worldwide. Incidence is higher in people who come from Africa, South or Central America (Panama), the Caribbean islands, Mediterranean countries (Turkey, Greece, and Italy), India, and Saudi Arabia.

Description

Carriers of the sickle cell gene are said to have sickle cell trait. Unlike sickle cell disease, sickle cell trait does not cause health problems. In fact, sickle cell trait is protective against **malaria**, a disease caused by blood-borne parasites transmitted through mosquito **bites**. According to a widely accepted theory, the genetic mutation associated with the sickle cell trait occurred thousands of years ago. Coincidentally, this mutation increased the likelihood that carriers would survive malaria infection. Survivors then passed the mutation to their offspring and the trait became established throughout areas where malaria was common. As populations migrated, so did the sickle cell trait.

The most common and well-known type of sickle cell disease is sickle cell anemia, also called SS disease. All types of sickle cell disease are caused by a genetic

change in hemoglobin, the oxygen-carrying protein inside the red blood cells. The red blood cells of affected individuals contain a predominance of a structural variant of the usual adult hemoglobin. This variant hemoglobin, called sickle hemoglobin (Hb-S), has a tendency to develop into rod-like structures that alter the shape of the usually flexible and round red blood cells. The cells take on a shape that resembles the curved blade of a sickle, an agricultural tool. Sickle cells have a shorter life span than normally-shaped red blood cells. This results in chronic anemia characterized by low levels of hemoglobin and decreased numbers of red blood cells. Sickle cells are also less flexible and more sticky than normal red blood cells and can become trapped in small blood vessels preventing blood flow. This compromises the delivery of oxygen, which can result in **pain** and damage to associated tissues and organs. Sickle cell disease presents with marked variability, even within families.

Risk factors

In each **pregnancy** of two parents who both have sickle cell trait, there is a 50% chance that the child will have the trait, a 25% chance that the child will have sickle cell disease, and a 25% chance that the child will have neither the trait nor the disease.

Causes and symptoms

Humans normally make several types of the oxygen-carrying protein hemoglobin. An individual's stage in development determines whether he or she makes primarily embryonic, fetal, or adult hemoglobins. All types of hemoglobin are made of three components: heme, alpha (or alpha-like) globin, and beta (or beta-like) globin. Sickle hemoglobin is the result of a genetic change in the beta globin component of normal adult hemoglobin. The beta globin gene is located on chromosome 11. The sickle cell form of the beta globin gene results from the substitution of a single DNA nucleotide, or genetic building-block. The change from adenine to thymine at codon (position) 6 of the beta globin gene leads to insertion of the amino acid valine—instead of glutamic acid—at this same position in the beta globin protein. As a result of this change, sickle hemoglobin has unique properties in comparison to the usual type of adult hemoglobin.

Most individuals have two normal copies of the beta globin gene, which make normal beta globin that is incorporated into adult hemoglobin. Individuals who have sickle cell trait (called sickle cell carriers) have one normal beta globin gene and one sickle cell gene. These individuals make both the usual adult hemoglobin and sickle hemoglobin in roughly equal proportions, so they do not experience any health problems as a result

of having the trait. Although traces of blood in the urine and difficulty in concentrating the urine can occur, neither represents a significant health problem as a result of sickle cell trait. Of the millions of people with sickle cell trait worldwide, a small handful of individuals have experienced acute symptoms. In these very rare cases, individuals were subject to very severe physical strain.

Other types of sickle cell disease include SC disease, SD disease, and S/beta **thalassemia**. These conditions are caused by the co-inheritance of the sickle cell gene and another altered beta globin gene. For example, one parent may have sickle cell trait and the other parent may have hemoglobin C trait (another hemoglobin trait that does not cause health problems). For this couple, there would be a 25% chance of SC disease in each pregnancy.

Symptoms

Normal adult hemoglobin transports oxygen from the lungs to tissues throughout the body. Sickled hemoglobin can also transport oxygen. However, once the oxygen is released, sickled hemoglobin tends to polymerize (line-up) into rigid rods that alter the shape of the red blood cell. Sickling of the red blood cell can be triggered by low oxygen, which occurs in organs with slow blood flow. It can also be triggered by cold temperatures and **dehydration**.

Sickle cells have a decreased life span in comparison to normal red blood cells. Normal red blood cells survive for approximately 120 days in the bloodstream; sickle cells last only 10–12 days. As a result, the bloodstream is chronically short of red blood cells and hemoglobin and the affected individual develops anemia.

Sickle cells can create other complications. Due to their shape, they do not fit well through small blood vessels. As an aggravating factor, the outside surfaces of sickle cells may have altered chemical properties that increase the cells' stickiness. These sticky sickle cells are more likely to adhere to the inside surfaces of small blood vessels, as well as to other blood cells. As a result of the sickle cells' shape and stickiness, blockages form in small blood vessels. Such blockages prevent oxygenated blood from reaching areas where it is needed, causing pain as well as organ and tissue damage.

The severity of symptoms cannot be predicted based solely on the genetic inheritance. Some individuals with sickle cell disease develop health- or life-threatening problems in infancy but others may have only mild symptoms throughout their lives. Individuals may experience varying degrees of health at different stages in the lifecycle. For the most part, this clinical

variability is unpredictable and the reasons for the observed variability cannot be determined. However, certain types of sickle cell disease (i.e. SC disease) tend to result in fewer and less severe symptoms on average than other types of sickle cell disease (i.e. SS disease). Some additional modifying factors are known. For example, elevated levels of fetal hemoglobin in a child or adult can decrease the quantity and severity of some symptoms and complications. Fetal hemoglobin is a normally occurring hemoglobin that usually decreases from over 90% of the total hemoglobin to under 1% during the first year of life. This change is genetically determined, although some individuals may experience elevated levels of fetal hemoglobin due to variation in the genes that control fetal hemoglobin production. Such individuals often experience a reduction in their symptoms and complications due to the ability of fetal hemoglobin to prevent the polymerization of sickle hemoglobin, which leads to sickling of the red blood cell.

There are several symptoms that warrant immediate medical attention, including the following:

- Signs of infection (fever 101°F or 38.3°C, coughs frequently or breathing trouble, unusual crankiness, feeding difficulties)
- Signs of severe anemia (pale skin or lips, yellowing of the skin or eyes, very tired, very weak)
- Signs indicating possible dehydration (vomiting, diarrhea, fewer wet diapers)
- Other signs (pain or swelling in the abdomen, swollen hands or feet, screams when touched).

These can be signs of various complications that occur in sickle cell disease.

INFECTIONS AND EFFECTS ON THE SPLEEN. Children with sickle cell disease who are under age three are particularly prone to life-threatening bacterial infections. *Streptococcus pneumoniae* is the most common offending bacteria and invasive infection from this organism leads to **death** in 15% of patients. The spleen, an organ that helps to fight bacterial infections, is particularly vulnerable to the effects of sickling. Sickled cells can impede blood flow through the spleen causing organ damage, which usually results in loss of spleen function by late childhood. The spleen can also become enlarged due to blockages and/or increased activity of the spleen. Rapid enlargement of the spleen may be a sign of another complication called *splenic sequestration*, which occurs mostly in young children and can be life-threatening. Widespread sickling in the spleen prevents adequate blood flow from the organ, removing

increasing volumes of blood from the circulation and leading to accompanying signs of severe anemia.

PAINFUL EVENTS. Painful events, also known as *vaso-occlusive events*, are a hallmark symptom of sickle cell disease. The frequency and duration of the pain can vary tremendously from person to person and over an individual's lifecycle. Painful events are the most common cause of hospitalizations in sickle cell disease. However, only a small portion of individuals with sickle cell disease experience frequent and severe painful events. Most painful events can be managed at home. Pain results when small blood vessel blockages prevent oxygen from reaching tissues. Pain can affect any area of the body although the extremities, chest, abdomen, and bones are frequently affected sites. There is some evidence that cold temperatures or infection can trigger a painful event but most events occur for unknown reasons. The hand-foot syndrome, or *dactylitis*, is a particular type of painful event. Most common in toddlers, dactylitis results in pain and swelling in the hands and feet, sometimes accompanied by a **fever**.

ANEMIA. Sickle cells have a high turnover rate leading to a deficit of red blood cells in the bloodstream. Common symptoms of anemia include **fatigue**, paleness, and a **shortness of breath**. A particularly severe form of anemia—aplastic anemia—occurs following infection with parvovirus. Parvovirus causes extensive destruction of the bone marrow, bringing production of new red blood cells to a halt. Bone marrow production resumes after 7 to 10 days; however, given the short lives of sickle cells, even a brief shutdown in red blood cell production can cause a rapid decline in hemoglobin concentrations.

DELAYED GROWTH. The energy demands of the bone marrow for red blood cell production compete with the demands of a growing body. Children with sickle cell anemia may have delayed growth and reach **puberty** at a later age than normal. By early adulthood, they catch up on growth and attain normal height; however, weight typically remains below average.

STROKE. Children with sickle cell disease have a significantly elevated risk of having a **stroke**, which can be one of the most concerning complications of sickle cell disease. Approximately 11% of individuals with sickle cell disease will have a recognizable stroke by the age of 20. **Magnetic resonance imaging** studies have found that 17% of children with sickle cell anemia have evidence of a previous stroke or clinically 'silent' stroke-like events called *transient ischemic events*. Stroke in sickle cell disease is usually caused by a blockage of a blood vessel but about one fourth of the time may be caused by a hemorrhage (or rupture) of a blood vessel.

Strokes result in compromised delivery of oxygen to an area of the brain. The consequences of stroke can range from life-threatening, to severe physical or cognitive impairments, to apparent or subtle learning disabilities, to undetectable effects. Common stroke symptoms include weakness or **numbness** that affects one side of the body, sudden behavioral changes, loss of vision, confusion, loss of speech or the ability to understand spoken words, **dizziness**, **headache**, seizures, **vomiting**, or even **coma**.

Approximately two-thirds of the children who have a stroke will have at least one more. Transfusions have been shown to decrease the incidence of a second stroke. A recent study showed that children at highest risk to experience a first stroke were 10 times more likely to have a stroke if untreated when compared to high-risk children treated with chronic blood **transfusion** therapy. High-risk children were identified using transcranial doppler ultrasound technology to detect individuals with increased blood flow speeds due to constricted intracranial blood vessels.

ACUTE CHEST SYNDROME. Acute chest syndrome (ACS) is a leading cause of death for individuals with sickle cell disease, and recurrent attacks can lead to permanent lung damage. Therefore, rapid diagnosis and treatment is of great importance. ACS can occur at any age and is similar but distinct from **pneumonia**. Affected persons may experience fever, **cough**, chest pain, and shortness of breath. ACS seems to have multiple causes including infection, sickling in the small blood vessels of the lungs, fat embolisms in the lungs, or a combination of factors.

PRIAPIST. Males with sickle cell anemia may experience **priapism**, a condition characterized by a persistent and painful erection of the penis. Due to blood vessel blockage by sickle cells, blood is trapped in the tissue of the penis. Priapism may be short in duration or it may be prolonged. Priapism can be triggered by low oxygen (hypoxemia), alcohol consumption, or sexual intercourse. Since priapism can be extremely painful and result in damage to this tissue causing **impotence**, rapid treatment is essential.

KIDNEY DISEASE. The environment in the kidney is particularly prone to damage from sickle cells. Signs of kidney damage can include blood in the urine, incontinence, and enlarged kidneys. Adults with sickle cell disease often experience insufficient functioning of the kidneys, which can progress to kidney failure in a small percentage of adults with sickle cell disease.

JAUNDICE AND GALLSTONES. **Jaundice** is indicated by a yellow tone in the skin and eyes and alone it is not a health concern. Jaundice may occur if bilirubin levels increase, which can occur with high levels of red blood

cell destruction. Bilirubin is the final product of hemoglobin degradation, and is typically removed from the bloodstream by the liver. Therefore, jaundice can also be a sign of a poorly functioning liver, which may also be evidenced by an enlarged liver. Increased bilirubin also leads to increased chance for **gallstones** in children with sickle cell disease. Treatment, which may include removal of the gall bladder, may be selected if the gallstones start causing symptoms.

RETINOPATHY. The blood vessels that supply oxygen to the retina—the tissue at the back of the eye—may be blocked by sickle cells, leading to a condition called retinopathy. This is one of the only complications that is actually more common in SC disease as compared to SS disease. Retinopathy can be identified through regular ophthalmology evaluations and effectively treated in order to avoid damage to vision.

Joint problems

Avascular necrosis of the hip and shoulder joints, in which bone damage occurs due to compromised blood flow due to sickling, can occur later in childhood. This complication can affect an individual's physical abilities and result in substantial pain.

Diagnosis

Examination

Sickle cell disease is typically diagnosed through genetic screening done when a baby is born. If sickle cell disease is diagnosed, the parents are referred to a doctor who specializes in blood disorders (hematologist) or a pediatric hematologist.

Tests

Testing for sickle cell is performed to identify the presence of hemoglobin S, and the presence of other abnormal hemoglobins. A **complete blood count** (CBC) will describe several aspects of an individual's blood cells. A person with sickle cell disease will have a lower than normal hemoglobin level, together with other characteristic red blood cell abnormalities. A **hemoglobin electrophoresis** is a test that can help identify the types and quantities of hemoglobin made by an individual. This test uses an electric field applied across a slab of gel-like material. Hemoglobins migrate through this gel at various rates and to specific locations, depending on their size, shape, and electrical charge. Although sickle hemoglobin (Hb S) and regular adult hemoglobin (called Hb A) differ by only one amino acid, they can be clearly separated using **hemoglobin electrophoresis**. **Isoelectric focusing** and **high-performance liquid chromatography (HPLC)** use similar principles to separate hemoglobins and can be used instead of or in various combinations

with hemoglobin electrophoresis to determine the types of hemoglobin present.

Another test, called the 'sickledex' can help confirm the presence of sickle hemoglobin, although this test cannot provide accurate or reliable diagnosis when used alone. When Hb S is present but there is an absence or only a trace of Hb A, sickle cell anemia is a likely diagnosis. Additional beta globin DNA testing, which looks directly at the beta globin gene, can be performed to help confirm the diagnosis and establish the exact genetic type of sickle cell disease. CBC and hemoglobin electrophoresis are also typically used to diagnosis sickle cell trait and various other types of beta globin traits.

Diagnosis of sickle cell disease can occur under various circumstances. If an individual has symptoms that are suggestive of this diagnosis, the above-described screening tests can be performed followed by DNA testing, if indicated. Screening at birth using HPLC or a related technique offers the opportunity for early intervention. More than 40 states include sickle cell screening as part of the usual battery of blood tests done for newborns. This allows for early identification and treatment. Hemoglobin trait screening is recommended for any individual of a high-risk ethnic background who may be considering having children. When both members of a couple are found to have sickle cell trait, or other related hemoglobin traits, they can receive **genetic counseling** regarding the risk of sickle cell disease in their future children and various testing options.

Sickle cell disease can be identified before birth through the use of prenatal diagnosis. **Chorionic villus sampling (CVS)** can be offered as early as 10 weeks of pregnancy and involves removing a sample of the placenta made by the baby and testing the cells. CVS carries a risk of causing a **miscarriage** that is between 5% to 1%.

Amniocentesis is generally offered between 16 and 18 weeks of pregnancy but can sometimes be offered earlier. Two to three tablespoons of the fluid surrounding the baby is removed. This fluid contains fetal cells that can be tested. This test carries a risk of causing a miscarriage, which is less than 1%. Pregnant woman and couples may choose prenatal testing in order to prepare for the birth of a baby that may have sickle cell disease.

Preimplantation genetic diagnosis (PGD) is a relatively new technique that involves in-vitro fertilization followed by **genetic testing** of one cell from each developing embryo. Only the embryos unaffected by sickle cell disease are transferred back into the uterus. PGD is currently available on a research basis only, and is relatively expensive.

KEY TERMS

Amino acid—Organic compounds that form the building blocks of protein. There are 20 types of amino acids (eight are “essential amino acids,” which the body cannot make and must therefore be obtained from food).

Anemia—A blood condition in which the level of hemoglobin or the number of red blood cells falls below normal values. Common symptoms include paleness, fatigue, and shortness of breath.

Bilirubin—A yellow pigment that is the end result of hemoglobin breakdown. This pigment is metabolized in the liver and excreted from the body through the bile. Bloodstream levels are normally low; however, extensive red cell destruction leads to excessive bilirubin formation and jaundice.

Bone marrow—A spongy tissue located in the hollow centers of certain bones, such as the skull and hip bones. Bone marrow is the site of blood cell generation.

Bone marrow transplantation—A medical procedure used to treat some diseases that arise from defective blood cell formation in the bone marrow. Healthy bone marrow is extracted from a donor to replace the marrow in an ailing individual. Proteins

on the surface of bone marrow cells must be identical or very closely matched between a donor and the recipient.

Globin—Protein component of hemoglobin. Normal adult hemoglobin has a pair each of alpha-globin and beta-globin molecules that each contain a heme group.

Heme—The iron-containing molecule in hemoglobin that serves as the site for oxygen binding. Normal hemoglobin has four hemes in four globin chains.

Hemoglobin—Iron-containing blood protein that carries oxygen to the cells and carries carbon dioxide away from the cells.

Hemoglobin A—Normal adult hemoglobin that contains four heme molecules, two alpha-globin molecules, and two beta-globin molecules.

Hemoglobin electrophoresis—A laboratory test that separates molecules based on their size, shape, or electrical charge.

Hemoglobin S—Hemoglobin produced in association with the sickle cell trait; the beta-globin molecules of hemoglobin S are defective.

Treatment

Traditional

There are several practices intended to prevent some of the symptoms and complications of sickle cell disease. These include preventative **antibiotics**, good hydration, immunizations, and access to comprehensive care. Maintaining good health through adequate **nutrition**, avoiding stresses and infection, and getting proper rest is also important. Following these guidelines is intended to improve the health of individuals with sickle cell disease.

As in any lifelong, chronic disease, comprehensive care is important. Assistance with the emotional, social, family-planning, economic, vocational, and other consequences of sickle cell disease can enable affected individuals to better access and benefit from their medical care.

Drugs

Infants are typically started on a course of penicillin that extends from infancy to age six. Use of this antibiotic is meant to ward off potentially fatal infections. Infections at any age are treated aggressively with

antibiotics. Vaccines for common infections, such as *pneumococcal pneumonia*, are also recommended.

Pain is one of the primary symptoms of sickle cell anemia and controlling it is an important concern. The methods necessary for pain control are based on individual factors. Some people can gain adequate pain control through over-the-counter oral painkillers (**analgesics**). Other individuals, or painful events, may require stronger methods, which can include administration of **narcotics**. Alternative therapies may be useful in avoiding or controlling pain, including relaxation, hydration, avoiding extremes of temperature, and the application of local warmth.

Emphasis is being placed on developing drugs that treat sickle cell anemia directly. The most promising of these drugs since the late 1990s has been hydroxyurea, a drug that was originally designed for anticancer treatment. Hydroxyurea has been shown to reduce the frequency of painful crises and acute chest syndrome in adults, and to lessen the need for blood transfusions. Hydroxyurea, and other related medications, seem to work by inducing a higher production of fetal hemoglobin. The major side effects of the drug include decreased production of platelets, red blood

Hydroxyurea—A drug that has been shown to induce production of fetal hemoglobin. Fetal hemoglobin has a pair of gamma-globin molecules in place of the typical beta-globins of adult hemoglobin. Higher-than-normal levels of fetal hemoglobin can prevent sickling from occurring.

Impotence—The inability to have a penile erection, which can be due to tissue damage resulting from sickling within the penis (priapism).

Iron overload—A side effect of frequent blood transfusions in which the body accumulates abnormally high levels of iron. Iron deposits can form in organs, particularly the heart, and cause life-threatening damage.

Jaundice—Yellowing of the skin or eyes due to excess of bilirubin in the blood.

Magnetic resonance imaging (MRI)—A technique that employs magnetic fields and radio waves to create detailed images of internal body structures and organs, including the brain.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease, and can be transmitted to offspring.

cells, and certain white blood cells. The effects of long-term hydroxyurea treatment are unknown.

Alternative

BLOOD TRANSFUSIONS. Blood transfusions are not usually given on a regular basis but are used to treat individuals with frequent and severe painful events, severe anemia, and other emergencies. In some cases blood transfusions are given as a preventative measure, for example to treat spleen enlargement or prevent a second stroke (or a first stroke in an individual shown to be at high risk).

Regular blood transfusions have the potential to decrease formation of hemoglobin S and reduce associated symptoms. However, there are limitations and risks associated with regular blood transfusions, including the risk of blood-borne infection and sensitization to proteins in the transfused blood that can make future transfusions very difficult. Most importantly, chronic blood transfusions can lead to iron overload. The body tends to store excess iron, such as that received through transfusions, in various organs. Over time, this iron

Narcotics—Strong, prescription medication that can be effective in treating pain, but have the potential to be habit-forming if their use is not supervised correctly.

Nucleic acid—A type of chemical used as a component for building DNA. The nucleic acids found in DNA are adenine, thymine, guanine, and cytosine.

Ophthalmology—The medical specialty of vision and the eye.

Placenta—The organ responsible for oxygen and nutrition exchange between a pregnant mother and her developing baby.

Red blood cell—Hemoglobin-containing blood cells that transport oxygen from the lungs to tissues. In the tissues, the red blood cells exchange their oxygen for carbon dioxide, which is brought back to the lungs to be exhaled.

Screening—Process through which carriers of a trait may be identified within a population.

Sickle cell—A red blood cell that has assumed a elongated shape due to the presence of hemoglobin S.

storage can cause damage to various tissues and organs, such as the heart and endocrine organs.

Some of this damage can be prevented by the administration of a medication called *desferrioxamine* that helps the body to eliminate excess iron through the urine. Alternately, some individuals receive a new, non-standard treatment called *erythrocytapheresis*. This involves the automated removal of sickle cells and is used in conjunction with a reduced number of regular transfusions. This treatment helps to reduce iron overload.

BONE MARROW TRANSPLANTATION. **Bone marrow transplantation** has been shown to cure sickle cell anemia in some cases. This treatment is reserved primarily for severely affected children with a healthy donor whose marrow proteins match those of the recipient, namely a brother or sister who has inherited the same tissue type. Indications for a bone marrow transplant are stroke, recurrent acute chest syndrome, and chronic unrelieved pain.

Bone marrow transplants tend to be the most successful in children; adults have a higher rate of

transplant rejection and other complications. There is approximately a 10% fatality rate associated with bone marrow transplants done for sickle cell disease. Survivors face potential long-term complications, such as chronic graft-versus-host disease (an immune-mediated attack by the donor marrow against the recipient's tissues), **infertility**, and development of some forms of **cancer**. A relatively recent advance in transplantation involves the use of donor stem cells obtained from *cord blood*, the blood from the placenta that is otherwise discarded following the birth of a new baby. Cord blood cells, as opposed to fully mature bone marrow cells, appear to be less likely to result in graft-versus-host disease in the recipient. This increases the safety and efficacy of the transplant procedure.

SURGERY. Certain surgical interventions are utilized in the treatment of specific sickle cell-related complications. Removal of a dysfunctional gallbladder or spleen can often lead to improvements in health. Investigations are currently underway to establish the efficacy of hip coring surgery in which a portion of affected bone is removed to treat avascular necrosis of the hip. The hope is that this may provide an effective treatment to alleviate some pain and restore function in the affected hip.

CLINICAL TRIALS. Clinical trials for the treatment of sickle cell anemia are currently sponsored by the National Institutes of Health (NIH) and other agencies. In 2009, NIH reported 198 on-going or recently completed studies. Some examples include the following:

- The evaluation of the effect of L-glutamine therapy on exercise endurance and breath by breath exercise response of sickle cell anemia patients. (NCT00586209)
- The study of how often people with sickle cell anemia develop pulmonary hypertension, a serious disease in which blood pressure in the artery to the lungs is elevated. (NCT00011648)
- The evaluation of the effectiveness of the nutritional supplement arginine at improving blood cell function and disease symptoms in people with sickle cell anemia. (NCT00513617)
- A study evaluating the iron overload in sickle cell anemia patients using clinical parameters and laboratory studies to determine cardiac and liver iron. (NCT00512564)

Clinical trial information is constantly updated by NIH and the most recent information on sickle cell anemia trials can be found at: <http://clinicaltrials.gov/> ct2/results?term=sickle+cell+disease.

Prognosis

Sickle cell disease is characteristically variable between and within affected individuals. Predicting the

course of the disorder based solely on genes is not possible. Several factors aside from genetic inheritance determine the prognosis for affected individuals, including the frequency, severity, and nature of specific complications in any given individual. The availability and access of comprehensive medical care also plays an important role in preventing and treating serious, acute complications, which cause the majority of sickle cell-related deaths. For those individuals who do not experience such acute events, life-expectancy is probably substantially greater than the average for all people with sickle cell disease. The impact of recent medical advances supports the hypothesis that current life-expectancies may be significantly greater than those estimated in the early 1990s. At that time, individuals with SS disease lived to the early-to mid-40s, and those with SC disease lived into the upper 50s on average. With early detection and comprehensive medical care, most people with sickle cell disease are in fairly good health most of the time. Most individuals can be expected to live well into adulthood, enjoying an improved quality of life including the ability to choose a variety of education, career, and family-planning options for themselves.

Prevention

Inheritance of sickle cell disease or trait cannot be prevented but it may be predicted. Screening is recommended for individuals in high-risk populations. In the United States, African Americans and Latino Americans have the highest risk of having the disease or trait. All 50 states in the United States have newborn screening programs for sickle cell disease.

Resources

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March of Dimes Foundation, 1275 Mamaroneck Avenue, White Plains, NY, 10605, (914) 428-7100, (888) MODIMES, (914) 428-8203, askus@marchofdimes.com, <http://www.marchofdimes.com>.

National Heart, Lung, and Blood Institute (NHLBI), Building 31, Room 5A52, 31 Center Drive MSC 2486, Bethesda, MD, 20892, (301) 592-8573, (240) 629-3246, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov>.

Sickle Cell Disease Association of America, Inc., 231 East Baltimore St., Suite 800, Baltimore, MD, 21202, (410) 528-1555, (800) 421-8453, (410) 528-1495, scdaa@sicklecelldisease.org, <http://www.sicklecelldisease.org>.

Sickle Cell Information Center, PO Box 109, Atlanta, GA, 30303, (404) 616-3572, (404) 616-5998, aplatt@emory.edu, <http://www.scinfo.org>.

Monique Laberge, PhD

Sideroblastic anemia

Definition

Sideroblastic anemia is a term used to describe a group of rare blood disorders characterized by the bone marrow's inability to manufacture normal red blood cells.

Description

Named for the Greek words for iron and germ, sideroblastic anemia is one of the principal types of iron-utilization anemia. Abnormal iron-saturated red cells are present in the blood of people who have this disease. Although the iron circulates normally from the plasma to the bone marrow, where new red blood cells are created, it is not properly incorporated into new red blood cells.

Sideroblastic anemia can be inherited but the disease is usually acquired as a result of illness or exposure to toxic substances.

Sideroblastic anemia is a disease of adults.

Causes and symptoms

The cause of sideroblastic anemia cannot always be identified. Drug toxicity, alcohol **abuse**, and **lead poisoning** are common causes of this condition.

Sideroblastic anemia is also associated with:

- leukemia
- lymphoma (cancer of the lymph glands)
- myeloma (cancer of the bone marrow)
- **rheumatoid arthritis**, and other inflammatory diseases

Symptoms of sideroblastic anemia are the same as symptoms of the disease that causes the condition, as well as anemia.

Complications

Possible complications of sideroblastic anemia include:

- congestive heart failure
- **diabetes mellitus**
- enlargement of the liver and spleen
- formation of liver nodules and scar tissue
- irregular heartbeat
- recurring inflammation of the sac that surrounds the heart
- secondary hypopituitarism (dwarfism)

- skin darkening
- underactivity of the thyroid gland

Diagnosis

Blood tests are used to examine the appearance and other characteristics of red cells and to measure the amount of iron in the blood. **Bone marrow biopsy** is also used.

Treatment

Acquired sideroblastic anemia may be cured when the condition that causes it is treated or removed.

If the cause of a patient's anemia cannot be determined, blood transfusions may be necessary. Medications are prescribed to stimulate excretion or excess iron that accumulates as a result of these transfusions.

In rare instances, treatment with oral pyridoxine (a B-complex vitamin) benefits patients whose sideroblastic anemia was present at birth. This treatment improves the condition of some patients but does not cure the anemia.

Prognosis

Sideroblastic anemia of unknown origin may lead to leukemia. It may take as long as 10 years for this disease progression to take place.

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ORGANIZATIONS

Leukemia and Lymphoma Society, 1311 Mamaroneck Avenue, Suite 310, White Plains, NY, 10605, (800) 955-4572, <http://www.leukemia-lymphoma.org>.

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

SIDS see **Sudden infant death syndrome**

Sigmoidoscopy

Definition

Sigmoidoscopy is a diagnostic and screening procedure for colorectal **cancer** and inflammatory bowel disease in which a rigid or flexible tube with a camera on the end (a sigmoidoscope) is inserted into the anus to examine the rectum and lower colon (bowel) for bowel disease, cancer, precancerous conditions, or causes of bleeding or **pain**. As of 2010, however, sigmoidoscopy is being used less frequently for screening than **colonoscopy**.

Purpose

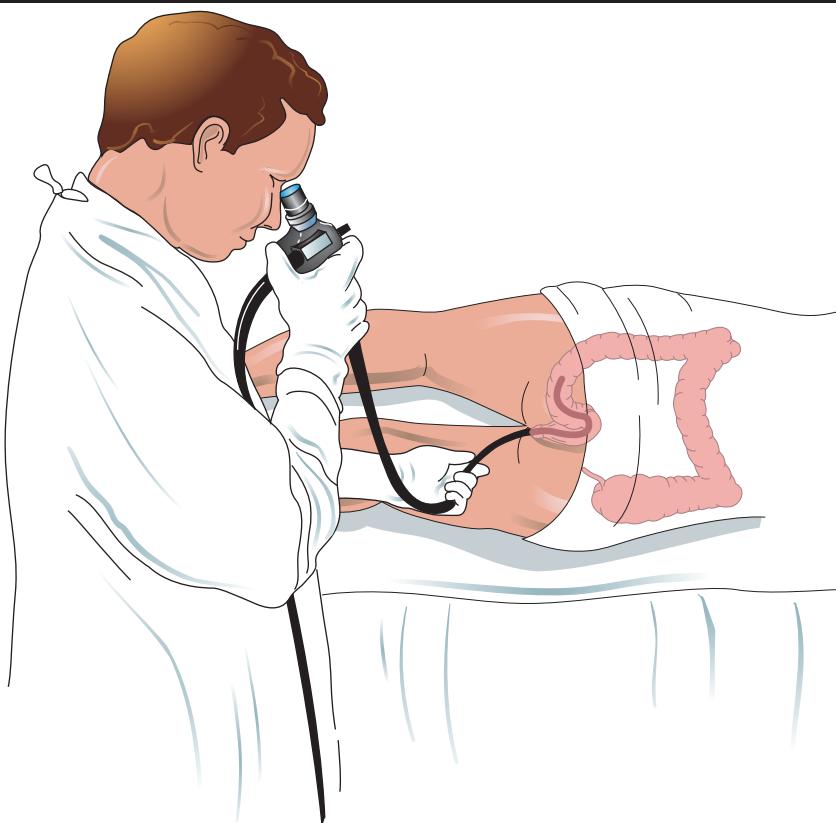
Sigmoidoscopy is used most often in screening for colorectal cancer or to determine the cause of rectal bleeding. It is also used in diagnosis of inflammatory bowel disease, microscopic and ulcerative **colitis**, and **Crohn's disease**.

Cancer of the rectum and colon is the second most common cancer in the United States. About 148,300 new cases are diagnosed annually. Between 55,000 and 60,000 Americans die each year of cancer in the colon or rectum. The lifetime risk of developing **colon cancer** in the United States is about 7%.

Experts recommend that people over 50 should be screened for colorectal cancer every one to two years. Various screening tests are used, ranging from fecal occult blood tests (FOBTs) and digital rectal examinations (DREs) in the doctor's office, to more invasive tests such as sigmoidoscopies and colonoscopies. Individuals with such inflammatory bowel conditions as Crohn's disease or ulcerative colitis, and thus at increased risk for colorectal cancer, may begin their screenings at a younger age, depending on when their disease was diagnosed. Screening should also be performed in people who have a family history of colon or **rectal cancer**, or small growths in the colon (polyps).

More and more physicians are performing invasive screening with a colonoscope, which allows them to see the entire colon, in contrast to a sigmoidoscope, which allows them to visualize only the rectum and the lower portion of the colon. Another newer option is virtual colonoscopy, also known as CT colonography, which uses computerized tomography (or **magnetic resonance imaging** in some cases) to obtain images of the interior of the colon and rectum. The patient must cleanse the bowel before this procedure but does not require **sedation** as virtual colonoscopy is not an invasive procedure.

Studies have shown that one-quarter to one-third of all precancerous or small cancerous growths can be



Sigmoidoscopy is a procedure most often used in screening for colorectal cancer and as a test in diagnosis of possible inflammatory bowel disease. As illustrated above, the physician can view the rectum and colon through a sigmoidoscope, a 12 inch (30 cm) or 24 inch (60 cm) flexible fiber-optic tube which contains a light source and a lens. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

seen with a sigmoidoscope. About one-half are found with a 1 ft (30 cm) scope, and two-thirds to three-quarters can be seen using a 2 ft (60 cm) scope.

In some cases, the sigmoidoscope can be used therapeutically in conjunction with such other equipment as electrosurgical devices to remove polyps and other lesions found during the sigmoidoscopy.

Demographics

According to the Centers for Disease Control and Prevention (CDC), approximately 2.8 million flexible sigmoidoscopies and 14.2 million colonoscopies are performed in an average year in the United States. This number includes most of the persons who are diagnosed with colon cancer each year, a greater number who are screened and receive negative results, persons who have been treated for colon conditions and receive a sigmoidoscopy as a follow-up procedure, and individuals who are diagnosed with other diseases of the large colon.

Description

Sigmoidoscopy may be performed using either a rigid or flexible sigmoidoscope, although rigid sigmoidoscopes are rarely used as of 2010. A sigmoidoscope is a thin tube with fiberscopes, electronics, a light source, and camera. A physician inserts the sigmoidoscope into the anus to examine the rectum (the first 1 ft [30 cm] of the colon) and its interior walls. If a 2 ft (60 cm) scope is used, the next portion of the colon can also be examined for any irregularities. The camera of the sigmoidoscope is connected to a viewing monitor, allowing the interior of the rectum and colon to be enlarged and viewed on the monitor. Images can then be recorded as still pictures or the entire procedure can be videotaped. The still pictures are useful for comparison purposes with the results of future sigmoidoscopic examinations.

If polyps, lesions, or other suspicious areas are found, the physician biopsies them for analysis. During the sigmoidoscopy, the physician may also use

KEY TERMS

Biopsy—The removal of a small portion of tissue during sigmoidoscopy to perform laboratory tests to determine if the tissue is cancerous.

Colonoscopy—A diagnostic endoscopic procedure that uses a long flexible tube called a colonoscope to examine the inner lining of the entire colon; may be used for colorectal cancer screening or for a more thorough examination of the colon.

Colorectal cancer—Cancer of the large intestine, or colon, including the rectum.

Electrosurgical device—A medical device that uses electrical current to cauterize or coagulate tissue during surgical procedures, often used in conjunction with laparoscopy, colonoscopy, or sigmoidoscopy.

Inflammatory bowel diseases—Ulcerative colitis or Crohn's disease: chronic conditions characterized by periods of diarrhea, bloating, abdominal cramps, and pain, sometimes accompanied by weight loss and malnutrition because of the inability to absorb nutrients.

Pathologist—A doctor who specializes in the diagnosis of disease by studying cells and tissues under a microscope.

Polyp—A small growth, usually not cancerous, but often precancerous when it appears in the colon.

Virtual colonoscopy—A noninvasive form of colonoscopy that uses computed tomography (CT scanning) to obtain images of the interior of the patient's colon and rectum. It is also called CT colonography.

forceps, graspers, snares, or electrosurgical devices to remove polyps, lesions, or tumors.

A typical sigmoidoscopy procedure requires 15 to 20 minutes to perform. Preparation begins one day before the procedure. There is some discomfort when the scope is inserted and throughout the procedure, similar to that experienced when a physician performs a rectal exam using a finger to test for occult blood in the stool. Individuals may also feel some minor cramping pain. There is rarely severe pain, except for persons with active inflammatory bowel disease.

Private insurance plans almost always cover the cost of sigmoidoscopy examinations for screening in healthy individuals over 50 or for diagnostic purposes. Medicare covers the cost for diagnostic exams, and may cover the costs for screening exams. Medicaid benefits vary by state, but sigmoidoscopy is not a covered procedure in

many states. Some community health clinics offer the procedure at reduced cost but this can only be done if a local gastroenterologist (a physician who specializes in treating stomach and intestinal disorders) is willing to donate personal time to perform the procedure.

Diagnosis/Preparation

The purpose of preparation for sigmoidoscopy is to cleanse the lower bowel of fecal material or stool so the physician can see the lining. Preparation begins 24 hours before the procedure when an individual must begin a clear liquid diet. Preparation kits are available in drug stores. In normal preparation, about 20 hours before the exam, a person begins taking a series of **laxatives**, which may be oral tablets or liquid. The individual must stop drinking any liquid four hours before the exam. An hour or two prior to the examination, the person uses an enema or laxative suppository to finish cleansing the lower bowel.

Patients must be careful about medications before having a sigmoidoscopy. They should not take **aspirin**, products containing aspirin, or products containing ibuprofen for one week prior to the exam, because these medications can exacerbate bleeding during the procedure. They should not take any iron or **vitamins** with iron for one week prior to the exam, since iron can cause color changes in the bowel lining that interfere with the examination. They should take any routine prescription medications but may need to stop certain medications. Prescribing physicians should be consulted regarding routine prescriptions and their possible effect(s) on sigmoidoscopy.

Individuals with renal insufficiency or congestive **heart failure** need to be prepared in an alternative way, and must be carefully monitored during the procedure.

Aftercare

There is no specific aftercare necessary following sigmoidoscopy. If a biopsy was taken, a small amount of blood may appear in the next stool. Persons should be encouraged to pass gas following the procedure to relieve any bloating or cramping that may occur after the procedure. In addition, an infection may develop following sigmoidoscopy. Persons should be instructed to call their physician if a **fever** or pain in the abdomen develops over the few days after the procedure.

Risks

There is a slight risk of bleeding from the procedure. This risk is heightened in individuals whose blood does not clot well, either due to disease or medication, and in those with active inflammatory bowel disease. Rarely,

trauma to the bowel or other organs can occur, resulting in an injury (perforation) that must be repaired, or **peritonitis**, which must be treated with medication.

Sigmoidoscopy may be contraindicated in persons with severe active colitis or toxic megacolon (an extremely dilated colon). In general, people experiencing continuous ambulatory peritoneal dialysis are not candidates due to a high risk of developing intraperitoneal bleeding.

Normal results

The results of a normal examination reveal a smooth colon wall with sufficient blood vessels for good blood flow.

Morbidity and mortality rates

For a cancer screening sigmoidoscopy, an abnormal result is one or more noncancerous or precancerous polyps, or clearly cancerous polyps. People with polyps have an increased risk of developing colorectal cancer in the future and may be required to undergo additional procedures such as colonoscopy or more frequent sigmoidoscopic examinations.

Small polyps can be completely removed. Larger polyps may require the physician to remove a portion of the growth for laboratory biopsy. Depending on the laboratory results, a person is then scheduled to have the polyp removed surgically, either as an urgent matter if it is cancerous, or as an elective procedure within a few months if it is noncancerous.

In a diagnostic sigmoidoscopy, an abnormal result shows signs of active inflammatory bowel disease, either a thickening of the intestinal lining consistent with ulcerative colitis or ulcerations or fissures consistent with Crohn's disease.

Mortality from a sigmoidoscopy examination is rare and is usually due to uncontrolled bleeding or perforation of the colon.

Alternatives

A screening examination for colorectal cancer is a test for fecal occult blood. A dab of fecal material from toilet tissue is smeared onto a card. The card is treated in a laboratory to reveal the presence of bleeding. This test is normally performed prior to a sigmoidoscopic examination.

A less invasive alternative to a sigmoidoscopic examination is an X-ray of the colon and rectum. Barium is used to coat the inner walls of the colon. This lower GI (gastrointestinal) X-ray may reveal the outlines of suspicious or abnormal structures. It has the disadvantage of

not allowing direct visualization of the colon. It is less costly than a sigmoidoscopic examination.

Another less invasive procedure is a virtual colonoscopy, described above. Virtual colonoscopy was endorsed by the American Cancer Society, the American College of Radiology and the U.S. Multisociety Task Force on Colorectal Cancer as an effective screening procedure for colorectal cancer in 2008.

A more invasive procedure is direct visualization of the colon during surgery. This procedure is rarely performed in the United States.

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ORGANIZATIONS

- American Academy of Family Physicians, 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2672, (913) 906-6000, (800) 274-2237, <http://www.aafp.org>.
- American College of Surgeons, 633 North Saint Claire Street, Chicago, IL, 60611, (312) 202-5000, <http://www.facs.org/>.
- American Society for Gastrointestinal Endoscopy, 1520 Kensington Road, Suite 202, Oak Brook, IL, 60523, (866) 353-ASGE, <http://www.asge.org>.
- Society of American Gastrointestinal Endoscopic Surgeons, 11300 West Olympic Boulevard, Suite 600, Los Angeles, CA, 90064, (310) 437-0544, (310) 437-0585, <http://www.sages.org>.

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Sildenafil citrate

Definition

Sildenafil citrate (Viagra and Revatio), was originally developed in 1991 to improve circulation to the heart and treat **angina**, or chest **pain**. While it was not effective for that purpose, it increased circulation in the lungs and penis and is used to treat arterial **hypertension** in the lungs and **erectile dysfunction** in men.

Purpose

By increasing blood flow to the penis during sexual stimulation, men with erectile dysfunction who take Viagra are able to achieve stronger erections and/or

maintain them longer. Erectile dysfunction can be caused by a host of emotional and psychological conditions, metabolic diseases, injuries to blood vessels and/or nerves supplying the penis, and by side effects of many medications.

For erectile dysfunction, Viagra is taken 30–60 minutes before intercourse. It should not be taken for this purpose more often than once every 24 hours.

Though it has not been clinically tested for this purpose, a significant number of men take Viagra in hopes of improving their sexual performance.

In pulmonary arterial hypertension, Revatio relaxes the blood vessels in the lung, increasing blood flow and improving **exercise tolerance**. For this purpose, it is taken more frequently and regularly.

Preparation

Viagra comes in 25, 50 and 100mg tablets.

Revatio comes in 20mg tablets.

Recommended dosage

The recommended starting dose of Viagra for treating erectile dysfunction is 50mg taken 30–60 minutes prior to sexual activity. If needed, the dose may be decreased to 25mg or increased to 100mg.

For treating **pulmonary hypertension**, 20mg of Revatio are taken three times daily.

Precautions

Viagra is not approved for use by women or children.

Sexual activity can stress the heart. A combination of high blood pressure and/or underlying heart or **vascular disease** plus Viagra and sexual activity may produce irregular heartbeat, **heart attack**, **stroke**, and/or sudden **death**.

Men who experience **shortness of breath**, **dizziness** or chest pain during sexual activity should not take Viagra and should consult a doctor.

Men who use nitrate medications to treat chest pain, like Isordil and nitroglycerine in any form, should not take Viagra. These drugs can act together to produce dangerous decreases in blood pressure.

Nitrate containing street drugs, poppers, should not be used with Viagra.

Many prescription and non-prescription medications, herbs, and **nutritional supplements** can alter the effects and toxicity of Viagra. Before taking Viagra, men should consult with a pharmacist or doctor and

discuss the possible effects of medications they take on one another and on Viagra.

Men with deformed penises, or who have experienced prolonged or painful erections should discuss these problems with a doctor before taking Viagra.

Men who have circulation problems involving their eyes or vision, or family members with inherited vision problems, like **retinitis pigmentosa**, should discuss these problems with an eye specialist before taking Viagra.

Men who have recently been ill and/or lost body fluids through **vomiting**, **diarrhea**, or sweating, are more likely to have side effects from Viagra.

Men who have stomach, liver or **kidney disease**, myeloma, leukemia or other blood disorders should discuss with a doctor whether or not it is safe to take Viagra.

Side effects

Men who experience these symptoms should consult their physician. Side effects may be reduced or eliminated by adjusting the dose of Viagra.

The most commonly side effects of Viagra are mild **headache**, flushing of the face, upset stomach, and nasal congestion.

Some side effects of Viagra can be serious:

- heartburn
- chest pain
- shortness of breath
- nosebleed
- numbness, burning or tingling in the arms, hands, feet or legs
- muscle aches
- vision problems, including sudden loss of vision, sensitivity to light, blurred vision, and a blue or green color tinge to vision
- ringing in the ears or sudden decrease or loss of hearing
- itching and burning during urination
- diarrhea
- dizziness or fainting
- rash

Resources

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James Waun, MD, RPh

Silent thyroiditis see **Thyroiditis**

Silicosis

Definition

Silicosis is a progressive disease that belongs to a group of lung disorders called pneumoconioses. Silicosis is marked by the formation of lumps (nodules) and fibrous scar tissue in the lungs. It is the oldest known occupational lung disease and is caused by exposure to inhaled particles of silica, mostly from quartz in rocks, sand, and similar substances.

Description

It is estimated that there are 2 million workers in the United States employed in occupations at risk for the development of silicosis. These include miners, foundry workers, stonecutters, potters and ceramics workers, sandblasters, tunnel workers, and rock drillers. Silicosis is mostly found in adults over 40 years of age. It has four forms:

- Chronic. Chronic silicosis may take 15 or more years of exposure to develop. There is only mild impairment of lung functioning. Chronic silicosis may progress to more advanced forms.
- Complicated. Patients with complicated silicosis have noticeable shortness of breath, weight loss, and extensive formation of fibrous tissue (fibrosis) in the lungs. These patients are at risk for developing **tuberculosis** (TB).
- Accelerated. This form of silicosis appears after 5–10 years of intense exposure. The symptoms are similar to those of complicated silicosis. Patients in this group often develop rheumatoid arthritis and other autoimmune disorders.
- Acute. Acute silicosis develops within 6 months to 2 years of intense exposure to silica. The patient loses a great deal of weight and is constantly short of breath. These patients are at severe risk of TB.

Causes and symptoms

The precise mechanism that triggers the development of silicosis is still unclear. What is known is that particles of silica dust get trapped in the tiny sacs (alveoli) in the lungs where air exchange takes place. White blood cells called macrophages in the alveoli ingest the silica and die. The resulting inflammation attracts other macrophages to the region. The nodule forms when the immune system forms fibrous tissue to seal off the reactive area. The disease process may stop at this point or speed up and destroy large areas of the lung. The fibrosis may continue even after the worker is no longer exposed to silica.

Early symptoms of silicosis include **shortness of breath** after exercising and a harsh, dry **cough**. Patients may have more trouble breathing and cough up blood as the disease progresses. Congestive **heart failure** can give their nails a bluish tint. Patients with advanced silicosis may have trouble sleeping and experience chest **pain**, hoarseness, and loss of appetite. Silicosis patients are at high risk for TB, and should be checked for the disease during the doctor's examination.

Diagnosis

Diagnosis of silicosis is based on:

- A detailed occupational history
- Chest x rays will usually show small round opaque areas in chronic silicosis; the round areas are larger in complicated and accelerated silicosis
- Bronchoscopy
- Lung function tests

It should be noted that the severity of the patient's symptoms does not always correlate with x-ray findings or lung function test results.

Treatment

Symptom management

There is no cure for silicosis. Therapy is intended to relieve symptoms, treat complications, and prevent respiratory infections. It includes careful monitoring for signs of TB. Respiratory symptoms may be treated with **bronchodilators**, increased fluid intake, steam inhalation, and **physical therapy**. Patients with severe breathing difficulties may be given **oxygen therapy** or placed on a mechanical ventilator. Acute silicosis may progress to complete **respiratory failure**. Heart-lung transplants are the only hope for some patients.

Patients with silicosis should call their doctor for any of the following symptoms:

- tiredness or mental confusion
- continued weight loss
- coughing up blood
- fever, chest pain, breathlessness, or new unexplained symptoms

Lifestyle changes

Patients with silicosis should be advised to quit **smoking**, prevent infections by avoiding crowds and persons with colds or similar infections, and receive vaccinations against **influenza** and **pneumonia**. They should be encouraged to increase their **exercise** capacity by keeping up regular activity and to learn to pace themselves with their daily routine.

KEY TERMS

Fibrosis—The development of excess fibrous connective tissue in an organ. Fibrosis of the lungs is a symptom of silicosis.

Pneumoconiosis (plural, pneumoconioses)—Any chronic lung disease caused by inhaling particles of silica or similar substances that lead to loss of lung function.

Silica—A substance (silicon dioxide) occurring in quartz sand, flint, and agate. It is used in making glass, scouring and grinding powders, pottery, etc.

Prognosis

Silicosis is currently incurable. The prognosis for patients with chronic silicosis is generally good. Acute silicosis, however, may progress rapidly to respiratory failure and **death**.

Prevention

Silicosis is a preventable disease. Preventive occupational safety measures include:

- Controls to minimize workplace exposure to silica dust.
- Substitution of substances—especially in sandblasting—that are less hazardous than silica.
- Clear identification of dangerous areas in the workplace.
- Informing workers about the dangers of overexposure to silica dust, training them in safety techniques, and giving them appropriate protective clothing and equipment.

Coworkers of anyone diagnosed with silicosis should be examined for symptoms of the disease. The state health department and the Occupational Safety and Health Administration (OSHA) or the Mine Safety and Health Administration (MSHA) must be notified whenever a diagnosis of silicosis is confirmed.

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ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

- Silo-filler's disease see **Lung diseases due to gas or chemical exposure**
 Simethicone see **Antigas agents**
 Singer's nodules see **Vocal cord nodules and polyps**

Single photon emission computed tomography

Definition

Single photon emission computed tomography (SPECT) is a type of imaging study used in nuclear medicine that uses radioactive materials injected through a vein that will pass into the brain or other organ generating a high-resolution brain image. SPECT relies on two technologies: computed tomography (CT) and the use of a radioactive material (radionuclide) to label a compound known as a tracer. Tracking the tracer's movement through body tissues and the rate of its radioactive decay allows the doctor to obtain 3-D images of blood flow in the heart, electrical activity in different areas of the brain, or to scan for tumors or bone disease by using a device called a gamma camera contained within the SPECT machine.

Purpose

SPECT is used to diagnose head trauma, **epilepsy**, **dementia**, and cerebrovascular disease. Development of a radiotracer called Tc99m (technetium-99) has increased the resolution of brain images generated from SPECT. The images yield very accurate spatial and contrast resolutions. Other radioactive isotopes used in SPECT are iodine-123, xenon-133, thallium-201, and fluorine-18. The resulting sharp images enable the clinician to visualize very small structures within the brain or other parts of the body. The accuracy of SPECT images makes it a very useful clinical and research tool.

Clinically, SPECT is useful for diagnosing the following disease states:

- Cerebrovascular disease or stroke: SPECT is useful to detect ischemia (reduced blood flow), determine causes of stroke, evaluate transient ischemia, determine prognosis, and monitor treatment.
- Such forms of dementia as Alzheimer's disease: SPECT studies can be used effectively to rule out other medical causes of dementia.
- Head trauma: Evidence suggests that SPECT is useful to detect greater number of lesions following the period

after head trauma. It seems that the high resolution and accurate brain images of SPECT can detect lesions in the brain that are not possible to visualize using other techniques such as positron emission tomography (PET) scanning. SPECT images can give clinicians important information concerning prognosis (also sometimes called outcome) and treatment of persons affected with head injury.

- Epilepsy: The radioactive material injected before SPECT imaging concentrates at the seizure locus (the region that contains nerve cells that generate an abnormal impulse). This can help identify the location of seizures and assist clinicians concerning management and outcomes.
- SPECT allows clinicians to visualize a specific area of the brain called the corpus striatum, which contains a neurotransmitter (a chemical that communicates nerve impulses from one nerve cell to another) called dopamine. Circuitry in the corpus striatum and interaction with dopamine can help provide valuable information concerning movement disorders, schizophrenia, mood disorders, and hormone diseases (since hormones require control and regulation from the brain in structures called the pituitary gland and hypothalamus).

As a research tool, SPECT imaging seems to be sensitive tool to measure blood flow through the brain (cerebral blood flow), in persons who have psychological disorder such as **obsessive-compulsive disorder** (higher blood flow) and **alcoholism** (lower blood flow).

More recently, SPECT has been used in myocardial perfusion imaging (MPI), which is a test done to evaluate patients for **coronary artery disease**. It is based on the understanding that diseased heart tissue under stress receives less blood than normal heart tissue (myocardium). A special form of technetium-99 known as Tc-sestamibi is injected. The patient's heart is then stressed by either **exercise** or by the administration of a drug, usually dobutamine, adenosine, or dipyridamole. SPECT imaging performed after the stress will reveal the distribution of the Tc-sestamibi within the different regions of the heart muscle. The patient is usually asked to return between one and seven days after the **stress test** for another set of SPECT images taken while he or she is at rest. Doctors then compare the two sets of images. If the images following the stress test are normal, the patient does not have to return for a second SPECT scan.

Other SPECT diagnostic indications and procedures are similar to other imaging studies such as computed tomography, **magnetic resonance imaging**, and **positron emission tomography**.

Precautions

Women who are pregnant or **breastfeeding** should not have SPECT scans because the radioactive tracer can be passed to the fetus or the nursing baby. Women of childbearing age may be asked to have a pregnancy test before a SPECT scan.

Description

In most cases, the SPECT scan involves injecting the patient with a compound containing the radioactive tracer or administering the tracer through an infusion given intravenously into a vein in the arm. In some cases, the patient may inhale the tracer through the nose. The patient is then asked to lie quietly in a room for 15–30 minutes while the radioactive tracer is absorbed by the body.

In the second phase of the scan, the patient is positioned by the health care team on a table in the room with the SPECT machine. The exact position depends on the part of the patient's body or the organ system that is being investigated. The SPECT machine itself contains a gamma camera, an imaging device that detects gamma rays given off by the radioactive tracer in the patient's body. The SPECT machine rotates around the patient while the gamma camera records a series of two-dimensional images of the patient's body organs. These images are then sent to a computer that produces 3-D images of the organs in question.

Preparation

Patients should wear comfortable clothing for a SPECT scan and expect to stay in the hospital for 1–3 hours. They do not need to fast beforehand or omit the medications they usually take.

Aftercare

Aftercare consists of drinking extra fluids to speed the excretion of the radioactive tracer in the urine. The tracer is usually flushed out in the patient's urine within a few hours after the scan. Any remaining tracer is broken down in the body within the next two days.

Risks

SPECT scans are generally safe and well tolerated by most patients. Some people, however, may experience bruising, bleeding, or **pain** at the point at which the needle was inserted into their vein. A small number of patients have allergic reactions to the radionuclide.

KEY TERMS

Gamma camera—A device inside the SPECT machine that forms images of the gamma rays emitted by the radionuclides used in tracers in nuclear medicine.

Gamma rays—Extremely short-wavelength electromagnetic radiation released during the process of radioactive decay.

Myocardium—The medical term for the specialized involuntary muscle tissue found in the walls of the heart.

Nuclear medicine—A branch of medicine that makes use of radioisotopes (also called radionuclides) to evaluate the rate of radioactive decay in diagnosing and treating various diseases.

Radionuclide—An atom with an unstable nucleus that emits gamma rays during the process of its radioactive decay. Radionuclides, also known as radioisotopes, are used to make the tracers used in SPECT. The most common radionuclides used in SPECT are iodine-123, technetium-99m, xenon-133, thallium-201, and fluorine-18.

Tracer—A substance containing a radioisotope, injected into the body and followed in order to obtain information about various metabolic processes in the body.

Normal results

Typical results of a SPECT scan show which parts of the patient's body or which areas within a specific organ absorbed larger amounts of the radionuclide and which absorbed less of the chemical. The images may be shown in different colors or in various shades of gray.

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Sinus endoscopy

Definition

An endoscope is a narrow flexible tube, which contains an optical device like a telescope or magnifying lens with a bright light. In sinus **endoscopy**, the endoscope is inserted into the nose and the interior of the nasal passages, sinuses, and throat is examined.

Purpose

Sinus endoscopy is used to help diagnose structural defects, infection or damage to the sinuses, or structures in the nose and throat. It may be used to view polyps and growths in the sinuses and to investigate causes of recurrent inflammation of the sinuses (**sinusitis**). During surgical procedures, an endoscope may be used to view the area to correct sinus-drainage problems or to remove polyps from the nose and throat.

Precautions

Insertion of the endoscope may cause a gag reflex and some discomfort, however, no special precautions are required to prepare for nasal endoscopy.

Description

This procedure can be done in a physician's office. The endoscope is inserted into a nostril and is threaded through the sinus passages to the throat. To make viewing of these areas easier, and to record the areas being examined, a camera, monitor, or other such viewing device is connected to the endoscope.

Preparation

For the procedure, the patient is usually awake and seated upright in a chair. A local anesthetic spray or liquid may be applied to the throat to make insertion of the endoscope less uncomfortable.

Aftercare

After the endoscope is removed, the patient can return to most normal activities. If an anesthetic was used, the patient may have to wait until the **numbness** wears off to be able to eat or drink.

Risks

The insertion and removal of the endoscope may stimulate a gag reflex and can cause some discomfort. The procedure may also irritate the tissues of the nose and throat, which can cause a **nosebleed** or coughing.

Normal results

Under normal conditions no polyps or growths are found in the sinuses. There should also be no evidence of infection, swelling, injury, or any structural defect that would prevent normal draining of the sinuses.

Abnormal results

Polyps, growths, infections, or structural defects of the nasal passages are considered abnormal.

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ORGANIZATIONS

American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Road, Alexandria, VA, 22314-2857, (703) 836-4444, <http://www.entnet.org>.
 EAR Foundation of Arizona, 668 North 44th Street, Suite 300, Phoenix, AZ, 85008, (602) 685-1050, (602) 239-5117, melissa@earfoundationaz.com, <http://www.earfoundationaz.com>.

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Sinus x ray see **Skull x rays**

Sinusitis

Definition

Sinusitis refers to an inflammation of the tissues that line the sinuses, which are air spaces within the bones of the face close to the nose. Sinusitis is most often caused by an infection within these spaces.

Sinusitis is usually classified as either acute or chronic. Acute sinusitis usually has a rapid onset; it is often a complication of the **common cold** but can also be triggered by **allergies**, bacterial infections, or fungal infections. Sinusitis that lasts longer than eight weeks, or keeps recurring (four or more times per year), is called chronic sinusitis.

Demographics

Acute sinusitis is a very common condition in Canada and the United States with 31–37 million cases reported each year and 200,000 surgical procedures performed to treat the disorder. It is possible that the actual number of cases is much higher because the symptoms of bacterial sinusitis often mimic those of colds or allergies, and many patients never see a doctor for proper diagnosis and treatment. About half of all diagnosed cases of acute sinusitis are caused by bacteria. The cost to the U.S. economy of acute sinusitis is estimated at \$3 billion per year.

About 90% of people will have an episode of sinusitis at some point in life. Most people diagnosed with sinusitis are young or middle-aged adults. Sinusitis is very rare in children younger than 18 months because the sinuses are not yet fully developed in infants.

Sinusitis is equally common in males and females and in all racial and ethnic groups.

Description

The sinuses are paired air pockets located within the bones of the face. They are:

- the frontal sinuses; located above the eyes, in the center region of each eyebrow
- the maxillary sinuses; located within the cheekbones, just to either side of the nose
- the ethmoid sinuses; located between the eyes, just behind the bridge of the nose.
- the sphenoid sinuses; located just behind the ethmoid sinuses and behind the eyes.

The sinuses are connected with the nose. They are lined with the same kind of skin found elsewhere within the respiratory tract. This skin has tiny little hairs projecting from it called cilia. The cilia beat constantly, to help move the mucus produced in the sinuses into the respiratory tract. The beating cilia sweeping the mucus along the respiratory tract help to clear the respiratory tract of any debris, or any organisms which may be present. When the lining of the sinuses is at all swollen, the swelling interferes with the normal flow of mucus. Trapped mucus can then fill the sinuses, causing an uncomfortable sensation of pressure and providing an excellent environment for the growth of infection-causing bacteria.

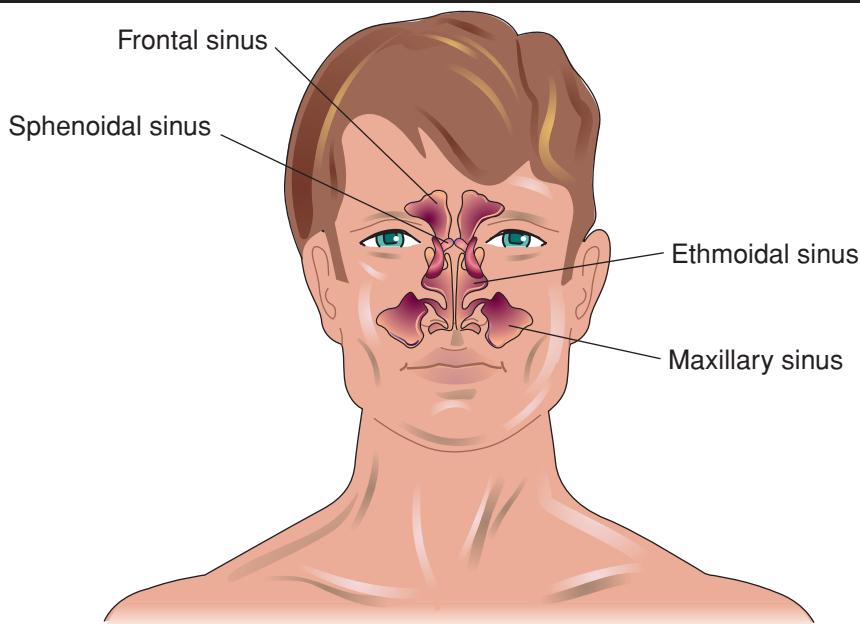
Risk factors

The risk factors for acute sinusitis and chronic sinusitis are similar. Risk factors for acute sinusitis include:

- A history of hay fever or other allergies affecting the nose.
- A structural abnormality in the nose, such as polyps or a deviated septum.
- Being a heavy smoker or being exposed to cigarette smoke in the home or at work.
- A history of cystic fibrosis, sarcoidosis, or gastroesophageal reflux disease (GERD).
- Having a disorder of the immune system or an antibody deficiency.
- Severe malnutrition, burns, liver disease, or cancer.
- Recent hospitalization, particularly in an intensive care unit.

In addition to these risk factors, there are two additional risk factors for chronic sinusitis:

- Asthma. About 20% of people with chronic sinusitis have asthma.
- Aspirin sensitivity that causes upper respiratory symptoms.



Sinusitis is the inflammation of the sinuses caused by a bacterial infection. Sometimes diagnosis may be problematic because the symptoms often mimic those of the common cold. Sinusitis is usually treated with antibiotics. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Causes and symptoms

Sinusitis is almost always due to an infection, although swelling from allergies can mimic the symptoms of pressure, **pain**, and congestion; and allergies can set the stage for a bacterial infection. Bacteria are the most common cause of sinus infection. *Streptococcus pneumoniae* causes about 33% of all cases, while *Haemophilus influenzae* causes about 25% of all cases. As of 2010, *Staphylococcus aureus* is seen more frequently as a cause of bacterial sinusitis in adults. About 2% of cases of viral sinusitis lead to secondary bacterial sinusitis. Some doctors think that nasal irritation from nose blowing leads to the secondary bacterial infection.

In some cases, sinusitis may result from a dental infection, a foreign body in the nose, **cocaine** abuse, or occupational exposure to such chemical irritants as chlorine gas.

Sinusitis in children may be caused by *Moraxella catarrhalis* (20%). In people with weakened immune systems (including patients with diabetes; acquired **immunodeficiency** syndrome or **AIDS**; and patients who are taking medications which lower their immune resistance, such as **cancer** and transplant patients), sinusitis may be caused by fungi such as *Aspergillus*, *Candida*, or *Mucorales*.

Acute sinusitis usually follows some type of upper respiratory tract infection or cold. Viral sinusitis

generally lasts from 7–10 days, while bacterial sinusitis tends to be more persistent. Instead of ending, the “cold” seems to linger on, with constant or even worsening congestion. Drainage from the nose often changes from a clear color to a thicker, yellowish-green discharge. There may be **fever**. **Headache** and pain over the affected sinuses may occur, as well as a feeling of pressure which may worsen when the patient bends over or lies down. There may be pain in the jaws or teeth. Some children, in particular, get upset stomachs from the infected drainage going down the back of their throats, and being swallowed into their stomachs. Some patients develop a **cough**.

Chronic sinusitis occurs when the problem has existed for at least eight weeks. There is rarely a fever with chronic sinusitis. Sinus pain and pressure is frequent, as is nasal congestion. Because of the nature of the swelling in the sinuses, they may not be able to drain out the nose. Drainage, therefore, drips constantly down the back of the throat, resulting in a continuously **sore throat** and **bad breath**.

Sinusitis in children is harder to distinguish from ordinary colds. Children younger than 6 years rarely develop headaches with sinusitis. However, swelling around the eyes or unusual irritability and **fatigue** are often associated with sinusitis in children, as are a discharge of yellowish or green mucus, sore throat,

KEY TERMS

Cilia (singular, cilium)—Tiny hair-like projections from a cell. Within the respiratory tract, the cilia act to move mucus along, in an effort to continually flush out and clean the respiratory tract.

Polyp—An abnormal growth of tissue projecting from a mucous membrane. Nasal polyps are tissue growths arising from the mucous membranes lining the nasal passages. They are a risk factor for sinusitis.

Septum—A structure comprised of cartilage and bony plates that separates the nasal cavity into two nostrils. A septum that is not in line with the center line of the nose is called a deviated septum and is a risk factor for sinusitis.

Sinus—An air-filled body cavity.

bad breath, fever above 100.4°F, and symptoms lasting longer than 10–14 days.

Diagnosis

Diagnosis of sinusitis can be made by a family doctor or by an otolaryngologist, a doctor who specializes in ear, nose, and throat disorders. In rare cases, the patient's dentist may be consulted to see whether a tooth infection is triggering the sinusitis.

Examination

Diagnosis of sinusitis is sometimes tricky because the symptoms so often resemble those of an uncomplicated cold. Sinusitis should be strongly suspected, however, when a cold lingers beyond a week's time. In some cases, the patient's history suggests the diagnosis, particularly a history of **asthma**, hay fever, **smoking**, or an occupational history of exposure to secondhand smoke or industrial chemicals. About 40% of cases of chronic sinusitis are associated with secondhand smoke.

Medical practitioners have differing levels of trust of certain basic examinations commonly conducted in the office. For example, tapping over the sinuses may cause pain in patients with sinusitis but it may not. A procedure called "sinus transillumination" may, or may not, also be helpful. Using a flashlight pressed up against the skin of the cheek, the practitioner will look in the patient's open mouth. When the sinuses are full of air (under normal conditions), the light will project through the sinus and will be visible on the roof of the mouth as a lit-up, reddened area. When the sinuses are full of mucus, the light will be blocked. While this simple

test can be helpful, it is certainly not a perfect way to diagnose or rule out the diagnosis of sinusitis.

Tests

Imaging tests can be useful in diagnosing sinusitis. X-ray pictures and CT scans of the sinuses are helpful for both acute and chronic sinusitis. People with chronic sinusitis may require an examination with a nasal endoscope to see whether any kind of anatomic obstruction is causing their illness. For example, the septum (the cartilage which separates the two nasal cavities from each other) may be slightly displaced, which is called a **deviated septum**. This can result in chronic obstruction, setting the person up for the development of an infection.

Procedures

In some cases a sample of tissue from the patient's nasal passages can be taken for biopsy and culture. Tissue culture is particularly useful in detecting fungal sinusitis.

If the doctor suspects that a previously undiagnosed allergy is triggering chronic sinusitis, he or she may recommend an allergy skin test to identify the specific allergens responsible for the sinusitis.

Treatment

Traditional

Drugs

Antibiotic medications are used to treat acute sinusitis. Suitable **antibiotics** include sulfa drugs, amoxicillin, and a variety of **cephalosporins**. These medications are usually given for about two weeks but may be given for even longer periods of time. **Decongestants**, or the short-term use of decongestant nose sprays, can be useful. **Acetaminophen** and ibuprofen can decrease the pain and headache associated with sinusitis. Also, running a humidifier can prevent mucus within the nasal passages from drying out uncomfortably and can help soothe any accompanying sore throat or cough.

Surgery

Chronic sinusitis is often treated initially with antibiotics. Steroid nasal sprays may be used to decrease swelling in the nasal passages. If an anatomic reason is found for chronic sinusitis, it may need to be corrected with surgery. If a surgical procedure is necessary, samples are usually taken at the same time to allow identification of any organisms present that may be causing infection.

Fungal sinusitis will require surgery to clean out the sinuses. Then, a relatively long course of a very strong antifungal medication called amphotericin B is given through a needle in the vein (intravenously).

Alternative

Chronic sinusitis is often associated with **food allergies**. An elimination/challenge diet is recommended to identify and eliminate allergenic foods. Irrigating the sinuses with a salt water solution is often recommended for sinusitis and allergies, in order to clear the nasal passages of mucus. Another solution for nasal lavage (washing) utilizes powdered goldenseal (*Hydrastis canadensis*). Other herbal treatments, taken internally, include a mixture made of eyebright (*Euphrasia officinalis*), goldenseal, yarrow (*Achillea millefolium*), horseradish, and ephedra (*Ephedra sinica*), or, when infection is present, a mixture made of **echinacea** (*Echinacea spp.*), wild indigo, and poke root (*Phytolacca decandra-American*a).

Homeopathic practitioners find a number of remedies useful for treating sinusitis. Among those they recommend are: *Arsenicum album*, *Kali bichromium*, *Nux vomica*, *Mercurius iodatus*, and *Silica*.

Acupuncture has been used to treat sinusitis, as have a variety of dietary supplements, including **vitamins A, C, and E**, and the mineral zinc. Contrast **hydrotherapy** (hot and cold compresses, alternating 3 minutes hot, 30 seconds cold, repeated 3 times always ending with cold) applied directly over the sinuses can relieve pressure and enhance healing. A direct inhalation of essential oils (2 drops of oil to 2 cups of water) using thyme, rosemary, and lavender can help open the sinuses and kill bacteria that cause infection.

Prognosis

Prognosis for sinus infections is usually excellent, although some individuals may find that they are particularly prone to contracting such infections after a cold. The chief risk for serious illness resulting from sinusitis is the closeness of the nasal passages to the central nervous system, the lymph nodes in the neck, and the blood vessels in the neck and throat. Complications of sinusitis can include **osteomyelitis**, an infection of the bone; **orbital cellulitis**, inflammation of the tissues surrounding the eye; and **meningitis**, inflammation of the membranes covering the brain and spinal cord. Fungal sinusitis, however, has a relatively high **death rate**.

Prevention

Prevention of sinusitis involves the usual standards of good hygiene to cut down on the number of colds an

individual catches. Quitting smoking or avoiding exposure to cigarette smoke, identifying and treating allergies, and avoiding deep dives in swimming pools may help prevent sinus infections. During the winter, it is a good idea to use a humidifier. Dry nasal passages may crack, allowing bacteria to enter. When allergies are diagnosed, a number of nasal sprays are available to try to prevent inflammation within the nasal passageways, thus allowing the normal flow of mucus.

The American Academy of Otolaryngology—Head and Neck Surgery adds the following suggestions for preventing sinusitis:

- Blowing the nose gently, blocking one nostril while blowing through the other.
- Avoiding air travel when other methods of transportation are available. Those who must travel by air should use a nasal spray decongestant before takeoff to prevent blockage of the sinuses.
- People with allergies should minimize their exposure to known allergens as well as using decongestants.

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American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Road, Alexandria, VA, 22314, (703) 836-4444, <http://www.entnet.org/>.

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

National Institute of Allergy and Infectious Diseases (NIAID), 6610 Rockledge Drive, MSC 6612, Bethesda, MD, 20892-6612, (301) 496-5717, (866) 284-4107, (301) 402-3573, <http://www3.niaid.nih.gov>.

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Situs inversus

Definition

Situs inversus is a condition in which the organs of the chest and abdomen are arranged in a perfect mirror image reversal of the normal positioning.

Description

Normal human development results in an asymmetrical arrangement of the organs within the chest and abdomen. Typically, the heart lies on the left side of the body (*levocardia*), the liver and spleen lie on the right, and the lung on the left has two lobes while the lung on the right has three lobes. This normal arrangement is known as *situs solitus*.

However, in about 1 in 8,500 people, the organs of the chest and abdomen are arranged in the exact opposite position: the heart is on the right (*dextrocardia*), as is the two-lobed lung, and the liver, spleen, and three-lobed lung are on the left. Yet because this arrangement, called *situs inversus*, is a perfect mirror image, the relationship between the organs is not changed, so functional problems rarely occur.

Causes and symptoms

Early in the normal development of an embryo, the tube-like structure that becomes the heart forms a loop toward the left, identifying the left/right axis along which the other organs should be positioned. Although the mechanism that causes the heart loop to go left is not fully understood, at least one gene has been identified to have a role in this process. However, it is thought that many factors may be involved in causing *situs inversus*. Rarely, *situs inversus* can run in families but most often it is an isolated and accidental event occurring in an individual for the first time in the family.

Most people with *situs inversus* have no medical symptoms or complications resulting from the condition. Although only 3–5% of people with *situs inversus* have any type of functional heart defect, this is higher than the rate of heart defects in the general population, which is less than 1%.

It is estimated that about 25% of people with *situs inversus* have an underlying condition called primary ciliary dyskinesia (PCD). PCD, also known as Kartagener's syndrome, is characterized as *situs inversus*, chronic sinus infections, increased mucous secretions from the lungs, and increased susceptibility to respiratory infections. PCD is caused by a defect in the cilia that impairs their normal movements.

Diagnosis

Situs inversus should be detected by a thorough **physical examination**. It is often picked up when a physician, using a stethoscope, hears otherwise normal heart sounds on the right side of the body instead of the left. To confirm the suspected diagnosis of *situs inversus*, imaging studies such as MRI, CT, or ultrasound may

KEY TERMS

Cilia—Tiny hairlike projections on certain cells within the body; cilia produce lashing or whipping movements to direct or cause motion of substances or fluids within the body.

CT—A special technique that uses a computer to create a cross-sectional image of the body from a series of x rays.

Gene—A single unit of genetic information, providing the body with instruction for a specific biological task.

MRI—An imaging study that uses magnetic forces to produce an image of the body's internal structures.

Ultrasound—An imaging study that uses high-frequency sound waves to form a visual image of the body's internal structures.

be ordered, and a referral may be made to a cardiologist or internist for completeness. Imaging studies will also rule out the possibility of random arrangement of the organs, or heterotaxy, which has a much higher risk for serious medical complications.

Treatment

There is no treatment for situs inversus. In the unlikely case that a heart defect is present, it should be treated accordingly by a cardiologist.

Individuals who have situs inversus should be sure to inform all physicians involved in their medical care. In addition to preventing unnecessary confusion, this will reduce the risk of missing a crucial diagnosis that presents with location-specific symptoms (such as **appendicitis**).

Alternative treatment

Not applicable.

Prognosis

The prognosis for an individual with situs inversus is good, and in the absence of a heart defect or other underlying diagnosis, life expectancy is normal.

Prevention

There is no known method of preventing situs inversus.

Resources

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ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

NIH/National Heart, Lung and Blood Institute, PO Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov>.

Stefanie B. N. Dugan, M.S.

Sitz bath

Definition

A sitz bath (also called a hip bath) is a type of bath in which only the hips and buttocks are soaked in water or saline solution. Its name comes from the German verb "sitzen," meaning "to sit."

Purpose

A sitz bath is used for patients who have had surgery in the area of the rectum, or to ease the **pain** of **hemorrhoids**, uterine cramps, prostate infections, painful ovaries, and/or testicles. It is also used to ease discomfort from infections of the bladder, prostate, or vagina. Inflammatory bowel diseases are also treated with sitz baths.

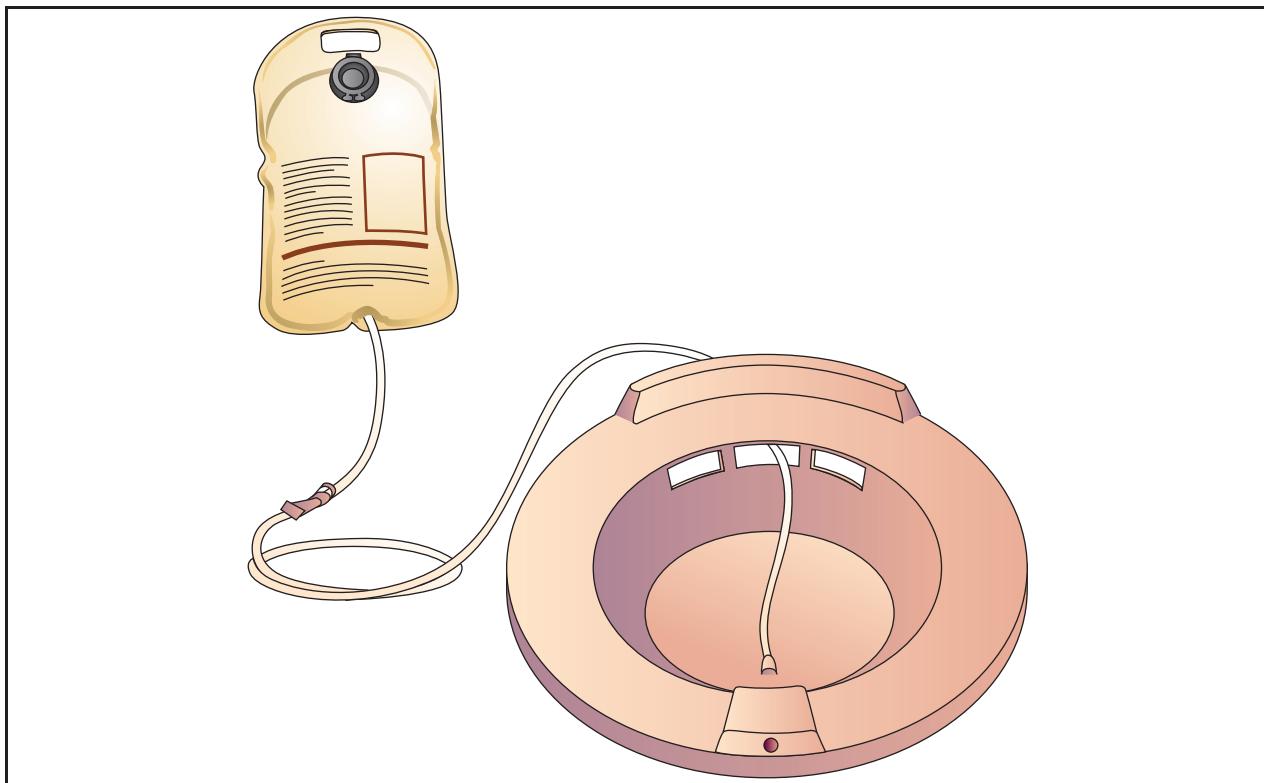
Precautions

Some patients may become dizzy when standing up after sitting in hot water; it is best to have someone else present when doing a contrast sitz bath.

Description

The sitz bath is a European tradition in which only the pelvis and abdominal area are placed in water, with the upper body, arms, legs, and feet out of the water. The water can be warm or cool and one or two tubs may be used.

Warm sitz baths are one of the easiest and most effective ways to ease the pain of hemorrhoids. A warm bath is also effective in lessening the discomfort



Equipment used for sitz baths. A sitz bath, in which only the hips and buttocks are soaked in water or saline solution, is used for patients who have had surgery in the rectal area or to ease discomfort from bladder, prostate, or vaginal infections. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

associated with **genital herpes**, uterine cramps, and other painful conditions in the pelvic area.

For prostate pain, patients should take two hot sitz baths a day, for about 15 minutes each.

To ease discomfort from a vaginal yeast infection, women should take a warm saline sitz bath. To prepare, fill the tub to hip height with warm water and add 1/2 cup of salt (enough to make the water taste salty) and 1/2 cup of vinegar. Sit in the bath for 20 minutes (or until the water gets cool). The vinegar will help bring the vaginal pH back to 4.5 (pH is a measurement of how acid or alkaline a fluid is).

A brief, cool sitz bath helps ease inflammation, **constipation**, and vaginal discharge. It can be used to tone the muscles in cases of bladder or bowel incontinence.

Other conditions respond to a “contrast bath” of both hot and cold. For this, a patient should have a tub of hot water (about 110°F/43°C) and one tub of ice water. The patient should sit in the hot water for 3–4 minutes and in the cold for 30–60 seconds. This is repeated 3–5 times, always ending with the cold water.

If two tubs are not handy, the patient may sit in a hot bath (up to the navel). Then the patient stands up in the water and pulls a cold towel between the legs and over the pelvis in front and back. The cold towel is held in place for up to 60 seconds. Then the patient should sit back into the hot bath, and repeat the process 3–5 times, ending with the cold towel.

Preparation

The bath should be filled with 3–4 in (8–10cm) of water. For most conditions, nothing else should be added (no bubble bath or oil).

Aftercare

The area should be carefully patted dry and, if necessary, clean **dressings** should be applied.

Risks

Sitz baths pose almost no risk. On rare occasions, patients can feel dizzy or experience rapid heart beat because of blood vessel dilation.

KEY TERMS

pH—A standard laboratory test that measures how acidic or alkaline a solution is.

Saline solution—Another word for salt water.

Normal results

Swelling goes down; discomfort is eased; healing is promoted.

Carol A. Turkington

Sjögren's syndrome

Definition

Sjögren's syndrome (SS) is a disorder in which the mouth and eyes become extremely dry. Sjögren's syndrome is often associated with other **autoimmune disorders**. It is named for Henrik Sjögren, a Swedish ophthalmologist.

Description

Like other autoimmune disorders, Sjögren's syndrome occurs when the body's immune system mistakenly begins treating parts of the body as foreign invaders. While the immune cells should attack and kill invaders like bacteria, viruses, and fungi, these cells should not attack the body itself. In autoimmune disorders, however, cells called antibodies see tissues of the body as foreign, and help to start a chain of events that results in damage and destruction of those tissues.

There are three types of Sjögren's syndrome. Primary Sjögren's syndrome occurs by itself, with no other associated disorders. Secondary Sjögren's syndrome occurs along with other autoimmune disorders, like **systemic lupus erythematosus, rheumatoid arthritis, scleroderma, vasculitis, or polymyositis**. When the disorder is limited to involvement of the eyes, with no other organ or tissue involvement evident, it is called sicca complex.

Women are about nine times more likely to suffer from Sjögren's syndrome than are men. SS affects all age groups, although most patients are diagnosed when they are between 40 and 55 years old. Sjögren's syndrome is commonly associated with other autoimmune disorders. In fact, 30% of patients with certain autoimmune disorders will also have Sjögren's syndrome.

SS is found in all races and ethnic groups. It is thought to affect between 0.1% and 3% of the population in the United States; this range reflects the lack of a uniform set of diagnostic criteria. According to the American College of Rheumatology, between 1 million and 4 million Americans have Sjögren's syndrome.

Causes and symptoms

The cause of Sjögren's syndrome has not been clearly defined but several causes are suspected. The syndrome sometimes runs in families. Other potential causes include hormonal factors (since there are more women than men with the disease) and viral factors. The viral theory suggests that the immune system is activated in response to a viral invader but then fails to turn itself off. Some other immune malfunction then causes the overly active immune system to begin attacking the body's own tissues. In 2004 a group of Greek researchers presented evidence that a coxsackievirus may be the disease organism that triggers SS.

The main problem in Sjögren's syndrome is dryness. The salivary glands are often attacked and slowly destroyed, leaving the mouth extremely dry and sticky. Swallowing and talking become difficult. Normally, the saliva washes the teeth clean. Saliva cannot perform this function in Sjögren's syndrome, so the teeth develop many cavities and decay quickly. The parotid glands produce the majority of the mouth's saliva. They are located lying over the jaw bones behind the area of the cheeks and in front of the ears, and may become significantly enlarged in Sjögren's syndrome.

The eyes also become extremely dry as the tear glands (called glands of lachrymation) are slowly destroyed. Eye symptoms include **itching**, burning, redness, increased sensitivity to light, and thick secretions gathering at the eye corners closest to the nose. The cornea may have small irritated pits in its surface (ulcerations).

Destruction of glands in other areas of the body may cause a variety of symptoms. In the nose, dryness may result in nosebleeds. In the rest of the respiratory tract, the rates of ear infection, hoarseness, **bronchitis**, and **pneumonia** may increase. Vaginal dryness can be quite uncomfortable. Rarely, the pancreas may slow production of enzymes important for digestion. The kidney may malfunction. About 33% of all patients with Sjögren's syndrome have other symptoms unrelated to gland destruction. These symptoms include **fatigue**, decreased energy, fevers, muscle aches and pains, and joint **pain**.

Many patients with SS also develop a variety of skin problems that include dry patches, vasculitis, and cutaneous B-cell lymphoma. These and other

dermatologic disorders are more common in SS than was previously thought.

Patients who also have other autoimmune diseases will suffer from the symptoms specific to those conditions.

In addition to physical symptoms, patients with SS appear to be at increased risk for depression and other **mood disorders**.

Diagnosis

Diagnosis of Sjögren's syndrome is based on the patient having at least three consecutive months of bothersome eye and/or mouth dryness. A variety of tests can then be done to determine the quantity of tears produced, the quantity of saliva produced, and the presence or absence of antibodies that could be involved in the destruction of glands.

Treatment

There is no cure for Sjögren's syndrome. Instead, treatment usually attempts to reduce the discomfort and complications associated with dryness of the eyes and mouth (and other areas). Artificial tears are available and may need to be used up to every 30 minutes. By using these types of products, the patient is more comfortable and avoids the complications associated with eyes that are overly dry. **Dry mouth** is treated by sipping fluids slowly but constantly throughout the day. Sugarless chewing gum can also be helpful. An artificial saliva is available for use as a mouthwash. Patients may also be given such drugs as pilocarpine (Salagen) or cevimeline (Evoxac) to increase saliva and tear secretions. Careful dental hygiene is important in order to avoid **tooth decay** and it is wise for patients to decrease sugar intake. Vaginal dryness can be treated with certain gel preparations. Steroid medications may be required when other symptoms of autoimmune disorders complicate Sjögren's syndrome. However, these medications should be avoided when possible because they may make the cornea thin and even more susceptible to injury.

Prognosis

The prognosis for patients with primary Sjögren's syndrome is particularly good; these patients have a normal life expectancy. Although the condition is quite annoying, serious complications rarely occur. The prognosis for patients with secondary Sjögren's syndrome varies since it depends on the prognosis for the accompanying autoimmune disorder.

KEY TERMS

Autoimmune disorder—A disorder in which the body's immune cells mistake the body's own tissues as foreign invaders; the immune cells then work to destroy tissues in the body.

Cornea—A transparent structure of the eye over the iris and pupil; light must pass through the cornea to make vision possible.

Coxsackievirus—Any of a group of enteroviruses that produce a disease in humans characterized by fever and rash. Coxsackieviruses are named for the town in upstate New York where they were first identified.

Immune system—The complex network of organs and blood cells that protect the body from foreign invaders, like bacteria, viruses, and fungi.

Prevention

Since the cause of Sjögren's syndrome is unknown, there are no known ways to prevent this syndrome.

Resources

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ORGANIZATIONS

American College of Rheumatology, 2200 Lake Boulevard NE, Atlanta, GA, 30319, (404) 633-3777, (404) 633-1870, acr@rheumatology.org, <http://www.rheumatology.org/>.

Rosalyn Carson-DeWitt, MD
Rebecca J. Frey, PhD

Skeletal traction see **Traction**;

Immobilization

Skin abrasion see **Skin resurfacing**

Skin allergy test see **Allergy tests**

Skin biopsy

Definition

A skin biopsy is a procedure in which a small piece of living skin is removed from the body for examination, usually under a microscope, to establish a precise diagnosis. Skin biopsies are usually brief, straightforward procedures performed by a skin specialist (dermatologist) or family physician.

Purpose

The word *biopsy* is taken from Greek words that mean "to view life." The term describes what a specialist in identifying diseases (pathologist) does with tissue obtained from a skin biopsy. The pathologist *visually* examines the tissue under a microscope.

A skin biopsy is used to make a diagnosis of many skin disorders. Information from the biopsy also helps the doctor choose the best treatment for the patient.

Doctors perform skin biopsies to:

- make a diagnosis
- confirm a diagnosis made from the patient's medical history and a physical examination
- check whether a treatment prescribed for a previously diagnosed condition is working
- check the edges of tissue removed with a tumor to make certain it contains all the diseased tissue

Skin biopsies also can serve a therapeutic purpose. Many skin abnormalities (lesions) can be removed completely during the biopsy procedure.

Precautions

A patient taking **aspirin** or another blood thinner (anticoagulant) may be asked to stop taking them a week or more before the skin biopsy. This adjustment in medication will prevent excessive bleeding during the procedure and allow for normal blood clotting.

Some patients are allergic to lidocaine, the numbing agent most frequently used during a skin biopsy. The doctor can usually substitute another anesthetic agent.

Description

The first part of the skin biopsy test is obtaining a sample of tissue that best represents the lesion being evaluated. Many biopsy techniques are available. The choice of technique and precise location from which to take the biopsy material are determined by factors such as the type and shape of the lesion. Biopsies can be classified as excisional or incisional. In excisional biopsy, the lesion is completely removed; in incisional biopsy, a portion of the lesion is removed.

The most common biopsy techniques are:

- Shave biopsy. A scalpel or razor blade is used to shave off a thin layer of the lesion parallel to the skin.
- Punch biopsy. A small cylindrical punch is screwed into the lesion through the full thickness of the skin and a plug of tissue is removed. A stitch or two may be needed to close the wound.
- Scalpel biopsy. A scalpel is used to make a standard surgical incision or excision to remove tissue. This technique is most often used for large or deep lesions. The wound is closed with stitches.
- Scissors biopsy. Scissors are used to snip off surface (superficial) skin growths and lesions that grow from a stem or column of tissue. Such growths are sometimes seen on the eyelids or neck.

After the biopsy tissue is removed, bleeding may be controlled by applying pressure or by burning with electricity or chemicals. **Antibiotics** often are applied to the wound to prevent infection. Stitches may be placed in the wound, or the wound may be bandaged and allowed to heal on its own.

The second part of the skin biopsy test is handling and examining the tissue sample. Drying and structural damage to the tissue sample must be prevented, so it should be placed immediately in an appropriate preservative, such as formaldehyde.

The pathologist can use a variety of laboratory techniques to process the biopsy tissue. Tissue stains and several different kinds of microscopes are used. Because there are many skin disorders (broadly called

KEY TERMS

Benign—Noncancerous.

Dermatitis—A skin disorder that causes inflammation, that is, redness, swelling, heat, and pain.

Dermatologist—A doctor who specializes in skin care and treatment.

Dermatoses—A noninflammatory skin disorder.

Lesion—An area of abnormal or injured skin.

Malignant—Cancerous.

Pathologist—A person who specializes in studying diseases. In particular, this person examines the structural and functional changes in the tissues and organs of the body that are caused by disease or that cause disease themselves.

dermatosis and **dermatitis**), the pathologist has extensive training in their accurate identification. Cases of melanoma, the most malignant kind of skin **cancer**, have almost tripled in the past 30 years. Because melanoma grows very rapidly in the skin, quick and accurate diagnosis is important.

Preparation

The area of the biopsy is cleansed thoroughly with alcohol or a disinfectant containing iodine. Sterile cloths (drapes) may be positioned, and a local anesthetic, usually lidocaine, is injected into the skin near the lesion. Sometimes the anesthetic contains epinephrine, a drug that helps reduce bleeding during the biopsy. Sterile gloves and surgical instruments are always used to reduce the risk of infection.

Aftercare

If stitches have been placed, they should be kept clean and dry until removed. Stitches are usually removed five to 10 days after the biopsy. Sometimes the patient is instructed to put protective ointment on the stitches before showering. **Wounds** that have not been stitched should be cleaned with soap and water daily until they heal. Adhesive strips should be left in place for two to three weeks. **Pain** medications usually are not necessary.

Risks

Infection and bleeding occur rarely after skin biopsy. If the skin biopsy may leave a scar, the patient usually is asked to give informed consent before the test.

Normal results

The biopsy reveals normal skin layers.

Abnormal results

The biopsy reveals a noncancerous (benign) or cancerous (malignant) lesion. Benign lesions may require treatment.

ORGANIZATIONS

American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168-4014, (847) 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

Collette L. Placek

Skin cancer see **Malignant melanoma**

Skin cancer, non-melanoma

Definition

Non-melanoma skin **cancer** is a malignant growth of the external surface or epithelial layer of the skin.

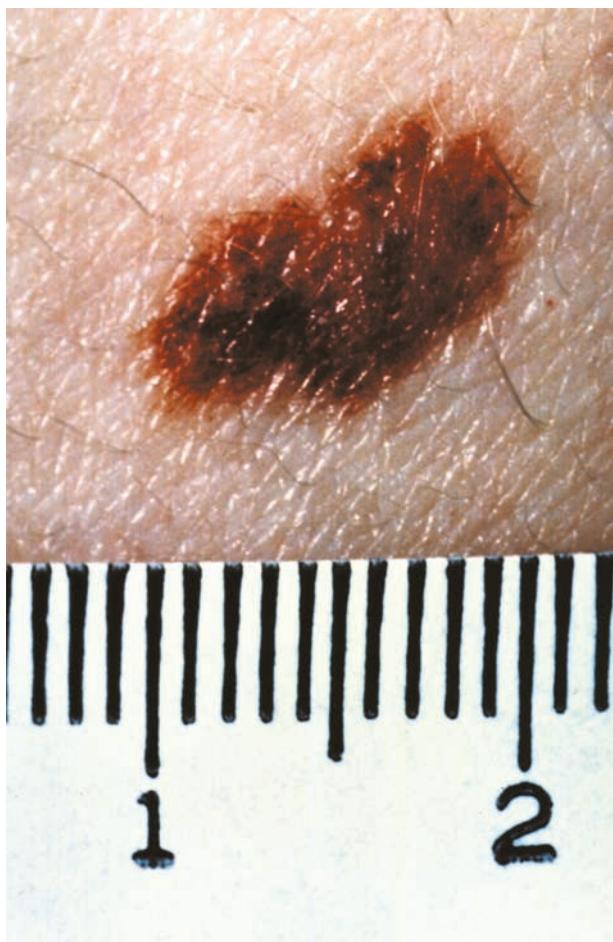
Demographics

Skin cancers are the most common type of cancer by far in the United States. Approximately 800,000 to 900,000 new cases of basal cell skin cancer are diagnosed each year. Squamous cell skin cancers are diagnosed less frequently with 200,000 to 300,000 new cases diagnosed annually. The number of new cases of non-melanoma skin cancers is increasing each year. This increase is attributed to improved detection capabilities, increased exposure to the sun, and increase in the lifespan of the general population. Most of the time, basal cell and squamous cell skin cancers are not fatal. The American Cancer Society reports a decline of about 30% in deaths from skin cancer over the last three decades.

Description

Risk factors

Exposure to sunlight is documented as the main cause of more than 1 million cases of non-melanoma skin cancers diagnosed each year in the United States. Incidence increases for those living where direct sunshine is plentiful, such as near the equator.



A close-up image of a precancerous mole that could develop into a melanoma. Melanomas arise from pigment-producing cells, while non-melanoma skin cancer arises from squamous cells or basal cells. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Ultraviolet B (UVB) rays are thought to cause most basal cell and squamous cell skin cancers. Ultraviolet A (UVA) rays may also directly cause some skin cancers. In addition to sunlight, overexposure to UVB rays can occur from the use of **tanning** booths and beds and from sunlamps.

People who are at highest risk for the development of skin cancer include individuals who have fair skin and light-colored eyes and who freckle or burn easily when exposed to UVB rays.

Other individuals at high risk include older adults because exposure increases over time. Males are two to three times as likely to develop skin cancer as females. Exposure to chemicals such as arsenic, industrial tar, coal, paraffin, and certain types of oil can lead to skin cancer. Other risk factors include a history of **smoking**,

a history of previous skin cancer, and history of illnesses, diseases, or conditions which impair immunity.

Skin cancer is the growth of abnormal cells capable of invading and destroying other associated skin cells. Skin cancer is often subdivided into either melanoma or non-melanoma. Melanoma is a dark-pigmented, usually malignant tumor arising from a skin cell capable of making the pigment melanin (a melanocyte). Melanomas can also develop from benign tissue such as **moles**. Non-melanoma skin cancer most often originates from the outermost skin surface as a squamous cell carcinoma or from cells in the basal layer, the deepest part of the epidermis. Cancers of the latter type are termed basal cell carcinomas. Basal cell and squamous cell skin cancers may also be referred to as keratinocyte cancers.

Other types of skin cancers which occur less frequently are: Merkel cell carcinoma, Kaposi sarcoma, cutaneous lymphoma, skin adenexal tumors, and various types of **sarcomas**. Combined, the incidence of all of these rarer types of non-melanoma skin cancer account for less than one percent of skin cancer.

Basal cell carcinoma affects the skin's basal layer and has the potential to grow progressively larger in size, although it rarely spreads to distant areas (metastasizes). Basal cell carcinomas account for 80% of skin cancers (excluding melanoma), whereas squamous cell cancer makes up about 20%. Basal cell cancer tends to recur, with approximately 50% of people diagnosed with basal cell cancer developing a new skin cancer within five years. Squamous cell carcinoma is a malignant growth of the external surface of the skin. Squamous cell cancers metastasize at a rate of 2–6%, with up to 10% of lesions affecting the ear and lip. Squamous cell carcinomas appear to be more aggressive than basal cell cancers.

Causes and symptoms

Cumulative sun exposure is considered a significant risk factor for non-melanoma skin cancer. There is evidence suggesting that early, intense exposure causing blistering **sunburn** in childhood may also play an important role in the cause of non-melanoma skin cancer. Basal cell carcinoma most frequently affects the skin of the face, with the next most common sites being the ears, the backs of the hands, the shoulders, and the arms. It is prevalent in both sexes and most common in people over 40.

About 1–2% of all skin cancers develop within burn **scars**; squamous cell carcinomas account for about 95% of these cancers, with 3% being basal cell carcinomas and the remainder malignant melanomas.

KEY TERMS

Autoimmune—Pertaining to an immune response by the body against one of its own tissues or types of cells.

Curettage—The removal of tissue or growths by scraping with a curette.

Dermatologist—A physician specializing in the branch of medicine concerned with skin.

Electrodesiccation—To make dry, dull, or lifeless with the use of electrical current.

Lesion—A patch of skin that has been infected or diseased.

Topical—Referring to a medication or other preparation applied to the skin or the outside of the body.

Basal cell carcinomas usually appear as small **skin lesions** that persist for at least three weeks. This form of non-melanomatous skin cancer looks flat and waxy with the edges of the lesion translucent and rounded. The edges also contain small fresh blood vessels. An ulcer in the center of the lesion gives it a dimpled appearance. Basal cell carcinoma lesions vary from four to six millimeters in size, but can slowly grow larger if untreated.

Squamous cell carcinoma also involves skin exposed to the sun, such as the face, ears, hands, or arms. This form of non-melanoma is also most common among people over 40. Squamous cell carcinoma presents as a small, scaling, raised bump on the skin with a crusting ulcer in the center, but without **pain** and **itching**. The lesion may also appear as flat, reddish, slow-growing patches.

Basal cell and squamous cell carcinomas can grow more easily when people have a suppressed immune system because they are taking immunosuppressive drugs or are exposed to radiation. Some people must take immunosuppressive drugs to prevent the rejection of a transplanted organ or because they have a disease in which the immune system attacks the body's own tissues (autoimmune illnesses); others may need **radiation therapy** to treat another form of cancer. Because of this, everyone taking immunosuppressive drugs or receiving radiation treatments should undergo complete skin examination at regular intervals. If proper treatment is delayed and the tumor continues to grow, tumor cells can spread (metastasize) to muscle, bone, nerves, and possibly the brain.

Diagnosis

Examination

To diagnose skin cancer, clinicians must carefully examine the lesion and ask the patient about how long it has been there, whether it itches or bleeds, and other questions about the patient's medical history. Lymph nodes in the vicinity of the suspicious lesion will be palpated.

The patient may be referred to a dermatologist for a more comprehensive examination. The dermatologist may use a device known as a dermatoscope to visualize spots on the skin more clearly.

Procedures

If skin cancer cannot be ruled out, a sample of tissue is removed and examined under a microscope (a biopsy). A definitive diagnosis of squamous or basal cell cancer can only be made with microscopic examination of the tumor cells. Once skin cancer has been diagnosed, the stage of the disease's development is determined. The information from the biopsy and staging allows the physician and patient to plan for treatment and possible surgical intervention.

Treatment

Traditional

A variety of treatment options are available for those diagnosed with non-melanoma skin cancer. Some carcinomas can be removed by cryosurgery, the process of freezing with liquid nitrogen. Uncomplicated and previously untreated basal cell carcinoma of the trunk and arms is often treated with curettage and electrodesiccation, which is the scraping of the lesion and the destruction of any remaining malignant cells with an electrical current. Removal of a lesion layer-by-layer down to normal margins (Mohs' surgery) is an effective treatment for both basal and squamous cell carcinoma. Removal of larger tumors may require **skin grafting** and **reconstructive surgery**.

Other treatments for non-melanoma skin cancer include **photodynamic therapy** (PDT), topical **chemotherapy** in which the anticancer drug is applied to the lesion as an ointment or as a cream, **laser surgery**, and the use of drugs such as imiquimod and interferon. These drugs are classified as immune response modifiers. The drugs work to boost the body's immune system to help decrease the size of the lesion and sometimes are effective in eliminating the skin cancer altogether.

Radiation therapy is best reserved for older, debilitated patients or when the tumor is considered inoperable.

Prognosis

Both squamous and basal cell carcinoma are curable with appropriate treatment, although basal cell carcinomas have a higher rate of recurrence. Early detection remains critical for a positive prognosis. Although it is rare for basal cell carcinomas to metastasize, metastases can rapidly lead to **death** if the tumor cells invade the eyes, ears, mouth, or the membranes covering the brain.

Prevention

Not all skin cancers can be prevented. However, there are ways to reduce risk for skin cancer. Avoiding exposure to the sun reduces the incidence of non-melanoma skin cancer. Sunscreen and sunblock preparations provide protection against both UVA and UVB rays. These preparations should also be rated with a sun protection factor (SPF) of 30 or higher. They should be applied 30 minutes before going outdoors and then reapplied every two hours and after swimming. Other recommended practices are to wear a hat, sunglasses, and clothing to shield the skin from sun damage. The lips should be protected by wearing lip balm with sunscreen.

Other strategies include avoiding the outdoors during times of maximum UV effects which is typically between the hours of 10 a.m. until 4 p.m. especially on days when the UV index is high. Check online at www.epa.gov/sunwise/uvindex.html to determine the UV index in your area on any particular day. Avoiding tanning beds, tanning booths, and sunlamps is also strongly recommended. Adults should consider applying protective wear for children. Such wear is designed to cover the child from the neck to the knees with sun-protective fabric.

People should examine their skin monthly for unusual lesions, especially if previous skin cancers have been experienced.

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ORGANIZATIONS

- American Academy of Dermatology (AAD), PO Box 4014, Schaumburg, IL, 60168, (866) 503-7546, <http://www.aad.org>.
- American Cancer Society (ACS), (800) 227-2345, <http://www.cancer.org>.
- Canadian Cancer Society (CCS), 10 Alcorn Ave., Suite 200, Toronto, ON, M4V 3B1, (416) 961-7223, <http://www.cancer.ca>.
- National Cancer Institute (NCI), 6116 Executive Blvd., Suite 300, Bethesda MD, 20892-8322, (800) 422-6237, <http://www.cancer.gov>.
- Skin Cancer Foundation (SCF), 149 Madison Ave., Suite 901, New York NY, 10016, (212) 725-5176, <http://www.skincancer.org>.

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Skin culture

Definition

A skin culture is a test that is done to identify the microorganism (bacteria, fungus, or virus) causing a skin infection and to determine the antibiotic or other treatment that will effectively treat the infection.

Purpose

Microorganisms can infect healthy skin but more often they infect skin already damaged by an injury or abrasion. Skin infections are contagious and, if left untreated, can lead to serious complications. A culture enables a physician to diagnose and treat a skin infection.

Description

Several groups of microorganisms cause skin infections: bacteria, fungi (molds and yeast), and viruses. Based on the appearance of the infection, the physician determines what group of microorganisms is likely causing the infection, then he or she collects a specimen for one or more types of cultures. A sample of material—such as skin cells, pus, or fluid—is taken from the infection site, placed in a sterile container, and sent to the laboratory. In the laboratory, each type of culture is handled differently.

Bacterial infections are the most common. Bacteria cause lesions, ulcers, **cellulitis**, and **boils**. Pyoderma are pus-containing skin infections, such as **impetigo**, caused by *Staphylococcus* or group A *Streptococcus* bacteria. To culture bacteria, a portion of material from the infection site is spread over the surface of a culture plate and placed in an incubator at body temperature for one to two days. Bacteria in the skin sample multiply and appear on the plates as visible colonies. They are identified by noting the appearance of their colonies and by performing biochemical tests and a Gram's stain.

The Gram's stain is done by smearing part of a colony onto a microscope slide. After it dries, the slide is colored with purple and red stains, then examined under a microscope. The color of stain picked up and retained by the bacteria (purple or red), their shape (such as round or rectangle), and their size provide valuable clues as to their identity.

A sensitivity test, also called antibiotic susceptibility test, is also done. The bacteria are tested against different **antibiotics** to determine which will effectively treat the infection by killing the bacteria.

Fungal cultures are done less frequently. A group of fungi called dermatophytes cause a skin infection called **ringworm**. Yeast causes an infection called thrush. These infections are usually diagnosed using a method other than culture, such as the **KOH test**. A culture is done only when specific identification of the mold or yeast is necessary. The specimen is spread on a culture plate designed to grow fungi, then incubated.

KEY TERMS

Pyoderma—A pus-containing skin infection, such as impetigo, caused by *Staphylococcus* or group A *Streptococcus* bacteria.

Sensitivity test—A test that determines which antibiotics will treat an infection by killing the bacteria.

Several different biochemical tests and stains are used to identify molds and yeasts.

Viruses, such as herpes, can also cause skin infections. Specimens for viral cultures are mixed with commercially-prepared animal cells in a test tube. Characteristic changes to the cells caused by the growing virus help identify the virus.

Results for bacterial cultures are usually available in one to three days. Cultures for fungi and viruses may take longer—up to three weeks. Cultures are covered by insurance.

Preparation

After cleaning the infected area with sterile saline and alcohol, the physician collects skin cells, pus, or fluid using a needle or swab. If necessary, the physician will open a lesion to collect the specimen. To collect a specimen for a fungal culture, the physician uses a scalpel to scrape skin cells into a sterile container.

Normal results

Many types of microorganisms are normally found on a person's skin. Presence of these microorganisms is noted on a skin culture report as "normal flora."

Abnormal results

A microorganism is considered to be a cause of the infection if it is either the only or predominant microorganism that grew, if it grew in large numbers, or if it is known to produce infection.

Resources

PERIODICALS

Carroll, John A. "Common Bacterial Pyodermas." *Post-graduate Medicine* September 1996: 311–322.

Nancy J. Nordenson

Skin grafting

Definition

Skin grafting is a surgical procedure by which skin or skin substitute is placed over a burn or non-healing wound to permanently replace damaged or missing skin or provide a temporary wound covering.

Purpose

Wounds such as third-degree **burns** must be covered as quickly as possible to prevent infection or loss of fluid. Wounds that are left to heal on their own can contract, often resulting in serious scarring; if the wound is large enough, the scar can actually prevent movement of limbs. Non-healing wounds, such as diabetic ulcers, venous ulcers, or pressure sores, can be treated with skin grafts to prevent infection and further progression of the wounded area.

Precautions

Skin grafting is generally not used for first- or second-degree burns, which generally heal with little or no scarring. Also, the tissue for grafting and the recipient site must be as sterile as possible to prevent later infection that could result in failure of the graft.

Description

The skin is the largest organ of the human body. It consists of two main layers: the epidermis is the outer layer, sitting on and nourished by the thicker dermis. These two layers are approximately 0.04–0.08 in (1–2 mm) thick. The epidermis consists of an outer layer of dead cells, which provides a tough, protective coating, and several layers of rapidly dividing cells called keratinocytes. The dermis contains the blood vessels, nerves, sweat glands, hair follicles, and oil glands. The dermis consists mainly of connective tissue, primarily the protein collagen, which gives the skin its flexibility and provides structural support. Fibroblasts, which make collagen, are the main cell type in the dermis.

Skin protects the body from fluid loss, aids in temperature regulation, and helps prevent disease-causing bacteria or viruses from entering the body. Skin that is damaged extensively by burns or non-healing wounds can compromise the health and well-being of the patient. More than 50,000 people are hospitalized for burn treatment each year in the United States, and 5,500 die. Approximately 4 million people suffer from non-healing wounds, including 1.5 million with venous ulcers and 800,000 with diabetic ulcers, which result in 55,000 amputations per year in the United States.

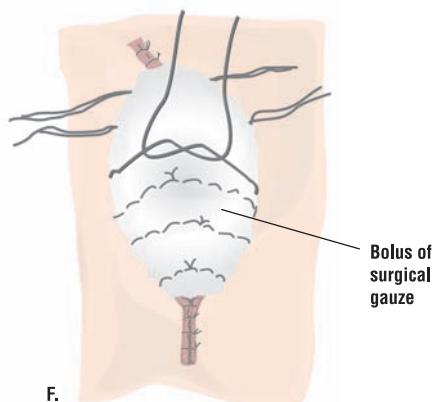
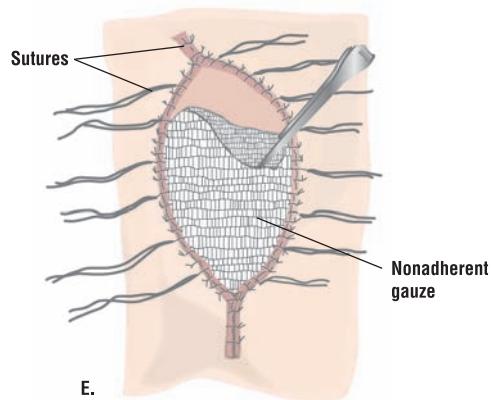
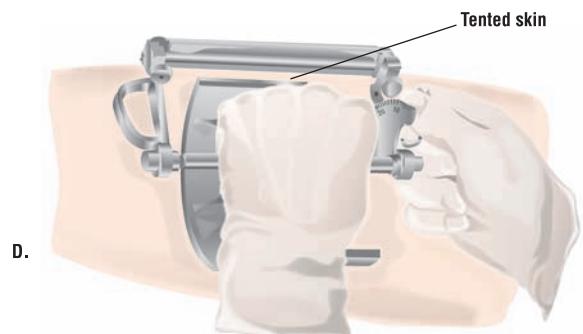
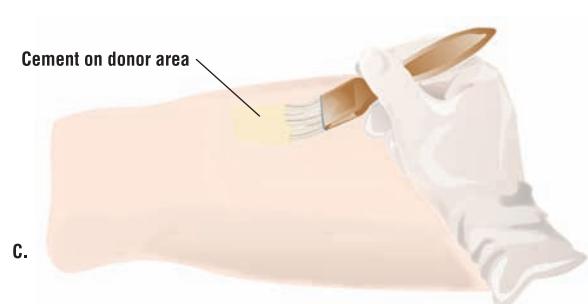
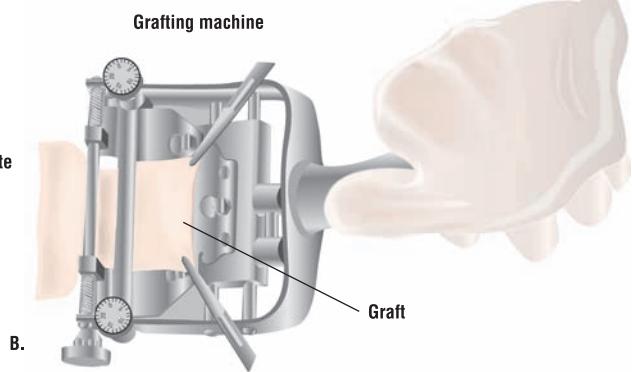
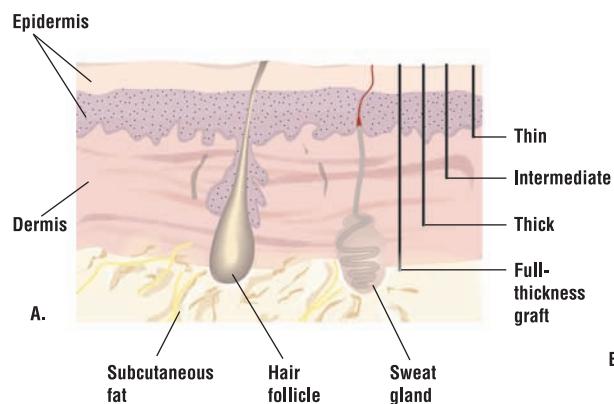
Skin for grafting can be obtained from another area of the patient's body, called an autograft, if there is enough undamaged skin available, and if the patient is healthy enough to undergo the additional surgery required. Alternatively, skin can be obtained from another person (donor skin from cadavers is frozen, stored, and available for use), called an allograft, or from an animal (usually a pig), called a xenograft. Allografts and xenografts provide only temporary covering—they are rejected by the patient's immune system within seven to 10 days and must be replaced with an autograft.

A split-thickness skin graft takes mainly the epidermis and a little of the dermis and usually heals within several days. The wound must not be too deep if a split-thickness graft is going to be successful, since the blood vessels that will nourish the grafted tissue must come from the dermis of the wound itself.

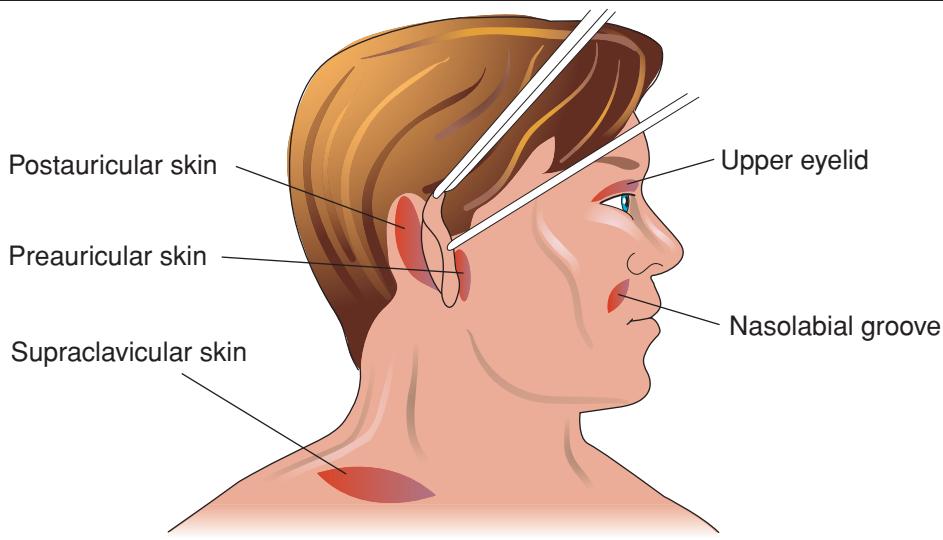
A full-thickness graft involves both layers of the skin. Full-thickness autografts provide better contour, more natural color, and less contraction at the grafted site. The main disadvantage of full-thickness skin grafts is that the wound at the donor site is larger and requires more careful management; often a split-thickness graft must be used to cover the donor site.

A composite skin graft is sometimes used, consisting of combinations of skin and fat, skin and cartilage, or dermis and fat. Composite grafts are used where three-dimensional reconstruction is necessary. For example, a wedge of ear containing skin and cartilage can be used to repair the nose.

Several artificial skin products are available for burns or non-healing wounds. Unlike allographs and xenographs, these products are not rejected by the patient's body and actually encourage the generation of new tissue. Artificial skin usually consists of a synthetic epidermis and a collagen-based dermis. This artificial dermis, the fibers of which are arranged in a lattice, acts as a template for the formation of new tissue. Fibroblasts, blood vessels, nerve fibers, and lymph vessels from surrounding healthy tissue cross into the collagen lattice, which eventually degrades as these cells and structures build a new dermis. The synthetic epidermis, which acts as a temporary barrier during this process, is eventually replaced with a split-thickness autograft or with an epidermis cultured in the laboratory from the patient's own epithelial cells. The cost for the synthetic products is about \$1,000 for a 40 in (100 cm) square piece of artificial skin, in addition to the costs of the surgery. This procedure is covered by insurance.

Skin grafting

Skin grafts may be used in several thicknesses (A). To begin the procedure, a special cement is used on the donor skin area (C). The grafting machine is applied to the area, and sample taken (D). After the graft is stitched to the recipient area, it is covered with nonadherent gauze (E) and a layer of fluffy surgical gauze held in place with sutures (F). (Illustration by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)



Skin grafting is a surgical procedure by which skin or a skin substitute is placed over a burn or non-healing wound to replace the damaged skin or provide a temporary wound covering. Skin for grafting can be obtained from another area of the patient's body, such as the face and neck, as shown in the illustration above. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Aftercare

Once a skin graft has been put in place, even after it has healed, it must be maintained carefully. Patients who have grafts on their legs should remain in bed for seven to 10 days with their legs elevated. For several months the patient should support the graft with an Ace bandage or Jobst stocking. Grafts in other areas of the body should be similarly supported after healing to decrease the amount of contracture.

Grafted skin does not contain sweat or oil glands, and should be lubricated daily for two to three months with a bland oil (e.g. mineral oil) to prevent drying and cracking.

Risks

The risks of skin grafting include those inherent in any surgical procedure that involves anesthesia. These include reactions to the medications, problems breathing, bleeding, and infection. In addition, the risks of an allograft procedure include transmission of **infectious disease**.

Normal results

A skin graft should provide significant improvement in the quality of the wound site, and may prevent the serious complications associated with burns or non-healing wounds.

KEY TERMS

Allograft—Tissue that is taken from one person's body and grafted to another person.

Autograft—Tissue that is taken from one part of a person's body and transplanted to a different part of the same person.

Collagen—A protein that provides structural support; the main component of connective tissue.

Dermis—The underlayer of skin, containing blood vessels, nerves, hair follicles, and oil and sweat glands.

Epidermis—The outer layer of skin, consisting of a layer of dead cells that perform a protective function and a second layer of dividing cells.

Fibroblasts—A type of cell found in connective tissue; produces collagen.

Keratinocytes—Cells found in the epidermis. The keratinocytes at the outer surface of the epidermis are dead and form a tough protective layer. The cells underneath divide to replenish the supply.

Xenograft—Tissue that is transplanted from one species to another (e.g. pigs to humans).

Abnormal results

Failure of a graft can result from poor blood flow, swelling, or infection.

ORGANIZATIONS

American Burn Association, 625 N. Michigan Ave., Suite 2550, Chicago, IL, 60611, (312) 642-9260, (312) 642-9130, info@ameriburn.org, http://www.ameriburn.org.
American Diabetes Association, 1701 North Beauregard Street, Alexandria, VA, 22311, (800) 342-2383, Ask ADA@diabetes.org, http://www.diabetes.org/.

Lisa Christenson, PhD

Skin lesion removal

Definition

Skin lesion removal employs a variety of techniques from relatively simple biopsies, to more complex surgical excisions, to remove lesions that range from benign growths to **malignant melanoma**.

Purpose

Sometimes the purpose of skin lesion removal is to excise an unsightly mole or other cosmetically unattractive skin growth. Other times, physicians will remove a skin lesion to make certain it is not cancerous and, if it proves cancerous, to prevent its spread to other parts of the body.

Precautions

Most skin lesion removal procedures require few precautions. The area to be treated is cleaned before the procedure with alcohol or another antibacterial preparation, but generally it is not necessary to use a sterile operating room. Most procedures are performed on an outpatient basis using a local anesthetic. Some of the more complex procedures may require specialized equipment available only in an outpatient surgery center. Most of the procedures are not highly invasive and, frequently, can be well-tolerated by young and old patients, as well as those with other medical conditions.

Description

A variety of techniques are used to remove **skin lesions**. The particular technique selected will depend on such factors as the seriousness of the lesion, its location, and the patient's ability to tolerate the procedure. Some of the simpler techniques, such as a biopsy or cryosurgery, can be performed by a primary care physician. Some of the more complex techniques, such as excision with a scalpel, electrosurgery, or **laser surgery**, are typically performed by a dermatologic

surgeon, plastic surgeon, or other surgical specialist. Often, the technique selected will depend on how familiar the physician is with the procedure and how comfortable he or she is with performing it.

Biopsy

In this procedure, the physician commonly injects a local anesthetic at the site of the skin lesion, then removes a sample of the lesion so that a definite diagnosis can be made. The sample is sent to a pathology laboratory, where it is examined under a microscope. Certain characteristic skin cells, and their arrangement in the skin, offer clues to the type of skin lesion, and whether it is cancerous or otherwise poses danger. Depending on the results of the microscopic examination, additional surgery may be scheduled.

A variety of methods are used to obtain a **skin biopsy**. The physician may use a scalpel to cut a piece or remove all of the lesion for examination. Lesions that are confined to the surface may be sampled with a shave biopsy, where the physician holds a scalpel blade parallel to the surface of the skin and slides the blade across the base of the lesion, removing a sample. Some physicians use a single-edge razor blade for this instead of a scalpel. A physician may also perform a punch biopsy, in which a small circular punch removes a plug of skin.

Excision

When excising a lesion, the physician attempts to remove it completely by using a scalpel to cut the shape of an ellipse around the lesion. Leaving an elliptical wound, rather than a circular wound, makes it easier to insert stitches. If a lesion is suspected to be cancerous, the physician will not cut directly around the lesion, but will attempt to also remove a healthy margin of tissue surrounding it. This is to ensure that no cancerous cells remain, which would allow the tumor to reappear. To prevent recurrence of basal and squamous cell skin cancers, experts recommend a margin of 0.08–0.16 in (2–4 mm) for malignant melanoma, the margin may be 1.2 in (3 cm) or more.

Destruction

Not all lesions need to be excised. A physician may simply seek to destroy the lesion using a number of destructive techniques. These techniques do not leave sufficient material to be examined by a pathologist, however, and are best used in cases where a visual diagnosis is certain.

- **Cryosurgery.** This technique employs an extremely cold liquid or instrument to freeze and destroy abnormal skin cells that require removal. Liquid

nitrogen is the most commonly used cryogen. It is typically sprayed on the lesion in several freeze-thaw cycles to ensure adequate destruction of the lesion.

- **Curettage.** In this procedure, an instrument with a circular cutting loop at the end is drawn across the lesion, starting at the middle and moving outward. With successive strokes, the physician scrapes portions of the lesion away. Sometimes a physician will use the curet to reduce the size of the lesion before turning to another technique to finish removing it.
- **Electrosurgery.** This utilizes an alternating current to selectively destroy skin tissue. Depending on the type of current and device used, physicians may use electro-surgical equipment to dry up surface lesions (electro-dessication), to burn off the lesion (electrocoagulation), or to cut the lesion (electrosection). One advantage of electrosurgery is that it minimizes bleeding.

Mohs' micrographic surgery

The real extent of some lesions may not be readily apparent to the eye, making it difficult for the surgeon to decide where to make incisions. If some **cancer** cells are left behind, for example, the cancer may reappear or spread. In a technique called Mohs' micrographic surgery, surgeons begin by removing a lesion and examining its margins under a microscope for evidence of cancer. If cancerous cells are found, the surgeon then removes another ring of tissue and examines the margins again. The process is repeated until the margins appear clear of cancerous cells. The technique is considered ideal for aggressive tumors in areas such as the nose or upper lip, where an excision with wide margins may be difficult to repair, and may leave a cosmetically poor appearance.

Lasers

Laser surgery is now applied to a variety of skin lesions, ranging from spider veins to more extensive blood vessel lesions called hemangiomas. Until recently, CO₂ lasers were among the more common laser devices used by physicians, primarily to destroy skin lesions. Other lasers, such as the Nd:YAG and flashlamp-pumped pulse dye laser have been developed to achieve more selective results when used to treat vascular lesions, such as hemangiomas, or pigmented lesions, such as café-au-lait spots.

Preparation

No extensive preparation is required for skin lesion removal. Most procedures can be performed on an outpatient basis with a local anesthetic. The lesion and surrounding area is cleaned with an

KEY TERMS

Curet—A surgical instrument with a circular cutting loop at one end. The curet is pulled over the skin lesion in repeated strokes to remove one portion of the lesion at a time.

Mohs' micrographic surgery—A surgical technique in which successive rings of skin tissue are removed and examined under a microscope to ensure that no cancer is left.

Shave biopsy—A method of removing a sample of skin lesion so it can be examined by a pathologist. A scalpel or razor blade is held parallel to the skin's surface and is used to slice the lesion at its base.

antibacterial compound before the procedure. A sterile operating room is not required.

Aftercare

The amount of aftercare will vary, depending on the skin lesion removal technique. For biopsy, curettage, cryosurgery, and electrosurgery procedures, the patient is told to keep the wound clean and dry. Healing will take at least several weeks, and may take longer, depending on the size of the wound and other factors. Healing times will also vary with excisions and with Mohs' micrographic surgery, particularly if a skin graft or skin flap is needed to repair the resulting wound. Laser surgery may produce changes in skin coloration that often resolve in time. **Pain** is usually minimal following most outpatient procedures, so pain medicines are not routinely prescribed. Some areas of the body, such as the scalp and fingers, can be more painful than others and a pain medicine may be required.

Risks

All surgical procedures present risk of infection. Keeping the wound clean and dry can minimize the risk. **Antibiotics** are not routinely given to prevent infection in skin surgery, but some doctors believe they have a role. Other potential complications include:

- bleeding below the skin, which may create a hematoma and sometimes requires the wound to be reopened and drained,
- temporary or permanent nerve damage resulting from excision in an area with extensive and shallow nerve branches,
- wounds that may reopen after they have been stitched closed, increasing the risk of infection and scarring.

Normal results

Depending on the complexity of the skin lesion removal procedure, patients can frequently resume their normal routine the day of surgery. Healing frequently will take place within weeks. Some excisions will require later reconstructive procedures to improve the appearance left by the original procedure.

Abnormal results

In addition to the complications outlined above, it is always possible that the skin lesion will reappear, requiring further surgery.

ORGANIZATIONS

American Academy of Dermatology, PO Box 4014, Schaumburg, IL, 60168-4014, (847) 240-1859, (866) 503-SKIN (7546), <http://www.aad.org>.

American Society for Dermatologic Surgery, 5550 Meadowbrook Dr., Suite 120, Rolling Meadows, IL, 60008, (847) 956-0900, (847) 956-0999, <http://www.asds.net/>.

American Society of Plastic Surgeons, 444 E. Algonquin Rd, Arlington Heights, IL, 60005, (847) 228-9900, <http://www.plasticsurgery.org/>.

Richard H. Camer

Skin lesions

Definition

A skin lesion is a superficial growth or patch of the skin that does not resemble the area surrounding it.

Description

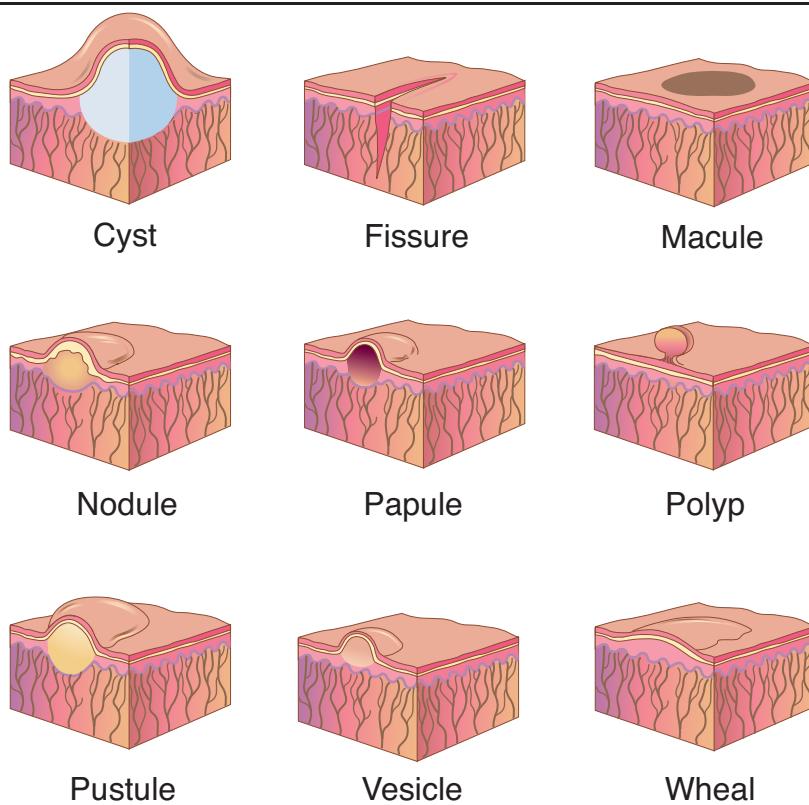
Skin lesions can be grouped into two categories: primary and secondary. Primary skin lesions are variations in color or texture that may be present at birth, such as **moles** or **birthmarks**, or that may be acquired during a person's lifetime, such as those associated with infectious diseases (e.g. **warts**, **acne**, or **psoriasis**), allergic reactions (e.g. **hives** or **contact dermatitis**), or environmental agents (e.g. **sunburn**, pressure, or temperature extremes). Secondary skin lesions are those changes in the skin that result from primary skin lesions, either as a natural progression or as a result of a person manipulating (e.g. scratching or picking at) a primary lesion.

The major types of primary lesions are:

- Macule. A small, circular, flat spot less than $\frac{2}{5}$ in (1 cm) in diameter. The color of a macule is not the same as that of nearby skin. Macules come in a variety of

shapes and are usually brown, white, or red. Examples of macules include freckles and flat moles. A macule more than $\frac{2}{5}$ in (1 cm) in diameter is called a patch.

- Vesicle. A raised lesion less than $\frac{1}{5}$ in (5 mm) across and filled with a clear fluid. Vesicles that are more than $\frac{1}{5}$ in (5 mm) across are called bullae or blisters. These lesions may be the result of sunburns, insect bites, chemical irritation, or certain viral infections, such as herpes.
 - Pustule. A raised lesion filled with pus. A pustule is usually the result of an infection, such as acne, impetigo, or boils.
 - Papule. A solid, raised lesion less than $\frac{2}{5}$ in (1 cm) across. A patch of closely grouped papules more than $\frac{2}{5}$ in (1 cm) across is called a plaque. Papules and plaques can be rough in texture and red, pink, or brown in color. Papules are associated with such conditions as warts, syphilis, psoriasis, seborrheic and actinic keratoses, lichen planus, and skin cancer.
 - Nodule. A solid lesion that has distinct edges and that is usually more deeply rooted than a papule. Doctors often describe a nodule as "palpable," meaning that, when examined by touch, it can be felt as a hard mass distinct from the tissue surrounding it. A nodule more than 2 cm in diameter is called a tumor. Nodules are associated with, among other conditions, keratinous cysts, lipomas, fibromas, and some types of lymphomas.
 - Wheal. A skin elevation caused by swelling that can be itchy and usually disappears soon after erupting. Wheals are generally associated with an allergic reaction, such as to a drug or an insect bite.
 - Telangiectasia. Small, dilated blood vessels that appear close to the surface of the skin. Telangiectasia is often a symptom of such diseases as rosacea or scleroderma.
- The major types of secondary skin lesions are:
- Ulcer. Lesion that involves loss of the upper portion of the skin (epidermis) and part of the lower portion (dermis). Ulcers can result from acute conditions such as bacterial infection or trauma, or from more chronic conditions, such as scleroderma or disorders involving peripheral veins and arteries. An ulcer that appears as a deep crack that extends to the dermis is called a fissure.
 - Scale. A dry, horny build-up of dead skin cells that often flakes off the surface of the skin. Diseases that promote scale include fungal infections, psoriasis, and seborrheic dermatitis.
 - Crust. A dried collection of blood, serum, or pus. Also called a scab, a crust is often part of the normal healing process of many infectious lesions.



A skin lesion is an abnormal growth or an area of skin that does not resemble the skin surrounding it. The illustrations above feature some of the different types of skin lesions. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

- Erosion. Lesion that involves loss of the epidermis.
- Excoriation. A hollow, crusted area caused by scratching or picking at a primary lesion.
- Scar. Discolored, fibrous tissue that permanently replaces normal skin after destruction of the dermis. A very thick and raised scar is called a keloid.
- Lichenification. Rough, thick epidermis with exaggerated skin lines. This is often a characteristic of scratch dermatitis and atopic dermatitis.
- Atrophy. An area of skin that has become very thin and wrinkled. Normally seen in older individuals and people who are using very strong topical corticosteroid medication.

Causes and symptoms

Skin lesions can be caused by a wide variety of conditions and diseases. A tendency toward developing moles, freckles, or birthmarks may be inherited. Infection of the skin itself by bacteria, viruses, fungi, or parasites is the most common cause of skin lesions. Acne, **athlete's foot** (*tinea pedis*), warts, and **scabies** are examples of skin infections that cause lesions.

Allergic reactions and sensitivity to outside environmental factors can also lead to the formation of skin lesions. Underlying conditions can also precipitate the appearance of skin lesions. For example, the decreased sensitivity and poor circulation that accompanies **diabetes mellitus** can contribute to the formation of extensive ulcers on extremities such as the feet. Infections of body's entire system can cause the sudden onset of skin lesions. For example, skin lesions are a hallmark symptom of such diseases as chicken pox, herpes, and small pox. Cancers affecting the skin, including **basal cell carcinoma**, squamous cell carcinoma, **malignant melanoma**, and **Kaposi's sarcoma**, are recognized by their lesions.

Diagnosis

Diagnosis of the underlying cause of skin lesions is usually based on patient history, characteristics of the lesion, and where and how it appears on the patient's body (e.g. pustules confined to the face, neck and upper back can indicate acne, while scales appearing on the scalp and face may indicate **seborrheic dermatitis**). To

determine the cause of an infection, doctors may also take scrapings or swab samples from lesions for examination under a microscope or for use in bacterial, fungal, or viral cultures. In cases where a fungal infection is suspected, a doctor may examine a patient's skin under ultraviolet light using a filter device called a Woods light—under these conditions, certain species will taken on specific fluorescent colors. Dermatologists may also use contrast lighting and subdued lighting to detect variations in the skin. When involvement of the immune system is suspected, doctors may order a immunofluorescence test, which detects antibodies to specific antigens using a fluorescent chemical. In cases of contact **dermatitis**, a condition in which a allergic reaction to something irritates the skin, doctors may use patch tests in which samples of specific antigens are introduced into the skin via a scratch or a needle prick, to determine what substances are provoking the reaction.

The vast majority of skin lesions are noncancerous. However, doctors will determine whether or not a particular lesion or lesions are cancerous based on observation and the results of an excisional or punch biopsy, in which a tissue sample is excised for microscopic analysis. Since early detection is a key to successful treatment, individuals should examine their skin on a monthly basis for changes to existing moles, the presence of new moles, or a change in a certain area of skin. When examining moles, factors to look for include:

- Asymmetry. A normal mole is round, whereas a suspicious mole is uneven.
- Border. A normal mole has a clear-cut border with the surrounding skin, whereas the edges of a suspect mole may be irregular.
- Color. Normal moles are uniformly tan or brown but cancerous moles may appear as mixtures of red, white, blue, brown, purple, or black.
- Diameter. Normal moles are usually less than $\frac{1}{5}$ in (5 mm) in diameter, a skin lesion greater than this may be suspected as cancerous.

Treatment

Treatment of skin lesions depends upon the underlying cause, what type of lesions they are, and the patient's overall health. If the cause of the lesions is an allergic reaction, removing the allergen from the patient's environment is the most effective treatment. Topical preparations can also be used to clean and protect irritated skin as well as to remove dead skin cells and scales. These may come in a variety of forms, including ointments, creams, lotions, and solutions. **Topical antibiotics**, fungicides, pediculicides (agents that kill lice), and scabicides (agents that kill the

KEY TERMS

Corticosteroid—A type of steroid medication that helps relieve itching (puritis) and reduce inflammation.

Fibroma—A usually benign tumor consisting of fibrous tissue.

Lesion—A possibly abnormal change or difference in a tissue or structure, such as the skin.

Lipoma—A usually benign tumor of fatty tissue.

Patch test—Test in which different antigens (substances that cause an allergic reaction) are introduced into a patient's skin via a needle prick or scratch and then observed for evidence of an allergic reaction to one or more of them. Also known as a scratch test.

Woods light—Device that allows only ultraviolet light to pass through it.

scabies parasite) can be applied to treat appropriate skin infections. Oral medications may be taken to address systemic infections or conditions. Deeply infected lesions may require minor surgery to lance and drain pus. Topical agents to sooth irritated skin and reduce inflammation may also be applied. **Corticosteroids** are particularly effective in reducing inflammation and **itching** (puritis). Oatmeal baths, baking soda mixtures, and calamine lotion are also recommended for the relief of these symptoms. A type of corticosteroid may be used to reduce the appearance of keloid **scars**. Absorbent powders may also be used to reduce moisture and prevent the spread of infection. In cases of ulcers that are slow to heal, pressure **dressings** may be used. At times, surgical removal of a lesion may be recommended—this is the usual course of therapy for skin **cancer**. Surgical removal usually involves a simple excision under local anesthetic but it may also be accomplished through freezing (**cryotherapy**) or **laser surgery**.

Prognosis

Skin lesions such as moles, freckles, and birthmarks are a normal part of skin and will not disappear unless deliberately removed by a surgical procedure. Lesions due to an allergic reaction often subside soon after the offending agent is removed. Healing of lesions due to infections or disorders depends upon the type of infection or disorder and the overall health of the individual. Prognosis for skin cancer primarily depends upon whether or not the lesion is localized and whether or not it has spread to other areas of the

body, such as the lymph nodes. In cases where the lesion is localized and has not spread to other parts of the body, the cure rate is 95–100%.

Prevention

Not all skin lesions are preventable; moles and freckles, for example, are benign growths that are common and unavoidable. However others can be avoided or minimized by taking certain precautions. Skin lesions caused by an allergic reaction can be avoided by determining what the offending agent is and removing it from the home or workplace or, if this is impossible, developing strategies for safely handling it, such as with gloves and protective clothing. Keeping the skin, nails, and scalp clean and moisturized can help reduce or prevent the incidence of infectious skin diseases, as can not sharing personal care items such as combs and make-up with others. Skin lesions associated with **sexually transmitted diseases** can be prevented by the use of **condoms**. Scratching or picking at existing lesions should be avoided since this usually serves only to spread infection and may result in scarring. Individuals who have systemic conditions, such as diabetes mellitus or poor circulation, that could lead to serious skin lesions should inspect their bodies regularly for changes in their skin's condition. Regular visual inspection of the skin is also a key to preventing or minimizing the occurrence of skin cancer, as is the regular use of sun screens with an SPF of 15 or more.

Resources

BOOKS

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Bridget Travers

Skin pigmentation disorders

Definition

Skin pigmentation disorders are conditions that cause the skin to appear lighter or darker than normal, or blotchy and discolored.

Demographics

People of all races have skin pigmentation disorders. Some disorders, like **albinism** (which affects one out of every 17,000 people) are rare. Others, such as age spots, are very common.

Description

Skin pigmentation disorders occur because the body produces either too much or too little melanin, a pigment responsible for the color of hair, skin, and eyes. Melanin protects the body by absorbing ultraviolet light.

The term hypopigmentation refers to instances in which the body does not produce enough melanin. Albinism, for example, is an inherited condition that causes a lack of pigment. So people with albinism typically have light skin, white or pale yellow hair, and light blue or gray eyes. Another condition called **vitiligo**, is responsible for the development of smooth, depigmented white spots on the skin. Vitiligo affects nearly 2% of the population but it strikes people between 10 and 30 years old more often than other age groups, and is more evident in people with darker skin.

In **hyperpigmentation**, the body produces too much melanin, causing skin to become darker than usual. **Lichen simplex chronicus** is a skin disorder with severe **itching** that causes thick, dark patches of skin to develop. Lamellar **ichthyosis** (fish scale disease) is an inherited disease that also is characterized by darkened, scaly, dry patches of skin.

Hyperpigmentation also occurs in melasma, a dark mask-like discoloration that covers the cheeks and bridge of the nose. Melasma can occur during the end of **pregnancy**. People with the autoimmune disease systemic lupus also may develop a similar butterfly-shaped mask on their faces. In addition, many people have **moles**, freckles, age spots, and **birthmarks**, ranging from red or brown to bluish or black, covering various parts of their bodies.

Causes and symptoms

Scientists are still studying the reasons why skin pigmentation disorders occur. In some cases there are tangible causes, such as sun exposure, drug reactions or genetic inheritance. In other cases, etiology is not as clear.

Albinism is an inherited recessive trait. Albinism has many different forms but most people who have this condition have pale skin, hair, and eyes. Melanin is also responsible for eye color, and serves as a filter that prevents too much light from entering the eye. Since they lack melanin in their eyes, many people with

KEY TERMS

Albinism—An inherited condition that causes a lack of pigment. People with albinism typically have light skin, white or pale yellow hair, and light blue or gray eyes.

Hypopigmentation—A skin condition that occurs when the body has too little melanin, or pigment.

Hypopigmentation—A skin condition that occurs when the body has too much melanin, or pigment.

Lamellar ichthyosis—Also called fish scale disease, this inherited condition is characterized by darkened, scaly, dry patches of skin.

Lichen simplex chronicus—A skin disorder accompanied by severe itching that causes thick, dark patches of skin to develop.

Melanin—A pigment that is responsible for the color of hair, skin and eyes. Melanin also protects the body by absorbing ultraviolet light.

Melanocytes—Cells that create melanin.

Melasma—A dark mask-like discoloration that covers the cheeks and bridge of the nose. Also called “the mask of pregnancy.”

Vitiligo—A skin disorder that is characterized by smooth, depigmented white spots on the skin.

albinism also have **visual impairment**. With little skin pigmentation, they also **sunburn** easily and are more prone to skin **cancer**.

The hypopigmentation spots associated with vitiligo sometimes form in places where a person has been cut or injured. Research has shown that the light patches associated with vitiligo do not contain melanocytes, the skin cells that create melanin. Some scientists believe vitiligo may be caused by an autoimmune disorder. It also has been linked to other conditions such as **hyperthyroidism** (too much thyroid hormone) and **Addison’s Disease**, which affects the adrenal gland.

Hyperpigmentation can be caused by many factors, from too much sunbathing to drug reactions or poor **nutrition**. **Wounds** and **scars** also can develop darker patches of skin. A psychological syndrome gives people with lichen simplex chronicus a compulsive need to scratch, which causes dark, leathery skin to form. This can lead to permanent scarring and infection if untreated. Scientists believe lamellar ichthyosis is caused by genetic factors.

The mask caused by melasma may be related to pregnancy hormones, and usually disappears after a woman gives birth. Birthmarks, moles, and **aging** spots usually are harmless. Some moles, however, can change in size, color, texture, or start bleeding, which could indicate possible skin cancer.

Diagnosis

Diagnostic tests vary for different types of skin pigmentation disorders. Physicians usually can diagnose albinism by looking carefully at a person’s hair, skin, and eyes. They may order blood tests and eye

exams as well. A visual exam also is enough to diagnose vitiligo.

For most hyperpigmentation disorders, doctors can make a diagnosis by looking at a person’s appearance. To detect conditions such as lichen simplex chronicus or lamellar ichthyosis, or skin cancer, they may also do a biopsy to remove some of the affected skin for further study under a microscope. Some physicians also use a wood’s lamp, or black light test, to diagnose skin conditions. Affected areas absorbs ultraviolet light and stands out with fluorescent colors in a darkened room.

Treatment

For albinism, healthcare providers advise people to cover exposed body parts, use sunscreen, and avoid excess sunlight to prevent skin cancer. People with albinism also must wear protective sunglasses and, in some cases, prescription corrective lenses. Surgery may be necessary to correct visual impairments.

To treat vitiligo, physicians may prescribe a combination of photo-sensitive medications like trimethylpsoralen and ultraviolet **light therapy** to darken the spots. If the person has depigmented patches covering more than 50% of the body, doctors also may be able to use skin bleaching agents like monobenzene to give the skin a lighter, more uniform appearance. Other options include cosmetic concealers and **skin grafting**.

Skin-lightening creams are available for hyperpigmentation disorders. Doctors also advise staying out of the sun. Counseling with a dietitian may help in cases caused by poor nutrition. For lichen simplex chronicus, doctors prescribe **antihistamines** and topical steroid creams to stop the itching. If a mole or

birthmark appears suspicious, physicians often will surgically remove it to prevent skin cancer.

Prognosis

Most skin pigmentation disorders do not affect a person's health, only the outward appearance.

Prevention

In most cases, doctors will recommend using sunscreen and avoiding too much sun exposure.

Resources

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 SkinCancerNet. [Accessed September 19, 2010] http://www.skincarephysicians.com/skin_cancernet/index.html.

ORGANIZATIONS

- American Academy of Dermatology (AAD), 930 E. Woodfield Rd., PO Box 4014, Schaumburg, IL, 60173-4014, (847) 330-0230, <http://www.aad.org>.
 National Organization for Albinism and Hypopigmentation (NOAH), PO Box 959, East Hampstead, NH, 03826-0959, (800) 473-2310, <http://www.albinism.org>.
 National Vitiligo Foundation, Inc. (NVFI), PO Box 23226, Cincinnati, OH, 45223, (513) 541-3903, <http://nvfi.org>.

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Skin resurfacing

Definition

Skin resurfacing employs a variety of techniques to change the surface texture and appearance of the skin. Common skin resurfacing techniques include chemical peels, dermabrasion, and laser resurfacing.

Purpose

Skin resurfacing procedures may be performed for cosmetic reasons, such as diminishing the appearance of wrinkles around the mouth or eyes. They may also be used as a medical treatment, such as removing large numbers of certain precancerous lesions called actinic keratoses. Physicians sometimes combine techniques, using dermabrasion or laser resurfacing on some areas of the face, while performing a chemical peel on other areas.

Precautions

As the popularity of skin resurfacing techniques has increased, many unqualified or inexperienced providers have entered the field. Patients should choose their provider with the same degree of care they take for any other medical procedure. Complications of skin resurfacing techniques can be serious, including severe infection and scarring.

Patient's with active herpes virus infections are not good candidates for resurfacing procedures. Persons who tend to scar easily may also experience poor results. Patients who have recently used the oral acne medication isotretinoin (Accutane) may be at higher risk of scarring following skin resurfacing.

Description

Chemical peel

Chemical peels employ a variety of caustic chemicals to selectively destroy several layers of skin. The peeling solutions are "painted on," area-by-area, to ensure that the entire face is treated. After the skin heals, discoloration, wrinkles, and other surface irregularities are often eliminated.

Chemical peels are divided into three types: superficial, medium-depth, and deep. The type of peel depends on the strength of the chemical used, and on how deeply it penetrates. Superficial peels are used for fine wrinkles, sun damage, acne, and rosacea. The medium-depth peel is used for more obvious wrinkles and sun damage as well as for precancerous lesions like actinic keratoses. Deep peels are used for the most severe wrinkling and sun damage.

Dermabrasion

Dermabrasion uses an abrasive tool to selectively remove layers of skin. Some physicians use a hand-held motorized tool with a small wire brush or diamond-impregnated grinding wheel at the end. Other physicians prefer to abrade the skin by hand with an abrasive pad or other instrument. Acne scarring is one of the

prime uses for dermabrasion. It also can be used to treat wrinkling, remove surgical **scars**, and obliterate **tattoos**.

Laser resurfacing

Laser resurfacing is the most recently developed technique for skin resurfacing. Specially designed, pulsed CO₂ lasers can vaporize skin layer-by-layer, causing minimal damage to other skin tissue. Special scanning devices move the laser light across the skin in predetermined patterns, ensuring proper exposure. Wrinkling around the eyes, mouth, and cheeks are the primary uses for laser resurfacing. Smile lines or those associated with other facial muscles tend to reappear after laser resurfacing. Laser resurfacing appears to achieve its best results as a spot treatment; patients expecting complete elimination of their wrinkles will not be satisfied.

Preparation

Chemical peel

Preparation for the chemical peel begins several weeks before the actual procedure. To promote turnover of skin cells, patients use a mild glycolic acid lotion or cream in the morning, and the acne cream tretinoin in the evening. They also use hydroquinone cream, a bleaching product that helps prevent later discoloration. To prevent reappearance of a herpes simplex virus infection, antiviral medicine is started a few days before the procedure and continues until the skin has healed.

Patients arrive for the procedure wearing no makeup. The physician “degreases” the patient’s face using alcohol or another cleanser. Some degree of **pain** accompanies all types of peels. For a superficial peel, use of a hand held fan to cool the face during the procedure is often sufficient. For medium-depth peels, the patient may take a sedative or **aspirin**. During the procedure, cold compresses and a hand-held fan can also reduce pain. Deep peels can be extremely painful. Some physicians prefer **general anesthesia** but local anesthetics combined with intravenous sedatives are frequently sufficient to control pain.

Dermabrasion

Dermabrasion does not require much preparation. It is usually performed under **local anesthesia**, although some physicians use intravenous **sedation** or general anesthesia. The physician begins by marking the areas to be treated and then chilling them with ice packs. In order to stiffen the skin, a spray refrigerant is applied to the area, which also helps control pain. Some physicians prefer to inject the area with a solution of saline and local anesthetic, which also leaves the skin’s surface

more solid. Since dermabrasion can cause quite a bit of bleeding, physicians and their assistants will wear gloves, gowns, and masks to protect themselves from possible blood-transmitted infection.

Laser resurfacing

Antiviral medications should be started several days before the procedure. Laser resurfacing is performed under local anesthesia. An oral sedative may also be taken. The patient’s eyes must be shielded, and the area surrounding the face should be shielded with wet drapes or crumpled foil to catch stray beams of laser light. The physician will mark the areas to be treated before beginning the procedure.

Aftercare

Chemical peel

Within a day or so following a superficial peel, the skin will turn faint pink or brown. Over the next few days, dead skin will peel away. Patients will be instructed to wash their skin frequently with a mild cleanser and cool water, then apply an ointment to the skin to keep it moist. After a medium-depth peel, the skin turns deep red or brown and crusts may form. Care is similar to that following a superficial peel. Redness may persist for a week or more. Deep-peeled skin will turn brown and crusty. There may also be swelling and some oozing of fluid. Frequent washing and ointments are favored over **dressings**. The skin typically heals in about two weeks, but redness may persist.

Dermabrasion

Following the procedure, an ointment may be applied and the wound will be covered with a dressing and mask. Patients with a history of herpesvirus infections will begin taking an antiviral medication to prevent a recurrence. After 24 hours, the dressing is removed and ointment is reapplied to keep the wound moist. Patients are encouraged to wash their face with plain water and reapply ointment every few hours. This relieves **itching** and pain and helps remove oozing fluid and other matter. Patients may require a pain medication. A steroid medication may be taken during the first few days to reduce swelling. The skin will take a week or more to heal, but may remain very red.

Laser resurfacing

The skin should be kept moist following laser resurfacing. This promotes more rapid healing and reduces the risk of infection. Some physicians favor

KEY TERMS

Actinic keratosis—A crusty, scaly skin lesion, caused by exposure to the sun, which can transform into skin cancer.

Herpesviruses—A family of viruses responsible for cold sores, chicken pox, and genital herpes.

Isotretinoin—A powerful vitamin A derivative used in the treatment of acne. It can promote scarring after skin resurfacing procedures.

application of ointments only to the skin; others prefer the use of dressings. In either case, care of the skin is similar to that given following a chemical peel. The face is washed with plain water to remove ooze and an ointment is reapplied. Healing will take approximately two weeks. Pain medications and a steroid to reduce swelling may also be taken.

Risks

All resurfacing procedures can lead to infection and scarring. It is also possible that skin coloration will be altered or that redness of the skin will be prolonged for many months. Some of the peeling agents used in deep chemical peels can affect the function of the heart.

Normal results

Depending on the resurfacing techniques selected, it is possible to improve the appearance of skin damaged by sun, age, or disease in many people. Skin resurfacing techniques address only the surface of the skin; procedures such as face-lift surgery or **blepharoplasty** may be needed to repair other age-related skin changes. All resurfacing procedures are accompanied by some pain, redness, and skin color changes. These may persist for several months following the procedure, but they usually resolve over time.

Abnormal results

As noted above, resurfacing procedures can reactivate herpesvirus infections or lead to new, sometimes serious infections. All resurfacing techniques intentionally create skin **wounds**, creating the possibility for scarring. Abnormal results such as these can be minimized with use of antiviral medications prior to the procedure and good wound care afterward. Selection of an experienced, reputable provider also is key.

ORGANIZATIONS

American Society for Dermatologic Surgery, 5550

Meadowbrook Dr., Suite 120, Rolling Meadows, IL, 60008, (847) 956-0900, (847) 956-0999, <http://www.asds.net/>.

American Society for Laser Medicine and Surgery, 2100

Stewart Ave., Suite 240, Wausau, WI, 54401, (715) 845-9283, (715) 848-2493, (877) 258-6028, information@aslms.org, <http://www.aslms.org/>.

American Society of Plastic Surgeons, 444 E. Algonquin Rd, Arlington Heights, IL, 60005, (847) 228-9900, <http://www.plasticsurgery.org/>.

Richard H. Camer

Skin traction see **Traction; Immobilization**

Skull x rays

Definition

Skull x rays are performed to examine the nose, sinuses, and facial bones. These studies may also be referred to as sinus x rays. X-ray studies produce films, also known as radiographs, by aiming x rays at soft bones and tissues of the body. X-ray beams are similar to light waves, except their shorter wavelength allows them to penetrate dense substances, producing images and shadows on film.

Purpose

Doctors may order skull x rays to aid in the diagnosis of a variety of diseases or injuries.

Sinusitis

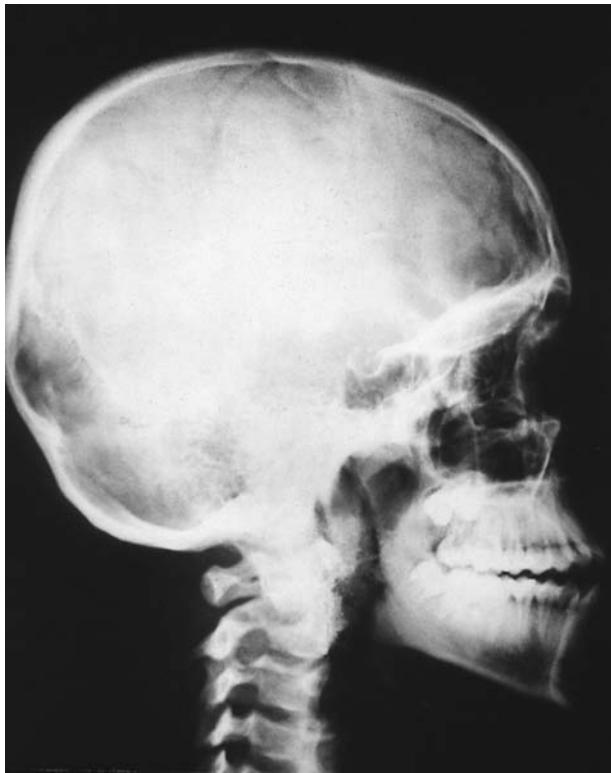
Sinus x rays may be ordered to confirm a diagnosis of **sinusitis**, or sinus infection.

Fractures

A skull x ray may detect bone **fractures** resulting from injury or disease. The skull x ray should clearly show the skull cap, jaw bones, and facial bones.

Tumors

Skull radiographs may indicate tumors in facial bones, tissues, or the sinuses. Tumors may be benign (not cancerous) or malignant (cancerous).



A skull x ray. (Photo Researchers, Inc.)

Other

Birth defects (referred to as congenital anomalies) may be detected on a skull x ray by changes in bone structure. Abnormal tissues or glands resulting from various conditions or diseases may also be shown on a skull radiograph.

Precautions

As with any x-ray procedure, women who may be pregnant are advised against having a skull x ray if it is not absolutely necessary. However, a lead apron may be worn across the abdomen during the procedure to protect the fetus. Children are also more sensitive to x-ray exposure. Children of both sexes should wear a protective covering (a lead apron) in the genital/reproductive area. In general, skull x-ray exposure is minimal and x-ray equipment and procedures are monitored to ensure radiation safety.

Description

Skull or sinus x rays may be performed in a doctor's office that has x-ray equipment and a technologist available. The exam may also be performed in an

outpatient radiology facility or a hospital radiology department.

In many instances, particularly for sinus views, the patient will sit upright in a chair, perhaps with the head held stable by a foam vise. A film cassette is located behind the patient. The x-ray tube is in front of the patient and may be moved to allow for different positions and views. A patient may also be asked to move his or her head at various angles and positions.

In some cases, technologists will ask the patient to lie on a table and will place the head and neck at various angles. In routine skull x rays, as many as five different views may be taken to allow a clear picture of various bones and tissues. The length of the test will vary depending on the number of views taken, but in general, it should last about 10 minutes. The technologist will usually ask a patient to wait while the films are being developed to ensure that they are clear before going to the radiologist.

Preparation

There is no preparation for the patient prior to arriving at the radiology facility. Patients will be asked to remove jewelry, dentures, or other metal objects that may produce artifacts on the film. The referring doctor or x-ray technologist can answer any questions regarding the procedure. Any woman who is, or may be, pregnant should tell the technologist.

Aftercare

There is no aftercare required following skull or sinus x-ray procedures.

Risks

There are no common side effects from skull or sinus x ray. The patient may feel some discomfort in the positioning of the head and neck, but will have no complications. Any x-ray procedure carries minimal radiation risk, and children and pregnant women should be protected from radiation exposure to the abdominal or genital areas.

Normal results

Normal results should indicate sinuses, bones, tissues, and other observed areas are of normal size, shape, and thickness for the patient's age and medical history. Results, whether normal or abnormal, will be provided to the referring doctor in a written report.

Abnormal results

Abnormal results may include:

KEY TERMS

Radiograph—The actual picture or film produced by an x-ray study.

X ray—A form of electromagnetic radiation with shorter wavelengths than normal light. X rays can penetrate most structures.

Sinusitis

Air in sinuses will show up on a radiograph as black but fluid will be cloudy or white (opaque). This helps the radiologist to identify trapped fluids in the sinuses. In chronic sinusitis, the radiologist may also note thickening or hardening of the bony wall of an infected sinus.

Fractures

Radiologists may recognize even tiny facial bone fractures as a line of defect.

Tumors

Tumors may be visible if the bony sinus wall is distorted or destroyed. Abnormal findings may result in follow-up imaging studies.

Other

Skull x rays may also detect disorders that show up as changes in bone structure, such as Paget's disease of the bone or acromegaly (a disorder associated with excess growth hormones from the pituitary gland). Areas of calcification, or gathering of **calcium** deposits, may indicate a condition such as an infection of bone or bone marrow (**osteomyelitis**).

ORGANIZATIONS

Long Island Head Injury Association, 65 Austin Blvd., Commack, NY, (631) 543-2245, (631) 543-2261, <http://www.lihia.org/>.

National Cancer Institute (National Institutes of Health), NCI Office of Communications and Education, 6116 Executive Blvd. Suite 300, Bethesda, MD, 20892-8322, 800 4-CANCER (422-6237), cancergovstaff@mail.nih.gov, <http://www.cancer.gov/>.

Radiological Society of North America, 820 Jorie Boulevard, Oak Brook, IL, 60523-2251, (630) 571-2670, (630) 571-7837, (800) 381-6660, radiologyinfo.org.

Teresa Odle

SLE see **Systemic lupus erythematosus**

Sleep apnea

Definition

Sleep apnea is a condition in which breathing stops for more than ten seconds during sleep. Sleep apnea is a major, though often unrecognized, cause of daytime sleepiness. It can have serious negative effects on a person's quality of life and is thought to be considerably underdiagnosed in the United States.

Demographics

Approximately 6–7% of the U.S. population, or 18 million Americans, are thought to have sleep apnea but only 10 million have symptoms, and only 0.6 million have yet been diagnosed. In Americans aged 30–60 years, obstructive sleep apnea affects nearly one in four men and one in ten women; men are twice as likely as women to have sleep apnea. As sleep apnea seldom occurs in premenopausal females, it is suggested that hormones may play some role in the disorder.

Other predisposing factors include age, as nearly 20–60% of the elderly may be affected; overweight status or **obesity**; or use of alcohol or sedatives. Some studies have demonstrated that elderly African-Americans are more than twice as likely as elderly whites to suffer from sleep apnea. Some families appear to have increased incidence of sleep apnea.

Description

A sleeping person normally breathes continuously and uninterrupted throughout the night. A person with sleep apnea, however, has frequent episodes (up to 400–500 per night) in which he or she stops breathing. This interruption of breathing is called "apnea." Breathing usually stops for about 30 seconds; then the person usually startles awake with a loud snort and begins to breathe again, gradually falling back to sleep.

There are two forms of sleep apnea. In obstructive sleep apnea (OSA), breathing stops because tissue in the throat closes off the airway. In central sleep apnea, (CSA), the brain centers responsible for breathing fail to send messages to the breathing muscles. OSA is much more common than CSA. It is thought that about 1–10% of adults are affected by OSA; only about one tenth of that number have CSA. OSA can affect people of any age and of either sex, but it is most common in middle-aged, somewhat overweight men, especially those who use alcohol.

Causes and symptoms

Obstructive sleep apnea

Obstructive sleep apnea occurs when part of the airway is closed off (usually at the back of the throat) while a person is trying to inhale during sleep. People whose airways are slightly narrower than average are more likely to be affected by OSA. Obesity, especially obesity in the neck, can increase the risk of developing OSA because the fat tissue tends to narrow the airway. In some people, the airway is blocked by enlarged tonsils, an enlarged tongue, jaw deformities, or growths in the neck that compress the airway. Blocked nasal passages may also play a part in some people.

When a person begins to inhale, the expansion of the lungs lowers the air pressure inside the airway. If the muscles that keep the airway open are not working hard enough, the airway narrows and may collapse, shutting off the supply of air to the lungs. OSA occurs during sleep because the neck muscles that keep the airway open are not as active then. Congestion in the nose can make collapse more likely, since the extra effort needed to inhale will lower the pressure in the airway even more. Drinking alcohol or taking tranquilizers in the evening worsens this situation because these cause the neck muscles to relax. (These drugs also lower the “respiratory drive” in the nervous system, reducing breathing rate and strength.)

People with OSA almost always snore heavily because the same narrowing of the airway that causes **snoring** can also cause OSA. Snoring may actually help cause OSA as well because the vibration of the throat tissues can cause them to swell. However, most people who snore do not go on to develop OSA.

Other risk factors for developing OSA include male sex; **pregnancy**; a family history of the disorder; and **smoking**. With regard to gender, it has been found that male sex hormones sometimes cause changes in the size or structure of the upper airway. The weight gain that accompanies pregnancy can affect a woman’s breathing patterns during sleep, particularly during the third trimester. With regard to family history, OSA is known to run in families even though no gene or genes associated with the disorder have been identified. Smoking increases the risk of developing OSA because it causes inflammation, swelling, and narrowing of the upper airway.

Some patients being treated for **head and neck cancer** develop OSA as a result of physical changes in the muscles and other tissues of the neck and throat. Doctors recommend prompt treatment of the OSA to improve the patient’s quality of life.

Central sleep apnea

In central sleep apnea, the airway remains open but the nerve signals controlling the respiratory muscles are not regulated properly. This can cause wide fluctuations in the level of carbon dioxide (CO_2) in the blood. Normal activity in the body produces CO_2 , which is brought by the blood to the lungs for exhalation. When the blood level of CO_2 rises, brain centers respond by increasing the rate of respiration, clearing the CO_2 . As blood levels fall again, respiration slows down. Normally, this interaction of CO_2 and breathing rate maintains the CO_2 level within very narrow limits. CSA can occur when the regulation system becomes insensitive to CO_2 levels, allowing wide fluctuations in both CO_2 levels and breathing rates. High CO_2 levels cause very rapid breathing (hyperventilation), which then lowers CO_2 so much that breathing becomes very slow or even stops. CSA occurs during sleep because when a person is awake, breathing is usually stimulated by other signals, including conscious awareness of breathing rate.

A combination of the two forms is also possible and is called mixed sleep apnea. Mixed sleep apnea episodes usually begin with a reduced central respiratory drive, followed by obstruction.

OSA and CSA cause similar symptoms. The most common symptoms are:

- daytime sleepiness
- morning headaches
- a feeling that sleep is not restful
- disorientation upon waking
- poor judgment
- personality changes

Sleepiness is caused not only by the frequent interruption of sleep but by the inability to enter long periods of deep sleep, during which the body performs numerous restorative functions. OSA is one of the leading causes of daytime sleepiness and is a major risk factor for motor vehicle accidents. Headaches and disorientation are caused by low oxygen levels during sleep from the lack of regular breathing.

Other symptoms of sleep apnea may include **sexual dysfunction**, loss of concentration, **memory loss**, intellectual impairment, and behavioral changes including **anxiety** and depression.

Sleep apnea is also associated with night sweats and nocturia, or increased frequency of urination at night. **Bedwetting** in children is also linked to sleep apnea.

Sleep apnea can also cause serious changes in the cardiovascular system. Daytime **hypertension** (high blood pressure) is common. An increase in

KEY TERMS

Continuous positive airway pressure (CPAP)—A ventilation system that blows a gentle stream of air into the nose to keep the airway open.

Genioplasty—An operation performed to reshape the chin. Genioplasties are often done to treat OSA because the procedure changes the structure of the patient's upper airway.

Mandible—The medical term for the lower jaw. One type of oral appliance used to treat OSA pushes the mandible forward in order to ease breathing during sleep.

Nocturia—Excessive need to urinate at night. Nocturia is a symptom of OSA and often increases the patient's daytime sleepiness.

Polysomnography—A group of tests administered to analyze heart, blood, and breathing patterns during sleep.

Tracheotomy—A surgical procedure in which a small hole is cut into the trachea, or windpipe, below the level of the vocal cords.

Uvulopalatopharyngoplasty (UPPP)—An operation to remove excess tissue at the back of the throat to prevent it from closing off the airway during sleep.

the number of red blood cells (polycythemia) is possible, as is an enlarged left ventricle of the heart (**cor pulmonale**), and left ventricular failure. In some people, sleep apnea causes life-threatening changes in the rhythm of the heart, including heartbeat slowing (bradycardia), racing (tachycardia), and other types of "arrhythmias." Sudden **death** may occur from such **arrhythmias**. Patients with the **Pickwickian syndrome** (named after a Charles Dickens character) are obese and sleepy, with right **heart failure**, **pulmonary hypertension**, and chronic daytime low blood oxygen (hypoxemia) and increased blood CO₂ (hypercapnia).

Diagnosis

Excessive daytime sleepiness is the complaint that usually brings a person to see the doctor. A careful medical history will include questions about alcohol or tranquilizer use, snoring (often reported by the person's partner), and morning headaches or disorientation. A physical exam will include examination of the throat to look for narrowing or obstruction. Blood pressure is also measured. Measuring heart rate or

blood levels of oxygen and CO₂ during the daytime will not usually be done since these are abnormal only at night in most patients.

In some cases the person's dentist may suggest the diagnosis of OSA on the basis of a dental checkup or evaluation of the patient for oral surgery.

Confirmation of the diagnosis usually requires making measurements while the person sleeps. These tests are called a **polysomnography** study and are conducted during an overnight stay in a specialized sleep laboratory. Important parts of the polysomnography study include measurements of:

- heart rate
- airflow at the mouth and nose
- respiratory effort
- sleep stage (light sleep, deep sleep, dream sleep, etc.)
- oxygen level in the blood, using a noninvasive probe (ear oximetry)

Simplified studies done overnight at home are also possible, and may be appropriate for people whose profile strongly suggests the presence of obstructive sleep apnea; that is, middle-aged, somewhat overweight men, who snore and have high blood pressure. The home-based study usually includes ear oximetry and cardiac measurements. If these measurements support the diagnosis of OSA, initial treatment is usually suggested without polysomnography. Home-based measurements are not used to rule out OSA, however, and if the measurements do not support the OSA diagnosis, polysomnography may be needed to define the problem further.

Treatment

Behavioral changes

Treatment of obstructive sleep apnea begins with reducing the use of alcohol or tranquilizers in the evening, if these have been contributing to the problem. Weight loss is also effective but if the weight returns, as it often does, so does the apnea. Changing sleeping position may be effective; snoring and sleep apnea are both most common when a person sleeps on his back. Turning to sleep on the side may be enough to clear up the symptoms. Raising the head of the bed may also help. Opening of the nasal passages can provide some relief. There are a variety of nasal devices such as clips, tapes, or holders which may help, though discomfort may limit their use. Nasal **decongestants** may be useful but should not be taken for sleep apnea without the consent of the treating physician.

Oxygen and drug therapy

Supplemental nighttime oxygen can be useful for some people with either central and obstructive sleep apnea. Tricyclic **antidepressant drugs** such as protriptyline (Vivactil) may help by increasing the muscle tone of the upper airway muscles but their side effects may severely limit their usefulness.

Mechanical ventilation

For moderate to severe sleep apnea, the most successful treatment is nighttime use of a ventilator, called a CPAP machine. CPAP (continuous positive airway pressure) blows air into the airway continuously, preventing its collapse. CPAP requires the use of a nasal mask. The appropriate pressure setting for the CPAP machine is determined by polysomnography in the sleep lab. Its effects are dramatic; daytime sleepiness usually disappears within one to two days after treatment begins. CPAP is used to treat both obstructive and central sleep apnea.

CPAP is tolerated well by about two-thirds of patients who try it. Bi-level positive airway pressure (BiPAP), is an alternative form of ventilation. With BiPAP, the ventilator reduces the air pressure when the person exhales. This is more comfortable for some.

Surgery

Surgery can be used to correct obstructions in the airways. The most common surgery is called UPPP, for uvulopalatopharyngoplasty. This surgery removes tissue from the rear of the mouth and top of the throat. The tissues removed include parts of the uvula (the flap of tissue that hangs down at the back of the mouth), the soft palate, and the pharynx. Tonsils and adenoids are usually removed in this operation. This operation significantly improves sleep apnea in slightly more than half of all cases.

Reconstructive surgery is possible for those whose OSA is due to constriction of the airway by lower jaw deformities. Genioplasty, which is a procedure that plastic surgeons usually perform to reshape a patient's chin to improve his or her appearance, is now being done to reshape the upper airway in patients with OSA.

When other forms of treatment are not successful, obstructive sleep apnea may be treated by a tracheostomy. In this procedure, an opening is made into the trachea (windpipe) below the obstruction, and a tube inserted to maintain an air passage. A tracheostomy requires a great deal of care to prevent infection of the tracheostomy site. In addition, since air is no longer being filtered and moistened by the nasal passages

before entering the lungs, the lower airways can become dry and susceptible to infection as well. Tracheostomy is usually reserved for those whose apnea has led to life-threatening heart arrhythmias, and who have not been treated successfully with other treatments.

Oral appliances

Another approach to treating OSA involves the use of oral appliances intended to improve breathing either by holding the tongue in place or by pushing the lower jaw forward during sleep to increase the air volume in the upper airway. The first type of oral appliance is known as a tongue retaining device or TRD. The second type is variously called an oral protrusive device (OPD) or mandibular advancement splint (MAS), because it holds the mandible, or lower jaw, forward during sleep. These oral devices appear to work best for patients with mild-to-moderate OSA, and in some cases can postpone or prevent the need for surgery. Their rate of patient compliance is about 50%; most patients who stop using oral appliances do so because their teeth are in poor condition. TRDs and OPDs can be fitted by dentists; however, most dentists work together with the patient's physician following a polysomnogram rather than prescribing the device by themselves.

Prognosis

The combination of behavioral changes, ventilation assistance, drug therapy, and surgery allow most people with sleep apnea to be treated successfully, although it may take some time to determine the most effective and least intrusive treatment. Polysomnography testing is usually required after beginning a treatment to determine how effective it has been.

Prevention

For people who snore frequently, weight control, avoidance of evening alcohol or tranquilizers, and adjustment of sleeping position may help reduce the risk of developing obstructive sleep apnea.

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- American Dental Association, 211 East Chicago Avenue, Chicago, IL, 60611, (312) 440-2500, <http://www.ada.org>.
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Richard Robinson
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Sleep deprivation

Definition

Sleep deprivation is an inadequate amount of sleep for a given individual.

Demographics

Sleep deprivation has become so widespread in industrialized societies that daytime drowsiness may no longer seem abnormal. Between 1998 and 2005 the number of American adults who reported getting eight or more hours of sleep on weekday nights fell from 35% to 26%. Driver **fatigue** causes 100,000 accidents and 1,500 deaths annually in the United States.

Sleep deprivation is considered to be a widespread chronic health problem among American teenagers. A 2006 poll found that only 20% of teens got adequate sleep on school nights: by the end of high school they averaged fewer than seven hours and most teens reported feeling tired during the day.

Although sleep disturbances and disorders do not necessarily result in sleep deprivation, they can contribute to it:

- About half of all people over age 65 suffer frequent sleep disturbances.
- About 60 million Americans suffer from frequent or extended periods of insomnia resulting in sleep

Recommended hours of sleep, by age group

Infants		
0–2 months	12–18 hours	
2–12 months	14–15 hours	
Toddlers/Children		
1–3 years	12–14 hours	
3–5 years	11–13 hours	
5–10 years	10–11 hours	
Adolescents		
10–17 years	8.5–9.25 hours	
Adults		
18+	7–9 hours	

SOURCE: National Sleep Foundation, "How Much Sleep Do We Really Need?" Available online at: <http://www.sleepfoundation.org/article/how-sleep-works/how-much-sleep-do-we-really-need> (accessed August 17, 2010).

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deprivation. The incidence of insomnia increases with age, affecting about 40% of women and 30% of men.

- An estimated 18 million Americans have sleep apnea, although it usually goes undiagnosed.
- Restless legs syndrome (RLS) is one of the most common sleep disorders that causes sleep deprivation, especially among older people. RLS is estimated to affect as many as 12 million Americans.
- Narcolepsy—which affects about 250,000 Americans—can cause nighttime insomnia, resulting in sleep deprivation.
- Most people with mental disorders—including depression and schizophrenia—have sleep disturbances that cause sleep deprivation.
- Many totally blind people have life-long sleeping problems, including insomnia and a type of permanent jetlag, which can result in sleep deprivation.

Description

Sleeping and wakefulness are controlled by neurotransmitters—chemical messengers in the brain—which act on different sets of nerve cells or neurons. The neurotransmitters serotonin and norepinephrine in the brainstem—the connection between the brain and the spinal cord—keep parts of the brain active during wakefulness and are switched off during sleep. Also during wakefulness an important chemical called adenosine builds up in the blood to the point where it eventually causes drowsiness and is broken down during sleep.

Humans normally cycle through five stages of sleep throughout the night, with one complete sleep cycle averaging 90–120 minutes:

- Stage 1 is drowsiness, drifting in and out of sleep, and being easily awakened.
- Stage 2 is light sleep, which accounts for about 50% of total sleeping time.
- Stage 3 is deep sleep.
- Stage 4 is slow-wave deep sleep.
- Stages 1, 3, and 4 together account for about 30% of sleeping time.
- Rapid eye movement (REM) sleep accounts for about 20% of total sleep time. During the first sleep cycles of the night, deep sleep is relatively long and REM sleep short. REM periods gradually increase in length as deep sleep shortens. Towards waking almost all sleep is stages 1, 2, and REM.

Sleep is essential for survival and sleep deprivation can eventually result in **death**. However scientists have only recently begun to understand the many functions of sleep:

- The brain is very active during sleep. During this period of low sensory input the brain consolidates recently acquired memories. Nerve-signaling patterns that are generated during the day are repeated during deep sleep. REM sleep is required for learning certain mental skills. Thus sleep appears to be necessary for encoding memories and learning.
- Sleep is required for proper nervous system functioning. Parts of the brain that are involved in emotions, decision-making, and social interactions are less active during sleep. These neurons may need sleep to repair and replenish themselves.
- Sleep may be necessary for growing new neurons.
- Sleep may be required for exercising neuronal functions that are less active during wakefulness.
- Many cells in the body produce more protein during deep and REM sleep. Thus sleep may be necessary for replenishing energy and repairing damage in cells throughout the body.
- Sleep is required for proper immune system function and cytokines—chemicals that fight infection—are produced during sleep.
- Growth hormones in children and young adults are released during deep sleep.

The amount of sleep required to prevent deprivation depends on a variety of factors, especially age and genetics:

- Infants need about 16 hours of sleep out of every 24 hours, with about 50% spent in REM.

- Toddlers need about 14 hours of sleep, which gradually decreases with age to a requirement of slightly over nine hours in teenagers.
- Most adults need 7–8 hours of sleep each night, although individual requirements may vary from four to 12 hours per night.
- Researchers have identified a gene called DEC2 that turns off some genes involved in controlling circadian rhythms—the internal clock that regulates the sleep-wake cycle. People with certain mutations in the DEC2 gene require only about six hours of sleep per night.
- Women often need several extra hours of sleep during the first three months of pregnancy.
- Although older people tend to sleep more lightly and for shorter periods, they need about the same amount of total sleep as when they were younger.

For most people sleep deprivation accumulates as a “sleep debt,” which must be made up. The ability to function relatively well—at least for short periods—under conditions of sleep deprivation appears to be genetically determined. Estimates suggest that 10–15% of people function adequately on little or no sleep, whereas another 10–15% cannot function at all without sleep. Most people cannot function at all after 48 hours without sleep; nor do humans appear to adapt to sleep deprivation. One study found that subjects who slept only 4–6 hours per night for 14 consecutive nights showed cognitive impairment equivalent to going without sleep for three consecutive days.

Although people may adjust to a sleep-depriving schedule, daily functioning and physical and mental health suffer:

- Sleep deprivation interferes with concentration, learning, and problem-solving.
- At least 6 hours of regular sleep are required for peak memory performance and sleep deprivation is directly linked to memory loss.
- Sleep deprivation interferes with work, school, and social interactions.
- Sleep deprivation can cause stress.
- Sleep deprivation slows reaction times. Sleep-deprived people perform at least as poorly on driving simulators and hand-eye coordination tasks as people who are intoxicated.
- Sleep deprivation easily disrupts the decision-making machinery of the brain, impairing judgment, increasing risky behaviors, and reducing sensitivity to loss.
- Sleep deprivation increases the risk of falls and accidents.

- Sleep deprivation can increase the risk for many health problems, including hypertension, cardiovascular disease, diabetes, obesity, and infections.
- Sleep deprivation can inhibit weight loss, even with proper exercise and diet.
- Sleep deprivation increases the effects of alcohol.
- Sleep deprivation can cause sleep paralysis—a rare but frightening condition in which a person temporarily loses the ability to speak or move while falling asleep or waking up.

Teenagers require an average of 9.25 hours of sleep per night for brain development, health, and optimal performance. Sleep-deprived teens are at risk for:

- impaired cognitive function and decision-making
- health problems
- poor grades and athletic performance
- emotional and behavioral problems
- depression
- substance abuse
- violence
- automobile accidents

Risk factors

Risk factors for sleep deprivation include:

- anxiety and stress
- careers with long or irregular working hours
- night or shift work
- work requiring long-distance travel
- multiple jobs
- combining full-time work and school
- being a family caregiver

Causes and symptoms

Sleep deprivation is most often caused by lifestyle choices or the requirements of work, school, or caregiving. Irregular sleep patterns that differ between weekdays and weekends can harm the quality of sleep. A new baby often results in sleep-deprived parents. Teenagers with hectic schedules of school, homework, athletics, after-school activities, jobs, and family and social obligations find themselves without enough hours for quality sleep. Furthermore, hormonal changes in adolescence set most teens’ biological clocks on later schedules than those of children and adults. Teens may be wide awake—albeit exhausted—at bedtime but still have to wake up early for school.

Foods and drugs that change the balance of neurotransmitters in the brain can cause sleep deprivation:

- Caffeinated drinks, such as coffee, and drugs, such as diet pills and decongestants, stimulate parts of the brain and can cause insomnia.
- Although alcohol induces and maintains light sleep, it deprives the brain of REM and deeper sleep.
- Many antidepressants suppress REM sleep.
- Heavy smokers often sleep lightly, are deprived of REM sleep, and wake up after three or four hours from nicotine withdrawal.

Changes in regions of the brain and in neurotransmitters can result in sleep deprivation. Sleeping problems in older people may be a normal part of **aging** or can be related to underlying medical conditions, medications, medical treatments, or sleep-disrupting hospital routines. **Anxiety** or chronic **pain** can cause sleep deprivation, which, in turn, can cause **anxiety disorders** or make it harder to cope with pain. Other conditions that can cause sleep deprivation include:

- menopause
- vision loss
- attention-deficit/hyperactivity disorder (AD/HD)
- head injury
- stroke
- cancer
- Alzheimer's disease

There are more than 70 known types of **sleep disorders**, which may or may not result in sleep deprivation. The most common include:

- insomnia, which can have various causes—including stress, jetlag, diet, or an underlying medical condition—and almost always affects next-day functioning
- sleep apnea—disrupted breathing during sleep—which causes frequent awakenings
- RLS—which is inherited or linked to conditions such as pregnancy, anemia, or diabetes—causes constant leg movement and insomnia
- periodic limb movement disorder (PLMD), which often accompanies RLS and causes repeated awakenings
- narcolepsy, which is characterized by brief attacks of daytime deep sleep and is usually caused by an inherited malfunction in the regulation of sleep-wake cycles

Symptoms of sleep deprivation include:

- difficulty awakening each morning
- daytime drowsiness
- microsleeps—very brief, often unnoticed—periods of sleep during waking hours

- falling asleep during school or work
- need for frequent naps
- routinely falling asleep within five minutes of lying down
- disrupted sleep
- parasomnias—uncontrollable actions during sleep, such as sleepwalking
- headaches
- poor school or work performance
- inability to concentrate
- inability to perform mathematical calculations
- impaired memory
- problems with decision-making
- clumsiness or impaired physical performance
- irritability or mood swings
- paranoia
- confusion
- hallucinations
- decreased consciousness

Although sleep deprivation can be an effective therapy for people with certain types of depression, it can cause depression in otherwise healthy people. Sleep deprivation can also trigger manic episodes of agitation and hyperactivity in people with **bipolar disorder** and seizures in patients with some types of **epilepsy**.

Diagnosis

Examination

Sleep deprivation is usually readily diagnosed from the symptoms accompanying the lack of sleep. Underlying medical problems resulting in sleep deprivation may require further diagnoses.

Procedures

Simple devices are available for detecting **sleep apnea**. Sleep apnea may be diagnosed at a specialized sleep center using **polysomnography** to record brain waves, heartbeat, and breathing for an entire night.

Treatment

Traditional

The usual treatment for sleep deprivation is sleep. Underlying conditions that result in sleep deprivation require more extensive treatments. For example, severe sleep apnea may require a mask—called a continuous positive airway pressure (CPAP) device—to

KEY TERMS

Adenosine—A nucleoside that plays multiple physiological roles in energy transfer and molecular signaling, as a component of RNA, and as an inhibitory neurotransmitter that promotes sleep.

Apnea—The transient cessation of breathing.

Circadian rhythm—A 24-hour cycle of physiological or behavioral activities.

Cytokines—A class of proteins, including interferons and interleukins, that are released by cells as part of the immune response and as mediators of intercellular communication.

Insomnia—Prolonged or abnormal inability to obtain adequate sleep.

Melatonin—A hormone involved in regulation of circadian rhythms.

Narcolepsy—A condition characterized by brief attacks of deep sleep.

Neurotransmitters—Chemicals that transmit nerve impulses from one nerve cell to another.

REM—Rapid eye movement; a stage of the normal sleep cycle characterized by rapid eye movements, increased forebrain and midbrain activity, and dreaming.

Restless legs syndrome (RLS)—A neurological disorder characterized by aching, burning, or creeping sensations in the legs and an urge to move the legs, often resulting in insomnia.

keep the airways open during sleep. Surgery may be required to correct an airway obstruction.

Drugs

Caffeine and other stimulants cannot overcome the effects of severe sleep deprivation. However various products are available to treat sleep disturbances that can result in sleep deprivation:

- Over-the-counter sleep aids usually contain antihistamines. Although they are sometimes effective, they have side effects and tolerance can develop after just a few days of use.
- RLS and PLMD are often relieved with drugs that affect the neurotransmitter dopamine.
- Daily melatonin supplements can improve nighttime sleep in blind patients.

The U.S. Food and Drug Administration (FDA) has approved several sleep aids—called sedatives/hypnotics—for indefinite use. However most sleeping pills are usually prescribed only for short-term **insomnia**, because they:

- usually become ineffective after several weeks of nightly use
- can cause insomnia with long-term use
- may be habit-forming
- can mask an underlying cause of sleep deprivation
- can interact with alcohol and other medications
- can cause next-day grogginess
- can cause bizarre behaviors, such as sleep binge-eating or sleep driving
- can prevent people with sleep apnea from waking up to breathe

Alternative

Alternative treatments for insomnia and other sleep disturbances include:

- cognitive-behavioral therapy (CBT)
- hypnosis
- melatonin, a hormone derived from the neurotransmitter serotonin
- tryptophan, an amino acid precursor of serotonin

Herbal remedies for insomnia include:

- lemon balm
- chamomile
- valerian root
- kava kava
- passionflower
- lavender
- St. John's wort

Home remedies

Short-term sleep deprivation may require only a night or two of additional sleep. Longer-term sleep deprivation may require a sleep vacation—a few days devoted to sleeping as much as needed. Mild sleep apnea can be treated effectively by weight loss or by not sleeping on one's back.

Prognosis

Sleep deprivation is usually readily reversible with adequate sleep.

Prevention

Sleep deprivation is preventable by getting as much sleep as an individual requires. Sleep deprivation caused by mild insomnia can often be prevented by:

- sleeping on a schedule—going to bed and rising at the same time every day, including weekends
- structuring daily activities
- exercising 20–30 minutes every day, especially 5–6 hours before sleep
- practicing stress management
- avoiding caffeine, nicotine, and alcohol
- relaxing before bed with activities such as reading or a warm bath that become routinely associated with sleep
- avoiding extreme temperatures that prevent falling or staying asleep
- avoiding lying awake in bed for more than 20 minutes since this can cause anxiety
- reading, watching television, listening to music, or performing an activity until drowsy
- sleeping until sunrise or waking with very bright lights to reset one's internal clock each day
- getting an hour of morning sun exposure

It is the responsibility of parents to ensure that their children and teens get adequate sleep. Some high schools have moved to a later start-time to address sleep deprivation in teenagers.

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ORGANIZATIONS

American Academy of Sleep Medicine, One Westbrook Corporate Center, Ste. 920, Westchester, IL, 60154, (708) 492-0930, (708) 492-0943, <http://www.aasmnet.org>.

National Institute of Neurological Disorders and Stroke (NINDS), NIH Neurological Institute, PO Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov/index.htm>.

National Sleep Foundation, 1522 K Street, NW, Suite 500, Washington, DC, 20005, (202) 347-3471, (202) 347-3472, nsf@sleepfoundation.org, <http://www.sleepfoundation.org>.

Margaret Alic, PhD

Sleep disorders

Definition

Sleep disorders are a group of syndromes characterized by disturbance in the patient's amount of sleep, quality or timing of sleep, or in behaviors or physiological conditions associated with sleep. There are about 81 different sleep disorders, according to the second edition of the *International Classification of Sleep Disorders*. To qualify for the diagnosis of sleep disorder, the condition must be a persistent problem, cause the patient significant emotional distress, and interfere with his or her social or occupational functioning.

Because sleep requirements vary from person to person, there is no specific amount of time spent sleeping that can be used as a cutoff to determine whether a person has a sleep disorder. Some healthy adults need



A patient suffering from acute sleep apnea is hooked up to monitors in preparation for a night's sleep at a Stanford University sleep lab. (Russell D. Curtis/Photo Researchers, Inc.)

as much as 10 hours of sleep per night whereas others need as little as 5 hours.

Demographics

Sleep disorders are a common problem in the general population of North America. Researchers estimate that 20–40% of adults report difficulty sleeping at some point each year. About a third of all Americans will have a sleep disorder at some point in their lives. Twenty percent of adults say that they have problems with chronic **insomnia** and 17% consider their sleeping problem to be serious.

As far as is known, sleep disorders are equally common in all racial and ethnic groups in Canada and the United States.

Description

Normal sleep

Although sleep is a basic behavior in animals as well as humans, researchers still do not completely understand all of its functions in maintaining health.

In the past 30 years, however, laboratory studies on human volunteers have yielded new information about the different types of sleep. Increasing interest in sleep disorders led to the recognition of sleep medicine as a distinct medical subspecialty with its own board certification procedures in 1978. Researchers have learned about the cyclical patterns of different types of sleep and their relationships to breathing, heart rate, brain waves, and other physical functions. These measurements are obtained by a technique called **polysomnography**.

There are five stages of normal human sleep. Four stages consist of non-rapid eye movement (NREM) sleep, with unique brain wave patterns and physical changes occurring. Dreaming occurs in the fifth stage, during rapid eye movement (REM) sleep.

- Stage 1 NREM sleep. This stage occurs while a person is falling asleep. It represents about 5% of a normal adult's sleep time.
- Stage 2 NREM sleep. In this stage, (the beginning of “true” sleep), the person's electroencephalogram

(EEG) will show distinctive wave forms called sleep spindles and K complexes. About 50% of sleep time is stage 2 REM sleep.

- Stages 3 and 4 NREM sleep. Also called delta or slow wave sleep, these are the deepest levels of human sleep and represent 10–20% of sleep time. They usually occur during the first 30–50% of the sleeping period.
- REM sleep. REM sleep accounts for 20–25% of total sleep time. It usually begins about 90 minutes after the person falls asleep, an important measure called REM latency. It alternates with NREM sleep about every hour and a half throughout the night. REM periods increase in length over the course of the night.

Sleep cycles vary with a person's age. Children and adolescents have longer periods of stage 3 and stage 4 NREM sleep than do middle aged or elderly adults. Because of this difference, the doctor will need to take a patient's age into account when evaluating a sleep disorder. Total REM sleep also declines with age.

The average length of nighttime sleep varies among different age groups. Infants typically need about 16 hours of sleep each day, while adolescents need about 9 hours. Most adults sleep between 7 and 9 hours a night, although pregnant women may need as many as 10 or 11 hours of sleep. This population average appears to be constant throughout the world. In temperate climates, however, people often notice that sleep time varies with the seasons. It is not unusual for people in North America and Europe to sleep about 40 minutes longer per night during the winter.

Primary sleep disorders

Sleep disorders are classified based on what causes them. Primary sleep disorders are distinguished from those that are not caused by other mental disorders, prescription medications, **substance abuse**, or medical conditions. The two major categories of primary sleep disorders are the dyssomnias and the parasomnias.

DYSSOMNIAS. Dyssomnias are primary sleep disorders in which the patient suffers from changes in the amount, restfulness, and timing of sleep. The most important dyssomnia is primary insomnia, which is defined as difficulty in falling asleep or remaining asleep that lasts for at least one month. It is estimated that 35% of adults in the United States experience insomnia during any given year but the number of these adults who are experiencing true primary insomnia is unknown. Primary insomnia can be caused by a traumatic event related to sleep or bedtime and it is often associated with increased physical or psychological arousal at night. People who experience primary insomnia are often anxious about not being able to

sleep. The person may then associate all sleep-related things (their bed, bedtime, etc.) with frustration, making the problem worse. The person then becomes more stressed about not sleeping. Primary insomnia usually begins when the person is a young adult or in middle age.

Hypersomnia is a condition marked by excessive sleepiness during normal waking hours. The patient has either lengthy episodes of daytime sleep or episodes of daytime sleep on a daily basis even though he or she is sleeping normally at night. In some cases, patients with primary hypersomnia have difficulty waking in the morning and may appear confused or angry. This condition is sometimes called sleep drunkenness and is more common in males. The number of people with primary hypersomnia is unknown, although 5–10% of patients in sleep disorder clinics have the disorder. Primary hypersomnia usually affects young adults between the ages of 15 and 30.

Nocturnal myoclonus and **restless legs syndrome** (RLS) can cause either insomnia or hypersomnia in adults. Patients with nocturnal myoclonus wake up because of cramps or twitches in the calves. These patients feel sleepy the next day. Nocturnal myoclonus is sometimes called periodic limb movement disorder (PLMD). RLS patients have a crawly or aching feeling in their calves that can be relieved by moving or rubbing the legs. RLS often prevents the patient from falling asleep until the early hours of the morning, when the condition is less intense.

Kleine-Levin syndrome is a recurrent form of hypersomnia that affects a person three or four times a year. Doctors do not know the cause of this syndrome. It is marked by two to three days of sleeping 18–20 hours per day, hypersexual behavior, compulsive eating, and irritability. Men are three times more likely than women to have the syndrome. Currently, there is no cure for this disorder.

Narcolepsy is a dyssomnia characterized by recurrent “sleep attacks” that the patient cannot fight. The sleep attacks are about 10–20 minutes long. The patient feels refreshed by the sleep, but typically feels sleepy again several hours later. Narcolepsy has three major symptoms in addition to sleep attacks: cataplexy, **hallucinations**, and sleep **paralysis**. Cataplexy is the sudden loss of muscle tone and stability (“drop attacks”). Hallucinations may occur just before falling asleep (hypnagogic) or right after waking up (hypnopompic) and are associated with an episode of REM sleep. Sleep paralysis occurs during the transition from being asleep to waking up. About 40% of patients with narcolepsy have or have had another mental disorder.

Although narcolepsy is often regarded as an adult disorder, it has been reported in children as young as three years old. Almost 18% of patients with narcolepsy are 10 years old or younger. It is estimated that 0.02–0.16% of the general population suffer from narcolepsy. Men and women are equally affected.

Breathing-related sleep disorders are syndromes in which the patient's sleep is interrupted by problems with his or her breathing. There are three types of breathing-related sleep disorders:

- Obstructive sleep apnea syndrome. This is the most common form of breathing-related sleep disorder, marked by episodes of blockage in the upper airway during sleep. It is found primarily in obese people. Patients with this disorder typically alternate between periods of snoring or gasping (when their airway is partly open) and periods of silence (when their airway is blocked). Very loud snoring is a clue to this disorder.
- Central sleep apnea syndrome. This disorder is primarily found in elderly patients with heart or neurological conditions that affect their ability to breathe properly. It is not associated with airway blockage and may be related to brain disease.
- Central alveolar hypoventilation syndrome. This disorder is found most often in extremely obese people. The patient's airway is not blocked, but his or her blood oxygen level is too low.
- Mixed-type sleep apnea syndrome. This disorder combines symptoms of both obstructive and central sleep apnea.

Circadian rhythm sleep disorders are dyssomnias resulting from a discrepancy between the person's daily sleep/wake patterns and demands of social activities, shift work, or travel. The term *circadian* comes from a Latin word meaning daily. There are three circadian rhythm sleep disorders. Delayed sleep phase type is characterized by going to bed and arising later than most people. **Jet lag** type is caused by travel to a new time zone. Shift work type is caused by the schedule of a person's job. People who are ordinarily early risers appear to be more vulnerable to jet lag and shift work-related circadian rhythm disorders than people who are "night owls." There are some patients who do not fit the pattern of these three disorders and appear to be the opposite of the delayed sleep phase type. These patients have an advanced sleep phase pattern and cannot stay awake in the evening, but wake up on their own in the early morning.

PARASOMNIAS. Parasomnias are primary sleep disorders in which the patient's behavior is affected by specific sleep stages or transitions between sleeping

and waking. They are sometimes described as disorders of physiological arousal during sleep.

Nightmare disorder is a parasomnia in which the patient is repeatedly awakened from sleep by frightening dreams and is fully alert on awakening. The actual rate of occurrence of nightmare disorder is unknown. Approximately 10–50% of children between three and five years old have nightmares. They occur during REM sleep, usually in the second half of the night. The child is usually able to remember the content of the nightmare and may be afraid to go back to sleep. More females than males have this disorder but it is not known whether the sex difference reflects a difference in occurrence or a difference in reporting. Nightmare disorder is most likely to occur in children or adults under severe or traumatic **stress**.

Sleep terror disorder is a parasomnia in which the patient awakens screaming or crying. The patient also has physical signs of arousal, like sweating, shaking, etc. It is sometimes referred to as *pavor nocturnus*. Unlike nightmares, sleep terrors typically occur in stage 3 or stage 4 NREM sleep during the first third of the night. The patient may be confused or disoriented for several minutes and cannot recall the content of the dream. He or she may fall asleep again and not remember the episode the next morning. Sleep terror disorder is most common in children 4 to 12 years old and is outgrown in adolescence. It affects about 3% of children. Fewer than 1% of adults have the disorder. In adults, it usually begins between the ages of 20 and 30. In children, more males than females have the disorder. In adults, men and women are equally affected.

Sleepwalking disorder, which is sometimes called somnambulism, occurs when the patient is capable of complex movements during sleep, including walking. Like sleep terror disorder, sleepwalking occurs during stage 3 and stage 4 NREM sleep during the first part of the night. If the patient is awakened during a sleepwalking episode, he or she may be disoriented and have no memory of the behavior. In addition to walking around, patients with sleepwalking disorder have been reported to eat, use the bathroom, unlock doors, or talk to others. It is estimated that 10–30% of children have at least one episode of sleepwalking. However, only 1–5% meet the criteria for sleepwalking disorder. The disorder is most common in children 8 to 12 years old. It is unusual for sleepwalking to occur for the first time in adults.

Unlike sleepwalking, REM sleep behavior disorder occurs later in the night and the patient can remember what they were dreaming. The physical activities of the patient are often violent.

Sleep disorders related to other conditions

In addition to the primary sleep disorders, there are three categories of sleep disorders that are caused by or related to substance use or other physical or mental disorders.

SLEEP DISORDERS RELATED TO MENTAL DISORDERS.

Many mental disorders, especially depression or one of the **anxiety disorders**, can cause sleep disturbances. Psychiatric disorders are the most common cause of chronic insomnia.

SLEEP DISORDERS DUE TO MEDICAL CONDITIONS.

Some patients with chronic neurological conditions like **Parkinson's disease** or **Huntington's disease** may develop sleep disorders. Sleep disorders have also been associated with viral **encephalitis**, brain disease, and hypo- or **hyperthyroidism**.

SUBSTANCE-INDUCED SLEEP DISORDERS. The use of drugs, alcohol, and **caffeine** frequently produces disturbances in sleep patterns. Alcohol **abuse** is associated with insomnia. The person may initially feel sleepy after drinking but wakes up or sleeps fitfully during the second half of the night. Alcohol can also increase the severity of breathing-related sleep disorders. With amphetamines or **cocaine**, the patient typically suffers from insomnia during drug use and hypersomnia during drug withdrawal. Opioids usually make short-term users sleepy. However, long-term users develop tolerance and may suffer from insomnia.

In addition to alcohol and drugs that are abused, a variety of prescription medications can affect sleep patterns. These medications include **antihistamines**, **corticosteroids**, **asthma** medicines, and drugs that affect the central nervous system.

Sleep disorders in children and adolescents

Pediatricians estimate that 20–30% of children have difficulties with sleep that are serious enough to disturb their families. Although sleepwalking and night terror disorder occur more frequently in children than in adults, children can also suffer from narcolepsy and **sleep apnea** syndrome.

Risk factors

Risk factors for sleep disorders include:

- Sex. Primary insomnia is more common in women than in men, while obstructive sleep apnea is twice as common in men as in women.
- Age. Older adults are more likely to develop sleep disorders; the rate rises from 5% of adults between 30 and 50 to 30% in those over 50. One reason for the greater risk of sleep disorders in seniors is that they

are more likely to have medical conditions that disturb sleep or to be taking medications that cause sleep disruption.

- Employment that requires frequent travel across time zones or frequent changes in work schedules.
- Environmental factors, including noise, high altitude, and abnormally hot or cold temperatures.
- Smoking. Heavy smokers often wake up after only a few hours of sleep due to nicotine withdrawal.
- High levels of emotional stress, whether job-related or associated with family or personal problems.
- Family history of sleep disorders. Sleepwalking is particularly likely to run in families.
- Having a disease or disorder that causes physical discomfort.
- Genetic factors. There is increasing evidence that obstructive sleep apnea, narcolepsy, and restless legs syndrome are associated with susceptibility genes for these disorders, although no specific genes have been identified as of 2009.

Causes and symptoms

The causes of sleep disorders have already been discussed with respect to the classification of these disorders.

The most important symptoms of sleep disorders are insomnia and sleepiness during waking hours. Insomnia is by far the more common of the two symptoms. It covers a number of different patterns of sleep disturbance. These patterns include inability to fall asleep at bedtime, repeated awakening during the night, and/or inability to go back to sleep once awakened.

Diagnosis

Diagnosis of sleep disorders usually requires a psychological history as well as a medical history. The patient's sex and age are useful starting points in assessing the problem. The doctor may also talk to other family members in order to obtain information about the patient's symptoms. The family's observations are particularly important to evaluate sleepwalking, kicking in bed, **snoring** loudly, or other behaviors that the patient cannot remember.

Examination

With the exception of sleep apnea syndromes, physical examinations are not usually revealing.

Tests

The doctor may order blood or urine tests to determine whether the patient's sleep disorder is associated

with anemia, thyroid dysfunction, **alcoholism**, or opioid abuse. The patient's blood oxygen level may be tested during sleep in order to determine whether sleep apnea or other types of sleep-disordered breathing are involved. Imaging tests are not routinely done for sleep disorders but may be ordered in some cases to rule out brain tumors or other medical conditions.

Sleep logs

Many doctors ask patients to keep a sleep diary or sleep log for a minimum of one to two weeks in order to evaluate the severity and characteristics of the sleep disturbance. The patient records medications taken as well as the length of time spent in bed, the quality of the sleep, and similar information. Some sleep logs are designed to indicate circadian sleep patterns as well as simple duration or restfulness of sleep.

Psychological testing

The doctor may use **psychological tests** or inventories to evaluate insomnia because it is frequently associated with mood or affective disorders. The **Minnesota Multiphasic Personality Inventory** (MMPI), the Millon Clinical Multiaxial Inventory (MCMII), the Beck Depression Inventory, and the Zung Depression Scale are the tests most commonly used in evaluating this symptom.

Self-report tests

The Epworth Sleepiness Scale, a self-rating form recently developed in Australia, consists of eight questions used to assess daytime sleepiness. Scores range from 0–24, with scores higher than 16 indicating severe daytime sleepiness.

Laboratory sleep studies

If the doctor is considering breathing-related sleep disorders, myoclonus, or narcolepsy as possible diagnoses, he or she may ask the patient to be tested in a sleep laboratory or at home with portable instruments.

POLYSOMNOGRAPHY. Polysomnography can be used to help diagnose sleep disorders as well as conduct research into sleep. In some cases the patient is tested in a special sleep laboratory. The advantage of this testing is the availability and expertise of trained technologists but it is expensive. As of 2009, however, portable equipment is available for home recording of certain specific physiological functions.

MULTIPLE SLEEP LATENCY TEST (MSLT). The multiple sleep latency test (MSLT) is frequently used to measure the severity of the patient's daytime sleepiness. The test measures sleep latency (the speed with which the patient

falls asleep) during a series of planned naps during the day. The test also measures the amount of REM sleep that occurs. Two or more episodes of REM sleep under these conditions indicates narcolepsy. This test can also be used to help diagnose primary hypersomnia.

REPEATED TEST OF SUSTAINED WAKEFULNESS (RTSW). The repeated test of sustained wakefulness (RTSW) is a test that measures sleep latency by challenging the patient's ability to stay awake. In the RTSW, the patient is placed in a quiet room with dim lighting and is asked to stay awake. As with the MSLT, the testing pattern is repeated at intervals during the day.

Treatment

Traditional

Treatment for a sleep disorder depends on what is causing the disorder. For example, if major depression is the cause of insomnia, then treatment of the depression with antidepressants should resolve the insomnia. In other cases, a change of environment or work schedule may help.

Drugs

Sedative or hypnotic medications are generally recommended only for insomnia related to a temporary stress (like surgery or grief) because of the potential for **addiction** or overdose. In general, these drugs are given for two weeks or less in order to reduce the risk of dependence. Trazodone, a sedating antidepressant, is often used for chronic insomnia that does not respond to other treatments. Sleep medications may also cause problems for elderly patients because of possible interactions with their other prescription medications. Among the safer hypnotic agents are lorazepam, temazepam, and zolpidem. Chloral hydrate is often preferred for short-term treatment in elderly patients because of its mildness. Short-term treatment is recommended because this drug may be habit forming.

Narcolepsy is treated with such stimulants as dextroamphetamine sulfate or methylphenidate. Nocturnal myoclonus has been successfully treated with clonazepam.

Children with sleep terror disorder or sleepwalking are usually treated with **benzodiazepines** because this type of medication suppresses stage 3 and stage 4 NREM sleep.

If the cause of insomnia is RLS, treatment includes massage, warm baths, and visualization techniques to distract from the discomfort. The only medication

KEY TERMS

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Benzodiazepines—A class of sedative drugs used to treat sleep disorders.

Cataplexy—Sudden loss of muscle tone (often causing a person to fall), usually triggered by intense emotion. It is regarded as a diagnostic sign of narcolepsy.

Circadian rhythm—Any body rhythm that recurs in 24-hour cycles. The sleep-wake cycle is an example of a circadian rhythm.

Cognitive behavioral therapy (CBT)—A type of psychotherapy that helps patients identify and change problematic thoughts and behaviors.

Dyssomnia—A primary sleep disorder in which the patient suffers from changes in the quantity, quality, or timing of sleep.

Electroencephalogram (EEG)—The record obtained by a device that measures electrical impulses in the brain.

Hypersomnia—An abnormal increase of 25% or more in time spent sleeping. Patients usually have excessive daytime sleepiness.

Hypnotic—A medication that makes a person sleep.

Hypopnea—Shallow or excessively slow breathing usually caused by partial closure of the upper airway during sleep, leading to disruption of sleep.

Insomnia—Difficulty in falling asleep or remaining asleep.

Jet lag—A temporary disruption of the body's sleep-wake rhythm following high-speed air travel across several time zones. Jet lag is most severe in people who have crossed eight or more time zones in 24 hours.

Kleine-Levin syndrome—A disorder that occurs primarily in young males, three or four times a year. The syndrome is marked by episodes of hypersomnia, hypersexual behavior, and excessive eating.

Melatonin—A hormone produced by the pineal gland that is associated with sleep, and that may be useful in the treatment of some sleep disorders.

Narcolepsy—A lifelong sleep disorder marked by four symptoms: sudden brief sleep attacks,

approved by the U.S. Food and Drug Administration (FDA) for the treatment of RLS is ropinirole (Requip), although drugs used for the treatment of Parkinson's disease, benzodiazepines, and anticonvulsant medications also may be effective for this disorder.

Psychotherapy

Psychotherapy is recommended for patients with sleep disorders associated with depression or other mental disorders. In many cases the patient's scores on the Beck or Zung inventories will suggest the appropriate direction of treatment.

Cognitive-behavioral therapy (CBT) is a form of psychotherapy that is often recommended for insomnia as a way of breaking the cycle of **anxiety** about sleep and sleeplessness associated with insomnia. The patient is typically advised to limit the amount of time they spend in bed and to change certain habits that may contribute to the insomnia. For example, some patients are clock watchers who check their bedside clocks frequently to see how little sleep they are getting. They will be advised to put the clock under the bed or in some other location where they can't see it during the night.

Sleep education

“Sleep hygiene” or sleep education for sleep disorders often includes instructing the patient in methods to enhance sleep. Patients are advised to:

- wait until they are sleepy before going to bed
- avoid using the bedroom for work, reading, or watching television
- get up at the same time every morning no matter how much or how little they slept
- avoid smoking and avoid drinking liquids with caffeine
- get some physical exercise early in the day every day
- limit fluid intake after dinner; in particular, avoid alcohol because it frequently causes interrupted sleep
- learn to meditate or practice relaxation techniques
- avoid tossing and turning in bed; instead, the patient can get up and listen to relaxing music or read

Lifestyle changes

Patients with sleep apnea or hypopnea are encouraged to stop **smoking**, avoid alcohol or drugs of abuse,

cataplexy, temporary paralysis, and hallucinations. The hallucinations are associated with falling asleep or the transition from sleeping to waking.

Nocturnal myoclonus—A disorder in which the patient is awakened repeatedly during the night by cramps or twitches in the calf muscles. Nocturnal myoclonus is sometimes called periodic limb movement disorder (PLMD).

Non-rapid eye movement (NREM) sleep—A type of sleep that differs from rapid eye movement (REM) sleep. The four stages of NREM sleep account for 75–80% of total sleeping time.

Parasomnia—A primary sleep disorder in which the person's physiology or behaviors are affected by sleep, the sleep stage, or the transition from sleeping to waking.

Pavor nocturnus—Another term for sleep terror disorder.

Polysomnography—Laboratory measurement of a patient's basic physiological processes during sleep. Polysomnography usually measures eye movement, brain waves, and muscular tension.

Primary sleep disorder—A sleep disorder that cannot be attributed to a medical condition, another mental disorder, or prescription medications or other substances.

Rapid eye movement (REM) sleep—A phase of sleep during which the person's eyes move rapidly beneath the lids. It accounts for 20–25% of sleep time. Dreaming occurs during REM sleep.

REM latency—After a person falls asleep, the amount of time it takes for the first onset of REM sleep.

Restless legs syndrome (RLS)—A disorder in which the patient experiences crawling, aching, or other disagreeable sensations in the calves that can be relieved only by movement. RLS is a frequent cause of difficulty falling asleep at night.

Sedative—A medication given to calm agitated patients; sometimes used as a synonym for hypnotic.

Sleep latency—The amount of time that it takes to fall asleep. Sleep latency is measured in minutes and is important in diagnosing depression.

Somnambulism—Another term for sleepwalking.

and lose weight in order to improve the stability of the upper airway.

In some cases, patients with sleep disorders related to jet lag or shift work may need to change employment or travel patterns. Patients may need to avoid rapid changes in shifts at work.

Children with nightmare disorder may benefit from limits on television or movies. Violent scenes or frightening science fiction stories appear to influence the frequency and intensity of children's nightmares.

Surgery

Although making a surgical opening into the windpipe (a tracheostomy) for sleep apnea or hypopnea in adults is a treatment of last resort, it is occasionally performed if the patient's disorder is life threatening and cannot be treated by other methods. In children and adolescents, surgical removal of the tonsils and adenoids is a fairly common and successful treatment for sleep apnea. Most sleep apnea patients are treated with continuous positive airway pressure (CPAP). Sometimes an oral prosthesis is used for mild sleep apnea.

Alternative

Some alternative approaches may be effective in treating insomnia caused by anxiety or emotional stress. **Meditation** practice, breathing exercises, and **yoga** can break the vicious cycle of sleeplessness, worry about inability to sleep, and further sleeplessness for some people. Yoga can also help some people to relax muscular tension in a direct fashion. The breathing exercises and meditation can keep some patients from obsessing about sleep.

Homeopathic practitioners recommend that people with chronic insomnia see a professional homeopath. They do, however, prescribe specific remedies for at-home treatment of temporary insomnia: *Nux vomica* for alcohol or substance-related insomnia, *Ignatia* for insomnia caused by grief, *Arsenicum* for insomnia caused by fear or anxiety, and *Passiflora* for insomnia related to mental stress.

Melatonin has also been used as an alternative treatment for sleep disorders. Melatonin is produced in the body by the pineal gland at the base of the brain. This substance is thought to be related to the body's circadian rhythms.

Practitioners of Chinese medicine usually treat insomnia as a symptom of excess yang energy. Cinnabar is recommended for chronic nightmares. Either magnetic magnetite or “dragon bones” is recommended for insomnia associated with **hysteria** or fear. If the insomnia appears to be associated with excess yang energy arising from the liver, the practitioner will give the patient oyster shells. **Acupuncture** treatments can help bring about balance and facilitate sleep.

Dietary changes like eliminating stimulant foods (coffee, cola, chocolate) and late-night meals or snacks can be effective in treating some sleep disorders. Nutritional supplementation with magnesium, as well as botanical medicines that calm the nervous system, can also be helpful. Among the botanical remedies that may be effective for sleep disorders are valerian (*Valeriana officinalis*), passionflower (*Passiflora incarnata*), and skullcap (*Scutellaria lateriflora*).

Home remedies

Warm milk before bedtime is a classic home remedy for insomnia. It is thought that this treatment works for many people because milk contains tryptophan, an amino acid that increases the brain's production of melatonin.

Prognosis

The prognosis depends on the specific disorder. Children usually outgrow sleep disorders. Patients with Kleine-Levin syndrome usually get better around age 40. Narcolepsy, however, is a lifelong disorder, although cataplexy can be successfully controlled with medication, and many people find that their symptoms naturally decrease after age 60. The prognosis for sleep disorders related to other conditions depends on successful treatment of the substance abuse, medical condition, or other mental disorder. The prognosis for primary sleep disorders is affected by many things, including the patient's age, sex, occupation, personality characteristics, family circumstances, neighborhood environment, and similar factors.

About 85% of people with insomnia find relief with a combination of sleep hygiene and medication. Although there is no cure for sleep apnea, treatment can reduce the associated risks of high blood pressure and heart disease.

Insomnia and other sleep disorders are not fatal in and of themselves; however, chronic insomnia is associated with an increased risk of depression and **suicide**. In addition, insufficient sleep increases a person's risk of accidents on the road and in the workplace, with the

possibility of serious injury or **death**. Driver **fatigue** is responsible for an estimated 100,000 motor vehicle accidents and 1500 deaths each year in the United States, according to the National Highway Traffic Safety Administration.

Prevention

Certain sleep disorders, such as insomnia, can sometimes be prevented by practicing good sleep hygiene. As mentioned, sleep hygiene involves going to bed at a regular time each night and avoiding stimulating activities, smoking, or heavy meals close to bedtime. Sleep apnea may be prevented in some cases by controlling body weight.

Resources

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ORGANIZATIONS

- American Academy of Sleep Medicine (AASM), One Westbrook Corporate Center, Suite 920, Westchester, IL, 60154, (708) 492-0930, (708) 492-0943, <http://www.aasmnet.org/>.
- American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209-3901, (703) 907-7300, apa@psych.org, <http://www.psych.org/>.
- American Sleep Association (ASA), 110 West Ninth Street, Suite 826, Wilmington, DE, 19801, (940) 234-3357, <http://www.sleepassociation.org/>.
- Anxiety Disorders Association of America (ADAA), 8730 Georgia Ave., Suite 600, Silver Spring, MD, 20910, (240) 485-1001, (240) 485-1035, information@adaa.org, <http://www.adaa.org/>.
- National Institute of Neurological Disorders and Stroke (NINDS), P.O. Box 5801, Bethesda, MD, 20824, (800) 352-9424, (301) 496-5751, <http://www.ninds.nih.gov/index.htm>.
- National Sleep Foundation (NSF), 1522 K Street, NW, Suite 500, Washington, DC, 20005, (202) 347-3471, (202) 347-3472, nsf@sleepfoundation.org, <http://www.sleepfoundation.org/>.

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Sleep study see **Polysomnography**

Sleeping drugs see **Anti-insomnia drugs**

Sleeping sickness

Definition

Sleeping sickness (also called trypanosomiasis) is an infection caused by *Trypanosoma* protozoa; it is passed to humans through the bite of the tsetse fly. If left untreated, the infection progresses to **death** within months or years.

Description

Protozoa are single-celled organisms considered to be the simplest life form in the animal kingdom. The protozoa responsible for sleeping sickness are a variety that bear numerous flagella (hair-like projections from the cell that help the cell to move). These protozoa exist only on the continent of Africa. The type of protozoa causing sleeping sickness in humans is referred to as the *Trypanosoma brucei* complex, which can be divided further into Rhodesian (Central and East African) and Gambian (Central and West African) subspecies.

The Rhodesian variety live within antelopes in savanna and woodland areas and they cause no problems with the antelope's health. The protozoa are then acquired by tsetse flies when they bite and suck the blood of an infected antelope or cow.

Within the tsetse fly, the protozoa cycle through several different life forms; ultimately they migrate to the salivary glands of the tsetse fly. Once the protozoa are harbored in the salivary glands, they are ready to be deposited into the bloodstream of the fly's next source of a blood meal.

Humans most likely to become infected by Rhodesian trypanosomes are people such as game wardens and visitors to game parks in East Africa, who may be bitten by a tsetse fly that has fed on game (antelope) carrying the protozoa. The Rhodesian variety of sleeping sickness causes a much more severe illness, with even greater likelihood of eventual death than the Gambian form.

The Gambian variety of *Trypanosoma* thrives in tropical rain forests throughout Central and West Africa; it does not infect game or cattle, and is primarily a threat to people dwelling in such areas, rarely infecting visitors.

Causes and symptoms

The first sign of infection with the trypanosome may be a sore appearing at the site of the tsetse fly bite about two to three days after having been bitten. Redness, **pain**, and swelling occur but are often ignored by the patient.

DAVID BRUCE (1855–1931)

David Bruce was born in Melbourne, Australia, on May 29, 1855, to Scottish immigrants. Bruce's family moved back to Scotland when he was five years old. Bruce attended the University of Edinburgh where he studied natural history and medicine. Following his graduation, he accepted a position working with a doctor. In 1883, Bruce married Mary Elizabeth Steele who would help him with his work throughout his life.

In 1884, Bruce began to study the disease "Malta, Mediterranean" when he and Mary were stationed in Malta with the Army Medical Service. Using a microscope, Bruce discovered that the disease was caused by a "micrococcus" that grew in the individual's spleen. The organism responsible for this disease was ultimately isolated by Bernhard L. F. Bang. In 1905, Bruce led a scientific team that discovered that the soldiers who contracted the disease had ingested the milk of infected goats. The disease disappeared when the soldiers quit drinking goat's milk. Many physicians began calling the disease "brucellosis" in honor of Bruce's discoveries. Bruce also conducted research in Africa where he found that the tsetse fly could infect humans, as well as animals, with the "nagana" disease. Ultimately, his work would prove that sleeping sickness was caused by the tsetse fly.

In 1903, Bruce became the director of the Royal Society's Sleeping Sickness Commission and, in 1908, he was knighted. He served as commandant of the Royal Army Medical College after he and his wife returned to England. Bruce died on November 20, 1931.

Stage I illness

Two to three weeks later, Stage I disease develops as a result of the protozoa being carried through the blood and lymph circulation of the host. This systemic (meaning that symptoms affect the whole body) phase of the illness is characterized by a **fever** that rises quite high, then falls to normal, then respikes (rises rapidly). A rash with intense **itching** may be present, and **headache** and mental confusion may occur. The Gambian form, in particular, includes extreme swelling of lymph tissue, with enlargement of both the spleen and liver, and greatly swollen lymph nodes. "Winterbottom's sign" is classic of Gambian sleeping sickness, and consists of a visibly swollen area of lymph nodes located behind the ear and just above the base of the neck. During this stage, the heart may be affected by a severe inflammatory reaction, particularly when the infection is caused by the Rhodesian variety of trypanosomiasis.

Many of the symptoms of sleeping sickness are actually the result of attempts by the patient's immune

system to get rid of the invading organism. The heightened activity of the cells of the immune system result in damage to the patient's own organs, anemia, and leaky blood vessels. These leaks in the blood vessels end up helping to further spread the protozoa throughout the afflicted person's body.

One reason for the intense reaction of the immune system to the presence of the trypanosomes is also the reason why the trypanosomes survive so well despite the efforts of the immune system to eradicate them. The protozoa causing sleeping sickness are able to rapidly change specific markers (unique proteins) on their outer coats. These kinds of markers usually serve to stimulate the host's immune system to produce immune cells that will specifically target the marker, allowing quick destruction of those cells bearing the markers. Trypanosomes, however, are able to express new markers at such a high rate of change that the host's immune system is constantly trying to catch up.

Stage II illness

Stage II sleeping sickness involves the nervous system. Gambian sleeping sickness, in particular, has a clearly delineated phase in which the predominant symptoms involve the brain. The patient's speech becomes slurred, mental processes slow, and the patient sits and stares for long periods of time, or sleeps. Other symptoms resemble Parkinson's disease, including imbalance when walking, slow and shuffling gait, trembling of the limbs, involuntary movements, muscle tightness, and increasing mental confusion. Untreated, these symptoms eventually lead to **coma** and then to death.

Diagnosis

Diagnosis of sleeping sickness can be made by microscopic examination of fluid from the original sore at the site of the tsetse fly bite. Trypanosomes will be present in the fluid for a short period of time following the bite. If the sore has already resolved, fluid can be obtained from swollen lymph nodes for examination. Other methods of trypanosome diagnosis involve culturing blood, lymph node fluid, bone marrow, or spinal fluid. These cultures are then injected into rats, which develop blood-borne protozoa infection that can be detected in blood smears within one to two weeks. However, this last method is effective only for the Rhodesian variety of sleeping sickness.

Treatment

Without treatment, sleeping sickness will lead to death. Unfortunately, however, those medications effective against the *Trypanosoma brucei* complex

KEY TERMS

Immune system—That network of tissues and cells throughout the body that is responsible for ridding the body of any invaders, such as viruses, bacteria, protozoa, etc.

Protozoa—Single-celled organisms considered to be the simplest life form in the animal kingdom.

protozoa all have significant potential side effects for the patient. Suramin, eflornithine, pentamidine, and several drugs that contain arsenic (a chemical which in higher doses is highly poisonous to humans), are all effective anti-trypansomal agents. Each of these drugs, however, requires careful monitoring to ensure that the drugs themselves do not cause serious complications such as fatal hypersensitivity (allergic) reaction, kidney or liver damage, or inflammation of the brain.

Prevention

Prevention of sleeping sickness requires avoiding contact with the tsetse fly. Insect repellents and clothing that covers the limbs to the wrists and ankles are advisable. Public health measures have included drug treatment of humans who are infected with one of the *Trypanosoma brucei* complex. There are currently no immunizations available to prevent the acquisition of sleeping sickness.

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ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

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Sleepwalking see **Sleep disorders**

Slipped disk see **Herniated disk**

Slit lamp examination see **Eye examination**

Small-for-gestational-age infant see

Intrauterine growth retardation

Small bowel follow-through (SBFT) see

Upper GI exam

Small cell lung cancer see **Lung cancer, small cell**

Small intestine biopsy

Definition

A biopsy is a diagnostic procedure in which tissue or cells are removed from a part of the body and specially prepared for examination under a microscope. When the tissue involved is part of the small intestine, the procedure is called a small-intestine (or small-bowel) biopsy.

Purpose

The small-bowel biopsy is used to diagnose and confirm disease of the intestinal mucosa (the lining of the small intestine). The test is most commonly done to test for tumors of the small bowel or malabsorption syndromes.

Precautions

Due to the slight risk of bleeding during or after this procedure, **aspirin**, aspirin-containing medications, **nonsteroidal anti-inflammatory drugs**, and anticoagulants and **antiplatelet drugs** should be withheld for at least five days before the test.

Description

The small intestine is approximately 21 ft (6.4 m). It has three sections: the duodenum (a short, curved segment fixed to the back wall of the abdomen), the jejunum, and the ileum (two larger, coiled, and mobile segments). Some digestion occurs in the stomach, but the small intestine is mainly responsible for digestion and absorption of foods.

Malabsorption syndromes occur when certain conditions result in impaired absorption of nutrients, **vitamins**, or **minerals** from the diet by the lining of the small intestine. For example, injury to the intestinal lining can interfere with absorption, as can infections, intestinal parasites, some drugs, blockage of the lymphatic vessels, poor blood supply to the intestine, or diseases like sprue.

Malabsorption is suspected when a patient not only loses weight but has **diarrhea** and nutritional deficiencies despite eating well (weight loss alone can have other causes). Laboratory tests like fecal fat, a measurement of fat in stool samples collected over 72 hours, are the most reliable tests for diagnosing fat malabsorption but abnormalities of the small intestine itself are diagnosed by small-intestine biopsy.

Several different methods are used to detect abnormalities of the small intestine. A tissue specimen can be obtained by using an endoscope (a flexible viewing tube) or by using a thin tube with a small cutting

instrument at the end. This latter procedure is ordered when specimens larger than those provided by endoscopic biopsy are needed because it allows removal of tissue from areas beyond the reach of an endoscope.

Several similar types of capsules are used for tissue collection. In each, a mercury-weighted bag is attached to one end of the capsule, while a thin polyethylene tube about 5 ft (1.5 m) long is attached to the other end. Once the bag, capsule, and tube are in place in the small bowel, suction on the tube draws the tissue into the capsule and closes it, cutting off the piece of tissue within. This is an invasive procedure but it causes little pain and complications are rare.

A newer method of obtaining diagnostic information about the small intestine was approved by the Food and Drug Administration (FDA) in 2001. Known as the M2A Imaging System, the device was developed by a company in Atlanta, Georgia. The M2A system consists of an imaging capsule, a portable belt-pack image receiver and recorder, and a specially modified computer. The patient swallows the capsule, which is the size of a large pill. A miniature lens in the capsule transmits images through an antenna/transmitter to the belt-pack receiver, which the patient wears under ordinary clothing as he or she goes about daily activities. The belt-pack recording device is returned after seven or eight hours to the doctor, who then examines the images recorded as a digital video. The capsule itself is simply allowed to pass through the digestive tract.

Preparation requires only **fasting** the night before the M2A examination and taking nothing but clear liquids for two hours after swallowing the capsule. After four hours the patient can eat food without interfering with the test. As of the early 2000s, the M2A system is used to evaluate gastrointestinal bleeding from unknown causes, inflammatory bowel disease, some malabsorption syndromes, and to monitor surgical patients following small-bowel transplantation. The system has shown good results in detecting **Crohn's disease** undiagnosed by conventional methods.

Small-intestine biopsy procedure

After application of a topical anesthetic to the back of the patient's throat, the capsule and the tube are introduced, and the patient is asked to swallow as the tube is advanced. The patient is then placed on the right side and the instrument tip is advanced another 20 in (51 cm) or so. The tube's position is checked by fluoroscopy or by instilling air through the tube and listening with a stethoscope for air to enter the stomach.

The tube is advanced 2–4 in (5.1–10 cm) at a time to pass the capsule through the stomach outlet (pylorus).

KEY TERMS

Sprue—A disorder of impaired absorption of nutrients from the diet by the small intestine (malabsorption), resulting in malnutrition. Two forms of sprue exist: tropical sprue, which occurs mainly in tropical regions; and celiac sprue, which occurs more widely and is due to sensitivity to the wheat protein gluten.

Whipple's disease—A disorder of impaired absorption of nutrients by the small intestine. Symptoms include diarrhea, abdominal pain, progressive weight loss, joint pain, swollen lymph nodes, abnormal skin pigmentation, anemia, and fever. The precise cause is unknown but it is probably due to an unidentified bacterial infection.

Wireless capsule endoscopy—A newer method of examining the small bowel by means of a capsule swallowed by the patient. The capsule contains a miniaturized lens and an antenna that transmits information to a belt-pack recorder worn by the patient during the day.

When fluoroscopy confirms that the capsule has passed the pylorus, small samples of small intestine tissue are obtained by the instrument's cutting edge, after which the instrument and tube are withdrawn. The entire procedure may be completed in minutes.

Preparation

This procedure requires tissue specimens from the small intestine through means of a tube inserted into the stomach through the mouth. The patient is to withhold food and fluids for at least eight hours before the test.

Aftercare

The patient should not have anything to eat or drink until the topical anesthetic wears off (usually about one to two hours). If intravenous sedatives were administered during the procedure, the patient should not drive for the remainder of the day. Complications from this procedure are uncommon, but can occur. The patient is to note any abdominal pain or bleeding and report either immediately to the doctor.

Risks

Complications from this procedure are rare, but can include bleeding (hemorrhage), bacterial infection with **fever** and pain, and bowel puncture (perforation).

The patient should immediately report any abdominal pain or bleeding to the physician in charge. Biopsy is contraindicated in uncooperative patients, those taking aspirin or anticoagulants, and in those with uncontrolled bleeding disorders.

Normal results

Normal results are no abnormalities seen on gross examination of the specimen(s) or under the microscope after tissue preparation.

Abnormal results

Small-intestine tissue exhibiting abnormalities may indicate Whipple's disease, a malabsorption disease; lymphoma, a group of cancers; and parasitic infections like **giardiasis**, strongyloidiasis, and coccidiosis. When biopsy indicates celiac sprue (a malabsorption disorder), infectious **gastroenteritis** (inflammation of the gastrointestinal tract), folate and B₁₂ deficiency, or **malnutrition**, confirmation studies are needed for conclusive diagnosis.

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Smallpox pustules on the arm of an Indian man. (© C. James Webb/Phototake. — All rights reserved.)

the complete eradication of the virus with the exception of samples of stored virus in two laboratories.

Description

Smallpox is strictly an infection of human beings. Animals and insects can neither be infected by smallpox, nor carry the virus in any form. Most infections are caused by contact with a person who has already developed the characteristic **skin lesions** (pox) of the disease, although a person who has a less severe infection (not symptomatic or diagnosable in the usual way) can unwittingly spread the virus.

Causes and symptoms

Smallpox is a relatively contagious disease, which accounts for its ability to cause massive epidemics. The variola virus is acquired from direct contact with individuals infected with the disease, from contaminated air droplets, and even from objects used by another

Smallpox

Definition

Smallpox is an infection caused by the variola virus, a member of the poxvirus family. Throughout history, smallpox has been a greatly feared disease because it was responsible for huge epidemics worldwide that resulted in large numbers of deaths. In 1980, the World Health Organization (WHO) announced that an extensive program of **vaccination** against the disease had resulted in

smallpox victim (books, blankets, utensils, etc.). The respiratory tract is the usual entry point for the variola virus into a human being.

After the virus enters the body, there is a 12–14 day incubation period during which the virus multiplies, although no symptoms are recognizable. After the incubation period, symptoms appear abruptly and include **fever**, chills, and muscle aches. Two to three days later, a bumpy rash begins appearing first on the face and forearms. The rash progresses, ultimately reaching the chest, abdomen, and back. Seven to ten days after the rash appears, the patient is most infectious. The individual bumps (papules) fill with clear fluid and eventually become pus-filled over the course of 10–12 days. These pox eventually scab over, each leaving a permanently scarred pock or pit when the scab drops off.

Initially, the smallpox symptoms and rash appear similar to **chickenpox**. However, unlike chickenpox, smallpox lesions develop at the same rate so that they are all visible in the same stage. Another major difference is that smallpox occurs primarily on the face and extremities, whereas chickenpox tends to be concentrate on the face and trunk area.

Complications such as bacterial infection of the open skin lesions, **pneumonia**, or bone infections are the major causes of **death** from smallpox. A very severe and quickly fatal form called “sledgehammer smallpox,” occurs in 5–10% of patients and results in massive, uncontrollable bleeding (hemorrhage) from the skin lesions, as well as from the mouth, nose, and other areas of the body. This form is very infectious and usually fatal five to seven days after onset.

Fear of smallpox comes from both the epidemic nature of the disease as well as from the fact that no therapies have ever been discovered to either treat the symptoms of smallpox, or shorten the course of the disease.

Diagnosis

In modern times, a diagnosis of smallpox is made using an electron microscope to identify virus in fluid from the papules, urine, or in the patient’s blood prior to the appearance of the papular rash.

Treatment

No treatments have been developed to halt the progression of the disease. Treatment for smallpox is only supportive, meaning that it is aimed at keeping a patient as comfortable as possible. **Antibiotics** are sometimes administered to prevent secondary bacterial infections.

Prognosis

Approximately one in three patients die from smallpox, with the more severe, hemorrhagic form nearly 100% fatal. Patients who survive smallpox infection nearly always have multiple areas of scarring where each pock has been.

Prevention

From about the tenth century in China, India, and the Americas, it is noted that individuals who had even a mild case of smallpox could not be infected again. Fascinating accounts appear in writings from all over the world of ways in which people tried to prevent smallpox. Material from people mildly ill with smallpox (fluid or pus from the papules, scabs over the pox) was scratched into the skin of people who had never had the illness, in an attempt to produce a mild reaction and its accompanying protective effect. These efforts often resulted in full-fledged smallpox, and probably served only to help effectively spread the infection throughout a community. In fact, such crude smallpox “vaccinations” were against the law in Colonial America.

In 1798, Edward Jenner published a paper in which he discussed his important observation that milkmaids who contracted a mild infection of the hands (called cowpox, and caused by a relative of the variola virus) appeared to be immune to smallpox. Jenner created an immunization against smallpox using the pus found in the lesions of cowpox infection. Jenner’s paper led to much work in the area of vaccinations and ultimately resulted in the creation of a very effective vaccination against smallpox that utilized the vaccinia virus, another close relative of variola. Indeed, the term vaccination is derived from *vaccine*, Latin for cow and related to the cowpox link. Later, the term was applied to other vaccinations.

In 1967, WHO began its attempt to eradicate the smallpox virus worldwide. The methods used in the program were simple:

- Careful surveillance for all smallpox infections worldwide, to allow for quick diagnosis and immediate quarantine of patients.
- Immediate vaccination of all contacts diagnosed with infection, in order to interrupt the virus’ usual pattern of infection.

The WHO’s program was extremely successful, and the virus was declared eradicated worldwide in May 1980.

KEY TERMS

Epidemic—A situation in which a particular infection is experienced by a very large percentage of the people in a given community within a given time frame.

Eradicate—To completely do away with something, eliminate it, end its existence.

Hemorrhage—Bleeding that is massive, uncontrollable, and often life-threatening.

Lesion—The tissue disruption or the loss of function caused by a particular disease process.

Papules—Firm bumps on the skin.

Pox—A pus-filled bump on the skin.

Vaccine—A preparation using a non-infectious element or relative of a particular virus or bacteria, and administered with the intention of halting the progress of an infection, or completely preventing it.

Future concerns

Today, two laboratories (the Centers for Disease Control and Prevention in Atlanta, Georgia, and the Russian State Centre for Research on Virology and Biotechnology in Koltsovo, Novosibirsk Region) officially retain samples of the smallpox virus. These samples, as well as stockpiles of the smallpox vaccine, are stored because some level of concern exists that another poxvirus could undergo genetic changes (mutate) and cause human infection. There is also the remote chance that smallpox virus could somehow escape from the laboratories where it is stored. For these reasons, surveillance continues of various animal groups that continue to be infected with viruses related to the variola virus, and large quantities of vaccine are stored in different countries around the world, so that response to any future threat by the smallpox virus could be prompt.

Of greatest concern is the potential use of smallpox as a biological weapon. Since 1980, when the WHO announced smallpox had been eradicated, essentially no one has been vaccinated against the disease. Those individuals vaccinated prior to 1980 are believed to be susceptible as well because immunity only lasts 15–20 years. These circumstances coupled with the nature of smallpox to spread quickly from person to person could lead to devastating consequences.

The United States and Russia are the only two countries to officially house remaining samples of the

virus. However, it is believed that other countries, such as Iraq, may have obtained samples of the smallpox virus during the Cold War through their association with Russia. It is also possible that scientists with access to the virus may have sold their services and knowledge to other governments.

On June 22 and 23, 2001, four U.S. organizations (CSIS—Center for Strategic and International Studies, Johns Hopkins Center for Civilian Biodefense Studies, ANSER—Analytic Services Inc., and MIPT—Memorial Institute for the Prevention of Terrorism) presented a fictitious scenario of the United States' response to a deliberate introduction of smallpox titled *Dark Winter*. This exercise demonstrated that if such an event were to occur, the United States would be ill prepared on several fronts. The primary concern is an inadequate supply of vaccine, which is essential to preventing disease development in exposed persons. Between 1997 and 2001, two companies were contracted to produce additional smallpox vaccines for both military and civilian use. Through these contracts, an additional 40 million doses would be made available for civilian use by 2005. In the meantime, studies are underway to determine if the existing vaccines can be diluted in order to increase the number of doses available for immediate use. Results from a very small group of volunteers tested in 2000 found that at one-tenth strength, the existing smallpox vaccines are approximately 70% effective. In late 2001, a new study began evaluating the effectiveness of the vaccine at one-fifth strength.

In the event that smallpox is reintroduced into the current population, it will be imperative that doctors immediately recognize the symptoms and isolate the individual to prevent further spread of the disease. Prompt vaccination of any persons who had contact with the patient is also necessary to prevent additional cases of smallpox from developing. Controlling and containing spread of this disease is critical for prevention of a world-wide epidemic that would have a devastating impact on current populations.

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Rosalyn Carson-DeWitt, MD

Smelling disorders

Definition

Smelling disorders are disturbances of the olfactory sense, which is known as the sense of smell. These nasal dysfunctions range from the total loss of smell (**anosmia**) to dysosmia, a distorted sense of smell.

Description

An awareness of how the olfactory system works is helpful for understanding how smelling disorders affect the sense of smell. People detect odors because sensory receptors located in the nose carry smell sensations to the brain. The receptors, which are nerve cell endings, are found in the mucous membrane in the roof of the nose. This section of the nose called the olfactory area is located just below the brain's frontal lobes.

In the olfactory area are millions of tiny olfactory cells. Each cell contains about 12 cilia, tiny hairs that extend into a mucus layer. The mucus moistens the cilia. Mucus also catches odor molecules, while receptors in the cilia stimulate the molecules and send nerve impulses to the brain.

Olfactory nerve fibers carry the impulse to two olfactory bulbs located in the brain. Information is processed in the bulbs and then sent to the cerebral cortex. Once the transmission is inside the smell center of the brain, a person experiences the sense of smell.

A person with a normal sense of smell (normosmia) is able to distinguish 10,000 odors. The sense of smell stimulates salivary glands. As a result, smelling disorders often affect the sense of taste. The olfactory sense allows people to experience pleasurable odors like the scent of roses; smell is also thought to contribute to sexual attraction.

A smelling disorder that affects the sense of smell is generally not life-threatening. However, it can be dangerous. Without a sense of smell, a person might eat spoiled food. Lack of a sense of smell could pose a health risk if a person has little appetite and fails to eat enough. Furthermore, without a sense of smell, a

person might not detect a gas leak or the smell of something burning. Loss of smell and the resulting loss of taste may lead to depression.

Types of smelling disorders

Smelling disorders differ in the way that the sense of smell is affected and how long a person has the disorder. For example, anosmia, the loss of the sense of smell, is often a temporary symptom of a cold or flu. However, a **head injury** could cause permanent anosmia. In addition, a head injury could produce dysosmia, the distorted sense of smell that could cause a person to hallucinate a foul odor.

Smelling disorders are categorized as:

- Anosmia, the loss of the sense of smell. It is the most common smelling disorder. This condition can be temporary or permanent.
- Dysosmia is a distorted sense of smell. A person senses non-existent unpleasant odors. It can be caused by medical and mental conditions.
- Hyperosmia is an increased sensitivity to smell. It can be a characteristic of someone with a neurotic or histrionic personality.
- Hyposmia is the diminished sense of smell. This is usually a temporary condition that a person may experience after a case of acute influenza. Sometimes this condition is referred to as partial anosmia.
- Presbyosmia refers to the lessening or loss of the olfactory sense that occurs when a person ages.

Smelling disorder demographics

Anosmia occurs in about 10% of head trauma injuries and head trauma is a leading cause of anosmia in young adults. In older adults, the disorder is generally caused by viral infection. **Aging** may also bring a loss of the sense of smell. In rare cases, anosmia is inherited. It is a symptom of male **hypogonadism** (Kallmann's syndrome).

Olfactory **hallucinations** known as dysosmia are generally associated with psychological conditions. In some cases, people may believe they are the source of foul odors.

Causes and symptoms

Anosmia is the most common type of smelling disorder. Loss of the olfactory sense is generally caused by nasal congestion or obstruction. Temporary partial anosmia often occurs when a person has a cold, the flu, or some types of **rhinitis**, especially hay **fever (allergic rhinitis)**. During these conditions, nasal mucus membranes become inflamed. Other causes for anosmia are:

- Nasal polyps and other disorders that prevent air from getting to the area in the nose where the smell receptors are found. Hay fever or an allergy may cause one or more polyps to show up.
- Viral upper respiratory infection.
- Atrophic rhinitis. This condition causes mucus membrane to waste away. The person may experience some level of permanent anosmia. One symptom of this condition is that a person expels a foul-smelling discharge.
- Hypertrophic rhinitis. Mucous membrane thickens, covering the olfactory nerve endings. If not treated, hypertrophic rhinitis can lead to permanent anosmia. This discharge could overpower other odors.
- Cigarettes. Smoking aggravates the nose's membrane and intensifies nasal polyp symptoms.
- A crooked nose or a deviated septum.
- When the olfactory bulbs, tracts, or central connections are destroyed. This can occur in situations such as head trauma, infections or nasal or sinus surgery.
- Head injury. If both olfactory nerves are torn during a head injury, permanent anosmia results.
- Medications such as antihistamines and decongestants, especially prolonged use of decongestants.
- Drugs like amphetamines, estrogen, naphazoline, phenothiazines, and reserpine.
- The aging process may cause the sense to lessen. In most cases, there is no other obvious cause for the disorder.
- A tumor behind the nose or in the membranes surrounding the brain.
- Lead poisoning.
- Exposure to insecticides or other chemicals.
- Radiation therapy.
- Nervous disorders.
- Idiopathic loss, which means there is no diagnosable cause for the condition.

Anosmia symptoms

Most people with anosmia can distinguish salty, sweet, bitter, and sour tastes since the tongue senses these tastes. However, people with anosmia cannot sense other tastes. Since taste is largely based on the olfactory sense, people complain of losing the sense of taste (ageusia).

Dysosmia

Infected nasal sinuses and damage to the olfactory bulbs can cause dysosmia, the distorted sense of smell. Head trauma can cause this disorder. Poor **oral hygiene** can lead to dysosmia. In these cases, a person

may also find that disagreeable odors are accompanied by the sensing of unpleasant tastes. In addition, brain-stem disease can cause smelling disorders. An epileptic seizure can include olfactory hallucinations.

Mental conditions such as depression and **schizophrenia** may be accompanied by dysosmia. In addition, when people who are severely dependent on alcohol quit drinking, they may experience dysosmia.

Diagnosis

If a smelling disorder is a symptom of a mental condition such as schizophrenia, diagnosis should be part of treatment for that condition.

When the condition is caused by a medical condition such as **allergies** or a viral infection, a person may notice that the olfactory sense is impaired during that condition. If the smelling disorder continues after the person is well, an appointment should be made with a primarily health care provider.

Diagnosis of smelling disorders begins with a health assessment to determine the cause of the olfactory impairment. The patient's primary care doctor will ask if the patient has a cold, allergies, **sinusitis**, or an upper respiratory infection.

Treatment of a head injury or follow-up medical appointment should address smelling disorders. In all cases, discussion of the symptoms covers issues such as when the smelling disorder started, if this has been an ongoing problem, and whether the disorder is becoming more intense. The assessment will include questions about whether the patient can taste food and if the disorder affects all odors or specific smells. The patient will also be asked about medications taken.

Physical examinations

The **physical examination** will include a thorough inspection of the nose, nasopharynx, and the examination of the upper respiratory tract. The examination could include sinus transillumination, placement of a light on the face to help determine if sinuses are full. **Skull x rays** may be required to determine the presence of tumors in the nose or brain.

The patient may be referred to a neurologist; an ear, nose, and throat specialist; or to a center that specializes in treatment of smelling disorders.

Other diagnostic tests could be required. These include:

- A CT scan (computed tomography scans) of the head. Also known as a CAT scan, this process provides a more detailed image than the x ray.

- Olfactory nerve testing.
- Nasal cytology, which involves the study of mucus under a microscope.
- Testing to determine the scope of smelling disorder. A basic smell test involves the patient trying to identify each one of a group of different odors. A variation of this is a scratch-and-sniff test. The patient may be asked to differentiate among concentrations of one odor. The alcohol sniff test that involves use of a material soaked in isopropyl alcohol. Patients close their eyes and the doctor moves around. Patients tell the doctor when they smell the alcohol.
- The patient may also take a taste test.

Medical costs

The costs for diagnosis and treatment vary because of the different types of smelling disorders, the range of causes for olfactory dysfunction, and the different types of treatment.

There are also differences in what health plans require in terms of patient co-pay. A health plan could cover treatments ranging from the initial appointment with a primary care provider to the surgery to remove brain tumors.

In addition, some health plans cover costs of treatment at specialized facilities like the Center for Smell and Taste Disorders at the University of Colorado Health Sciences Center in Denver. A series of tests including a taste-and-smell test cost \$250 in May of 2001.

Treatment

Treating a condition that causes a smelling disorder can sometimes restore the olfactory sense. Treatments for smelling disorders are as varied as the olfactory dysfunctions. Treatment for smelling disorders ranges from lifestyle changes to surgery. Treatment of mental conditions could affect the smelling disorder. In some cases, the disorder can't be treated, and the person must adjust to the loss of the sense of smell. Anosmia associated with aging is not treatable.

Basic treatments for anosmia

The sense of smell should return after a condition like a cold or the flu ends. **Decongestants** such as Sudafed help reduce congestion related to colds, allergies, and sinus conditions. Manufacturer's dosage recommendations should be followed. If anosmia is related to excessive use of nasal decongestants, a person should discontinue use of those medications.

Saline sprays can be used to clean the interior of the nose.

If **smoking** causes anosmia, a person should quit smoking.

The sense of smell may return after treatment of allergic or bacterial rhinitis and sinusitis. An over-the-counter antihistamine such as Actifed may provide relief.

If allergies cause anosmia, adjustments should be made to avoid allergens. If dust causes allergies, care should be taken to clean areas such as the bedroom.

Antibiotics may be prescribed for infections.

Other medications prescribed for smelling disorders include **steroids** such as Prednisone. It should only be used for a short time since longterm use could lessen resistance to infection.

Surgical treatment

Removal of **nasal polyps** and benign tumors may cause the sense of smell to return. Polyp removal is an uncomplicated surgery. Generally, only a local anesthetic is needed.

Septoplasty straightens the nasal passage. It is generally an outpatient surgery, with local or **general anesthesia** required. **Rhinoplasty** straightens the structure of the nose. This surgery could be combined with septoplasty.

Endoscopic sinus surgery opens sinus drainage channels. This outpatient surgery is an option after a person sees no improvement after trying treatments such as medications.

Surgical treatment may not be effective in conditions that result in the destruction of the olfactory nerve or its central passages. However, regeneration of those tissues may cause the sense to return.

Enhancing taste

Without a sense of smell, most people can still taste salt and flavors that are sweet, sour, and bitter. People with anosmia could distinguish other tastes by adding spices such as pepper to food. These spices stimulate facial nerves that also sense flavors.

Alternative treatment

Alternative treatments for smelling disorder center around the theory that zinc supplements help improve the sense of smell. The supplement is said to be effective when the olfactory sense is impaired by conditions such as a head injury or an upper respiratory infection. A person should take 50 mg of zinc picolinate each day after eating. This procedure might be effective in the case of head injury. However, it may be several months

KEY TERMS

Histrionic—A behavior characterized by an excitable nature and the constant desire for stimulation.

Nasopharynx—The passage that connects the nasal cavity to the top of the throat.

Neurotic—Behavior characterized by neurosis, mental functional orders with symptoms such as anxiety, depression, compulsions, and phobias.

Polyp—A benign growth in areas such as the nasal passage.

Rhinitis—The inflammation of the mucous membrane in the nose.

Septum—A sheet of cartilage and bone that separates the nostrils.

Sinusitis—Inflammation of the paranasal sinuses because of allergic reactions or viral, bacterial, or functional infections.

before results are seen. **Acupuncture** may also produce results.

If polyps cause a smelling disorder, a change in diet could be helpful. A person should avoid dairy products, take supplements such as garlic, and follow other recommendations from a health care practitioner. A daily dosage of 5,000–10,000 mg of vitamin C could cut back on the amount of polyps. **Vitamins** should not be taken all at once. A multi-vitamin and mineral complex could also help.

Prognosis

In cases where smelling disorders are treatable, the outcome is positive because the olfactory sense is restored. In those cases where the sense of smell is lost, the person must make adjustments to adapt to life without that sense. Those adjustments include using spices like pepper to stimulate taste buds.

Since a person with anosmia can no longer smell food to determine whether it is safe to eat, care should be taken. The person who lives with other people can ask them if food smells fresh. People who live alone should discard food if there is a chance that it has spoiled. Other home safety measures include installing smoke alarms and gas detectors. Cooking on an electric stove is preferable to a gas stove.

Furthermore, people with smelling disorders can find support groups. These are often associated with smell and taste clinics. In addition, there are

on-line bulletin board where people can share experiences. One site contains descriptions of how things smell. Those words provide a connection to a missing sense in the same way that sign language allows the hearing-impaired to understand the spoken word.

Prevention

Not all causes of smelling disorders can be prevented. However, people with a disorder should not smoke and should ask those around them not to smoke. Those with smelling disorders related to allergies should be taken to avoid allergens. Since head trauma injuries can lead to smelling disorders, people should wear protective helmets when bicycling or participating in sports like football.

Resources

BOOKS

Devere, Ronald, and Marjorie Calvert. *Navigating Smell and Taste Disorders*. New York: Demos Health, 2011.

ORGANIZATIONS

American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Road, Alexandria, VA, 22314-2857, (703) 836-4444, <http://www.entnet.org>.

The Smell and Taste Center, University of Pennsylvania, 5 Ravidin Pavilion 3400 Spruce Street, Philadelphia, PA, 19104-4283, (215) 662-6580, (215) 349-5266, Geraldine.Fischer@uphs.upenn.edu, <http://www.med.upenn.edu>.

Liz Swain

Smoke inhalation

Definition

Smoke inhalation is breathing in the harmful gases, vapors, and particulate matter contained in smoke.

Demographics

Smoke inhalation is estimated to be the cause of 60–80% of all fire-related deaths in the United States each year. People who are trapped in fires may suffer from smoke inhalation independent of receiving skin **burns**; however, the incidence of smoke inhalation increases with the percentage of total body surface area burned. Children under age 11 and adults over age 70 are most vulnerable to the effects of smoke inhalation. Children younger than 11 years and adults older than 70 are estimated to make up about 40% of all people who die

from fire-related causes each year. Two to three times more people die of smoke inhalation than die of burns each year.

Description

Smoke inhalation typically occurs in victims or firefighters caught in structural fires. However, cigarette **smoking** also causes similar damage to the lungs on a much smaller scale over a longer period of time.

Causes and symptoms

Dangerous gasses and smoke are produced by both combustion (burning of material) and pyrolysis (the breakdown of material by heat in the absence of adequate amounts of oxygen to support combustion).

The harmful materials given off by combustion and pyrolysis injure the airways and lungs in three ways: heat damage, tissue irritation, and oxygen **starvation** of tissues (asphyxiation). Signs of heat damage are singed nasal hairs, burns around and inside the nose and mouth, and internal swelling of the throat.

Tissue irritation of the throat and lungs may appear as noisy breathing, coughing, hoarseness, black or gray sputtle, and fluid in the lungs. Certain materials such as PVP pipe, wool, silk, nylon, and polyurethane give off poisonous gasses when they burn. When inhaled, these gasses can cause tissue irritation and chemical burns in the throat and lungs.

Asphyxiation is apparent from **shortness of breath** and blue-gray or cherry-red skin color. Carbon monoxide (CO), a colorless, odorless gas, is produced in large quantities in incomplete combustion or pyrolysis. The CO binds with hemoglobin molecules that normally carry oxygen to the body. Because hemoglobin bonded to CO cannot carry oxygen, oxygen starvation or asphyxiation occurs.

Diagnosis

In addition to looking for the signs of heat damage, tissue irritation, and asphyxiation, the physician will assess the patient's breathing by the respiratory rate (number of breaths per minute) and motion of the chest as the lungs inflate and deflate. The patient's circulation is also evaluated by the pulse rate (number of heartbeats per minute) and blood pressure. Blood tests will indicate the levels of oxygen and byproducts of poisonous gases. Chest x rays are too insensitive to show damage to delicate respiratory tissues, but can show fluid in the lungs (**pulmonary edema**).

The physician may perform a **bronchoscopy**, a visual examination in which the airways and lungs are seen through a fiber optic tube inserted down the patient's windpipe (trachea). Other **pulmonary function tests** may be performed to measure how efficiently the lungs are working.

Treatment

Treatment varies with the severity of the damage caused by smoke inhalation. The primary focus of treatment is to maintain an open airway and provide an adequate level of oxygen. If the airway is open and stable, the patient may be given high-flow humidified 100% oxygen by mask. If swelling of the airway tissues is closing off the airway, the patient may require the insertion of an endotracheal tube to artificially maintain an open airway.

Oxygen is often the only medication necessary. However, patients who have a **cough** with **wheezing** (bronchospasm), indicating that the bronchial airways are narrowed or blocked, may be given a bronchodilator to relax the muscles and increase ventilation. There are also antidotes for specific poisonous gases in the blood; dosage is dependent upon the level indicated by blood tests. **Antibiotics** are not given unless sputum and blood cultures confirm the presence of a bacterial infection.

In institutions where it is available, hyperbaric **oxygen therapy** may be used to treat smoke inhalation resulting in severe carbon monoxide or cyanide **poisoning**. This treatment requires a special chamber in which the patient receives pure oxygen at three times the normal atmospheric pressure, thus receiving more oxygen faster to overcome loss of consciousness, altered mental state, cardiovascular dysfunction, pulmonary **edema**, and severe neurological damage.

Alternative treatment

Following immediate treatment by conventional medicine, herbal medicines may help to maintain open airways and heal damaged mucous membranes. They can also help support the entire respiratory system. **Acupuncture** and homeopathic treatment can provide support to the whole person who has suffered a traumatic injury such as smoke inhalation.

Prognosis

Although the outcome depends of the severity of the smoke inhalation and the severity of any accompanying burns or other injuries, with prompt medical treatment, the prognosis for recovery is good. However, some patients may experience chronic pulmonary

KEY TERMS

Asphyxiation—Oxygen starvation of tissues. Chemicals such as carbon monoxide prevent the blood from carrying sufficient oxygen to the brain and other organs. As a result, the person may lose consciousness, stop breathing, and die without artificial respiration (assisted breathing) and other means of elevating the blood oxygen level.

Hyperbaric oxygen therapy—Pure oxygen is administered to the patient in a special chamber at three times the normal atmospheric pressure. The patient gets more oxygen faster to overcome severe asphyxiation.

Pulmonary—Pertaining to the lungs.

Pulmonary edema—The filling of the lungs with fluid as the body's response to injury or infection.

problems following smoke inhalation, and those with **asthma** or other chronic respiratory conditions prior to smoke inhalation may find their original conditions have been aggravated by the inhalation injury.

Prevention

Smoke inhalation is best avoided by preventing structural fires. This includes inspection of wiring, safe use and storage of flammable liquids, and maintenance of clean, well-ventilated chimneys, wood stoves, and space heaters. Properly placed and working smoke detectors in combination with rapid evacuation plans will minimize a person's exposure to smoke in the event of a fire. When escaping a burning building, a person should move close to the floor where there is more cool, clear air to breathe because hot air rises, carrying gases and particulate matter upward. Finally, firefighters should wear proper protective gear.

Resources

OTHER

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ORGANIZATIONS

American Association for Respiratory Care, 9425 N. MacArthur Boulevard, Suite 100, Irving, TX, 75063-4706, (972) 243-2272, www.aarc.org.

American Lung Association, 1301 Pennsylvania Ave., NW Suite 800, Washington, DC, 20004, (212) 315-8700, (800) LUNG-USA [(800) 548-8252], <http://www.lungusa.org>.

National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573; TTY: (240) 629-3255, (240) 629-3246, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov>.

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Smoking-cessation drugs

Definition

Smoking cessation drugs are medicines that help people stop smoking cigarettes or using other forms of tobacco.

Purpose

The known major health risks associated with smoking have led to efforts to dramatically reduce the number of people who do smoke and to encourage individuals, particularly young people, to not begin smoking. These efforts include restricting access to tobacco products to minors, substantially raising the costs of tobacco products, and using taxes which smokers pay on tobacco products to fund community-based tobacco reduction and cessation education and intervention programs. In addition, more and more communities and states have adopted smoke-free laws and regulations.

Although most smokers state they would like to quit smoking, people who smoke cigarettes or use other forms of tobacco often have a difficult time when they try to stop smoking. The difficulty is partly psychological; individuals get in the habit of using tobacco at certain times of day or while they are doing certain things, such as having a cup of coffee or reading the newspaper. But the habit is also hard to break for physical reasons. Tobacco contains nicotine, a drug that is as addictive as **cocaine** or heroin. Of those who have ever tried even a single cigarette, about a third become nicotine-dependent. A person who is addicted to nicotine has withdrawal symptoms, such as irritability, **anxiety**, difficulty concentrating, and craving for tobacco when he or she stops using the product.

Some people can stop smoking through willpower alone but most do better if they have support from friends, family, a physician or pharmacist, or a formal stop-smoking program. Heavy tobacco users may find that smoking cessation products also help by easing their withdrawal symptoms. Most smoking cessation products contain nicotine but the nicotine is delivered

Symptom	Cause	Duration	Relief
Craving for cigarette	Nicotine craving	Begins in first week and can linger for months	Distract yourself with other activities (e.g., exercise, hobbies, etc.)
Coughing, dry throat, nasal drip	Body ridding itself of mucus in lungs and airways	Several weeks	Drink plenty of fluids, use cough drops
Irritability, impatience	Nicotine craving	2–4 weeks	Exercise, practice relaxation techniques, avoid caffeine
Lack of concentration	Lack of nicotine stimulation	A few weeks	Reduce workload, avoid stress
Fatigue	Lack of nicotine stimulation	2–4 weeks	Practice relaxation techniques, nap
Insomnia	Nicotine craving temporarily reduces deep sleep	2–4 weeks	Avoid caffeine after 6 p.m.
Hunger	Cigarette cravings confused with hunger pangs	Up to several weeks	Drink water or low-calorie drinks, eat low-calorie snacks
Constipation, gas	Intestinal movement decreases with lack of nicotine	1–2 weeks	Drink plenty of fluids, add fiber to diet, exercise

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

in small, steady doses spread out over many hours. In contrast, when a person inhales a cigarette, nicotine enters the lungs and then travels to the brain within seconds, delivering the “rush” that smokers crave. Another difference is that smoking cessation products do not contain the tar, carbon monoxide, and other toxins that make cigarettes so harmful to people’s health.

Description

U.S. brand names

The brand names of nicotine replacement products which are sold either as over-the-counter (OTC) or non-prescription products or by prescription only in the United States are as follows:

- Nicotine patches: Habitrol (prescription), Nicoderm CQ (OTC), Nicotrol (OTC), ProStep (prescription)
- Nicotine Polacrilex Gums: Nicorette (OTC) and Nicorette DS (OTC)
- Nicotine Lozenges: Commit (OTC)
- Nicotine Inhaler: Nicotrol Inhaler (prescription)
- Nicotine Nasal Spray: Nicotrol NS (prescription)

Nicotine replacement products

Smoking cessation drugs that contain nicotine are also called nicotine substitution products or nicotine replacement therapy (NRT). Five forms are approved by the Food and Drug Administration (FDA) as of 2010—gum, skin patch, nasal spray, inhaler, and lozenges. Results of numerous research studies

conducted in the United States and other countries have validated that nicotine replacement therapy (NRT) is an effective and safe approach when used as a first-line pharmacological treatment for tobacco reduction and/or cessation.

The nasal spray and inhaler are available only with a prescription, but gum, lozenges and some brands of the patch can be bought over-the-counter (without a prescription). People who buy the nonprescription products should check with a physician before starting to use them.

Other medications

Another type of smoking cessation drug, buproprion (Zyban), also reduces craving and withdrawal symptoms, although it is not a nicotine replacement product. Bupropion is an antidepressant medication that is thought to help people stop smoking by mimicking some of the effects of tobacco on brain tissue. Bupropion can be used together with nicotine replacement products. Several studies indicate that the combination helps more smokers quit than either method by itself. Bupropion is the only antidepressant currently approved by the FDA for use as a smoking cessation product.

Another non-nicotine product is the drug varenicline (Chantix). Varenicline is available only by prescription and is classified as a nicotinic receptor partial agonist. This drug works by targeting nicotine receptors in the brain. The drug attaches itself to the nicotine receptors and blocks nicotine from reaching the

KEY TERMS

Acupuncture—A Chinese medical practice that treats illness or addictions by the insertion of very thin steel needles at specified points along the body's energy channels.

Bupropion—An antidepressant medication given to smokers for nicotine withdrawal symptoms. It is sold under the trade name Zyban.

Buspirone—An antianxiety medication that is also given for withdrawal symptoms. It is sold under the trade name BuSpar.

Nicotine—A colorless, oily chemical found in tobacco that makes people physically dependent on smoking. It is poisonous in large doses.

Withdrawal symptoms—A group of physical or mental symptoms that may occur when a person suddenly stops using a drug on which he or she has become dependent.

receptors. This action blocks the pleasurable sensation smokers derive from smoking.

Alternative approaches

Other approaches that have been used to help smokers quit include hypnosis and **acupuncture**. The evidence for the usefulness of hypnosis is largely anecdotal. It appears to be most helpful when used in combination with nicotine replacement products or bupropion. Although acupuncture has been used in Western countries since the 1970s to help people quit smoking, it does not appear to be particularly effective in this regard.

Recommended dosage

The recommended dosage of nicotine replacement products depends on the method of administration. Each form of this medicine comes with detailed instructions for its use. Following directions exactly is very important. For example, nicotine gum should not be chewed like regular chewing gum. It must be chewed very slowly until it has a slight taste or causes a slight **tingling** sensation in the mouth; then "parked" between the cheek and gum until the taste and tingling goes away; then chewed and parked in the same way for about 30 minutes. Nicotine patches and other products also must be used correctly to be effective. Some patches are meant to be worn only during the day and removed at night; others are worn 24 hours a day.

Smokers who are heavily dependent on nicotine may want to ask their doctors about using a combination of nicotine replacement products. Some study results indicate that combining the transdermal patch with either the gum or the nasal spray helps more smokers quit than any of the three products by itself. Authorities believe that the higher success rate is due to the different rates of speed at which these products deliver nicotine to the body. The nasal spray delivers nicotine very rapidly, and can be used to relieve intense cravings at times of the day when the smoker is accustomed to having a cigarette, while the patch delivers a smaller dosage of nicotine to the body at a steadier rate.

Precautions

Seeing a physician regularly while using smoking cessation drugs is important. The physician will check to make sure the medicine is working as it should and will watch for unwanted side effects.

Some side effects of smoking cessation drugs include:

- nausea
- vomiting
- severe pain in the stomach or abdomen
- severe diarrhea
- severe dizziness
- fainting
- convulsions (seizures)
- low blood pressure
- fast, weak, or irregular heartbeat
- hearing or vision problems
- severe breathing problems
- severe watering of the mouth or drooling
- cold sweat
- severe headache
- confusion
- severe weakness

Keep these drugs, including thrown-away patches and gum, out of the reach of children and pets. Even a small amount of nicotine can seriously harm a child or animal.

Nicotine in any form should not be used during **pregnancy**, as it may harm the fetus or cause **m miscarriage**. Women who may become pregnant should use effective birth control while taking smoking cessation drugs. Women who become pregnant while taking this medicine should stop taking it immediately and check with their physicians.

Nicotine passes into breast milk and may cause problems for nursing babies. Women who are **breast-feeding** and want to use smoking cessation drugs may need to stop breastfeeding during treatment.

Anyone who has had unusual reactions to nicotine in the past should let his or her physician know before using a smoking cessation drug. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances. People who have had a rash or irritation from adhesive **bandages** should check with a physician before using a nicotine patch.

Smoking cessation patches, gum, and other products may make certain medical problems worse. Before using a smoking cessation drug, people with any of these medical problems should make sure their physicians are aware of their conditions:

- heart or blood vessel disease
- high blood pressure
- diabetes
- overactive thyroid
- skin rash or irritation
- stomach ulcer
- pheochromocytoma (pcc) (a tumor of the adrenal medulla)
- dental problems or mouth sores
- sore throat
- jaw pain or temporomandibular joint disorder (TMJ)

There are also precautions to take with bupropion and buspirone. Bupropion should not be taken by patients with a history of seizures, high blood pressure, anorexia, or **bulimia nervosa**. People taking buspirone should be careful about driving or operating heavy machinery until they can tell whether the drug makes them drowsy as a side effect. Although buspirone does not interact with alcohol as intensely as most tranquilizers do, patients should still use alcohol cautiously if they are taking buspirone.

In 2009, the FDA directed the makers of the drugs varenicline (Chantix) and bupropion (Zyban) to include new warnings on the drug labels of their products related to the potential for the development of serious mental health side effects in individuals taking these drugs. Individuals with known psychiatric or mental health problems are at particularly high risk for the development of these mental health side effects, which include depression, agitation, and increased risk of **suicide**. Individuals without pre-existing mental health problems may also experience these serious side-effects

Side effects

Each type of smoking cessation product may cause minor side effects that usually go away as the body adjusts to the drug. These side effects usually do not need medical attention unless they continue or they interfere with normal activities. For example, nicotine gum may cause belching, jaw aches, or sore mouth or throat. Nicotine patches may cause redness, **itching**, or burning where the patch is applied. The nasal spray may irritate the nose and sinuses, while the inhaler may cause throat irritation or coughing.

If nicotine gum injures the mouth, teeth, or dental work, check with a dentist or physician as soon as possible. Other side effects are possible. Anyone who has unusual symptoms while using smoking cessation drugs should get in touch with his or her physician.

The side effects of bupropion include **dry mouth** and difficulty sleeping. The possible side effects of buspirone include headaches and drowsiness.

Bupropion and varenicline are associated with serious mental health side-effects including depression, agitation, thoughts of suicide, hostility, and attempted suicide. These side effects tend to occur shortly after the patient begins to take the medication and typically stop once the patient stops taking the drug. Patients experiencing these symptoms should be closely monitored by a physician until the side-effects stop.

Interactions

People taking certain drugs may need to change their doses when they stop smoking. Anyone who uses a smoking cessation drug should let the physician know all other medicines he or she is taking and should ask whether the doses need to be changed. Examples of drugs that may be affected when a person stops smoking are:

- insulin
- airway opening drugs (bronchodilators) such as aminophylline (Somophyllin), oxtriphylline (Choledyl) and theophylline (Somophyllin-T)
- opioid (narcotic) pain relievers such as propoxyphene (Darvon)
- the beta blocker propranolol (Inderal)

Other drugs may also interact with smoking cessation drugs. Be sure to check with a physician or pharmacist before combining smoking cessation drugs with any other prescription or nonprescription (over-the-counter) medicine.

Bupropion should not be used by patients who are also taking monoamine oxidase inhibitor (MAOI) medications. These include such drugs as furazolidone,

isocarboxazid, and phenelzine. Bupropion may also interact with phenytoin, carbamazepine, and levodopa. Buspirone also interacts with MAOIs, as well as with trazadone and haloperidol.

Resources

PERIODICALS

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ORGANIZATIONS

- American Association for Respiratory Care (AARC), 9425 N. MacArthur Blvd., Suite 100, Irving, TX, 75063-4706, (972) 243-2272, <http://www.aarc.org>.
- American Cancer Society (ACS), (800) ACS-2345, (404) 329-7530, <http://www.cancer.org>.
- American Lung Association (ALA), 1301 Pennsylvania Ave. NW, Suite 800, Washington, DC, 20004, (202) 785-3355, (201) 452-1805, <http://www.lungusa.org>.
- Office on Smoking and Health. Centers for Disease Control and Prevention (CDC-OSH), 4770 Buford Hwy, NE. MS K-50, Atlanta, GA, 30341-3717, (800) CDC-INFO (232-4636), <http://www.cdc.gov/tobacco>.

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Smoking

Definition

Smoking is the inhalation of the smoke of burning tobacco encased in cigarettes, pipes, and cigars. Casual smoking is the act of smoking only occasionally, usually in a social situation or to relieve **stress**. A smoking habit is a physical **addiction** to tobacco products. Many health experts now regard habitual smoking as a psychological addiction, too, and one with serious health consequences.

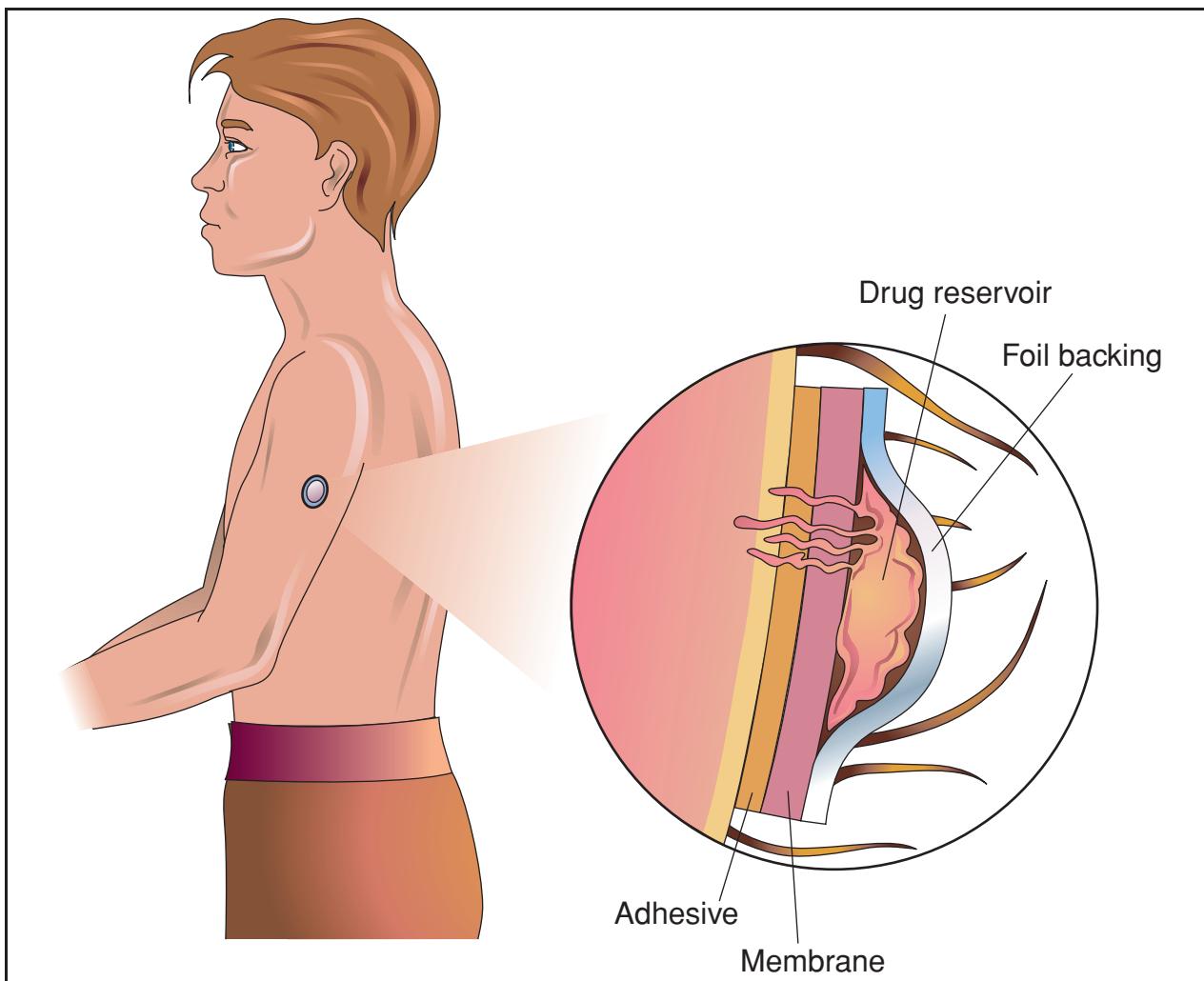
Description

The U.S. Food and Drug Administration has asserted that cigarettes and smokeless tobacco should be considered nicotine delivery devices. Nicotine, the active ingredient in tobacco, is inhaled into the lungs, where most of it stays. The rest passes into the bloodstream, reaching the brain in about 10 seconds and dispersing throughout the body in about 20 seconds.

Depending on the circumstances and the amount consumed, nicotine can act as either a stimulant or tranquilizer. This can explain why some people report that smoking gives them energy and stimulates their mental activity, while others note that smoking relieves **anxiety** and relaxes them. The initial "kick" results in



The wheals on the arm of this patient was caused by an allergic reaction to nicotine patches used to help subdue the urge to smoke. (Custom Medical Stock Photo, Inc. Reproduced by permission.)



The nicotine patch is a type of transepidermal patch designed to deliver nicotine, the addictive substance contained in cigarettes, directly through the skin and into the blood stream. The patch contains a drug reservoir sandwiched between a nonpermeable back layer and a permeable adhesive layer that attaches to the skin. The drug leeches slowly out of the reservoir, releasing small amounts of the drug at a constant rate for up to 24 hours. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

part from the drug's stimulation of the adrenal glands and resulting release of epinephrine into the blood. Epinephrine causes several physiological changes—it temporarily narrows the arteries, raises the blood pressure, raises the levels of fat in the blood, and increases the heart rate and flow of blood from the heart. Some researchers think epinephrine contributes to smokers' increased risk of high blood pressure.

Nicotine, by itself, increases the risk of heart disease. However, when a person smokes, he or she is ingesting a lot more than nicotine. Smoke from a cigarette, pipe, or cigar is made up of many additional toxic chemicals, including tar and carbon monoxide. Tar is a sticky substance that forms into deposits in the lungs, causing lung **cancer** and respiratory distress.

Carbon monoxide limits the amount of oxygen that the red blood cells can convey throughout your body. Also, it may damage the inner walls of the arteries, which allows fat to build up in them.

Besides tar, nicotine, and carbon monoxide, tobacco smoke contains 4,000 different chemicals. More than 200 of these chemicals are known to be toxic. Nonsmokers who are exposed to tobacco smoke also take in these toxic chemicals. They inhale the smoke exhaled by the smoker as well as the more toxic *sidestream smoke*—the smoke from the end of the burning cigarette, cigar, or pipe.

Here's why sidestream smoke is more toxic than exhaled smoke: When a person smokes, the smoke he

or she inhales and then breathes out leaves harmful deposits inside the body. But because lungs partially cleanse the smoke, exhaled smoke contains fewer poisonous chemicals. This is why exposure to tobacco smoke is dangerous even for a nonsmoker.

Causes and symptoms

No one starts smoking to become addicted to nicotine. It is not known how much nicotine may be consumed before the body becomes addicted. However, once smoking becomes a habit, the smoker faces a lifetime of health risks associated with one of the strongest addictions known to man.

About 70% of smokers in the United States would like to quit; in any given year, however, only about 3.6% of the country's 47 million smokers quit successfully. In 2008, the Centers for Disease Control and Prevention (CDC) reported that the prevalence of smoking in the United States fell in 2007 to 19.8%, almost a full percentage point decline from 20.8% in 2006.

Researchers conjecture that genetic factors contribute substantially to developing a smoking habit. Several twin studies have led to estimates of 46–84% heritability for smoking. It is thought that some genetic variations affect the speed of nicotine metabolism in the body and the activity level of nicotinic receptors in the brain.

Smoking risks

Smoking is recognized as the leading preventable cause of **death**, causing or contributing to the deaths of approximately 440,000 Americans each year. Anyone with a smoking habit has an increased chance of lung, cervical, and other types of cancer; respiratory diseases such as **emphysema**, **asthma**, and chronic **bronchitis**; and cardiovascular disease, such as **heart attack**, high blood pressure, **stroke**, and **atherosclerosis** (narrowing and hardening of the arteries). The risk of stroke is especially high in women who take birth control pills.

Smoking can damage fertility, making it harder to conceive, and it can interfere with the growth of the fetus during **pregnancy**. It accounts for an estimated 14% of premature births and 10% of infant deaths. There is some evidence that smoking may cause **impotence** in some men.

Because smoking affects so many of the body's systems, smokers often have vitamin deficiencies and suffer oxidative damage caused by free radicals. Free radicals are molecules that steal electrons from other molecules, turning the other molecules into free radicals and destabilizing the molecules in the body's cells.

Smoking is recognized as one of several factors that might be related to a higher risk of **hip fractures** in older adults.

Studies reveal that the more a person smokes, the more likely he is to sustain illnesses such as cancer, chronic bronchitis, and emphysema. But even smokers who indulge in the habit only occasionally are more prone to these diseases.

Some brands of cigarettes are advertised as "low tar" but no cigarette is truly safe. If a smoker switches to a low-tar cigarette, he or she is likely to inhale longer and more deeply to get the chemicals his body craves. A smoker has to quit the habit entirely in order to improve his health and decrease the chance of disease.

Though some people believe chewing tobacco is safer, it also carries health risks. People who chew tobacco have an increased risk of heart disease and mouth and throat cancer. Pipe and cigar smokers have increased health risks as well, even though these smokers generally do not inhale as deeply as cigarette smokers do. These groups haven't been studied as extensively as cigarette smokers but there is evidence that they may be at a slightly lower risk of cardiovascular problems but a higher risk of cancer and various types of circulatory conditions.

Recent research reveals that passive smokers, or those who unavoidably breathe in second-hand tobacco smoke, have an increased chance of many health problems such as lung cancer and asthma, and in children, **sudden infant death syndrome**.

Smokers' symptoms

Smokers are likely to exhibit a variety of symptoms that reveal the damage caused by smoking. A nagging morning **cough** may be one sign of a tobacco habit. Other symptoms include **shortness of breath**, **wheezing**, and frequent occurrences of respiratory illness, such as bronchitis. Smoking also increases **fatigue** and decreases the smoker's sense of smell and taste. Smokers are more likely to develop poor circulation, with cold hands and feet and premature wrinkles.

Sometimes the illnesses that result from smoking come on silently with little warning. For instance, **coronary artery disease** may exhibit few or no symptoms. At other times, there will be warning signs, such as bloody discharge from a woman's vagina, a sign of cancer of the cervix. Another warning sign is a hacking cough, worse than the usual smoker's cough, that brings up phlegm or blood—a sign of lung cancer.

Withdrawal symptoms

A smoker who tries to quit may expect one or more of these withdrawal symptoms: **nausea, constipation or diarrhea**, drowsiness, loss of concentration, **insomnia, headache**, nausea, and irritability.

Diagnosis

It is not easy to quit smoking, which is why it may be wise for a smoker to turn to his physician for help. For the greatest success in quitting and to help with the withdrawal symptoms, the smoker should talk over a treatment plan with his doctor or alternative practitioner. He should have a general **physical examination** to gauge his general health and uncover any deficiencies. He should also have a thorough evaluation for some of the serious diseases that smoking can cause.

Treatment

Research shows that most smokers who want to quit benefit from the support of other people. It helps to quit with a friend or to join a group such as those organized by the American Cancer Society. These groups provide support and teach behavior modification methods that can help the smoker quit. The smoker's physician can often refer him to such groups.

Other alternatives to help with the withdrawal symptoms of kicking the habit include nicotine replacement therapy in the form of gum, patches, nasal sprays, and oral inhalers. These are available by prescription or over the counter. A physician can provide advice on how to use them. They slowly release a small amount of nicotine into the bloodstream, satisfying the smoker's physical craving. Over time, the amount of gum the smoker chews is decreased and the amount of time between applying the patches is increased. This helps wean the smoker from nicotine slowly, eventually beating his addiction to the drug. But there's one important caution: If the smoker lights up while taking a nicotine replacement, a nicotine overdose may cause serious health problems.

The prescription drug Zyban (bupropion hydrochloride) has shown some success in helping smokers quit. This drug contains no nicotine and was originally developed as an antidepressant. It isn't known exactly how bupropion works to suppress the desire for nicotine. A five-year study of bupropion reported that the drug has a very good record for safety and effectiveness in treating tobacco dependence. Its most common side effect is insomnia, which can also result from nicotine withdrawal.

Researchers are investigating two new types of drugs as possible treatments for tobacco dependence. The first is an alkaloid known as 18-methoxycoronaridine (18-MC), which selectively blocks the nicotinic receptors in brain tissue. Another approach involves developing drugs that inhibit the activity of cytochrome P450 2A6 (CYP2A6), which controls the metabolism of nicotine.

Expected results

Research on smoking shows that most smokers desire to quit. But smoking is so addictive that fewer than 20% of the people who try ever successfully kick the habit. Still, many people attempt to quit smoking over and over again, despite the difficulties—the cravings and withdrawal symptoms, such as irritability and restlessness.

For those who do quit, the benefits to health are well worth the effort. The good news is that once a smoker quits the health effects are immediate and dramatic. After the first day, oxygen and carbon monoxide levels in the blood return to normal. At two days, nerve endings begin to grow back and the senses of taste and smell revive. Within two weeks to three months, circulation and breathing improve. After one year of not smoking, the risk of heart disease is reduced by 50%. After 15 years of abstinence, the risks of health problems from smoking virtually vanish. A smoker who quits for good often feels a lot better too, with less fatigue and fewer respiratory illnesses.

Alternative treatment

There are a wide range of alternative treatments that can help a smoker quit the habit, including **hypnotherapy**, herbs, **acupuncture**, and **meditation**. For example, a controlled trial demonstrated that self-massage can help smokers crave less intensely, smoke fewer cigarettes, and in some cases completely give them up.

Hypnotherapy

Hypnotherapy helps the smoker achieve a trance-like state, during which the deepest levels of the mind are accessed. A session with a hypnotherapist may begin with a discussion of whether the smoker really wants to and truly has the motivation to stop smoking. The therapist will explain how hypnosis can reduce the stress-related symptoms that sometimes come with kicking the habit.

Often the therapist will discuss the dangers of smoking with the patient and begin to "reframe" the patient's thinking about smoking. Many smokers are convinced they can't quit and the therapist can help

KEY TERMS

Antioxidant—Any substance that reduces the damage caused by oxidation, such as the harm caused by free radicals.

Chronic bronchitis—A smoking-related respiratory illness in which the membranes that line the bronchi, or the lung's air passages, narrow over time. Symptoms include a morning cough that brings up phlegm, breathlessness, and wheezing.

Cytochrome—A substance that contains iron and acts as a hydrogen carrier for the eventual release of energy in aerobic respiration.

Emphysema—An incurable, smoking-related disease, in which the air sacs at the end of the lung's bronchi become weak and inefficient. People with emphysema often first notice shortness of breath, repeated wheezing and coughing that brings up phlegm.

Epinephrine—A nervous system hormone stimulated by the nicotine in tobacco. It increases heart rate and may raise smokers' blood pressure.

Flavonoid—A food chemical that helps to limit oxidative damage to the body's cells, and protects against heart disease and cancer.

Free radical—An unstable molecule that causes oxidative damage by stealing electrons from surrounding molecules, thereby disrupting activity in the body's cells.

Nicotine—The addictive ingredient of tobacco, it acts on the nervous system and is both stimulating and calming.

Nicotine replacement therapy—A method of weaning a smoker away from both nicotine and the oral fixation that accompanies a smoking habit by giving the smoker smaller and smaller doses of nicotine in the form of a patch or gum.

Sidestream smoke—The smoke that is emitted from the burning end of a cigarette or cigar, or that comes from the end of a pipe. Along with exhaled smoke, it is a constituent of second-hand smoke.

persuade them that they can change this behavior. These suggestions are then repeated while the smoker is under hypnosis. The therapist may also suggest while the smoker is under hypnosis that his feelings of worry, anxiety, and irritability will decrease.

In a review of 17 studies of the effectiveness of hypnotherapy, the percentage of people treated by hypnosis who still were not smoking after six months ranged from 4–8%. In programs that included several hours of treatment, intense interpersonal interaction, individualized suggestions, and follow-up treatment, success rates were above 50%.

Aromatherapy

One study demonstrated that inhaling the vapor from black pepper extract can reduce symptoms associated with smoking withdrawal. Other essential oils can be used for relieving the anxiety a smoker often experiences while quitting.

Herbs

A variety of herbs can help smokers reduce their cravings for nicotine, calm their irritability, and even reverse the oxidative cellular damage done by smoking. Lobelia, sometimes called Indian tobacco, has historically been used as a substitute for tobacco. It contains a substance called lobeline, which decreases

the craving for nicotine by bolstering the nervous system and calming the smoker. In high doses, lobelia can cause **vomiting** but the average dose—about 10 drops per day—should pose no problems.

Herbs that can help relax a smoker during withdrawal include wild oats and kava kava.

To reduce the oral fixation supplied by a nicotine habit, a smoker can chew on licorice root—the plant, not the candy. Licorice is good for the liver, which is a major player in the body's **detoxification** process. Licorice also acts as a tonic for the adrenal system, which helps reduce stress. And there's an added benefit: If a smoker tries to light up after chewing on licorice root, the cigarette tastes like burned cardboard.

Other botanicals that can help repair free-radical damage to the lungs and cardiovascular system are those high in flavonoids, such as hawthorn, gingko biloba, and bilberry, as well as **antioxidants** such as vitamin A, vitamin C, zinc, and selenium.

Acupuncture

This ancient Chinese method of healing is used commonly to help beat addictions, including smoking. The acupuncturist will use hair-thin needles to stimulate the body's *qi*, or healthy energy. Acupuncture is a sophisticated treatment system based on revitalizing *qi*, which supposedly flows through the body in defined pathways

called meridians. During an addiction like smoking, qi isn't flowing smoothly or gets stuck, the theory goes.

Points in the ear and feet are stimulated to help the smoker overcome his addiction. Often the acupuncturist will recommend keeping the needles in for five to seven days to calm the smoker and keep him balanced.

Vitamins

Smoking seriously depletes vitamin C in the body and leaves it more susceptible to infections. Vitamin C can prevent or reduce free-radical damage by acting as an antioxidant in the lungs. Smokers need additional C, in higher dosage than nonsmokers. Fish in the diet supplies **Omega-3 fatty acids**, which are associated with a reduced risk of **chronic obstructive pulmonary disease** (emphysema or chronic bronchitis) in smokers. Omega-3 fats also provide cardiovascular benefits as well as an anti-depressive effect. Vitamin therapy doesn't reduce craving but it can help beat some of the damage created by smoking. Vitamin B₁₂ and **folic acid** may help protect against smoking-induced cancer.

Prevention

How do you give up your cigarettes for good and never go back to them again?

Here are a few tips from the experts:

- Have a plan and set a definite quit date.
- Get rid of all the cigarettes and ashtrays at home or in your desk at work.
- Don't allow others to smoke in your house.
- Tell your friends and neighbors that you're quitting. Doing so helps make quitting a matter of pride.
- Chew sugarless gum or eat sugar-free hard candy to redirect the oral fixation that comes with smoking. This will prevent weight gain, too.
- Eat as much as you want but only low-calorie foods and drinks. Drink plenty of water. This may help with the feelings of tension and restlessness that quitting can bring. After eight weeks, you'll lose your craving for tobacco so it's safe then to return to your usual eating habits.
- Stay away from social situations that prompt you to smoke. Dine in the nonsmoking section of restaurants.
- Spend the money you save not smoking on an occasional treat for yourself.

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- American Association of Oriental Medicine. 5530 Wisconsin Avenue, Suite 1210, Chevy Chase MD, 20815, (301) 941-1064, (888) 500-7999, <http://www.aaom.org>.
- American Cancer Society. Contact the local organization or call (800) 227-2345, <http://www.cancer.org>.
- American Lung Association. 1740 Broadway, New York NY, 10019, (800) 586-4872, (212) 315-8700, <http://www.lungusa.org>.
- Herb Research Foundation. 1007 Pearl St., Suite 200, Boulder CO, 80302, (303) 449-2265, <http://www.herbs.org>.
- National Heart, Lung, and Blood Institute (NHLBI). Building 31, Room 5A52, 31 Center Drive, MSC 2486,

Bethesda MD, 20892, (301) 592-8573, <http://www.nhlbi.nih.gov>.

Smoking, Tobacco, and Health Information Line. Centers for Disease Control and Prevention. Mailstop K-50, 4770 Buford Highway NE, Atlanta GA, 30341-3724, (800) 232-1311, <http://www.cdc.gov/tobacco>.

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Barbara Boughton

Snoring

Definition

Snoring is a sound generated during sleep by vibration of loose tissue in the upper airway.

Description

Snoring is one symptom of a group of disorders known as sleep disordered breathing. It occurs when the soft palate, uvula, tongue, tonsils, and/or muscles in the back of the throat rub against each other and generate a vibrating sound during sleep. 20% of all adults are chronic snorers and 45% of normal adults snore occasionally. As people grow older, their chance

of snoring increases. Approximately half of all individuals over 60 snore regularly.

In some cases, snoring is a symptom of a more serious disorder called obstructed **sleep apnea** (OSA). OSA occurs when part of the airway is closed off (usually at the back of the throat) while a person is trying to inhale during sleep, and breathing stops for more than 10 seconds before resuming again. These breathless episodes can occur as many as several hundred times a night.

People with OSA almost always snore heavily because the same narrowing of the airway that causes snoring can also cause OSA. Snoring may actually attribute to OSA as well, because the vibration of the throat tissues which occurs in snoring can cause the tissue to swell.

Snoring is associated with physical problems as well as social **stress**. People who do not suffer from OSA may be diagnosed with socially unacceptable snoring (SUS), which refers to snoring that is loud enough to prevent the sleeper’s bed partner or roommate from sleeping. SUS is a factor in the breakup of some marriages and other long-term relationships. Moreover, a study published in 2002 indicates that people who snore are at increased risk of developing type 2 diabetes. Snoring appears to be a risk factor that is independent of body weight or a family history of diabetes.

Causes and symptoms

There are several major causes of snoring, including:

- Excessively relaxed throat muscles. Alcohol, drugs, and sedatives can cause the throat muscles to become lax, and/or the tongue to pull back into the airway.
- Large uvula. The piece of tissue that hangs from the back of the throat is called the uvula. Individuals with a large or longer than average uvula can suffer from snoring when the uvula vibrates in the airway.
- Large tonsils and/or adenoids. The tonsils (tissue at the back of either side of the throat) can also vibrate if they are larger than normal, as can the adenoids.
- Excessive weight. Overweight people are more likely to snore. This is frequently caused by the extra throat and neck tissue they are carrying around.
- Nasal congestion. Colds and allergies can plug the nose, creating a vacuum in the throat that results in snoring as airflow increases.
- Cysts and tumors. Cysts and/or tumors of the throat can trigger snoring.
- Structural problems of the nose. A deviated septum or other nasal problems can also cause snoring.

Diagnosis

A patient interview, and possibly an interview with the patient's spouse or anyone else in the household who has witnessed the snoring, is usually enough for a diagnosis of snoring. A medical history that includes questions about alcohol or tranquilizer use; past ear, nose, and throat problems; and the pattern and degree of snoring will be completed, and a physical exam will be performed to determine the cause of the problem. This will typically include examination of the throat to look for narrowing, obstruction, or malformations. If the snoring is suspected to be a symptom of a more serious disorder such as obstructive sleep apnea, the patient will require further testing. This testing is called a **polysomnography** study, and is conducted during an overnight stay in a specialized sleep laboratory. The polysomnography study include measurements of heart rate, airflow at the mouth and nose, respiratory effort, sleep stage (light sleep, deep sleep, dream sleep, etc.), and oxygen level in the blood.

In some cases the patient may be referred to a dentist or orthodontist for evaluation of the jaw structure and dentition.

In addition, the patient may be examined by sleep **endoscopy**. In this procedure, the patient is given a medication (midazolam) to induce sleep. His or her throat and nasal passages are then examined with a flexible laryngoscope. In many cases, sleep endoscopy reveals obstructions that are not apparent during a standard **physical examination** of the throat. Many patients are found to have obstructions at more than one level in their breathing passages.

Treatment

Several surgical procedures are available for treating chronic snoring. These include:

- Uvulopalatopharyngoplasty (UPPP), a surgical procedure which involves removing excess throat tissues (e.g., tonsils, parts of the soft palate) to expand the airway.
- Laser-assisted uvulopalatoplasty (LAUP) uses a surgical laser to remove part of the uvula and palate.
- Palatal stiffening is a minimally-invasive surgical technique where a laser or a cauterizer is used to produce scar tissue in the soft palate in order to stop the vibrations that produce snoring.
- Radiofrequency ablation is another technique which uses scarring to shrink the uvula and/or soft palate. A needle electrode is used to shrink and scar the mouth and throat tissues.

Alternative treatment

There are a number of remedies for snoring, but few are proven clinically effective. Popular treatments include:

- Mechanical devices. Many splints, braces, and other devices are available which reposition the nose, jaw, and/or mouth in order to clear the airways. Other devices are designed to wake an individual when snoring occurs. Patients should consult a dentist or orthodontist about these devices, as most require custom fitting. In addition, persons with certain types of gum disease or dental problems should not be fitted with oral appliances to stop snoring.
- Nasal strips. Nasal strips that attach like an adhesive bandage to the bridge of the nose are available at most drugstores and can help stop snoring in some individuals by opening the nasal passages.
- Continuous positive airway pressure (CPAP). Some chronic snorers find relief by sleeping with a nasal mask which provides air pressure to the throat.
- Decongestants. Snoring caused by nasal congestion may be successfully treated with decongestants. Some effective herbal remedies that clear the nasal passages include golden rod (*Solidago virgaurea*) and golden seal (*Hydrastis canadensis*). Steam inhalation of essential oils of eucalyptus blue gum (*Eucalyptus globulus*) or peppermint (*Mentha x piperata*) can also relieve congestion.
- Weight loss. Snoring thought to be caused by excessive weight may be curtailed by a sensible weight loss and exercise program.
- Sleep position. Snoring usually worsens when an individual sleeps on his or her back, so sleeping on one's side may alleviate the problem. Those who have difficulty staying in a side sleeping position may find sleeping with pillows behind them helps them maintain the position longer. Other devices include a new vest designed to prevent the sleeper from lying on his or her back.
- Bed adjustments. For some people, raising the head of the bed solves their snoring problem. A slight incline can prevent the tongue from retracting into the back of the throat. Bricks, wooden blocks, or specially designed wedges can be used to elevate the head of the bed approximately 4–16 in (10–41 cm).

Alternative treatments that have been reported to be effective for patients whose snoring is caused by colds or **allergies** include **acupuncture**, homeopathy, and **aromatherapy** treatments. Aromatherapy treatments for snoring typically make use of marjoram

KEY TERMS

Ablation—The removal of abnormal tissue growths by surgery.

Cauterize—To seal tissue or blood vessels using a heat or electrical source.

Continuous positive airway pressure (CPAP)—A ventilation device that blows a gentle stream of air into the nose during sleep to keep the airway open.

Deviated septum—A hole or perforation in the septum, the wall that divides the two nasal cavities.

Endoscope—A slender optical instrument that allows a doctor to examine the inside of the throat or other hollow organ. Sleep endoscopy is a technique that allows the doctor to detect previously unsuspected obstructions in the patient's nose and throat.

Obstructive sleep apnea (OSA)—A potentially life-threatening condition characterized by episodes of breathing cessation during sleep alternating with snoring or disordered breathing. The low levels of oxygen in the blood of patients with OSA may eventually cause heart problems or stroke.

Polysomnography—A technique for diagnosing sleep disorders with the use of a machine that records the pulse, breathing rate and other variables while the patient sleeps.

Soft palate—The structure at the roof of the mouth that separates the mouth and the pharynx.

oil, which is thought to be particularly effective in clearing the nasal passages.

Prevention

Adults with a history of snoring may be able to prevent snoring episodes with the following measures:

- avoid alcohol and sedatives before bedtime
- remove allergens from the bedroom
- use a decongestant before bed
- sleep on the side, not the back

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ORGANIZATIONS

- American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Road, Alexandria, VA, 22314-2857, (703) 836-4444, <http://www.entnet.org>.
- American Academy of Sleep Medicine (AASM), 2510 N. Frontage Road, Darien, IL, 60561, (630) 737-9700, (630) 737-9790, inquiries@aasmnet.org, <http://www.aasmnet.org>.
- American Dental Association, 211 E. Chicago Ave., Chicago, IL, 60611-2678, (312) 440-2500, <http://www.ada.org>.

American Sleep Apnea Association, 6856 Eastern Avenue, NW, Suite 203, Washington, DC, 20012, (202) 293-3650, (202) 293-3656, <http://www.sleepapnea.org/>. National Sleep Foundation, 1522 K St. NW, Suite 500, Washington, DC, 20005, (202) 347-3471, (202) 347-2472, <http://www.sleepfoundation.org>.

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Sodium

Definition

Sodium is a mineral that exists in the body as the ion Na⁺. Sodium is acquired through diet, mainly in the form of salt (sodium chloride, NaCl). Regulating the amount of Na⁺ in the body is absolutely critical to life and health.

Purpose

Sodium is possibly the most important mineral in the body. It plays a major role in controlling the distribution of fluids, maintaining blood pressure and blood

Sodium	
Age	Adequate Intake (mg)
Children 0–6 mos.	120
Children 7–12 mos.	370
Children 1–3 yrs.	1,000
Children 4–8 yrs.	1,200
Children 9–13 yrs.	1,500
Adolescents 14–18 yrs.	1,500
Adults 19–50 yrs.	1,500
Adults 51–70 yrs.	1,300
Adults 71+ yrs.	1,200
Pregnant women	1,500
Breastfeeding women	1,500
Food	Sodium (mg)
Table salt, 1 tsp.	2,300
Dill pickle, 1 large	1,731
Chicken noodle soup, canned, 1 cup	850–1,100
Ham, 3 oz.	1,000
Sauerkraut, ½ cup	780
Pretzels, 1 oz.	500
Turkey breast, deli, 1 oz.	335
Soy sauce, 1 tsp.	304
Potato chips, 1 oz.	165–185

mg = milligram

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volume, creating an electrical gradient that allows nerve transmission and muscle contraction to occur, maintaining the mechanisms that allow wastes to leave cells, and regulating the acidity (pH) of the blood. Many different organs working together, including the kidneys, endocrine glands, and brain, tightly control the level of Na⁺ in the body. Researchers estimate that between 20% and 40% of an adult's resting energy use goes toward regulating sodium. Sodium affects every cell in the body and a major failure of sodium regulatory mechanisms means **death**.

Description

In the body, sodium exists as electrolyte. Electrolytes are ions that form when salts dissolve in water or fluids. These ions have an electric charge. Positively charged ions are called cations. Negatively charged ions are called anions. Electrolytes are not evenly distributed within the body and their uneven distribution allows many important metabolic reactions to occur. Sodium (Na⁺), potassium (K⁺), **calcium** (Ca²⁺), magnesium (Mg²⁺), chloride (Cl⁻), phosphate (HPO₄²⁻), bicarbonate (HCO₃⁻), and sulfate (SO₄²⁻) are important electrolytes in humans.

Na⁺ is ten times more concentrated in fluid outside cells (i.e. extracellular fluid and blood) than it is in fluid inside cells. This difference in concentration is maintained through the expenditure of cellular energy, and it is critical to many metabolic functions, including maintaining the proportion of water that exists inside and outside of cells. (See the entry on electrolytes for a more detailed explanation of how this occurs.) When Na⁺ is too high or too low, it is almost never because an individual has eaten too much or too little salt. Instead, it is because organs such as the kidneys or endocrine glands that regulate the conservation or removal of sodium from the body have broken down.

Sodium requirements

Researchers estimate that humans can remain healthy taking in only 500 mg of sodium daily. Salt is 40% sodium by weight and 500 mg is slightly less than the amount of sodium found in 1/4 teaspoon of salt. Humans almost never take in too little salt; their health problems result from too much salt in the diet.

The United States Institute of Medicine (IOM) of the National Academy of Sciences has developed values called Dietary Reference Intakes (DRIs) for many **vitamins** and **minerals**, including sodium. The DRIs consist of three sets of numbers. The Recommended Dietary Allowance (RDA) defines the average daily amount of the nutrient needed to meet the health needs of 97–98%

of the population. The Adequate Intake (AI) is an estimate set when there is not enough information to determine an RDA. The Tolerable Upper Intake Level (UL) is the average maximum amount that can be taken daily without risking negative side effects. The DRIs are calculated for children, adult men, adult women, pregnant women, and **breastfeeding** women.

The IOM has not set RDAs for sodium, but instead it has set AI levels for all age groups based on observed and experimental information about the amount of sodium needed to replace what is lost by a moderately active individual each day. Sodium is lost in both urine and sweat. IAs for sodium are measured in milligrams (mg). UL levels have not been set. However, the IOM recommends that adults limit their sodium intake to less than 2,400 mg per day, and the American Heart Association recommends an adult daily intake of 1,500–2,300 mg.

The following list gives the recommended daily AI levels of sodium for each age group.

- children birth–6 months: AI 120 mg
- children 7–12 months: AI 370 mg
- children 1–3 years: AI 1,000 mg
- children 4–8 years: AI 1,200 mg
- children 9–13 years: AI 1,500 mg
- adolescents 14–18 years: IA 1,500 mg
- adults age 19–50: AI 1,500 mg
- adults ages 50–70: 1,300 mg
- adults 71 years or older: AI 1,200 mg
- pregnant women: IA 1,500 mg
- breastfeeding women: AI 1,500 mg

Sources of sodium

Many people think that the main source of salt in their diet is what they add to food when they are cooking or at the table while eating. In reality, more than three-quarters of the sodium in the average American's diet is added to food during processing. Another 12% is already naturally in the food. For example, 1 cup of low-fat milk contains 110 mg of sodium. About 6% of sodium in the diet is added as salt during cooking and another 5% from salting food while eating.

Although most sodium in diet comes from salt, other sources of sodium include preservatives and flavor enhancers added during processing. Sodium content is required to be listed on food labels of processed foods. Some common "hidden" sources of sodium include:

- baking soda
- baking powder

- disodium phosphate
- monosodium glutamate (MSG)
- sodium nitrate or sodium nitrite

Below are some common foods and their sodium content.

- table salt, 1 teaspoon: 2,300 mg
- dill pickle, large: 1731 mg
- canned chicken noodle soup, 1 cup: 850–1,100 mg
- ham, 3 ounces: 1,000 mg
- sauerkraut, 1/2 cup: 780 mg
- pretzels, 1 ounce: 500 mg
- potato chips, 1 ounce: 165–185 mg
- soy sauce, 1 teaspoon: 304 mg
- deli turkey breast, 1 ounce: 335 mg

Fresh fruits, vegetables, unsalted nuts, and rice, dried beans and peas are examples of foods that are low in sodium.

Sodium and health

Too high a concentration of sodium in the blood causes a condition called **hyponatremia**. Too much sodium in the diet almost never causes Hyponatremia. Causes include excessive water loss (e.g. severe **diarrhea**), restricted water intake, untreated diabetes (causes water loss), **kidney disease**, and hormonal imbalances. Symptoms include signs of **dehydration** such as extreme thirst, dark urine, sunken eyes, **fatigue**, irregular heart beat, muscle twitching, seizures, and **coma**.

Too low a concentration of sodium in the blood causes **hypertension**. Hypotension is not usually a problem in healthy individuals, although it has been known to occur in endurance athletes such as ultramarathoners. It is common in seriously ill individuals and can result from **vomiting** or diarrhea (extreme loss of sodium), severe **burns**, taking certain drugs that cause the kidney to selectively excrete sodium, extreme overconsumption of water (water intoxication, a problem among the elderly with **dementia**), hormonal imbalances, kidney failure, and liver damage. Symptoms include **nausea**, **vomiting**, **headache**, tissue swelling (**edema**), confusion, mental disorientation, **hallucinations**, muscle trembling, seizures, and coma.

Hyponatremia and hypotension are at the extreme ends of sodium imbalance. However, high dietary intake of salt can cause less visible health damage in the form of high blood pressure (**hypertension**). Hypertension silently damages the heart, blood vessels, and kidney and increases the risk of **stroke**, **heart attack**, and kidney damage. A low-salt diet significantly lowers blood pressure in 30–60% of people with high blood pressure and a quarter to half of people with normal

blood pressure. Some individuals are more sensitive to sodium than others. Those people who are most likely to see a rise in blood pressure with increased sodium intake include people who are obese, have type 2 diabetes, are elderly, female, and African American.

The American Heart Association recommends reducing sodium in the diet to between 1,500 mg and 2,300 mg daily. Below are some suggestions for cutting down on salt.

- Eat more fresh fruits and vegetables.
- Look for processed foods that say “no salt added”
- Limit or eliminate salty snacks such as chips and pretzels.
- Restrict the amount processed meats such as hot dogs, pepperoni, and deli meats.
- Avoid high salt canned soups; choose heart-healthy lower salt soups instead.
- Use spices instead of salt to give foods flavor.

Precautions

People who are salt-sensitive may need to keep their salt intake at levels below the suggested daily amounts to control their blood pressure.

Interactions

Certain drugs cause large amounts of sodium to be excreted by the kidneys and removed from the body in urine. **Diuretics** (“water pills”) are among the best known of these drugs. Other types of drugs that may cause low sodium levels, especially in ill individuals, include non-steroidal anti-inflammatory drugs (NSAIDs) such as Advil, Motrin, and Aleve, opiates such as codeine and morphine, selective serotonin-reuptake inhibitors (SSRIs) such as Prozac or Paxil, and **tricyclic antidepressants** such as Elavil and Tofranil.

Complications

Health concerns about sodium have been discussed above. Most problems related to high blood pressure are chronic, slow to develop disorders that do not cause serious complications until the second half of an individual’s lifetime. Kidney failure, heart attack, and stroke are all complications of high blood pressure and potentially of high sodium intake.

Parental concerns

Salt is an acquired taste. Parents can help their children control their salt intake and discourage the development of a craving for salt by substituting low-salt foods for high-salt foods.

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ORGANIZATIONS

American Heart Association, 7272 Greenville Avenue, Dallas TX, 75231, (800) 242-8721, <http://www.americanheart.org>.

International Food Information Council, 1100 Connecticut Avenue, NW Suite 430, Washington, DC, 20036, (202) 296-6540, (202) 296-6547, <http://ific.org>.

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Sodium imbalance see **Hyponatremia; Hyponatremia**

Somatization disorder see **Somatoform disorders**

Somatoform disorders

Definition

The somatoform disorders are a group of mental disturbances placed in a common category on the basis of their external symptoms. These disorders are characterized by physical complaints that appear to be medical in origin but that cannot be explained in terms of a physical disease, the results of **substance abuse**, or by another mental disorder. In order to meet the criteria for a somatoform disorder, the physical symptoms must be serious enough to interfere with the patient's employment or relationships, and must be symptoms that are not under the patient's voluntary control.

It is helpful to understand that the present classification of these disorders reflects recent historical changes in the practice of medicine and psychiatry. When psychiatry first became a separate branch of medicine at the end of the nineteenth century, the term *hysteria* was commonly used to describe mental disorders characterized by altered states of consciousness (for example, sleepwalking or trance states) or physical symptoms (for example, a "paralyzed" arm or leg with no neurologic cause) that could not be fully explained by a medical disease. The term *dissociation* was used for the psychological mechanism that allows the mind to split off uncomfortable feelings, memories, or ideas so that they are lost to conscious recall. Sigmund Freud and other pioneering psychoanalysts thought that the hysterical patient's symptoms resulted from dissociated thoughts or memories re-emerging through bodily functions or trance states. Prior to the categorization all mental disorders that were considered to be forms of **hysteria** were grouped together on the basis of this theory about their cause. Since 1980, however, the somatoform disorders and the so-called **dissociative disorders** have been placed in separate categories on the basis of their chief symptoms. In general, the somatoform disorders are characterized by disturbances in the patient's physical sensations or ability to move the limbs or walk, while the dissociative disorders are marked by disturbances in the patient's sense of identity or memory.

Description

As a group, the somatoform disorders are difficult to recognize and treat because patients often have long histories of medical or surgical treatment with several different doctors. In addition, the physical symptoms are not under the patient's conscious control so that he or she is not intentionally trying to confuse the doctor

or complicate the process of diagnosis. Somatoform disorders are, however, a significant problem for the health care system because patients with these disturbances overuse medical services and resources.

Somatization disorder (Briquet's syndrome)

Somatization disorder was formerly called Briquet's syndrome, after the French physician who first recognized it. The distinguishing characteristic of this disorder is a group or pattern of symptoms in several different organ systems of the patient's body that cannot be accounted for by medical illness. The criteria for this disorder require four symptoms of **pain**, two symptoms in the digestive tract, one symptom involving the sexual organs, and one symptom related to the nervous system. Somatization disorder usually begins before the age of 30. It is estimated that 0.2% of the United States population will develop this disorder in the course of their lives. Another researcher estimates that 1% of all women in the United States have symptoms of this disorder. The female-to-male ratio is estimated to range between 5:1 and 20:1.

Somatization disorder is considered to be a chronic disturbance that tends to persist throughout the patient's life. It is also likely to run in families. Some psychiatrists think that the high female-to-male ratio in this disorder reflects the cultural pressures on women in North American society and the social "permission" given to women to be physically weak or sickly.

Conversion disorder

Conversion disorder is a condition in which the patient's senses or ability to walk or move are impaired without a recognized medical or neurological disease or cause and in which psychological factors (such as **stress** or trauma) are judged to be temporarily related to onset or exacerbation. The disorder gets its name from the notion that the patient is converting a psychological conflict or problem into an inability to move specific parts of the body or to use the senses normally. An example of a conversion reaction would be a patient who loses his or her voice in a situation in which he or she is afraid to speak. The symptom simultaneously contains the **anxiety** and serves to get the patient out of the threatening situation. The resolution of the emotion that underlies the physical symptom is called the patient's *primary gain*, and the change in the patient's social, occupational, or family situation that results from the symptom is called a *secondary gain*. Doctors sometimes use these terms when they discuss the after-effects of conversion disorder or of other somatoform disorders on the patient's emotional adjustment and lifestyle.

The specific physical symptoms of conversion disorder may include a loss of balance or **paralysis** of an arm or leg; the inability to swallow or speak; the loss of touch or pain sensation; going blind or deaf; seeing double; or having **hallucinations**, seizures, or convulsions.

Unlike somatization disorder, conversion disorder may begin at any age, and it does not appear to run in families. It is estimated that as many as 34% of the population experiences conversion symptoms over a lifetime but that the disorder is more likely to occur among less educated or sophisticated people. Conversion disorder is not usually a chronic disturbance; 90% of patients recover within a month, and most do not have recurrences. The female-to-male ratio is between 2:1 and 5:1. Male patients are likely to develop conversion disorders in occupational settings or military service.

Pain disorder

Pain disorder is marked by the presence of severe pain as the focus of the patient's concern. This category of somatoform disorder covers a range of patients with a variety of ailments, including chronic headaches, back problems, arthritis, muscle aches and cramps, or pelvic pain. In some cases the patient's pain appears to be largely due to psychological factors, but in other cases the pain is derived from a medical condition as well as the patient's psychology.

Pain disorder is relatively common in the general population, partly because of the frequency of work-related injuries in the United States. This disorder appears to be more common in older adults, and the sex ratio is nearly equal, with a female-to-male ratio of 2:1.

Hypochondriasis

Hypochondriasis is a somatoform disorder marked by excessive fear of or preoccupation with having a serious illness that persists in spite of medical testing and reassurance. It was formerly called hypochondriacal neurosis.

Although hypochondriasis is usually considered a disorder of young adults, it is now increasingly recognized in children and adolescents. It may also develop in elderly people without previous histories of health-related fears. The disorder accounts for about 5% of psychiatric patients and is equally common in men and women. Hypochondriasis may persist over a number of years but usually occurs as a series of episodes rather than continuous treatment-seeking. The flare-ups of the disorder are often correlated with stressful events in the patient's life.

Body dysmorphic disorder

Body dysmorphic disorder is a new category of somatoform disorders. It is defined as a preoccupation with an imagined or exaggerated defect in appearance. Most cases involve features on the patient's face or head, but other body parts—especially those associated with sexual attractiveness, such as the breasts or genitals—may also be the focus of concern.

Body dysmorphic disorder is regarded as a chronic condition that usually begins in the patient's late teens and fluctuates over the course of time. It was initially considered to be a relatively unusual disorder but may be more common than was formerly thought. It appears to affect men and women with equal frequency. Patients with body dysmorphic disorder frequently have histories of seeking or obtaining **plastic surgery** or other procedures to repair or treat the supposed defect. Some may even meet the criteria for a delusional disorder of the somatic type.

Somatoform disorders in children and adolescents

The most common somatoform disorders in children and adolescents are conversion disorders, although body dysmorphic disorders are being reported more frequently. Conversion reactions in this age group usually reflect stress in the family or problems with school rather than long-term psychiatric disturbances. Some psychiatrists speculate that adolescents with conversion disorders frequently have overprotective or overinvolved parents with a subconscious need to see their child as sick; in many cases the son or daughter's symptoms become the center of family attention. The rise in body dysmorphic disorders in adolescents is thought to reflect the increased influence of media preoccupation with physical perfection.

Causes and symptoms

The causes of somatoform disorders include several different factors and they are categorized on the basis of symptom patterns.

Family stress

Family stress is believed to be one of the most common causes of somatoform disorders in children and adolescents. Conversion disorders in this age group may also be connected with physical or **sexual abuse** within the family of origin.

Parental modeling

Somatization disorder and hypochondriasis may result in part from the patient's unconscious reflection or imitation of parental behaviors. This "copycat" behavior is particularly likely if the patient's parent derived considerable secondary gain from his or her symptoms.

Cultural influences

Cultural influences appear to affect the gender ratios and body locations of somatoform disorders as well as their frequency in a specific population. Some cultures (for example, Greek and Puerto Rican) report higher rates of somatization disorder among men than is the case for the United States. In addition, researchers found lower levels of somatization disorder among people with higher levels of education. People in Asia and Africa are more likely to report certain types of physical sensations (for example, burning hands or feet, or the feeling of ants crawling under the skin) than are Westerners.

Biological factors

Genetic or biological factors may also play a role. For example, people who suffer from somatization disorder may also differ in how they perceive and process pain.

Diagnosis

Accurate diagnosis of somatoform disorders is important to prevent unnecessary surgery, laboratory tests, or other treatments or procedures. Because somatoform disorders are associated with physical symptoms, patients are often diagnosed by primary care physicians as well as by psychiatrists. In many cases the diagnosis is made in a general medical clinic. Children and adolescents with somatoform disorders are most likely to be diagnosed by pediatricians. Diagnosis of somatoform disorders requires a thorough physical workup to exclude medical and neurological conditions, or to assess their severity in patients with pain disorder. A detailed examination is especially necessary when conversion disorder is a possible diagnosis because some neurological conditions—including **multiple sclerosis** and myasthenia gravis—have on occasion been misdiagnosed as conversion disorder. Some patients who receive a diagnosis of somatoform disorder ultimately go on to develop neurologic disorders.

In addition to ruling out medical causes for the patient's symptoms, a doctor who is evaluating a patient for a somatization disorder will consider the possibility of other psychiatric diagnoses or of

overlapping psychiatric disorders. Somatoform disorders often coexist with **personality disorders** because of the chicken-and-egg relationship between physical illness and certain types of character structure or personality traits. At one time, the influence of Freud's theory of hysteria led doctors to assume that the patient's hidden emotional needs "cause" the illness. But in many instances, the patient's personality may have changed over time due to the stresses of adjusting to a chronic disease. This gradual transformation is particularly likely in patients with pain disorder. Patients with somatization disorder often develop panic attacks or **agoraphobia** together with their physical symptoms. In addition to anxiety or personality disorders, the doctor will usually consider major depression as a possible diagnosis when evaluating a patient with symptoms of a somatoform disorder. Pain disorders may be associated with depression and body dysmorphic disorder may be associated with obsessive-compulsive disease.

Treatment

Relationship with primary care practitioner

Because patients with somatoform disorders often have lengthy medical histories, a long-term relationship with a trusted primary care practitioner (PCP) is a safeguard against unnecessary treatments as well as a comfort to the patient. Many PCPs prefer to schedule brief appointments on a regular basis with the patient and keep referrals to specialists to a minimum. This practice also allows them to monitor the patient for any new physical symptoms or diseases. However, some PCPs work with a psychiatric consultant.

Medications

Patients with somatoform disorders are sometimes given **antianxiety drugs** or **antidepressant drugs** if they have been diagnosed with a coexisting mood or anxiety disorder. In general, however, it is considered better practice to avoid prescribing medications for these patients since they are likely to become psychologically dependent on them. However, body dysmorphic disorder has been successfully treated with **selective serotonin reuptake inhibitors** (SSRI) antidepressants.

Psychotherapy

Patients with somatoform disorders are not considered good candidates for **psychoanalysis** and other forms of insight-oriented **psychotherapy**. They can benefit, however, from supportive approaches to treatment that are aimed at symptom reduction and stabilization of the patient's personality. Some patients with pain disorder benefit from **group therapy** or support

KEY TERMS

Briquet's syndrome—Another name for somatization disorder.

Conversion disorder—A somatoform disorder characterized by the transformation of a psychological feeling or impulse into a physical symptom. Conversion disorder was previously called hysterical neurosis, conversion type.

Dissociation—A psychological mechanism in which the mind splits off certain aspects of a traumatic event from conscious awareness. Dissociation can affect the patient's memory, sense of reality, and sense of identity.

Hysteria—The earliest term for a psychoneurotic disturbance marked by emotional outbursts and/or disturbances of movement and sense perception. Some forms of hysteria are now classified as somatoform disorders and others are grouped with the dissociative disorders.

Hysterical neurosis—An older term for conversion disorder or dissociative disorder.

Primary gain—The immediate relief from guilt, anxiety, or other unpleasant feelings that a patient derives from a symptom.

Repression—A unconscious psychological mechanism in which painful or unacceptable ideas, memories, or feelings are removed from conscious awareness or recall.

Secondary gain—The social, occupational, or interpersonal advantages that a patient derives from symptoms. A patient's being relieved of his or her share of household chores by other family members would be an example of secondary gain.

Somatoform disorder—A category of psychiatric disorder characterized by physical complaints that appear to be medical in origin but that cannot be explained in terms of a physical disease, the results of substance abuse, or by another mental disorder.

groups, particularly if their social network has been limited by their pain symptoms. **Cognitive-behavioral therapy** is also used sometimes to treat pain disorder.

Family therapy is usually recommended for children or adolescents with somatoform disorders, particularly if the parents seem to be using the child as a focus to divert attention from other difficulties. Working with families of chronic pain patients also helps avoid reinforcing dependency within the family setting.

Hypnosis is a technique that is sometimes used as part of a general psychotherapeutic approach to conversion disorder because it may allow patients to recover memories or thoughts connected with the onset of the physical symptoms.

Alternative treatment

Patients with somatization disorder or pain disorder may be helped by a variety of alternative therapies including **acupuncture**, **hydrotherapy**, therapeutic massage, **meditation**, botanical medicine, and homeopathic treatment. Relief of symptoms, including pain, can occur on the physical level, as well as on the mental, emotional, and spiritual levels.

Prognosis

The prognosis for somatoform disorders depends, as a rule, on the patient's age and whether the disorder

is chronic or episodic. In general, somatization disorder and body dysmorphic disorder rarely resolve completely. Hypochondriasis and pain disorder may resolve if there are significant improvements in the patient's overall health and life circumstances, and people with both disorders may go through periods when symptoms become less severe (remissions) or become worse (exacerbations). Conversion disorder tends to be rapidly resolved but may recur in about 25% of all cases.

Prevention

Generalizations regarding prevention of somatoform disorders are difficult because these syndromes affect different age groups, vary in their symptom patterns and persistence, and result from different problems of adjustment to the surrounding culture. In theory, allowing expression of emotional pain in children, rather than regarding it as "weak," might reduce the secondary gain of physical symptoms that draw the care or attention of parents.

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Rebecca J. Frey, PhD

Somatotrophic hormone test see **Growth hormone tests**

Sore throat

Definition

Sore throat, also called pharyngitis, is a painful inflammation of the mucous membranes lining the pharynx. It is a symptom of many conditions, but most often is associated with colds or **influenza**. Sore throat may be caused by either viral or bacterial infections or environmental conditions. Most sore throats heal without complications but they should not be ignored because some develop into serious illnesses. A chronic sore throat with hoarseness is one of the seven warning signs of **cancer**.

Demographics

Almost everyone gets a sore throat at one time or another, although children in child care or grade school have them more often than adolescents and adults. Sore throats are most common during the winter months when upper respiratory infections (colds) are more frequent.

About 10% of children who go to the doctor each year have pharyngitis. 40% of the time that children are taken to the doctor with a sore throat, the sore throat is diagnosed as viral. An antibiotic cannot help to cure a virus; a virus has to be left to run its course.

In about 30% of the cases for which children are taken to the doctor, bacteria are found to be responsible for the sore throat. Many of these bacterial sore throats are cases of **strep throat**. Sore throats caused by bacteria can be successfully treated with **antiotics**. In about 40% of these cases of pharyngitis, it is never clear what caused the sore throat. In these cases it is possible that the virus or bacteria was not identified or that other factors such as environment or post-nasal drip may have been responsible.



Sore throat caused by a viral infection. (© Scott Camazine/Photo Researchers, Inc.)

Description

Sore throats can be either acute or chronic. Acute sore throats are the more common. They appear suddenly and last from three to about seven days. A chronic sore throat lasts much longer and is a symptom of an unresolved underlying condition or disease, such as a sinus infection.

Causes and symptoms

Sore throats have many different causes and may or may not be accompanied by cold symptoms, **fever**, or swollen lymph glands. Proper treatment depends on understanding the cause of the sore throat.

Viral sore throat

Viruses cause 90–95% of all sore throats. Cold and flu viruses are the main culprits. These viruses cause an inflammation in the throat and occasionally the tonsils (**tonsillitis**). Cold symptoms almost always accompany

a viral sore throat. These can include a runny nose, **cough**, congestion, hoarseness, **conjunctivitis**, and fever. The level of throat **pain** varies from uncomfortable to excruciating, when it is painful for the patient to eat, breathe, swallow, or speak.

Another group of viruses that cause sore throat are the adenoviruses. These may also cause infections of the lungs and ears. In addition to a sore throat, symptoms that accompany an adenovirus infection include cough, runny nose, white bumps on the tonsils and throat, mild **diarrhea**, **vomiting**, and a rash. The sore throat lasts about one week.

A third type of virus that can cause severe sore throat is the coxsackie virus. It can cause a disease called herpangina. Although anyone can get herpangina, it is most common in children up to age ten and is more prevalent in the summer or early autumn. Herpangina is sometimes called summer sore throat.

Three to six days after being exposed to the virus, an infected person develops a sudden sore throat that is accompanied by a substantial fever usually between 102–104°F (38.9–40°C). Tiny grayish-white blisters form on the throat and in the mouth. These fester and become small ulcers. Throat pain is often severe, interfering with swallowing. Children may become dehydrated if they are reluctant to eat or drink because of the pain. In addition, people with herpangina may vomit, have abdominal pain, and generally feel ill and miserable.

One other common cause of a viral sore throat is mononucleosis. Mononucleosis occurs when the **Epstein-Barr virus** infects one specific type of lymphocyte. The infection spreads to the lymphatic system, respiratory system, liver, spleen, and throat. Symptoms appear 30–50 days after exposure.

Mononucleosis, sometimes called the kissing disease, is extremely common. It is estimated that by the age of 35–40, 80–95% of Americans will have had mononucleosis. Often, symptoms are mild, especially in young children, and are diagnosed as a cold. Since symptoms are more severe in adolescents and adults, more cases are diagnosed as mononucleosis in this age group. One of the main symptoms of mononucleosis is a severe sore throat.

Although a runny nose and cough are much more likely to accompany a sore throat caused by a virus than one caused by a bacteria, there is no absolute way to tell what is causing the sore throat without a laboratory test. Viral sore throats are contagious and are passed directly from person to person by coughing and sneezing.

Bacterial sore throat

From 5–10% of sore throats are caused by bacteria. The most common bacterial sore throat results

from an infection by group A *Streptococcus*. This type of infection is commonly called strep throat. Anyone can get strep throat but it is most common in school age children.

Pharyngeal **gonorrhea**, a sexually transmitted bacterial disease, causes a severe sore throat. Gonorrhea in the throat is transmitted by having oral sex with an infected person.

Noninfectious sore throat

Not all sore throats are caused by infection. Post-nasal drip can irritate the throat and make it sore. It can be caused by hay fever and other **allergies** that irritate the sinuses. Environmental and other conditions, such as heavy **smoking** or breathing secondhand smoke, heavy alcohol consumption, breathing polluted air or chemical fumes, or swallowing substances that burn or scratch the throat can also cause pharyngitis. Dry air, like that in airplanes or from forced hot air furnaces, can make the throat sore. People who breathe through their mouths at night because of nasal congestion often get sore throats that improve as the day progresses. Sore throat caused by environmental conditions is not contagious.

Diagnosis

It is easy for people to tell if they have a sore throat but difficult to know what has caused it without laboratory tests. Most sore throats are minor and heal without any complications. A small number of bacterial sore throats do develop into serious diseases. Because of this, it is advisable to see a doctor if a sore throat lasts more than a few days or is accompanied by fever, **nausea**, or abdominal pain.

Diagnosis of a sore throat by a doctor begins with a **physical examination** of the throat and chest. The doctor will also look for signs of other illness, such as a sinus infection or **bronchitis**. Since both bacterial and viral sore throat are contagious and pass easily from person to person, the doctor will seek information about whether the patient has been around other people with flu, sore throat, colds, or strep throat. If it appears that the patient may have strep throat, the doctor will do laboratory tests.

If mononucleosis is suspected, the doctor may do a mono spot test to look for antibodies indicating the presence of the Epstein-Barr virus. The test is inexpensive, takes only a few minutes, and can be done in a physician's office. An inexpensive blood test can also determine the presence of antibodies to the mononucleosis virus.

Treatment

Effective treatment varies depending on the cause of the sore throat. As frustrating as it may be to the patient, viral sore throat is best left to run its course without drug treatment. Antibiotics have no effect on a viral sore throat. They do not shorten the length of the illness nor do they lessen the symptoms.

Sore throat caused by a streptococci or another bacteria must be treated with antibiotics. Penicillin is the preferred medication. Oral penicillin must be taken for 10 days. Patients need to take the entire amount of antibiotic prescribed even after symptoms of the sore throat improve. Stopping the antibiotic early can lead to a return of the sore throat. Occasionally a single injection of long-acting penicillin G is given instead of 10 days of oral treatment. These medications generally cost under \$15.

Because mononucleosis is caused by a virus, there is no specific drug treatment available. Rest, a healthy diet, plenty of fluids, limiting heavy **exercise** and competitive sports, and treatment of aches with **acetaminophen** (Datril, Tylenol, Panadol) or ibuprofen (Advil, Nuprin, Motrin, Medipren) will help the illness pass. Nearly 90% of mononucleosis infections are mild. The infected person does not normally get the disease again.

In the case of chronic sore throat, it is necessary to treat the underlying disease to heal the sore throat. If a sore throat caused by environmental factors, the aggravating stimulus should be eliminated from the sufferer's environment.

Home care for sore throat

Regardless of the cause of a sore throat, there are some home care steps that people can take to ease their discomfort. These include:

- taking acetaminophen or ibuprofen for pain; aspirin should not be given to children because of its association with increased risk for Reye's Syndrome, a serious disease
- gargling with warm double strength tea or warm salt water made by adding 1 tsp of salt to 8 oz (237 mL) of water
- drinking plenty of fluids, but avoiding acid juices like orange juice, which can irritate the throat (sucking on popsicles is a good way to get fluids into children)
- eating soft, nutritious foods like noodle soup and avoiding spicy foods
- refraining from smoking

KEY TERMS

Antigen—A foreign protein to which the body reacts by making antibodies

Conjunctivitis—An inflammation of the membrane surrounding the eye; also known as pinkeye.

Lymphocyte—A type of white blood cell. Lymphocytes play an important role in fighting disease.

Pharynx—The pharynx is the part of the throat that lies between the mouth and the larynx or voice box.

Toxin—A poison. In the case of scarlet fever, the toxin is secreted as a byproduct of the growth of the streptococcus bacteria and causes a rash.

- resting until the fever is gone, then resuming strenuous activities gradually
- a room humidifier may make sore throat sufferers more comfortable
- antiseptic lozenges and sprays may aggravate the sore throat rather than improve it

Alternative treatment

Alternative treatment focuses on easing the symptoms of sore throat using herbs and botanical medicines.

- Aromatherapists recommend inhaling the fragrances of essential oils of lavender (*Lavandula officinalis*), thyme (*Thymus vulgaris*), eucalyptus (*Eucalyptus globulus*), sage (*Salvia officinalis*), and sandalwood.
- Ayurvedic practitioners suggest gargling with a mixture of water, salt, and turmeric (*Curcuma longa*) powder or astringents such as alum, sumac, sage, and bayberry (*Myrica* spp.).
- Herbalists recommend taking osha root (*Ligusticum porteri*) internally for infection or drinking ginger (*Zingiber officinale*) or slippery elm (*Ulmus fulva*) tea for pain.
- Homeopaths may treat sore throats with superdilute solutions *Lachesis*, *Belladonna*, *Phytolacca*, yellow jasmine (*Gelsemium*), or mercury.
- Nutritional recommendations include zinc lozenges every two hours along with vitamin C with bioflavonoids, vitamin A, and beta-carotene supplements.

Prognosis

Sore throat caused by a viral infection generally clears up on its own within one week with no

complications. The exception is mononucleosis. 90% of cases of mononucleosis clear up without medical intervention or complications, so long as **dehydration** does not occur. In young children the symptoms may last only a week but in adolescents the symptoms last longer. Adults over age 30 have the most severe and long lasting symptoms. Adults may take up to six months to recover. In all age groups **fatigue** and weakness may continue for up to six weeks after other symptoms disappear.

In rare cases of mononucleosis, breathing may be obstructed because of swollen tonsils, adenoids, and lymph glands. If this happens, the patient should immediately seek emergency medical care.

Patients with bacterial sore throat begin feeling better about 24 hours after starting antibiotics. Untreated strep throat has the potential to cause **scarlet fever**, kidney damage, or **rheumatic fever**. Scarlet fever causes a rash and can cause high fever and convulsions. Rheumatic fever causes inflammation of the heart and damage to the heart valves. Taking antibiotics within the first week of a strep infection will prevent these complications. People with strep throat remain contagious until after they have been taking antibiotics for 24 hours.

Prevention

There is no way to prevent a sore throat; however, the risk of getting one or passing one on to another person can be minimized by:

- washing hands well and frequently
- avoiding close contact with someone who has a sore throat
- not sharing food and eating utensils with anyone
- not smoking
- staying out of polluted air

Resources

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Tish Davidson, A.M.
Karl Finley

Sotalol see **Antiarrhythmic drugs**

Sound therapy see **Music therapy**

South American blastomycosis

Definition

South American **blastomycosis** is a potentially fatal, chronic fungus infection that occurs more often in men. The infection may affect different parts of the body, including the lungs or the skin, and may cause ulcers of the mouth, voicebox, and nose.

Description

South American blastomycosis occurs primarily in Brazil, although cases crop up in Mexico, Central America, or other parts of South America. It affects men between ages 20 and 50 about 10 times more often than women.

The disease is far more serious than its North American variant (North American blastomycosis), which is endemic to the eastern United States, southern Canada, and the midwest.

South American blastomycosis is known medically as paracoccidioidal granuloma, or paracoccidioidomycosis. The infection has a very long incubation period (at least five years).

Causes and symptoms

South American blastomycosis is caused by the yeast-like fungus *Paracoccidioides brasiliensis* that is acquired by breathing in the spores of the fungus, which is commonly found in old wood and soil. It may appear very similar to **tuberculosis**; in fact, both diseases may infect a patient at the same time.

Symptoms include ulcers in the mouth, larynx and nose, in addition to large, draining lymph nodes, **cough**, chest **pain**, swollen lymph glands, weight loss, and lesions on the skin, genitals, and intestines. There may also be lesions in the liver, spleen, intestines, and adrenal glands.

Diagnosis

A physician can diagnose the condition by microscopic examination of a smear prepared from a lesion or sputum (spit). Biopsy specimens may also reveal the infection. While blood tests are helpful, they cannot

KEY TERMS

Amphotericin B—A drug used to treat fungal infections.

Sulfonamide drugs—A group of antibacterial drugs used to treat infections of the lungs and skin, among other things.

determine the difference between past and active infection.

Treatment

The primary goal of treatment is to control the infection. The best treatment has been amphotericin B. Sulfonamide drugs have been used and can stop the progress of the infection but they do not kill the fungus.

Scientists are studying new treatments for the fungal infection including ketoconazole, fluconazole, and itraconazole, which appear to be equally effective as amphotericin B, according to research.

Prognosis

The disease is chronic and often fatal. Because blastomycosis may be recurrent, patients should continue follow-up care for several years.

Prevention

There is no way to prevent the disease.

ORGANIZATIONS

National Institute of Allergies and Infectious Diseases, 6610 Rockledge Drive, MSC 6612, Bethesda, MD, 20892-6612, (301) 496-5717, (301) 402-3573, (866) 284-4107, ocpostoffice@niaid.nih.gov, <http://www.niaid.nih.gov>.

National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Carol A. Turkington

Space medicine see **Aviation medicine**

Spanish flu see **Influenza**

Spastic colitis see **Irritable bowel syndrome**

Spastic colon see **Irritable bowel syndrome**

Speech disorders

Definition

According to the American Speech-Language-Hearing Association (ASHA), a language disorder is an impairment in comprehension use of the spoken, written, or other symbol system.

Description

Speech disorders affect the language and mechanics, the content of speech, or the function of language in communication. Because speech disorders affect a person's ability to communicate effectively, every aspect of the person's life can be affected, for example, the person's ability to make friends, and to communicate at school or at work.

Amyotrophic lateral sclerosis (ALS)

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a neurological disease that attacks the nerve cells in the brain that control voluntary muscles. ALS causes motor neurons to die so that the brain and spinal cord are unable to send messages to the muscles telling them to move. Because the muscles are not functioning, they begin to atrophy. Muscles in the face and jaw can be affected, and thereby affecting a person's speech.

Aphasia

Aphasia results from damage to the language centers of the brain, which affects a person's ability to communicate through speaking, listening, and writing.

Persons with aphasia have trouble with expressive language, what is said, or receptive language, what is understood. Not only are speech and understanding speech affected, but also reading and writing is affected. The severity of aphasia varies from person to person, but in the most severe cases, a person may not be able to understand speech at all. Persons with mild aphasia may only become confused when speech becomes lengthy and complicated.

Developmental apraxia of speech

Developmental apraxia is a disorder that affects the nervous system and affects a person's ability to sequence and say sounds, syllables, and words. The brain does not send the correct messages to the mouth and jaw so that the person can say what he or she wants to say.

Children who are suffering from this disorder do not babble as an infant and first words are delayed.

Older children may have more difficulty with longer phrases and may appear to be searching for words to express a thought. Listeners will likely have a difficult time understanding the child.

Laryngeal cancer

Laryngeal cancer is characterized by a malignant growth in the larynx, or the voice box, which sometimes requires removal of the larynx or part of it.

Cancer anywhere in the throat affects speech, swallowing, and chewing. Depending on the size of the growth, a person may have trouble moving the mouth and lips. Therefore, speech sounds and eating will be affected and a person will have trouble communicating.

Orofacial myofunctional disorders

Orofacial myofunctional disorder (OMD) causes the tongue to move forward in an exaggerated manner while a person is speaking or swallowing. The tongue also may protrude when resting in the mouth.

Because heredity contributes to the size and shape of a person's mouth, there may be genetic reasons for the disorder. **Allergies** also affect the mouth and face muscles, which make it difficult to breathe because of nasal congestion. Because a person may sleep with the tongue protruding, lip muscles weaken. Enlarged tonsils also can block airways, creating the same breathing problems. Additionally, thumb-sucking, nail-biting, and teeth-clenching and grinding also can contribute to the disorder.

Stuttering

Stuttering is a disorder of speech fluency that frequently interrupts the flow of speech.

Because children typically stumble and confuse their words as speech develops, stuttering is not immediately evident. It is usually when children become older and continue to stumble that stuttering becomes evident.

Causes and symptoms

Amyotrophic lateral sclerosis (ALS)

Initial symptoms include weakness in any part of the body, and appendages begin to tire easily. Occasionally the disease affects only one appendage rather than both at the same time. Persons with ALS may have trouble maintaining balance and may stumble or have difficulty with tasks that require manual dexterity, such as buttoning a shirt or tying a shoe.

Eventually, the diaphragm and chest wall become so weak that a person cannot breathe on his or her own and needs the help of a ventilator. Because of the lack of muscle strength, a person with ALS will experience difficulty speaking loudly and clearly until the person is unable to speak at all using the vocal cords. The person will have difficulty pronouncing words and have difficulty completing lengthy sentences.

Along with the difficulty in speaking also comes difficulty in chewing and swallowing. Food can be broken down and pureed to make it easier to chew and swallow. However, a person eventually will have difficulty chewing and swallowing foods that are broken down or pureed. When ability to eat is affected, proper **nutrition** and body weight also are affected, and medical professionals may decide that it is best to put in a feeding tube.

Aphasia

Stroke is the most common cause of aphasia, although other injuries, such as a **brain tumor** or gunshot wound, also can cause aphasia.

Developmental apraxia of speech

Developmental apraxia is a disorder that affects the nervous system and affects a person's ability to sequence and say sounds, syllables, and words. The brain does not send the correct messages to the mouth and jaw so that the person can say what he or she wants to say.

Children who are suffering from this disorder don't babble as an infant and first words are delayed. Older children may have more difficulty with longer phrases, and may appear to be searching for words to express a thought. Listeners will likely have a difficult time understanding the child.

There is no known cause for developmental apraxia of speech. Symptoms include weakness of the jaw, tongue, and lips, and delayed speech development. Persons with the disorder also may have trouble identifying an object in the mouth using the sense of touch, which is known as oral-sensory perception.

Laryngeal cancer

Any kind of **smoking** of cigarettes, cigars, or tobacco and alcohol **abuse** contribute to oral cancer, including smokeless tobacco. Persons with laryngeal cancer or another type of oral cancer may have a red or white patch or lump in the mouth. Symptoms also include difficulty chewing, swallowing, or chewing.

Stuttering

There is no known cause for stuttering, although poor muscle coordination and the rate of language development are believed to contribute to it.

Stuttering is characterized by repetition of sounds, syllables, portions of a word, words, and complete phrases; stretching the sounds and syllables; hesitation between words; words spoken in spurts; tense muscles in the jaw and mouth; and a feeling of loss of control.

Diagnosis

Amyotrophic lateral sclerosis (ALS)

About 20,000 people in the United States have ALS at any given time with 5,000 new cases diagnosed every year. ALS is in the same family of disorders as **multiple sclerosis**, Parkinson's disease, and **muscular dystrophy**. Persons of all races and ethnic groups are afflicted by the disease, although men are more likely to have it than women.

Aphasia

About 700,000 persons in the United States have strokes every year, and 1 million are estimated to have aphasia.

Developmental apraxia of speech

A child suspected to have apraxia should first have his or her hearing tested to determine if the child has any deafness. Muscle development in the face and jaw should be evaluated and speech exercises tested. Articulation of words should be tested as well as the person's expressive and receptive language skills.

Laryngeal cancer

It is likely that a dentist or physician will first detect signs of possible cancer. Oral cancer makes up about 2–5% of all cancers, and about 30,000 cases are diagnosed each year. Twice as many men than women are diagnosed with cancer typically between the ages of 50 and 70.

Orofacial myofunctional disorders

The diagnosis of orofacial myofunctional disorder affects speech sounds because of weak tongue tip muscles, although a person's speech may not be affected at all.

Stuttering

Stuttering is a problem that most likely will manifest itself during childhood rather than adulthood.

Treatment

Amyotrophic lateral sclerosis (ALS)

In addition to treatments such as a feeding tube, a person with ALS would likely enlist the help of a speech therapist to help him or her determine ways in which he or she can maintain vocal control. A person also may enlist the help of an occupational therapist, a medical professional trained to help persons who have trouble with activities of daily living such as dressing, bathing, and eating.

Aphasia

A speech-language pathologist can perform drills and exercises with a person that include practice in naming objects and following directions to try to improve skills. The person learns the best way to express himself or herself. **Group therapy** also is an option, which focuses on structured discussions.

Developmental apraxia of speech

Treatment should focus on the coordination of motor movements necessary during speech production, which includes controlling breathing. A speech-language pathologist teaches exercises to a person with apraxia that will strengthen the jaws, lips, and tongue to improve coordination during speech. The therapist uses tactile, auditory, and visual feedback to direct the brain to move the muscles used during speech.

Laryngeal cancer

Depending on when the cancer is first detected, and depending on the size of the cancer, the entire larynx may not need to be removed. Radiation, **chemotherapy**, or partial removal can be done in lieu of complete removal. In these cases, the voice may be preserved although the quality likely will be affected.

Orofacial myofunctional disorders

In cases where speech is affected, a speech pathologist should be consulted to help control breathing problems and work on speech articulation. The lip, palate, tongue, and facial muscles should be evaluated so that errors in speech can be detected. Therapy includes increasing awareness of the mouth and facial muscles, as well as the posture of the mouth and tongue. Muscle **exercise** can be done to increase strength and control.

Stuttering

A treatment plan by a speech therapist includes improving fluency and ease with which a person speaks. Strategies include reducing the rate of speech and using slower speech movements; articulating

lightly; and starting air flow for speech before any other muscle movement.

Alternative treatment

Developmental apraxia of speech

Some persons with apraxia may decide to use alternative communication systems, such as a computer that transcribes and “speaks” what a person is directing it to say. These augmentative systems should only be used when a person is so severely impaired that effective speech or communication isn’t possible.

Laryngeal cancer

In cases of a full **laryngectomy**, a hole is made in the neck and, rather than using the mouth and nose to talk and breath, the person must use the hole.

Once the larynx is removed, the person needs to develop a new speech system without a voice. A speech pathologist should follow one of three plans: esophageal speech, artificial larynx, or tracheoesophageal puncture (TEP).

- Esophageal speech. Without a larynx, a person is no longer able to exhale air from the lungs through the mouth to speak. Using esophageal speech, the person inhales and traps the air in the throat, causing the esophagus to vibrate and create sound.
- Artificial larynx. A mechanical instrument can be used that produces sound for some speech. These devices can be held against the neck or used by inserting a tube in the mouth.
- Tracheoesophageal puncture. This is a popular method in restoring speech production. During surgery, a hole is made between the trachea and esophagus and a valve is inserted into the hole. The person breathes air into the lungs and then covers the hole in the throat. During exhalation, the esophagus vibrates and creates speech.

Stuttering

A person suffering from stuttering may employ distraction strategies to help him or her stop stuttering. Typically, a person stuttering becomes frustrated and embarrassed; subsequently, encouraging the person to think of something or do something else may break the stuttering cycle.

Prognosis

Amyotrophic lateral sclerosis (ALS)

ALS patients often die of **respiratory failure** within three to five years of being diagnosed, although

KEY TERMS

Neurons—Nerve cells in the brain, brain stem, and spinal cord that connect the nervous system and the muscles.

some persons have been known to survive as many as 10 years or longer.

Aphasia

Persons with aphasia can improve and eventually function in more typical public settings, and possibly return to school or work.

Developmental apraxia of speech

With proper treatment, apraxia can be brought under control and the person will be able to function normally as an adult.

Laryngeal cancer

Full removal of the larynx removes the risk of a cancer relapse, although other parts of the throat and mouth can be affected.

Orofacial myofunctional disorders

A person can learn to control this disorder with proper treatment and maintain normal speech and breathing patterns.

Stuttering

With proper **speech therapy**, stuttering can be controlled or eliminated.

Prevention

Laryngeal cancer

Persons should not engage in smoking or drug abuse to decrease the risk of oral cancer.

Orofacial myofunctional disorders

In cases where the cause is evident, such as allergies or enlarged tonsils, a person should first remedy that problem; perhaps have the tonsils removed and treat allergies with medication.

Resources

BOOKS

Damico, Jack Samuel, Nicole Muller, and Martin J Ball. *The Handbook of Language and Speech Disorders*. Chichester, UK; Malden, MA: Wiley-Blackwell, 2010.

ORGANIZATIONS

American Speech Language Hearing Association, 2200 Research Boulevard , Rockville, MD, 20850-3289, (301) 296-5700, (301) 296-8580, (800) 638-8255, actioncenter @asha.org, http://asha.org/.

Meghan Gourley

Speech disturbance see **Aphasia**

Speech therapy

Definition

Speech therapy is the diagnosis and treatment of a speech disorder, expressive or receptive language disorder, or certain **swallowing disorders** by a trained speech-language pathologist (SLP). SLPs commonly are called speech therapists.

Purpose

The purpose of speech therapy is to improve communication and/or the understanding of language and/or to remediate swallowing difficulties. In addition to oral communication, speech therapy may include sign language, picture communication, and the use of assistive devices to help augment speech or serve as an alternate form of communication (augmented and alternative communication [AAC]).

Demographics

According to the Bureau of Labor Statistics, in 2008 there were about 119,300 practicing SLPs in the United States, with the number expected to grow to about 141,400 by 2018. Of these, about 48% practice in schools, 9% are self-employed in private practice, and the remainder practice at hospitals, nursing homes, other health care facilities, or provide in-home health care services, usually through Medicare and Medicaid programs.

Description

Speech therapy addresses problems with speech production, language disorders, and swallowing. Problems with speech production include issues of articulation, speech rhythm, fluency, voice production, resonance, tone, and accent.

There are two basic categories of language disorders: expressive language disorders, which involves problems producing language and receptive language

disorders, which involves problems understanding language. Individuals with expressive language disorders have difficulty in using language at the level expected for their age group. Often children with this type of disorder have lower than expected vocabularies, form sentences with a simpler structure than is expected, and have more difficulties expressing themselves in writing than other children their age. In some cases, (e.g. after a **stroke**) only one area will show a deficiency, such as an individual who has a good vocabulary but difficulty forming complex sentences. In other cases, all areas of language production are affected.

Individuals with receptive language disorders have difficulty understanding and processing language. This can affect comprehension of spoken or written language, or both. People who have difficulty understanding and following directions, responding to questions, or following a conversation may have a receptive language disorder. Individuals who can read words on the page but are unable to process the meaning of what they read have a receptive language disorder.

Speech therapy also addresses problems of swallowing that originate in the mouth and throat. Infants with **birth defects** of the mouth and individuals who have had a stroke or who have certain diseases such as **multiple sclerosis** are most likely to have swallowing problems.

Speech therapy is individualized. SLPs use a variety of techniques to overcome speech and language disorders based on the age of the client and the type of problem. Much work with children involves playing with them and using various toys and visual aids to encourage them to speak, along with modeling correct articulation and speech patterns. Therapy may be done one-on-one or with small groups of children. With older children and adults, the SLP may use a mirror to help the individual see how to move the muscles of their face to correctly form certain sounds. Exercises to strengthen certain facial muscles may be prescribed for people who have trouble swallowing. For individuals who have disabilities such as deafness or **cerebral palsy**, the SLP may teach the individual how to use assistive devices or alternate communication methods such as sign language.

Depending on the problems being addressed, the individual may have a speech therapy sessions as infrequently as once a week or as often as every day. There is no standard length of time an individual remains in speech therapy. Practice is the key to making progress with speech and language disorders. Parents and caregivers play an important role in the success of speech

KEY TERMS

Articulation—The ability to pronounce a word correctly. A lisp is an example of an articulation problem.

Dysphagia—Difficult or painful swallowing.

Fluency—The ability to produce a flow of words and not get “stuck.” Stuttering is an example of a fluency problem.

Stroke—Irreversible damage to the brain caused by insufficient blood flow to the brain as the result of a blocked artery. Damage can include loss of speech or vision, paralysis, cognitive impairment, and death.

therapy, as they usually are expected to both model good articulation and speech, to reinforce the lessons taught during therapy sessions, and to supervise practice of any home exercises the SLP prescribes.

Benefits

Speech therapy improves expressive communication and language understanding and can reduce or eliminate certain swallowing problems. Improved communication leads to reduced frustration and greater safety, both for language-impaired individuals and for those who live and work with them.

Precautions

As of 2009, 47 states in the United States had licensing requirements for SLPs. When choosing a speech therapist, families should make sure that the therapist is licensed. Not only is this a sensible precaution to assure appropriate therapy, but many government programs and private insurers will not pay for speech therapy performed by an unlicensed individual.

Many SLPs develop specialties, such as working with stroke victims, working with preschoolers, or working with autistic individuals, so it is important to find a speech therapist who has experience in the specific speech and language problem being treated. SLPs also should be willing to work closely with healthcare personnel and educators, as needed, to ensure maximum benefit from the therapy.

Preparation

No special preparation is needed to begin speech therapy. An evaluation of the speech-language-swallowing problem will be done at the first session.

Aftercare

Practice and repetition are key to the success of speech therapy. Individuals often are given speech, language and/or muscle exercises to perform regularly at home. Caregivers will need to supervise these exercises and reinforce what is accomplished at the therapy sessions.

Risks

No specific risks are associated with speech therapy.

Training and certification

Certification varies from country to country and in the United States, from state to state. All SLPs are required to have a college degree. In the United States, most states require a master's degree in speech-language pathology from an accredited college or university, passing a national examination offered through the Praxis Series of the Educational Testing Service, and a minimum of 300 hours of supervised clinical experience. Most states require the SLP to earn continuing education credits to maintain their license. The American Speech-Language-Hearing Association (ASHA) offers a voluntary Certificate of Clinical Competence (CCC), which imposes higher standards than the general standards mentioned above. Individuals who earn this certificate will indicate it by putting the letters CCC-SLP after their name. The ASHA also offers voluntary certification in certain speech-language therapy specialties. Certification and licensing requirements are quite similar in other countries in the English-speaking world.

Resources

BOOKS

Feit, Debbie. *The Parent's Guide to Speech and Language Problems*. New York: McGraw-Hill, 2007.

PERIODICALS

“Screening for Speech and Language Delay in Preschool Children: Recommendation Statement.” *Pediatrics* v117 i2, (February 2006, pp. 497–502).

OTHER

“Self-Help Groups for Speech, Language, and Swallowing Disorders.” American Speech-Language-Hearing Association Undated. [Accessed January 10, 2010]. http://www.asha.org/public/speech/speech_self-help.htm.

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 Speech Therapy Web. [Accessed January 10, 2010]. <http://www.speechtherapyweb.com>.

ORGANIZATIONS

American Speech-Language-Hearing Association (ASHA),
 2200 Research Boulevard, Rockville, MD, 20850-3289,
 (301) 296-5700, TTY (301) 296-5650, (800) 638-8255,
 (301) 296-8580, actioncenter@asha.org, <http://www.asha.org>.

National Institute on Deafness and Other Communication Disorders, 31 Center Drive, MSC 2320, Bethesda, MD, 20892-2320, nidcdinfo@nidcd.nih.gov, <http://www.nidcd.nih.gov>.

Stuttering Foundation of America, 3100 Walnut Grove Road, Suite 603; P.O. Box 11749, Memphis, TN, 38111-0749, (901) 452-7343, (800) 992-9392, (901) 452-3931, info@stutteringhelp.org, <http://www.stutteringhelp.org>.

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 Brenda Lerner

Sperm count see **Semen analysis**

Spina bifida

Definition

Spina bifida belongs to a group of disorders known as neural tube defects (NTDs). It is a serious birth abnormality characterized by the incomplete development of the brain, spinal cord, and/or meninges.

Demographics

According to the National Institute of Neurological Disorders and Stroke (NINDS), spina bifida is the most common neural tube defect (NTD) in the United States, affecting 1,500 to 2,000 of the more than 4 million babies born each year. The Center for Disease Control (CDC) reports that NTDs are more common among white women than black women and more common among Hispanic women than non-Hispanic women.

Spina bifida occurs worldwide but there has been a steady downward trend in occurrence rates over the past 50–70 years, particularly in regions of high prevalence. The highest prevalence rates, about one in 200 pregnancies, have been reported from certain northern



An infant with spina bifida. (Biophoto Associates/Photo Researchers, Inc.)

provinces in China. Intermediate prevalence rates, about one in 1,000 pregnancies, have been found in Central and South America. The lowest prevalence rates, less than one in 2,000 pregnancies, have been found in the European countries.

Description

Spina bifida is also known by the name spinal dysraphism. Spina bifida may appear in the body midline anywhere from the neck to the buttocks. In its most severe form, termed spinal rachischisis, the entire spinal canal is open, exposing the spinal cord and nerves. More commonly, the abnormality appears as a localized mass on the back that is covered by skin or by the meninges, the three-layered membrane that surrounds the spinal cord. Spina bifida is usually readily apparent at birth because of the malformation of the back and **paralysis** below the level of the abnormality.

Various forms of spina bifida are known as meningocele, myelomeningocele, spina bifida aperta, open spina bifida, myelodysplasia, spinal dysraphism, spinal rachischisis, myelocele, and meningocele. The term meningocele is used when the spine malformation contains only the protective covering (meninges) of the spinal cord. The other terms indicate involvement of the spinal cord and nerves in the malformation. A related term, spina bifida occulta, indicates that one or more of the bony bodies in the spine are incompletely hardened but that there is no abnormality of the spinal cord itself.

Risk factors

In the United States, 95% of neural tube defects (NTDs) occur in women with no family history of these conditions. The CDC outlines some of the risk factors associated with NTDs and by extension, with spina bifida. They include the occurrence of a previous NTD-affected **pregnancy**, or of maternal insulin-dependent diabetes as well as the use of certain anticonvulsant medications (such as Valproic acid/Depakene, and Carbamazepine/Tegretol). Medically diagnosed **obesity** is also considered a risk factor. The recurrence risk after the birth of an infant with isolated spina bifida is 3–5%. Recurrence may be for spina bifida or another type of spinal abnormality.

Causes and symptoms

Spina bifida occurs because the neural tube, around the area of the spine, fails to close during fetal development. Spina bifida may occur as an isolated abnormality or in the company of other malformations. As an isolated abnormality, spina bifida is

caused by the combination of genetic factors and environmental influences that bring about malformation of the spine and spinal column. The specific genes and environmental influences that contribute to the many-factored causes of spina bifida are not precisely known. An insufficiency of **folic acid** is known to be one influential nutritional factor. Changes (mutations) in genes involving the metabolism of folic acid are believed to be significant genetic risk factors.

Spina bifida may arise because of chromosome abnormalities, single gene mutations, or specific environmental insults such as maternal **diabetes mellitus** or prenatal exposure to certain **anticonvulsant drugs**. The recurrence risk varies with each of these specific causes.

In most cases, spina bifida is obvious at birth because of malformation of the spine. The spine may be completely open, exposing the spinal cord and nerves. More commonly, the spine abnormality appears as a mass on the back covered by membrane (meninges) or skin. Spina bifida may occur anywhere from the base of the skull to the buttocks. About 75% of abnormalities occur in the lower back (lumbar) region. In rare instances, the spinal cord malformation may occur internally, sometimes with a connection to the gastrointestinal tract.

In spina bifida, many complications arise, dependent in part on the level and severity of the spine malformation. As a rule, the nerves below the level of the abnormality develop in a faulty manner and fail to function, resulting in paralysis and loss of sensation below the level of the spine malformation. Since most abnormalities occur in the lumbar region, the lower limbs are paralyzed and lack sensation. Furthermore, the bowel and bladder have inadequate nerve connections, causing an inability to control bowel and bladder function. Most infants also develop hydrocephaly, an accumulation of excess fluid in the four cavities of the brain. At least one of every seven cases develop findings of Chiari II malformation, a condition in which the lower part of the brain is crowded and may be forced into the upper part of the spinal cavity.

There are a number of mild variant forms of spina bifida, including multiple vertebral abnormalities, skin dimples, tufts of hair, and localized areas of skin deficiency over the spine. Two variants, lipomeningocele and lipomyelomeningocele, typically occur in the lower back area (lumbar or sacral) of the spine. In these conditions, a tumor of fatty tissue becomes isolated among the nerves below the spinal cord, which may result in tethering of the spinal cord and complications similar to those with open spina bifida.

KEY TERMS

Anticonvulsant—Group of medications used in the treatment of epileptic seizures.

Chiari II anomaly—A structural abnormality of the lower portion of the brain (cerebellum and brain stem) associated with spina bifida. The lower structures of the brain are crowded and may be forced into the foramen magnum, the opening through which the brain and spinal cord are connected.

Fetus—The term used to describe a developing human infant from approximately the third month of pregnancy until delivery. The term embryo is used prior to the third month.

Folic acid—One of the B vitamins important for healthy growth of the fetus. It is essential to the normal development of a baby's spine, brain and skull, especially during the first four weeks of pregnancy.

Hydrocephalus—The excess accumulation of cerebrospinal fluid around the brain, often causing enlargement of the head.

Meninges—The protective covering around the brain and spinal cord.

Neural tube defect (NTD)—Defective neural tube, the narrow sheath that closes to form the brain and spinal cord of the embryo.

Diagnosis

Examination

Few disorders are to be confused with open spina bifida. The diagnosis is usually obvious based on the external findings at birth. Paralysis below the level of the abnormality and fluid on the brain (hydrocephaly) may contribute to the diagnosis. Other spine abnormalities such as congenital **scoliosis** and **kyphosis**, or soft tissue tumors overlying the spine, are not likely to have these accompanying findings. In cases in which there are no external findings, the diagnosis is more difficult and may not become evident until neurological abnormalities or hydrocephaly develop weeks, months, or years following birth.

Tests

Prenatal diagnosis may be made in most cases with ultrasound examination after 12–14 weeks of pregnancy. Ultrasounds cannot identify every structural problem in a developing baby, so some cases of spina bifida (especially mild forms) may be missed. However, it is a risk-free method to use that gives immediate results.

Prenatal blood screening is often offered to women between 15 and 21 weeks in a pregnancy. This screening measures the levels of various chemicals naturally found in a mother's blood, including alpha-fetoprotein (AFP). For this reason, the screening is often called AFP screening. AFP is a protein normally made by a developing fetus, so it is naturally present in maternal serum and called MS-AFP. When a fetus has spina bifida, the levels of MS-AFP may be higher than usual because it leaks out of the hole in the

spine. If a woman's AFP screen comes back abnormal with a high MS-AFP value, she often is at a higher risk for having a baby with spina bifida. This may prompt her physician to offer her a detailed ultrasound, as well as other medical options that might give her more information about the baby.

Once spina bifida is seen outwardly, imaging scans like x-rays, ultrasound, **magnetic resonance imaging** (MRI), or computed tomography (CT) can be helpful to see the extent of it. It is also a good way to identify whether someone has associated neurological complications like **hydrocephalus**.

Some **genetic testing**, like chromosome studies, may identify a diagnosis or cause for the spina bifida. Abnormal genetic test results cannot be changed or reversed, but may provide answers about why the spina bifida occurred.

Procedures

One option to find spina bifida is a procedure called **amniocentesis**. Amniocentesis involves removing a small amount of fluid from around the baby, using a fine needle. This fluid naturally contains AFP, which may also be elevated if the baby has spina bifida. There is a small risk of **miscarriage**, about 1 in 200, with this procedure. As such, every women usually receives proper counseling through their doctor or a genetic counselor before having the test done.

Treatment

As of 2009, there is no known cure for spina bifida. Treatment primarily focuses on dealing with symptoms as they arise, since they vary so greatly from person to person.

Traditional

Aggressive surgical and medical management have improved the survival and function of infants with spina bifida. Initial surgery may be carried out during the first days of life, providing protection against injury and infection. Subsequent surgery is often necessary to protect against excessive curvature of the spine, and in the presence of hydrocephaly, to place a mechanical shunt to decrease the pressure and amount of cerebrospinal fluid in the cavities of the brain. Because of weakness or paralysis below the level of the spine abnormality, most children will require **physical therapy**, bracing, and other orthopedic assistance to enable them to walk. A variety of approaches including periodic bladder catheterization, surgical diversion of urine, and **antibiotics** are used to protect urinary function.

Although most individuals with spina bifida have normal intellectual function, learning disabilities or mental impairment has occurred. This may result, in part, from hydrocephaly and/or infections of the nervous system. Children so affected may benefit from early educational intervention, physical therapy, and **occupational therapy**. Counseling to improve self-image and lessen barriers to socialization becomes important in late childhood and adolescence.

Drugs

Medications are widely available to treat those who develop seizures and these may need periodic adjustments. Those who have problems with bowel or bladder control may also require medications.

Alternative

Open fetal surgery has been performed for spina bifida during the last half of pregnancy. After direct closure of the spine malformation, the fetus is returned to the womb. By preventing chronic intrauterine exposure to mechanical and chemical trauma, **prenatal surgery** improves neurological function and leads to fewer complications after birth. Fetal surgery is considered experimental, and results have been mixed.

Prognosis

Prognosis in spina bifida is extremely varied and unpredictable. Years ago with far less intervention and fewer treatments available, someone with severe spina bifida had a high chance to die from complications. Today, more than 80% of infants born with spina bifida survive with surgical and medical management. Although complications from paralysis, hydrocephaly, Chiari II malformation, and urinary tract deterioration threaten the well-being of the survivors, the outlook for normal intellectual function is good.

Prevention

Prevention of isolated spina bifida and other spinal abnormalities has become possible during recent decades. The major prevention is through the use of a B vitamin, folic acid, for several months prior to and following conception. The CDC and Prevention recommend the intake of 400 micrograms of synthetic folic acid every day for all women of childbearing years.

Resources

BOOKS

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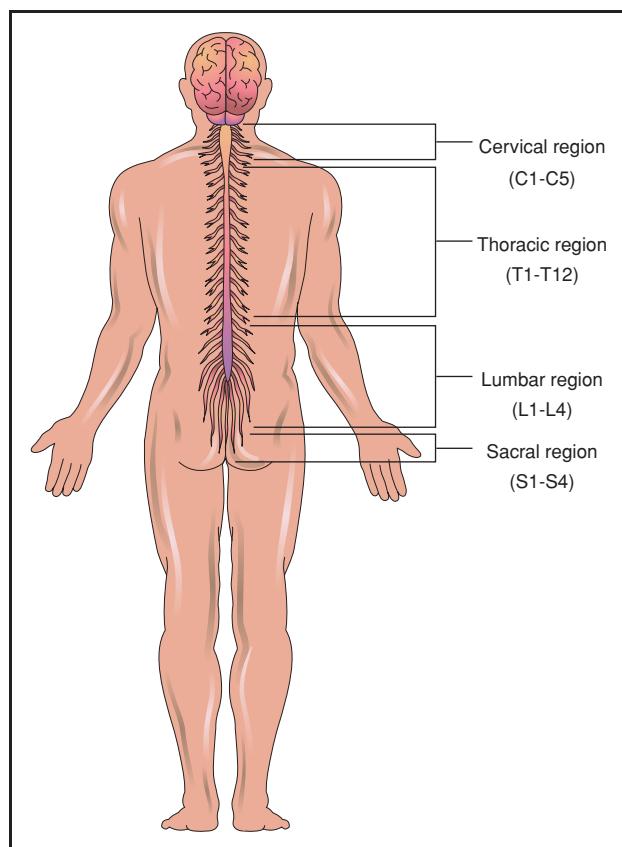
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ORGANIZATIONS

- Disabled Sports USA, 451 Hungerford Drive, Suite 100, Rockville, MD, 20850, (301) 217-0960, (301) 217-0968, dsusa@dsusa.org, <http://www.dsusa.org>.
- March of Dimes Foundation, 1275 Mamaroneck Avenue, White Plains, NY, 10605, (914) 428-7100, (888) MODIMES, (914) 428-8203, askus@marchofdimes.com, <http://www.marchofdimes.com>.
- National Dissemination Center for Children with Disabilities (NICHCY), PO Box 1492, Washington, DC, 20013-1492, (800) 695-0285, (202) 884-8441, nichcy@aed.org, <http://www.nichcy.org>.
- National Institute of Neurological Disorders and Stroke (NINDS), PO Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov>.
- Spina Bifida Association of America, 4590 MacArthur Blvd. NW, Suite 250, Washington, DC, 20007-4266, (202) 944-3285, (800) 621-3141, (202) 944-3295, sbaa@sbaa.org, <http://www.spinabifidaassociation.org>.

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Spina bifida occulta see **Spina bifida**



The extent of sensory and motor loss resulting from a spinal cord injury depends on the level of the injury because nerves at different levels control sensation and movement in different parts of the body. The distribution is as follows: C1–C4: head and neck; C3–C5: diaphragm; C5–T1: shoulders, arms, and hands; T2–T12: chest and abdomen (excluding internal organs); L1–L4: abdomen (excluding internal organs), buttocks, genitals, upper legs; L4–S3: legs; S2–S4: genitals, muscles of the perineum. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

Spinal cord injury

Definition

Spinal cord injury is damage to the spinal cord that causes loss of sensation and motor control.

Description

Approximately 10,000 new spinal cord injuries (SCIs) occur each year in the United States. About 250,000 people are currently affected. Spinal cord injuries can happen to anyone at any time of life. The typical patient, however, is a man between the ages of 19 and 26, injured in a motor vehicle accident (about 50% of all SCIs), a fall (20%), an act of violence (15%), or a sporting accident (14%). Alcohol or other drug abuse plays an important role in a large percentage of all spinal cord injuries. Six percent of people who receive injuries to the lower spine die within a year and 40% of people who receive the more frequent higher injuries die within a year.

Short-term costs for hospitalization, equipment, and home modifications are approximately \$140,000 for an SCI patient capable of independent living. Lifetime costs may exceed \$1 million. Costs may be three to four times higher for the SCI patient who needs long-term institutional care. Overall costs to the American economy in direct payments and lost productivity are more than \$10 billion per year.

Causes and symptoms

Causes

The spinal cord is about as big around as the index finger. It descends from the brain down the back through hollow channels of the backbone. The spinal cord is

made of nerve cells (neurons). The nerve cells carry sensory data from the areas outside the spinal cord (periphery) to the brain and they carry motor commands from brain to periphery. Peripheral neurons are bundled together to make up the 31 pairs of peripheral nerve roots. The peripheral nerve roots enter and exit the spinal cord by passing through the spaces between the stacked vertebrae. Each pair of nerves is named for the vertebra from which it exits. These are known as:

- C1–8. These nerves enter from the eight cervical or neck vertebrae.
- T1–12. These nerves enter from the thoracic or chest vertebrae.
- L1–5. These nerves enter from the lumbar vertebrae of the lower back.
- S1–5. These nerves enter through the sacral or pelvic vertebrae.
- Coccygeal. These nerves enter through the coccyx or tailbone.

Peripheral nerves carry motor commands to the muscles and internal organs and they carry sensations from these areas and from the body's surface. (Sensory data from the head, including sight, sound, smell, and taste, do not pass through the spinal cord and are not affected by most SCIs.) Damage to the spinal cord interrupts these signals. The interruption damages motor functions that allow the muscles to move, sensory functions such as feeling heat and cold, and autonomic functions such as urination, sexual function, sweating, and blood pressure.

Spinal cord injuries most often occur where the spine is most flexible, in the regions of C5–C7 of the neck, and T10–L2 at the base of the rib cage. Several physically distinct types of damage are recognized. Sudden and violent jolts to nearby tissues can jar the cord. This jarring causes a temporary spinal **concussion**. Concussion symptoms usually disappear completely within several hours. A spinal contusion or bruise is bleeding within the spinal column. The pressure from the excess fluid may kill spinal cord neurons. Spinal compression is caused by some object, such as a tumor, pressing on the cord. Lacerations or tears cause direct damage to cord neurons. Lacerations can be caused by bone fragments or missiles such as bullets. Spinal transection describes the complete severing of the cord. Most spinal cord injuries involve two or more of these types of damage.

Symptoms

PARALYSIS AND LOSS OF SENSATION. The extent to which movement and sensation are damaged depends on the level of the spinal cord injury. Nerves leaving

the spinal cord at different levels control sensation and movement in different parts of the body. The distribution is roughly as follows:

- C1–C4: head and neck.
- C3–C5: diaphragm (chest and breathing).
- C5–T1: shoulders, arms and hands.
- T2–T12: chest and abdomen (excluding internal organs).
- L1–L4: abdomen (excluding internal organs), buttocks, genitals, and upper legs.
- L4–S1: legs.
- S2–S4: genitals and muscles of the perineum.

Damage below T1, which lies at the base of the rib cage, causes **paralysis** and loss of sensation in the legs and trunk below the injury. Injury at this level usually does no damage to the arms and hands. Paralysis of the legs is called paraplegia. Damage above T1 involves the arms as well as the legs. Paralysis of all four limbs is called quadriplegia or tetraplegia. Cervical or neck injuries not only cause quadriplegia but also may cause difficulty in breathing. Damage in the lower part of the neck may leave enough diaphragm control to allow unassisted breathing. Patients with damage at C3 or above, just below the base of the skull, require mechanical assistance to breathe.

Symptoms also depend on the extent of spinal cord injury. A completely severed cord causes paralysis and loss of sensation below the wound. If the cord is only partially severed, some function will remain below the injury. Damage limited to the front portion of the cord causes paralysis and loss of sensations of **pain** and temperature. Other sensation may be preserved. Damage to the center of the cord may spare the legs but paralyze the arms. Damage to the right or left half causes loss of position sense, paralysis on the side of the injury, and loss of pain and temperature sensation on the opposite side.

DEEP VENOUS THROMBOSIS. Blood does not flow normally to a paralyzed limb that is inactive for long periods. The blood pools in the deep veins and forms clots, a condition known as **deep vein thrombosis**. A clot or thrombus can break free and lodge in smaller arteries in the brain, causing a **stroke**, or in the lungs, causing **pulmonary embolism**.

PRESSURE ULCERS. Inability to move also leads to pressure ulcers or bed sores. Pressure ulcers form where skin remains in contact with a bed or chair for a long time. The most common sites of pressure ulcers are the buttocks, hips, and heels.

SPASTICITY AND CONTRACTURE. A paralyzed limb is incapable of active movement, but the muscle still has tone, a constant low level of contraction. Normal muscle tone requires communication between the muscle and the brain. Spinal cord injury prevents the brain from telling the muscle to relax. The result is prolonged muscle contraction or spasticity. Because the muscles that extend and those that bend a joint are not usually equal in strength, the involved joint is bent, often severely. This constant pressure causes deformity. As the muscle remains in the shortened position over several weeks or months, the tendons remodel and cause permanent muscle shortening or contracture. When muscles have permanently shortened, the inner surfaces of joints, such as armpits or palms, cannot be cleaned and the skin breaks down in that area.

HETEROtopic OSSIFICATION. Heterotopic ossification is an abnormal deposit of bone in muscles and tendons that may occur after injury. It is most common in the hips and knees. Initially heterotopic ossification causes localized swelling, warmth, redness, and stiffness of the muscle. It usually begins one to four months after the injury and is rare after one year.

AUTONOMIC DYSREFLEXIA. Body organs that regulate themselves, such as the heart, gastrointestinal tract, and glands, are controlled by groups of nerves called autonomic nerves. Autonomic nerves emerge from three different places: above the spinal column, in the lower back from vertebrae T1–L4, and from the lowest regions of the sacrum at the base of the spine. In general, these three groups of autonomic nerves operate in balance. Spinal cord injury can disrupt this balance, a condition called autonomic dysreflexia or autonomic hyperreflexia. Patients with injuries at T6 or above are at greatest risk.

In autonomic dysreflexia, irritation of the skin, bowel, or bladder causes a highly exaggerated response from autonomic nerves. This response is caused by the uncontrolled release of norepinephrine, a hormone similar to adrenaline. Uncontrolled release of norepinephrine causes a rapid rise in blood pressure and a slowing of the heart rate. These symptoms are accompanied by throbbing **headache**, **nausea**, **anxiety**, sweating, and goose bumps below the level of the injury. The elevated blood pressure can rapidly cause loss of consciousness, seizures, cerebral hemorrhage, and **death**. Autonomic dysreflexia is most often caused by an overfull bladder or bladder infection, impaction or hard impassable fecal mass in the bowel, or skin irritation from tight clothing, **sunburn**, or other irritant. Inability to sense these irritants before the autonomic reaction begins is a major cause of dysreflexia.

LOSS OF BLADDER AND BOWEL CONTROL. Bladder and bowel control require both motor nerves and the autonomic nervous system. Both of these systems may be damaged by SCI. When the autonomic nervous system triggers an urge to urinate or defecate, continence is maintained by contracting the anal or urethral sphincters. A sphincter is a ring of muscle that contracts to close off a passage or opening in the body. When the neural connections to these muscles are severed, conscious control is lost. In addition, loss of feeling may prevent sensations of fullness from reaching the brain. To compensate, the patient may help empty the bowel or bladder by using physical maneuvers that stimulate autonomic contractions before they would otherwise begin. However, the patient may not be able to relax the sphincters. If the sphincters cannot be relaxed, the patient will retain urine or feces.

Retention of urine may cause muscular changes in the bladder and urethral sphincter that make the problem worse. **Urinary tract infection** is common. Retention of feces can cause impaction. Symptoms of impaction include loss of appetite and nausea. Untreated impaction may cause perforation of the large intestine and rapid overwhelming infection.

SEXUAL DYSFUNCTION. Men who have sustained SCI may be unable to achieve an erection or ejaculate. Sperm formation may be abnormal too, which reduces fertility. Fertility and the ability to achieve orgasm are less impaired for women. Women may still be able to become pregnant and deliver vaginally with proper medical care.

Diagnosis

The location and extent of spinal cord injury is determined with **computed tomography scans** (CT scans), **magnetic resonance imaging** (MRI) scans, and x rays. X rays may be enhanced with an injected contrast dye.

Treatment

A person who may have a spinal cord injury should not be moved. Treatment of SCI begins with **immobilization**. This strategy prevents partial injuries of the cord from severing it completely. Use of splints to completely immobilize suspected SCI at the scene of the injury has helped reduce the severity of spinal cord injuries in the last two decades. Intravenous methylprednisolone, a steroid anti-inflammatory drug, is given during the first 24 hours to reduce inflammation and tissue destruction.

Rehabilitation after spinal cord injury seeks to prevent complications, promote recovery, and make the most of remaining function. Rehabilitation is a complex and long-term process. It requires a team of

professionals, including a neurologist, psychiatrist or rehabilitation specialist, physical therapist, and occupational therapist. Other specialists who may be needed include a respiratory therapist, vocational rehabilitation counselor, social worker, speech-language pathologist, nutritionist, special education teacher, recreation therapist, and clinical psychologist. Support groups provide a critical source of information, advice, and support for SCI patients.

As of early 2008, scientists were experimenting with stem cells to treat spinal cord injuries. Research in rats that used embryonic stem cells may lead to new therapies in humans. Embryonic stem cells have the potential to become any cell type in the body depending on what chemical signals they get when they mature. Researchers hope that by triggering embryonic stem cells to become nerve cell precursors and then transplanting these precursor cells into the injured area they can promote healing of the spinal cord. This research is still in its infancy and is controversial because of its use of stem cells from embryos, many of which are obtained from aborted fetuses.

Paralysis and loss of sensation

Some limited mobility and sensation may be recovered but the extent and speed of this recovery cannot be predicted. Experimental electrical stimulation has been shown to allow some control of muscle contraction in paraplegia. This experimental technique offers the possibility of unaided walking. Further development of current control systems will be needed before useful movement is possible outside the laboratory.

The physical therapist focuses on mobility, to maintain range of motion of affected limbs and reduce contracture and deformity. **Physical therapy** helps compensate for lost skills by using those muscles that are still functional. It also helps to increase any residual strength and control in affected muscles. A physical therapist suggests adaptive equipment such as braces, canes, or wheelchairs.

An occupational therapist works to restore ability to perform the activities of daily living, such as eating and grooming, with tools and new techniques. The occupational therapist also designs modifications of the home and workplace to match the individual impairment.

A pulmonologist or respiratory therapist promotes airway hygiene through instruction in assisted coughing techniques and postural drainage. The respiratory professional also prescribes and provides

instruction in the use of ventilators, facial or nasal masks, and tracheostomy equipment where necessary.

Pressure ulcers

Pressure ulcers are prevented by turning in bed at least every two hours. The patient should be turned more frequently when redness begins to develop in sensitive areas. Special mattresses and chair cushions can distribute weight more evenly to reduce pressure. Electrical stimulation is sometimes used to promote muscle movement to prevent pressure ulcers.

Spasticity and contracture

Range of motion (ROM) exercises help to prevent contracture. Chemicals can be used to prevent **contractures** from becoming fixed when ROM exercise is inadequate. Phenol or alcohol can be injected onto the nerve or botulinum toxin directly into the muscle. Botulinum toxin is associated with fewer complications, but it is more expensive than phenol and alcohol. Contractures can be released by cutting the shortened tendon or transferring it surgically to a different site on the bone where its pull will not cause as much deformity. Such tendon transfers may also be used to increase strength in partially functional extremities.

Heterotopic ossification

Etidronate disodium (Didronel), a drug that regulates the body's use of **calcium**, is used to prevent heterotopic ossification. Treatment begins three weeks after the injury and continues for 12 weeks. Surgical removal of ossified tissue is possible.

Autonomic dysreflexia

Autonomic dysreflexia is prevented by bowel and bladder care and attention to potential irritants. It is treated by prompt removal of the irritant. Drugs to lower blood pressure are used when necessary. People with SCI should educate friends and family members about the symptoms and treatment of dysreflexia, because immediate attention is necessary.

Loss of bladder and bowel control

Normal bowel function is promoted through adequate fluid intake and a diet rich in fiber. Evacuation is stimulated by deliberately increasing the abdominal pressure, either voluntarily or by using an abdominal binder.

Bladder care involves continual or intermittent catheterization. The full bladder may be detected by feeling its bulge against the abdominal wall. Urinary

KEY TERMS

Autonomic nervous system—The part of the nervous system that controls involuntary functions such as sweating and blood pressure.

Botulinum toxin—Any of a group of potent bacterial toxins or poisons produced by different strains of the bacterium *Clostridium botulinum*.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct images of internal structures.

Motor—Of or pertaining to motion, the body apparatus involved in movement, or the brain functions that direct purposeful activity.

Motor nerve—Motor or efferent nerve cells carry impulses from the brain to muscle or organ tissue.

Peripheral nervous system—The part of the nervous system that is outside the brain and spinal cord. Sensory, motor, and autonomic nerves are included.

Postural drainage—The use of positioning to drain secretions from the bronchial tubes and lungs into the trachea or windpipe.

Range of motion (ROM)—The range of motion of a joint from full extension to full flexion (bending) measured in degrees like a circle.

Sensory nerves—Sensory or afferent nerves carry impulses of sensation from the periphery or outward parts of the body to the brain. Sensations include feelings, impressions, and awareness of the state of the body.

Voluntary—An action or thought undertaken or controlled by a person's free will or choice.

tract infection is a significant complication of catheterization and requires frequent monitoring.

Sexual dysfunction

Counseling can help in adjusting to changes in sexual function after spinal cord injury. Erection may be enhanced through the same means used to treat **erectile dysfunction** in the general population.

Prognosis

The prognosis of SCI depends on the location and extent of injury. Injuries of the neck above C4 with significant involvement of the diaphragm hold the gravest prognosis. Respiratory infection is one of the leading causes of death in long-term SCI. Overall, 85% of SCI patients who survive the first 24 hours are alive 10 years after their injuries. Recovery of function is impossible to predict. Partial recovery is more likely after an incomplete wound than after the spinal cord has been completely severed.

Prevention

Risk of spinal cord injury can be reduced through prevention of the accidents that lead to it. Chances of injury from automobile accidents, the major cause of SCIs, can be significantly reduced by driving at safe speeds, avoiding alcohol while driving, and using seat belts.

Resources

BOOKS

Boyles, Carolyn. *A Complete Plain-English Guide to Living with a Spinal Cord Injury: Valuable Information From a Survivor*. Lincoln, NE: iUniverse, Inc., 2007.

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Stiens, Steven, et al. "Healthy Aging After SCI: Growing Older Brings Certain Problems, but You Can Take Important Steps to Remain Active." *Paraplegia News* (December 2007): 52–4.

Tai, David, and David White. "Spinal Cord Injury: Management by Acupuncture." *The Journal of Chinese Medicine* (June 2006): 11–7.

ORGANIZATIONS

Canadian Paraplegic Association, 1101 Prince of Wales Drive, Suite 230, Ottawa, Canada, Ontario, K2C 3W7, (613) 723-1913, (613) 723-1060, info@canparaplegic.org, http://www.canparaplegic.org.

National Spinal Cord Injury Association, 1 Church St., Suite 600, Rockville, MD, 20850, (866) 387-2196, (800) 962-9629, info@spinalcord.org, http://www.spinalcord.org. Spinal Cord Injuries Australia, 1 Jennifer Street, Little Bay, Australia, NSW 2036, (800) 819-775, office@scia.org.au, http://www.scia.org.au.

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Spinal cord tumors

Definition

A spinal cord tumor is a benign or cancerous growth in the spinal cord, between the membranes covering the spinal cord, or in the spinal canal. A tumor in this location can compress the spinal cord or its nerve roots. As a result, even a noncancerous growth can be disabling unless properly treated.

Spinal cord tumors can be classified according to the origin of their location. Spinal cord tumors, which arise from inside the cord, are termed intramedullary tumors while tumors which originate outside of the cord are classified as extramedullary tumors.

Demographics

Spinal cord tumors are rare and account for about 15% of all tumors of the central nervous system. Primary spinal cord tumors (tumors originating in the spinal cord) are most likely to affect individuals between the ages of 30 and 50 years. Most spinal cord tumors are caused by metastatic lesions that originate from **cancer** elsewhere in the body. Up to 10% of patients diagnosed with cancer will be affected by metastasis of their primary cancer to the spinal cord.

Description

The spinal cord contains bundles of nerves that carry messages between the brain and the body. Because the spinal cord is rigidly encased in bone, any tumor that grows on or near it can compress the nerves, and interfere in this communication.

Newly formed tumors that begin within the spinal cord (primary spinal cord tumors) are unusual, especially among children and the elderly. The most common types of primary spinal cord tumors are classified as astrocytomas and ependymomas.

More typically, tumors originate elsewhere in the body and move through the bloodstream (metastasize)

KEY TERMS

Computed tomography scans (CT scan)—The CT scan combines an x ray with a computer to create a detailed picture of the spinal cord. It may help to determine the type of tumor, locate swelling or bleeding, and check results of treatment.

Magnetic resonance imaging (MRI)—MRI is an imaging technique that uses a magnetic field to scan the body's tissues and structures. It gives a better picture of tumors located near bone than does a CT scan, without the risk of radiation, and can provide a three-dimensional image of the tumor.

Myelogram—A myelogram is an x-ray exam of the spinal cord, nerves, and other tissues within the spinal cord that are highlighted by injected contrast dye.

to the spinal cord. Cancers which tend to metastasize to the spinal cord include cancers of the breast, prostate, kidney, and lung as well as lymphoma, sarcoma, and **multiple myeloma**. Intramedullary metastases are very rare.

Causes and symptoms

The cause of primary spinal cord tumors is unknown.

Initially symptoms of a spinal cord tumor, whether primary or metastatic, may be vague and may include symptoms such as **pain** or stiffness. As the lesion grows however, the tumor places increasing pressure on the spinal cord resulting in symptoms including:

- back pain
- severe or burning pain in other parts of the body
- numbness or cold
- progressive loss of muscle strength or sensation in the legs
- loss of bladder or bowel control

A tumor in the top of the spinal column can cause pain radiating from the arms or neck; a tumor in the lower spine may cause leg or back pain. If there are several tumors in different areas of the spinal cord at the same time, it may cause symptoms in a variety of locations in the body.

Pain may not be associated with spinal cord tumors diagnosed in children. The most common symptom in children affected by intramedullary spinal cord tumors is often disturbance in gait.

Diagnosis

Diagnosis of primary spinal cord tumors includes imaging studies such as MRI. There are no blood tests that can specifically detect tumors in the spinal cord.

Suspected spinal cord compression, by tumor, is a medical emergency. Prompt intervention may prevent **paralysis** and other neurologic complications.

If a **neurological exam** and review of symptoms suggest a spinal cord tumor, the doctor may order the following tests:

- MRI, the procedure of choice
- CT and nuclear medicine bone scans
- intrathecal contrast-enhanced myelography
- blood tests such as a complete blood test (CBC), erythrocyte sedimentation rate (ESR), clotting studies, and metabolic profile including tests to determine calcium levels and liver function tests
- x rays of the spine

Lumbar puncture is often contraindicated when a spinal tumor is suspected.

Treatment

Primary spinal cord tumors are often slow-growing tumors, which are typically contained to an anatomic site. Therefore, when possible, surgical removal of the tumor is the treatment of choice. Because these tumors are slow-growing, **radiation therapy** and **chemotherapy** are not considered to be as effective on this tumor type. Some of the more aggressive tumor types may be treated using radiation therapy. Currently, chemotherapy is considered experimental therapy for the treatment of spinal cord tumors.

Treatment of a metastatic lesion to the spinal cord is initiated rapidly and is considered a medical emergency to prevent permanent neurologic damage. Treatment is typically initiated using **corticosteroids** such as dexamethasone (Decadron, Hexadrol). Corticosteroids have anti-inflammatory properties and may help to preserve neurologic function in spinal cord compression. Other treatments that may be used include radiation therapy to the site, chemotherapy, and surgical decompression.

Prognosis

The prognosis for patients diagnosed with primary spinal cord tumors is dependent upon several factors including tumor type, size of the tumor, location of the tumor, effect of the tumor on neurologic function prior to surgery, success of the surgery in removing most or all

of the cancer, and age of the patient. Patients younger than age 60, with tumors affecting only one level of the spinal cord that are able to be totally removed generally, have the most favorable prognoses.

Patients with metastatic lesions to the spinal cord whose lesions are detected and treated promptly derive substantial benefit. However, the long-term prognosis is ultimately dependent upon the success of treating the primary tumor.

Prevention

At this time, since it is not known what causes most cases of primary spinal cord tumors, it is not possible to prevent this type of cancer.

Since metastatic spinal cord tumors are the result of a cancer that has first appeared elsewhere in the body, early detection and treatment of cancer in other organs may prevent the spread of the cancer to the spinal cord in some people. However, even though these primary cancers may be detected and treated promptly, it may not be possible to stop the spread of some of these cancers to the spinal cord.

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Spinal fluid analysis see **Cerebrospinal fluid (CSF) analysis**

Spinal fusion see **Disk removal**

Spinal instrumentation

Definition

Spinal instrumentation is a method of straightening and stabilizing the spine after spinal fusion, by surgically attaching hooks, rods, and wire to the spine in a way that redistributes the stresses on the bones and keeps them in proper alignment.

Purpose

Spinal instrumentation is used to treat instability and deformity of the spine. Instability occurs when the spine no longer maintains its normal shape during movement. Such instability results in nerve damage, spinal deformities, and disabling **pain**. Spinal deformities may be caused by:

- birth defects
- fractures
- marfan syndrome
- neurofibromatosis
- neuromuscular diseases
- severe injuries
- tumors

Curvature of the spine (**scoliosis**) is usually treated with spinal fusion and spinal instrumentation. Scoliosis is a disorder of unknown origin. It causes bending and twisting of the spine that eventually results in distortion of the chest and back. About 85% of cases occur in girls between the ages of 12 and 15, who are experiencing adolescent growth spurt.

Spinal instrumentation serves three purposes. It provides a stable, rigid column that encourages bones to fuse after spinal-fusion surgery. Second, it redirects the stresses over a wider area. Third, it restores the spine to its proper alignment.

Different types of spinal instrumentation are used to treat different spinal problems. Several common types of spinal instrumentation are explained below. Although the details of the insertion of rods, wires, and hooks varies, the purpose of all spinal instrumentation is the same—to correct and stabilize the backbone.

Harrington rod

The Harrington Rod is one of the oldest and most proven forms of spinal instrumentation. It is used to straighten and stabilize the spine when curvature is greater than 60 degrees. It is an appropriate treatment for scoliosis.

Advantages of the Harrington rod are its relative simplicity of installation, the low rate of complications, and a proven record of reducing curvature of the spine. The main disadvantage is that the patient must remain in a body cast for about six months, then wear a brace for another three to six months while the bone fusion solidifies.

Luque rod

Luque rods are custom contoured metal rods that are fixed to each segment (vertebra) in the affected part of the spine. The main advantage is that the patient may not need to wear a cast or brace after the procedure. The main disadvantage is that the risk of injury to the nerves and spinal cord is higher than with some other forms of instrumentation. This is because wires must be threaded through each vertebra near the spinal column, increasing the risk of such damage. Luque rods are sometimes used to treat scoliosis.

Drummond instrumentation

Drummond instrumentation, also called Harrington-Drummond instrumentation, uses a Harrington rod on the concave side of the spine and a Luque rod on the convex side. The advantage is that each vertebra segment is fixed, with the risk of nerve injury decreased over Luque rod instrumentation. The disadvantage is that, like Harrington rod instrumentation, the patient must wear a cast and a brace after surgery.

Cotrel-Dubousset instrumentation

Cotrel-Dubousset instrumentation uses hooks and rods in a cross-linked pattern to realign the spine and redistribute the biomechanical stress. The main advantage of Cotrel-Dubousset instrumentation is that because of the extensive cross-linking, the patient may have to wear a cast or brace after surgery. The disadvantage is the complexity of the operation and the number of hooks and cross-links that may fail.

Zeilke instrumentation

Zeilke instrumentation is similar to Cotrel-Dubousset instrumentation but is used to treat double curvature of the spine. It requires wearing a brace for many months after surgery.

Other forms of instrumentation

The Kaneda device is used to treat fractured thoracic or lumbar vertebrae when it is suspected that bone fragments are present in the spinal canal. Variations on the basic forms of spinal instrumentation, such as Wisconsin instrumentation, are being refined as technology improves. A physician chooses the proper type of instrumentation based on the type of disorder, the age and health of the patient, and on the physician's experience.

Precautions

Since the hooks and rods of spinal instrumentation are anchored in the bones of the back, spinal instrumentation should not be performed on people with serious **osteoporosis**. To overcome this limitation, techniques are being explored that help anchor instrumentation in fragile bones.

Description

Spinal instrumentation is performed by a neuro and/or orthopedic surgical team with special experience in spinal operations. The surgery is done in a hospital under **general anesthesia**. It is done at the same time as spinal fusion.

The surgeon strips the muscles away from the area to be fused. The surface of the bone is peeled away. A piece of bone is removed from the hip and placed alongside the area to be fused. The stripping of the bone helps the bone graft to fuse.

After the fusion site is prepared, the rods, hooks, and wires are inserted. There is some variation in how this is done based on the spinal instrumentation chosen. In general, Harrington rods are the simplest instrumentation to install, and Cotrel-Dubousset instrumentation is the most complex and risky. Once the rods are in place, the incision is closed.

Preparation

Spinal fusion with spinal instrumentation is a major surgery. The patient will undergo many tests to determine the nature and exact location of the back problem. These tests are likely to include x rays, **magnetic resonance imaging (MRI)**, **computed tomography scans** (CT scans), and myelograms. In addition, the patient will undergo a battery of blood and urine tests, and possibly an electrocardiogram to provide the surgeon and anesthesiologist with information that will allow the operation to be performed safely. In Harrington rod instrumentation, the patient may be

KEY TERMS

Lumbar vertebrae—The vertebrae of the lower back below the level of the ribs.

Marfan syndrome—A rare hereditary defect that affects the connective tissue.

Neurofibromatosis—A rare hereditary disease that involves the growth of lesions that may affect the spinal cord.

Osteoporosis—A bone disorder, usually seen in the elderly, in which the bones become increasingly less dense and more brittle.

Spinal fusion—An operation in which the bones of the lower spine are permanently joined together using a bone graft obtained usually from the hip.

Thoracic vertebrae—The vertebrae in the chest region to which the ribs attach.

placed in **traction** or an upper body cast to stretch contracted muscles before surgery.

Aftercare

After surgery, the patient will be confined to bed and a catheter will be inserted so that the patient can urinate without getting up. Vital signs will be monitored and the patient's position will be changed frequently so that **bedsores** do not develop.

Recovery from spinal instrumentation can be a long, arduous process. Movement is severely limited for a period of time. In certain types of instrumentation, the patient is put in a cast to allow the realigned bones to stay in position until healing takes place. This can be as long as six to eight months. Many patients will need to wear a brace after the cast is removed.

During the recovery period, the patient is taught respiratory exercises to help maintain respiratory function during the time of limited mobility. Physical therapists assist the patient in learning self-care and in performing strengthening and range of motion exercises. Length of hospital stay depends on the age and health of the patient as well as the specific problem that was corrected. The patient can expect to remain under a physician's care for many months.

Risks

Spinal instrumentation carries a significant risk of nerve damage and **paralysis**. The skill of the surgeon can affect the outcome of the operation so patients

should look for a hospital and surgical team that has a lot of experience doing spinal procedures.

After surgery there is a risk of infection or an inflammatory reaction due to the presence of the foreign material in the body. Serious infection of the membranes covering the spinal cord and brain can occur. In the long-term, the instrumentation may move or break, causing nerve damage and requiring a second surgery. Some bone grafts do not heal well, lengthening the time the patient must spend in a cast or brace, or necessitating additional surgery. Casting and wearing a brace may take an emotional toll, especially on young people. Patients who have had spinal instrumentation must avoid contact sports and, for the rest of their lives, eliminate situations that will abnormally put stress on their spines.

Normal results

Many young people with scoliosis heal with significantly improved alignment of the spine. Results of spinal instrumentation done for other conditions vary widely.

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National Scoliosis Foundation, 5 Cabot Place, Stoughton, MA, 20724, (800) 673-6922, NSF@scoliosis.org, <http://www.scoliosis.org>.

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Spinal meningitis see **Meningitis**

Spinal stenosis

Definition

Spinal stenosis is any narrowing of the spinal canal that causes compression of the spinal nerve cord. Spinal stenosis causes **pain** and may cause loss of some body functions.

Description

Spinal stenosis is a progressive narrowing of the opening in the spinal canal. The spine is a long series of bones called vertebrae. Between each pair of vertebra

is a fibrous intervertebral disk. Collectively, the vertebrae and disks are called the backbone. Each vertebra has a hole through it. These holes line up to form the spinal canal. A large bundle of nerves called the spinal cord runs through the spinal canal. This bundle of 31 nerves carries messages between the brain and the various parts of the body. At each vertebra, some smaller nerves branch out from these nerve roots to serve the muscles and tissue in the immediate area. When the spinal canal narrows, nerve roots in the spinal cord are squeezed. Pressure on the nerve roots causes chronic pain and loss of control over some functions because communication with the brain is interrupted. The lower back and legs are most affected by spinal stenosis. The nerve roots that supply the legs are near the bottom of the spinal cord. The pain gets worse after standing for a long time and after some forms of **exercise**. The posture required by these physical activities increases the **stress** on the nerve roots. Spinal stenosis usually affects people over 50 years of age. Women have the condition more frequently than men do.

Cervical spinal stenosis is a narrowing of the vertebrae of the neck (cervical vertebrae). The disease and its effects are similar to stenosis in the lower spine. A narrower opening in the cervical vertebrae can also put pressure on arteries entering the spinal column, cutting off the blood supply to the remainder of the spinal cord.

Causes and symptoms

Spinal stenosis causes pain in the buttocks, thigh, and calf and increasing weakness in the legs. The patient may also have difficulty controlling bladder and bowel functions. The pain of spinal stenosis seems more severe when the patient walks downhill. Spinal stenosis can be congenital, acquired, or a combination. Congenital spinal stenosis is a birth defect. Acquired spinal stenosis develops after birth. It is usually a consequence of tissue destruction (degeneration) caused by an **infectious disease** or a disease in which the immune system attacks the body's own cells (autoimmune disease). The two most common causes of spinal stenosis are birth defect and progressive degeneration of the tissue of the joints (**osteoarthritis**). Other causes include improper alignment of the vertebrae as in spondylolisthesis, destruction of bone tissue as in Paget's disease, or an overgrowth of bone tissue as in diffuse idiopathic skeletal hyperostosis. The spinal canal is usually more than 0.5 in (12 mm) in diameter. A smaller diameter indicates stenosis. The diameter of the cervical spine ranges is 0.6–1 in (15–12 mm). Any opening under 0.5 in (13 mm) in diameter is considered evidence of stenosis. Acquired spinal stenosis usually begins with degeneration of the intervertebral disks or the surfaces of the vertebrae or both. In trying to heal this degeneration, the body builds

KEY TERMS

Computed tomography (CT) Scans—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures.

Congenital—Present before birth. The term is used to describe disorders that developed in the fetal stage.

Doppler scanning—A procedure in which ultrasound images are used to watch a moving structure such as the flow of blood or the beating of the heart.

Electromyography—A test that uses electrodes to record the electrical activity of muscle. The information gathered is used to find disorders of the nerves that serve the muscles.

Evoked potential—A test of nerve response that uses electrodes placed on the scalp to measure brain reaction to a stimulus such as a touch.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct images of internal structures.

Nerve conduction velocity test—A test that measures the time it takes a nerve impulse to travel a specific distance over the nerve after electronic stimulation.

Stenosis—The narrowing or constriction of a channel or opening.

up the spinal column. In the process, the spinal canal can become narrower.

Diagnosis

The physician must determine that the symptoms are caused by spinal stenosis. Conditions that can cause similar symptoms include a slipped (herniated) intervertebral disk, spinal tumors, and disorders of the blood flow (circulatory disorders). Spinal stenosis causes back and leg pain. The leg pain is usually worse when the patient is standing or walking. Some forms of spinal stenosis are less painful when the patient is riding an exercise bike because the forward tilt of the body changes the pressure in the spinal column. Doppler scanning can trace the flow of blood to determine whether the pain is caused by circulatory problems. X-ray images, **computed tomography scans** (CT scans), and **magnetic resonance imaging** (MRI) scans can reveal any narrowing of the spinal canal. **Electromyography**, nerve conduction velocity, or **evoked potential studies** can locate problems in the muscles indicating areas of spinal cord compression.

Treatment

Mild cases of spinal stenosis may be treated with rest, **nonsteroidal anti-inflammatory drugs** (such as **aspirin**), and **muscle relaxants**. Spinal stenosis can be a progressive disease, however, and the source of pressure may have to be surgically removed (surgical decompression) if the patient is losing control over bladder and bowel functions. The surgical procedure removes bone and other tissues that have entered the

spinal canal or put pressure on the spinal cord. Two vertebrae may be fused, to eliminate improper alignment, such as that caused by spondylolisthesis. For surgery, patients lie on their sides or in a modified kneeling position. This position reduces bleeding and places the spine in proper alignment. Alignment is especially important if vertebrae are to be fused. Surgical decompression can eliminate leg pain and restore control of the legs, bladder, and bowels, but usually does not eliminate lower back pain. **Physical therapy** and massage can help reduce the symptoms of spinal stenosis. An exercise program should be developed to increase flexibility and mobility. A brace or corset may be worn to improve posture. Activities that place stress on the lower back muscles should be avoided.

Prognosis

Surgical decompression does not stop the degenerative processes that cause spinal stenosis and the condition can develop again. Nevertheless, most patients achieve good results with surgical decompression. The patient will probably continue to have lower back pain after the surgical procedure.

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Spinal tap see Cerebrospinal fluid (CSF) analysis

Spirometry

Definition

Spirometry is the measurement of air flow into and out of the lungs.

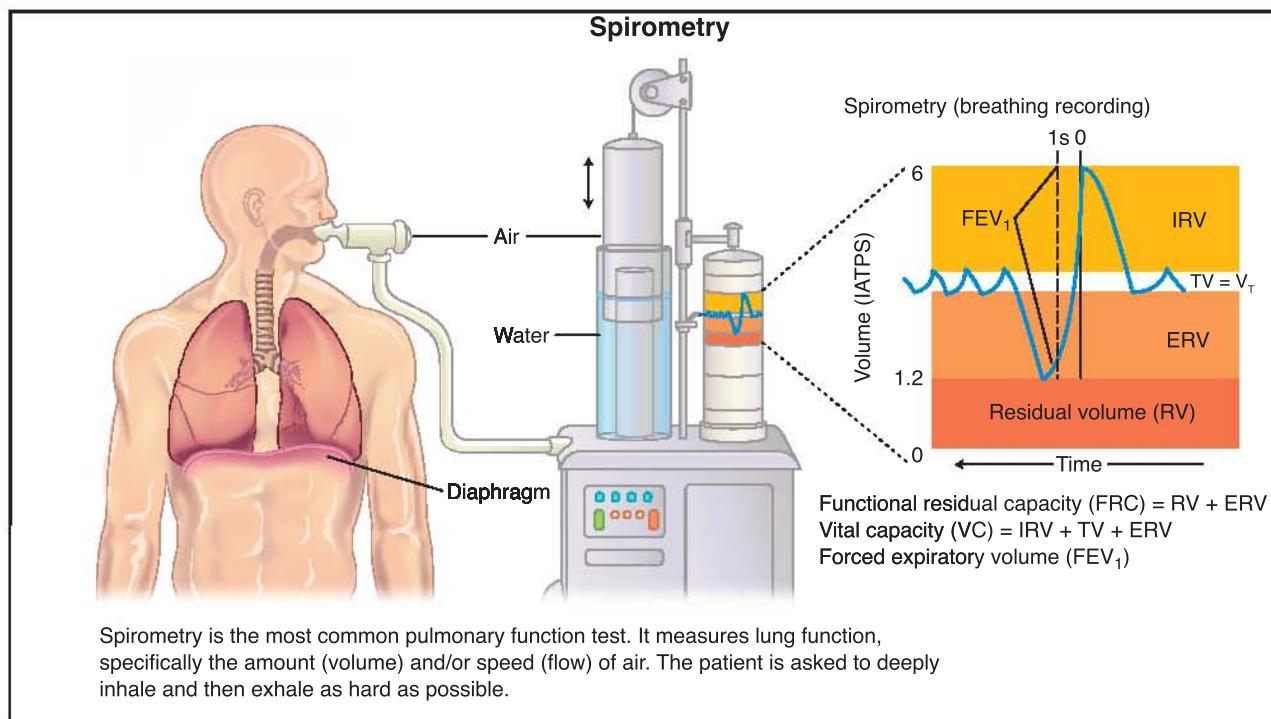
Description

Spirometry requires that the nose is pinched off as the patient breathes through a mouthpiece attached to the spirometer. The patient is instructed on how to breathe during the procedure. Three breathing maneuvers are practiced before recording the procedure, and the highest of three trials is used for evaluation of breathing. This procedure measures air flow by electronic or

mechanical displacement principles and uses a microprocessor and recorder to calculate and plot air flow.

The test produces a recording of the patient's ventilation under conditions involving both normal and maximal effort. The recording, called a spirogram, shows the volume of air moved and the rate at which it travels into and out of the lungs. Spirometry measures several lung capacities. Accurate measurement is dependent upon the patient performing the appropriate maneuver properly. The most common measurements are:

- Vital capacity (VC). This is the amount of air (in liters) moved out of the lung during normal breathing. The patient is instructed to breathe in and out normally to attain full expiration. Vital capacity is usually about 80% of the total lung capacity. Because of the elastic nature of the lungs and surrounding thorax, a small volume of air will remain in the lungs after full exhalation. This volume is called the residual volume (RV).
- Forced vital capacity (FVC). After breathing out normally to full expiration, the patient is instructed to breathe in with a maximal effort and then exhale as forcefully and rapidly as possible. The FVC is the volume of air that is expelled into the spirometer following a maximum inhalation effort.
- Forced expiratory volume (FEV). At the start of the FVC maneuver, the spirometer measures the volume of air delivered through the mouthpiece at timed



(Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

intervals of 0.5, 1.0, 2.0, and 3.0 seconds. The sum of these measurements normally constitutes about 97% of the FVC measurement. The most commonly used FEV measurement is FEV-1, which is the volume of air exhaled into the mouthpiece in one second. The FEV-1 should be at least 70% of the FVC.

- Forced expiratory flow 25–75% (FEF 25–75). This is a calculation of the average flow rate over the center portion of the forced expiratory volume recording. It is determined from the time in seconds at which 25% and 75% of the vital capacity is reached. The volume of air exhaled in liters per second between these two times is the FEF 25–75. This value reflects the status of the medium and small sized airways.
- Maximal voluntary ventilation (MVV). This maneuver involves the patient breathing as deeply and as rapidly as possible for 15 seconds. The average air flow (liters per second) indicates the strength and endurance of the respiratory muscles.

Normal values for FVC, FEV, FEF, and MVV are dependent on the patient's age, gender, and height.

Purpose

Spirometry is the most commonly performed pulmonary function test (PFT). The test can be performed at the bedside, in a physician's office, or in a pulmonary laboratory. It is often the first test performed when a problem with lung function is suspected. Spirometry may also be suggested by an abnormal x ray, arterial blood gas analysis, or other diagnostic pulmonary test result. The National Lung Health Education Program recommends that regular spirometry tests be performed on persons over 45 years old who have a history of smoking. Spirometry tests are also recommended for persons with a family history of lung disease, chronic respiratory ailments, and advanced age.

Spirometry measures ventilation and the movement of air into and out of the lungs. The spirogram will identify two different types of abnormal ventilation patterns, obstructive and restrictive.

Common causes of an obstructive pattern are **cystic fibrosis**, **asthma**, **bronchiectasis**, **bronchitis**, and **emphysema**. These conditions may be collectively referred to by using the acronym CABBE. Chronic bronchitis, emphysema, and asthma result in dyspnea (difficulty breathing) and ventilation deficiency, a condition known as **chronic obstructive pulmonary disease** (COPD). COPD is the fourth leading cause of death among Americans.

Common causes of a restrictive pattern are **pneumonia**, heart disease, **pregnancy**, lung fibrosis, **pneumothorax** (collapsed lung), and **pleural effusion** (compression caused by chest fluid).

KEY TERMS

Bronchodilator—A drug, usually self-administered by inhalation, that dilates the airways.

Forced expiratory volume (FEV)—The volume of air exhaled from the beginning of expiration to a set time (usually 0.5, 1, 2, and 3 seconds).

Forced vital capacity (FVC)—The volume of air that can be exhaled forcefully after a maximal inspiration.

Hemoptysis—Spitting up of blood derived from the lungs or bronchial tubes as a result of pulmonary or bronchial hemorrhage.

Thrombosis—Formation or presence of a thrombus; clotting within a blood vessel that may cause infarction of tissues supplied by the vessel.

Thrombotic—Relating to, caused by, or characterized by thrombosis.

Vital capacity (VC)—The volume of air that can be exhaled following a full inspiration.

Obstructive and restrictive patterns can be identified on spirographs using both a "y" and "x" axis. Volume (liters) is plotted on the y-axis versus time (seconds) on the x-axis. A restrictive pattern is characterized by a normal shape showing reduced volumes for all parameters. The reduction in volumes indicates the severity of the disease. An obstructive pattern produces a spirogram with an abnormal shape. Inspiration volume is reduced. The volume of air expelled is normal but the air flow rate is slower, causing an elongated tail to the FVC.

A flow-volume loop spirogram is another way of displaying spirometry measurements. This requires the FVC maneuver followed by a forced inspiratory volume (FIV). Flow rate in liters per second is plotted on the y-axis and volume (liters) is plotted on the x-axis. The expiration phase is shown on top and the inspiration phase on the bottom. The flow-volume loop spirogram is helpful in diagnosing upper airway obstruction and differentiating some types of restrictive patterns.

Some conditions produce specific signs on the spirogram. Irregular inspirations with rapid frequency are caused by hyperventilation associated with **stress**. Diffuse fibrosis of the lung causes rapid breathing of reduced volume, which produces a repetitive pattern known as the penmanship sign. Serial reduction in the FVC peaks indicates air trapped inside the lung. A

notch and reduced volume in the early segments of the FVC is consistent with airway collapse. A rise at the end of expiration is associated with airway resistance.

Spirometry is used to assess lung function over time and often to evaluate the efficacy of bronchodilator inhalers such as albuterol. It is important for the patient to refrain from using a bronchodilator prior to the evaluation. Spirometry is performed before and after inhaling the bronchodilator. In general, a 12% or greater improvement in both FVC and FEV-1, or an increase in FVC by 0.2 liters, is considered a significant improvement for an adult patient.

Precautions

The patient should inform the physician of any medications he or she is taking, or of any medical conditions that are present; these factors may affect the validity of the test. The patient's smoking habits and history should be thoroughly documented. The patient must be able to understand and respond to instructions for the breathing maneuvers. Therefore, the test may not be appropriate for very young, unresponsive, or physically impaired persons.

Spirometry is contraindicated in patients whose condition will be aggravated by forced breathing, including:

- hemoptysis (spitting up blood from the lungs or bronchial tubes)
- pneumothorax (free air or gas in the pleural cavity)
- recent heart attack
- unstable angina
- aneurysm (cranial, thoracic, or abdominal)
- thrombotic condition (such as clotting within a blood vessel)
- recent thoracic or abdominal surgery
- nausea or vomiting

The test should be terminated if the patient shows signs of significant head, chest, or abdominal **pain** while the procedure is in progress.

Spirometry is dependent upon the patient's full compliance with breathing instructions, especially his or her willingness to extend a maximal effort at forced breathing. Therefore, the patient's emotional state must be considered.

Preparation

The patient's age, gender, and race are recorded, and height and weight are measured before the procedure begins. The patient should not have eaten heavily within three hours of the test. He or she should be instructed to wear clothing that fits loosely over the

chest and abdominal area. The respiratory therapist or other testing personnel should explain and demonstrate the breathing maneuvers to the patient. The patient should practice breathing into the mouthpiece until he or she is able to duplicate the maneuvers successfully on two consecutive attempts.

Aftercare

In most cases, special care is not required following spirometry. Occasionally, a patient may become light-headed or dizzy. Such patients should be asked to rest or lie down, and should not be discharged until after the symptoms subside. In rare cases, the patient may experience pneumothorax, intracranial **hypertension**, chest pain, or uncontrolled coughing. In such cases, additional care directed by a physician may be required.

Normal results

The results of spirometry tests are compared to predicted values based on the patient's age, gender, and height. For example, a young adult in good health is expected to have the following FEV values:

- FEV-0.5—50–60% of FVC
- FEV-1—75–85% of FVC
- FEV-2—95% of FVC
- FEV-3—97% of FVC

In general, a normal result is 80–100% of the predicted value. Abnormal values are:

- mild lung dysfunction—60–79%
- moderate lung dysfunction—40–59%
- severe lung dysfunction—below 40%

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ORGANIZATIONS

National Lung Health Education Program (NLHEP), 1850 High Street, Denver, CO, 80218, <http://www.nlhep.org>.

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Spleen, enlarged see **Hypersplenism**
Spleen removal see **Splenectomy**

Splenectomy

Definition

Splenectomy is the surgical removal of the spleen, which is an organ that is part of the lymphatic system. The spleen is a dark-purple, bean-shaped organ located in the upper left side of the abdomen, just behind the bottom of the rib cage. In adults, the spleen is about 4.8 X 2.8 X 1.6 in (12 X 7 X 4 cm) in size and weighs about 4–5 oz (113–14 zg). Its functions include a role in the immune system; filtering foreign substances from the blood; removing worn-out blood cells from the blood; regulating blood flow to the liver; and sometimes storing blood cells. The storage of blood cells is called sequestration. In healthy adults, about 30% of blood platelets are sequestered in the spleen.

Purpose

Splenectomies are performed for a variety of different reasons and with different degrees of urgency. Most splenectomies are done after the patient has been diagnosed with **hypersplenism**. Hypersplenism is not a specific disease but a group of symptoms, or syndrome, that can be produced by a number of different disorders. It is characterized by enlargement of the spleen (splenomegaly), defects in the blood cells, and an abnormally high turnover of blood cells. It is almost always associated with splenomegaly caused by specific disorders such as **cirrhosis** of the liver or certain cancers. The decision to perform a splenectomy depends on the severity and prognosis of the disease that is causing the hypersplenism.

Splenectomy always necessary

There are two diseases for which splenectomy is the only treatment—primary cancers of the spleen and a blood disorder called hereditary spherocytosis (HS). In HS, the absence of a specific protein in the red

blood cell membrane leads to the formation of relatively fragile cells that are easily damaged when they pass through the spleen. The cell destruction does not occur elsewhere in the body and ends when the spleen is removed. HS can appear at any age, even in newborns, although doctors prefer to put off removing the spleen until the child is five or six years old.

Splenectomy usually necessary

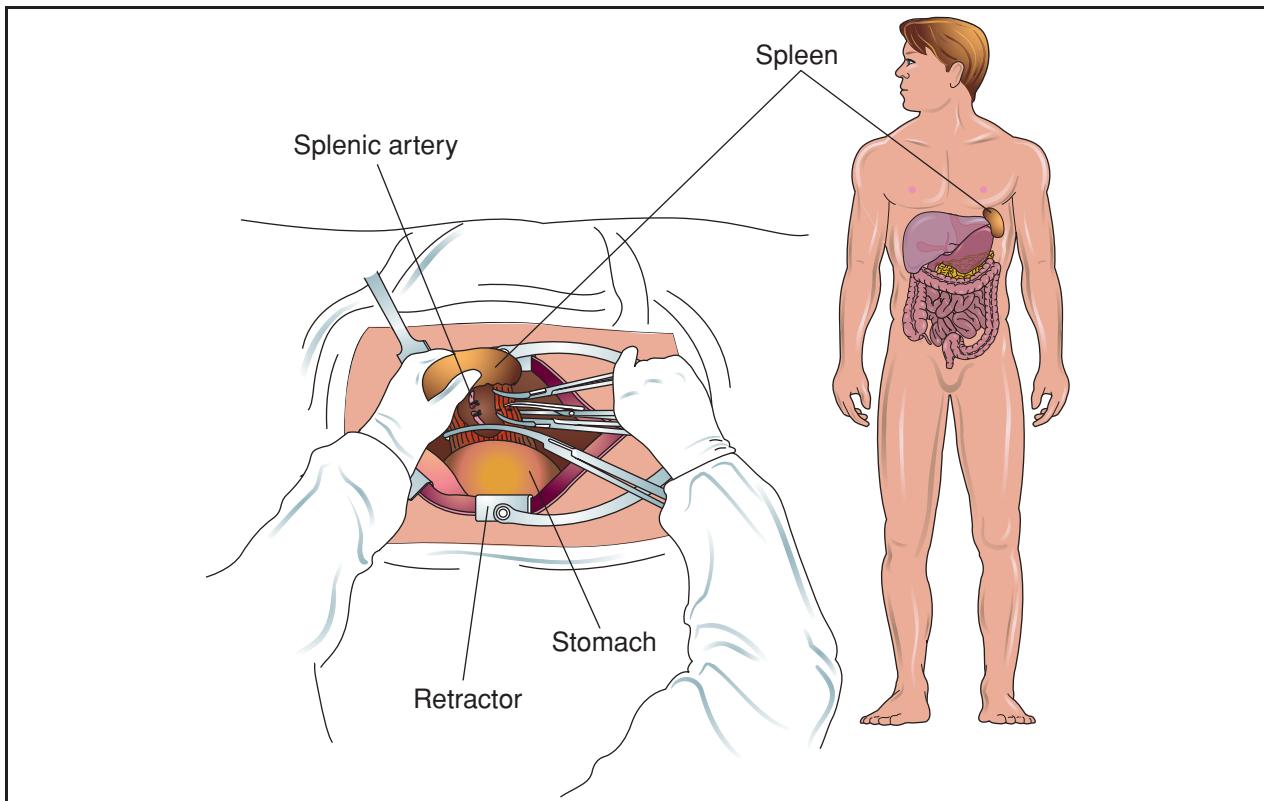
There are some disorders in which splenectomy is usually recommended. They include:

- Immune (idiopathic) thrombocytopenic purpura (ITP). ITP is a disease involving platelet destruction. Splenectomy has been regarded as the definitive treatment for this disease and is effective in about 70% of chronic ITP cases. More recently, however, the introduction of new drugs in the treatment of ITP has reopened the question as to whether splenectomy is always the best treatment option.
- Trauma. The spleen can be ruptured by blunt as well as penetrating injuries to the chest or abdomen. Car accidents are the most common cause of blunt traumatic injury to the spleen.
- Abscesses in the spleen. These are relatively uncommon but have a high mortality rate.
- Rupture of the splenic artery. Rupture sometimes occurs as a complication of pregnancy.
- Hereditary elliptocytosis. This is a relatively rare disorder. It is similar to HS in that it is characterized by red blood cells with defective membranes that are destroyed by the spleen.

Splenectomy sometimes necessary

In other disorders, the spleen may or may not be removed.

- Hodgkin's disease, a serious form of cancer that causes lymph nodes to enlarge. Splenectomy is often performed in order to find out how far the disease has progressed.
- Thrombotic thrombocytopenic purpura (TTP). TTP is a rare disorder marked by fever, kidney failure, and an abnormal decrease in the number of platelets. Splenectomy is one part of treatment for TTP.
- Autoimmune hemolytic disorders. These disorders may appear in patients of any age but are most common in patients over 50. The red blood cells are destroyed by antibodies produced by the patient's own body (autoantibodies).
- Myelofibrosis. Myelofibrosis is a disorder in which bone marrow is replaced by fibrous tissue. It produces



Splenectomy is the surgical removal of the spleen. This procedure is performed as a last result in most diseases involving the spleen. In some cases, however, splenectomy does not address the underlying causes of splenomegaly or other conditions affecting the spleen. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

severe and painful splenomegaly. Splenectomy does not cure myelofibrosis but may be performed to relieve pain caused by the swollen spleen.

- Thalassemia. Thalassemia is a hereditary form of anemia that is most common in people of Mediterranean origin. Splenectomy is sometimes performed if the patient's spleen has become painfully enlarged.

Precautions

Patients should be carefully assessed regarding the need for a splenectomy. Because of the spleen's role in protecting people against infection, it should not be removed unless necessary. The operation is relatively safe for young and middle-aged adults. Older adults, especially those with cardiac or pulmonary disease, are more vulnerable to post-surgical infections. Thromboembolism following splenectomy is another complication for this patient group, which has about 10% mortality following the surgery. Splenectomies are performed in children only when the benefits outweigh the risks.

The most important part of the assessment is the measurement of splenomegaly. The normal spleen cannot be felt when the doctor examines the patient's abdomen. A spleen that is large enough to be felt indicates splenomegaly. In some cases the doctor will hear a dull sound when he or she thumps (percusses) the patient's abdomen near the ribs on the left side. Imaging studies that can be used to demonstrate splenomegaly include ultrasound tests, technetium-99m sulfur colloid imaging, and CT scans. The rate of platelet or red blood cell destruction by the spleen can be measured by tagging blood cells with radioactive chromium or platelets with radioactive indium.

Description

Complete splenectomy

REMOVAL OF ENLARGED SPLEEN. Splenectomy is performed under general anesthesia. The most common technique is used to remove greatly enlarged spleens. After the surgeon makes a cut (incision) in the abdomen, the artery to the spleen is tied to prevent blood loss and reduce the spleen's size. It also helps

prevent further sequestration of blood cells. The surgeon detaches the ligaments holding the spleen in place and removes it. In many cases, tissue samples will be sent to a laboratory for analysis.

REMOVAL OF RUPTURED SPLEEN. When the spleen has been ruptured by trauma, the surgeon approaches the organ from its underside and fastens the splenic artery.

In some cases, the doctor may prefer conservative (nonsurgical) management of a ruptured spleen, most often when the patient's blood pressure is stable and there are no signs of other abdominal injuries. In the case of multiple abdominal trauma, however, the spleen is usually removed.

Partial splenectomy

In some cases the surgeon removes only part of the spleen. This procedure is considered by some to be a useful compromise that reduces **pain** from an enlarged spleen while leaving the patient less vulnerable to infection. Long-term follow-up of the results of partial splenectomies has not yet been done.

Laparoscopic splenectomy

Laparoscopic splenectomy, or removal of the spleen through several small incisions, has been more frequently used in recent years. Laparoscopic surgery involves the use of surgical instruments, with the assistance of a tiny camera and video monitor. Laparoscopic procedures reduce the length of hospital stay, the level of post-operative pain, and the risk of infection. They also leave smaller **scars**. Laparoscopic splenectomy is not, however, the best option for many patients.

Laparoscopic splenectomy is gaining increased acceptance in the early 2000s as an alternative to open splenectomy for a wide variety of disorders, although splenomegaly still presents an obstacle to laparoscopic splenectomy; massive splenomegaly has been considered a contraindication. In patients with enlarged spleens, however, laparoscopic splenectomy is associated with less morbidity, decreased **transfusion** rates, and shorter hospital stays than when the open approach is used. Patients with enlarged spleens usually have more severe hematologic diseases related to greater morbidity; therefore, laparoscopic splenectomy has potential advantages.

The most frequent serious complication following laparoscopic splenectomy is damage to the pancreas. Application of a hydrogel sealant to the pancreas during surgery, however, appears to significantly reduce the risk of leakage from the pancreas.

Splenic embolization

Splenic embolization is an alternative to splenectomy that is used in some patients who are poor surgical risks. Embolization involves plugging or blocking the splenic artery to shrink the size of the spleen. The substances that are injected during this procedure include polyvinyl alcohol foam, polystyrene, and silicone. Embolization is a technique that needs further study and refinement.

Preparation

Preoperative preparation for nonemergency splenectomy includes:

- Correction of abnormalities of blood clotting and the number of red blood cells.
- Treatment of any infections.
- Control of immune reactions. Patients are usually given protective vaccinations about a month before surgery. The most common vaccines used are Pneumovax or Pnu-Imune 23 (against pneumococcal infections) and Menomune-A/C/Y/W-135 (against meningococcal infections).

Aftercare

Immediately following surgery, patients should follow the physician's instructions and take all medications intended to prevent infection. Blood transfusions may be indicated for some patients to replace defective blood cells. The most important part of aftercare, however, is long-term caution regarding vulnerability to infection. Patients should see their doctor at once if they have a **fever** or any other sign of infection, and avoid travel to areas where exposure to **malaria** or similar diseases is likely. Children with splenectomies may be kept on antibiotic therapy until they are 16 years old. All patients can be given a booster dose of pneumococcal vaccine 5 to 10 years after splenectomy.

Risks

The chief risk following splenectomy is overwhelming bacterial infection, or postsplenectomy **sepsis**. This vulnerability results from the body's decreased ability to clear bacteria from the blood, and lowered levels of a protein in blood plasma that helps to fight viruses (immunoglobulin M). The risk of dying from infection after splenectomy is highest in children, especially in the first two years after surgery. The risk of postsplenectomy sepsis can be reduced by vaccinations before the

KEY TERMS

Embolization—An alternative to splenectomy that involves injecting silicone or similar substances into the splenic artery to shrink the size of the spleen.

Hereditary spherocytosis (HS)—A blood disorder in which the red blood cells are relatively fragile and are damaged or destroyed when they pass through the spleen. Splenectomy is the only treatment for HS.

Hypersplenism—A syndrome marked by enlargement of the spleen, defects in one or more types of blood cells, and a high turnover of blood cells.

Immune or idiopathic thrombocytopenic purpura (ITP)—A blood disease that results in destruction of platelets, which are blood cells involved in clotting.

Laparoscope—An instrument used to view the abdominal cavity through a small incision and perform surgery on a small area, such as the spleen.

Pneumovax—A vaccine that is given to splenectomy patients to protect them against bacterial infections. Other vaccines include Pnu-Imune and Menomune.

Sepsis—A generalized infection of the body, most often caused by bacteria.

Sequestration—A process in which the spleen withdraws some normal blood cells from circulation and holds them in case the body needs extra blood in an emergency. In hypersplenism, the spleen sequesters too many blood cells.

Splenomegaly—Abnormal enlargement of the spleen.

Thromboembolism—A clot in the blood that forms and blocks a blood vessel. It can lead to infarction, or death of the surrounding tissue due to lack of blood supply.

operation. Some doctors also recommend a two-year course of penicillin following splenectomy or long-term treatment with ampicillin.

Other risks following splenectomy include inflammation of the pancreas and collapse of the lungs. In some cases, splenectomy does not address the underlying causes of splenomegaly or other conditions. Excessive bleeding after the operation is an additional possible complication, particularly for ITP patients. Infection immediately following surgery may also occur.

Normal results

Results depend on the reason for the operation. In blood disorders, the splenectomy will remove the cause of the blood cell destruction. Normal results for patients with an enlarged spleen are relief of pain and of the complications of splenomegaly. It is not always possible, however, to predict which patients will respond well or to what degree.

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National Heart Lung and Blood Institute Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, (240) 629-3246, <http://www.nhlbi.nih.gov>.

Teresa Odle
Rebecca J. Frey, PhD

Splenic trauma

Definition

Splenic trauma is physical injury to the spleen, the lymphatic organ located in the upper left side of the abdomen just under the rib cage. The spleen weighs between 75 and 150 grams (between 0.16 and 0.33 pounds) in adults.

Description

The spleen is an organ that produces white blood cells, filters the blood (10–15% of the total blood supply every minute), stores red blood cells and platelets, and destroys those that are **aging**. It is located near the stomach on the left side of the abdomen. A direct blow to the abdomen may bruise, tear or shatter the spleen. Trauma to the spleen can cause varying degrees of damage, the major problem associated with internal bleeding. Mild splenic subcapsular hematomas are injuries in which bleeding is limited to small areas on and immediately around the spleen. Splenic contusions refer to bruising and bleeding on and around larger areas of the spleen. Lacerations (tears) are the most common splenic trauma injuries. Tears tend to occur on the areas between the three main blood vessels of the spleen. Because of the abundant blood supply, splenic trauma may cause serious internal bleeding. Most injuries to the spleen in children heal spontaneously. Severe trauma can cause the spleen or its blood vessels to rupture or fragment.

Splenic trauma is more common in children than in adults. In general, children are prone to abdominal injuries due to accidents and falls and because their abdominal organs are less protected by bone, muscle and fat. Abdominal injuries including splenic trauma are the most common cause of preventable deaths in children.

Causes and symptoms

The most common cause of injury to the spleen is blunt abdominal trauma. Blunt trauma is often caused by a direct blow to the belly, car and motorcycle accidents, falls, sports mishaps, and fights. The spleen is the most commonly injured organ in blunt abdominal trauma; splenic injury occurs in nearly 25% of injuries of this type. Penetrating injuries such as those from stabbing, gunshot **wounds**, and accidental impaling also account for cases of splenic trauma, although far less frequently than blunt trauma.

In adults, ruptured spleens may have been preceded by conditions causing rapid splenic enlargement,

such as infections, particularly those caused by the **Epstein-Barr virus** (EBV); **cancer**; immune system disorders; diseases of the spleen; or circulatory problems. In a very few cases the spleen may be injured by a spell of violent coughing. This type of rupture is known as an atraumatic rupture.

A spleen that has become enlarged and fragile from disease is sometimes ruptured by a doctor or medical student in the course of palpating (feeling) the patient's abdomen, or damaged by a surgeon in the course of an operation on other abdominal organs.

Damage to the spleen may cause localized or general abdominal **pain**, tenderness, and swelling. Fractured ribs may be present. Splenic trauma may cause mild or severe internal bleeding, leading to **shock** and for which symptoms include rapid heartbeat, **shortness of breath**, thirst, pale or clammy skin, weak pulse, low blood pressure, **dizziness**, **fainting**, sweating. **Vomiting** blood, blood in the stools or urine, deterioration of vital signs, and loss of consciousness are other symptoms.

Diagnosis

The goal of diagnosis of all abdominal traumas is to detect and treat life-threatening injuries as quickly as possible. The physician will determine the extent of organ damage and whether surgery will be necessary while providing appropriate emergency care. Initial diagnosis consists of detailing all circumstances of the injury from the patient and bystanders as well as the close **physical examination** of the patient and measurement of vital signs. Blood tests, **urinalysis**, stool samples and x rays of the chest and abdomen are usually performed. Plain x rays may show abdominal air pockets that indicate internal ruptures, but are rarely helpful because they do not show splenic and intra-abdominal damage.

Several other diagnostic tests may be used for the noninvasive and accurate assessment of splenic damage: **computed tomography scans** (CT), of **magnetic resonance imaging** (MRI), radionuclide scanning, and ultrasonography. Ultrasonography—particularly focused abdominal sonographic technique (FAST)—has now become a standard bedside technique in many hospitals to check for bleeding in the abdomen. Imaging tests allow doctors to determine the necessity and type of surgery required. The CT scan has been shown to be the most available and accurate test for abdominal trauma. MRI tests are accurate but costly and less available in some hospitals, while radionuclide scanning requires more time and patient stability. Peritoneal lavage is another diagnostic technique in which

the abdominal cavity is entered and flushed to check for bleeding. When patients exhibit shock, infection, or prolonged internal bleeding, exploratory **laparoscopy** is used for emergency diagnosis.

Treatment

Not long ago nearly all cases of splenic trauma were treated by laparoscopy, opening the abdomen, and by **splenectomy**, the surgical removal of the spleen. This approach resulted from the difficulty in assessing the severity of the injury, the potential dangers of shock and **death**, and the beliefs that the spleen healed poorly and that it was not an important organ. Nowadays, improved techniques of diagnosis and monitoring (particularly the introduction of CT scans), as well as understanding that removal of the spleen creates future risk of a lowered capacity to fight infection, has modified treatment approaches. Research over the past two decades has shown that the spleen has high healing potential, and confirmed that children are more susceptible to infection after splenectomy (post splenectomy **sepsis**, PSS). PSS has a mortality rate of over 50% and standard procedure now avoids splenectomy as much as possible. Adult splenic trauma is treated by splenectomy more often than children's; for unknown reasons, the adult spleen more frequently spontaneously ruptures after injury. Adults are also less susceptible to PSS.

Nonoperative treatment

In nonoperative therapy, splenic trauma patients are monitored closely, often in intensive care units for several days. Fluid and blood levels are observed and maintained by intravenous fluid and possible blood transfusions. Follow-up scans may be used to observe the healing process.

Operative treatment

Splenic trauma patients require surgery when nonoperative treatment fails, when major or prolonged internal bleeding exists and for gunshot and many stab wounds. Whenever possible, surgeons try to preserve at least part of the spleen and try to repair its blood vessels.

Prognosis

The ample blood supply to the spleen can promote rapid healing. Studies have shown that intra-abdominal bleeding associated with splenic trauma stops without surgical intervention in up to two out of three cases in children. When trauma patients stabilize during nonoperative therapy, chances are high that surgery will be

KEY TERMS

Computed tomography (CT) scan—Computer-aided x-ray examination that allows cross-sectional views of organs and tissues.

Laparoscope—An optical or fiberoptic instrument that is inserted by incision in the abdominal wall and is used to view the interior of the peritoneal cavity.

Laparoscopy—Procedure using a laparoscope to view organs, obtain tissue samples and perform surgery.

Magnetic resonance imaging (MRI)—Imaging technique using magnets and radio waves to provide internal pictures of the body.

Radionuclide scanning—Diagnostic test in which a radioactive dye is injected into the bloodstream and photographed to display internal vessels, organs and tissues.

Ultrasonography—Imaging test using sound waves to view internal organs and tissues.

avoided and that spleen injuries will heal themselves. Splenic trauma patients undergoing diagnostic tests such as CT and MRI scans have improved chances of avoiding splenectomy and retaining whole or partial spleens.

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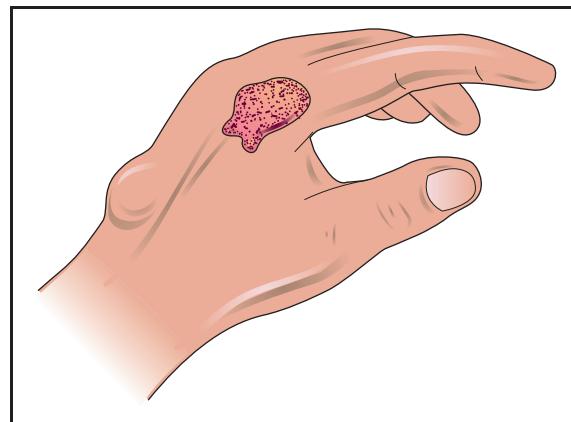
American Trauma Society, 8903 Presidential Pkwy Suite 512, Upper Marlboro, MD, 20227, (800) 556-7890.

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Split personality see **Multiple personality disorder**

Spontaneous abortion see **Miscarriage**

Sporothrix schenckii infection see **Sporotrichosis**



Sporotrichosis

Definition

Sporotrichosis is a chronic infection caused by the microscopic fungus *Sporothrix schenckii*. The disease causes ulcers on the skin that are painless but do not heal, as well as nodules or knots in the lymph channels near the surface of the body. Infrequently, sporotrichosis affects the lungs, joints, or central nervous system and can cause serious illness.

Description

The fungus that causes sporotrichosis is found in sphagnum moss, soil, and rotting vegetation. Anyone can get sporotrichosis but it is most common among nursery workers, farm laborers, and gardeners handling sphagnum moss, roses, or barberry bushes. Cases have also been reported in workers whose jobs took them under houses into crawl spaces contaminated with the fungus. Children who played on baled hay have also gotten the disease. Sporotrichosis is sometimes called sphagnum moss disease or alcoholic rose gardener's disease.

Causes and symptoms

The fungus causing sporotrichosis enters the body through scratches or cuts in the skin. Therefore, people who handle plants with sharp thorns or needles, like roses, barberry, or pines, are more likely to get sporotrichosis. Sporotrichosis is not passed directly from person to person, so it is not possible to catch sporotrichosis from another person who has it.

The first signs of sporotrichosis are painless pink, red, or purple bumps usually on the finger, hand, or arm where the fungus entered the body. These bumps may appear anywhere from one to 12 weeks after infection, but usually appear within three weeks. Unlike many

Sporotrichosis is a chronic infection caused by the microscopic fungus *Sporothrix schenckii*. It produces ulcers on the skin that are painless but do not heal, and nodules or knots in the lymph channels near the surface of the body.

(Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

other fungal infections sporotrichosis does not cause **fever** or any feelings of general ill health.

The reddish bumps eventually expand and fester, creating skin ulcers that do not heal. In addition, the infection often moves to nearby lymph nodes. Although most cases of sporotrichosis are limited to the skin and lymph channels, occasionally the joints, lungs, and central nervous system become infected. In rare cases, **death** may result.

People who have weakened immune systems, either from a disease such as acquired immune deficiency syndrome (**AIDS**) or leukemia, or as the result of medications they take (**corticosteroids, chemotherapy** drugs), are more likely to get sporotrichosis and are more at risk for the disease to spread to the internal organs. Alcoholics and people with **diabetes mellitus** or a pre-existing lung disease are also more likely to become infected. Although sporotrichosis is painless, it is important for people with symptoms to see a doctor and receive treatment.

Diagnosis

The preferred way to diagnose sporotrichosis is for a doctor to obtain a sample of fluid from a freshly opened sore and send it to a laboratory to be cultured. The procedure is fast and painless. It is possible to confirm the presence of advanced sporotrichosis through a blood test or a biopsy. Doctors may also take a blood sample to perform tests that rule out other fungal infections or diseases such as **tuberculosis** or bacterial **osteomyelitis**.

Dermatologists and doctors who work with AIDS patients are more likely to have experience in diagnosing sporotrichosis. In at least one state, New York, the laboratory test to confirm this disease is provided free through the state health department. In other cases, diagnosis should be covered by health insurance at the same level as other diagnostic laboratory tests.

Treatment

When sporotrichosis is limited to the skin and lymph system, it is usually treated with a saturated solution of potassium iodine that the patient dilutes with water or juice and drinks several times a day. The iodine solution can only be prescribed by a physician. This treatment must be continued for many weeks. Skin ulcers should be treated like any open wound and covered with a clean bandage to prevent a secondary bacterial infection. The drug itraconazol (Sporanox), taken orally, is also available to treat sporotrichosis.

In serious cases of sporotrichosis, when the internal organs are infected, the preferred treatment is the drug amphotericin B. Amphotericin B is a strong anti-fungal drug with potentially severe toxic side effects. It is given intravenously so hospitalization is required for treatment. The patient may also receive other drugs to minimize the side effects of the amphotericin B.

Alternative treatment

Alternative treatment for fungal infections focuses on maintaining general good health and eating a diet low in dairy products, sugars, including honey and fruit juice, and foods, such as beer, that contain yeast. This is complemented by a diet high in raw food. Supplements of vitamins C, E, and A, B complex, and pantothenic acid may also be added to the diet, as may *Lactobacillus acidophilus*, bifidobacteria, and garlic capsules.

Fungicidal herbs such as myrrh (*Commiphora molmol*), tea tree oil (*Melaleuca* spp.), citrus seed extract, pau d'arco tea, and garlic (*Allium sativum*) may also be applied directly to the infected skin.

Prognosis

Most cases of sporotrichosis are confined to the skin and lymph system. With treatment, skin sores begin healing in one to two months but complete recovery often takes six months or more. People who have AIDS are also more likely to have the fungus spread throughout the body, causing a life-threatening infection. In people whose bones and joints are infected or who have pulmonary lesions, surgery may be necessary.

KEY TERMS

Acidophilus—The bacteria *Lactobacillus acidophilus*, usually found in yogurt.

Bacterial osteomyelitis—An infection of the bone or bone marrow that is caused by a bacterium.

Bifidobacteria—A group of bacteria normally present in the intestine. Commercial supplements are available.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Lymph channels—The vessels that transport lymph throughout the body. Lymph is a clear fluid that contains cells important in forming antibodies that fight infection.

Prevention

Since an opening in the skin is necessary for the sporotrichosis fungus to enter the body, the best way to prevent the disease is to avoid accidental scrapes and cuts on the hands and arms by wearing gloves and long sleeves while gardening. Washing hands and arms well after working with roses, barberry, sphagnum moss, and other potential sources of the fungus may also provide some protection.

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Tish Davidson, A.M.

Sports injuries

Definition

Sports injuries result from acute trauma or repetitive **stress** associated with athletic activities. Sports injuries can affect bones or soft tissue (ligaments, muscles, tendons).

Professional dancers are increasingly recognized as performing athletes, and many of the treatments



A soccer player grips his shin after being injured during a match. (Manuel Queimadelos Alonso/Getty Images.)

and preventive measures utilized in sports medicine are now applied to dance-related injuries.

It is also important to remember that many types of injuries that affect athletes may also occur in workers in certain occupations; for example, many people in the building trades develop **tennis elbow** or golfer's elbow. The principles of sports medicine can be applied in the treatment of most common musculoskeletal injuries.

Description

Adults are less likely to suffer sports injuries than children, whose vulnerability is heightened by immature reflexes, an inability to recognize and evaluate risks, and underdeveloped coordination.

In 2002, about 20.3 million Americans suffered a sports injury. Of those, 53% were minor enough to be self-treated or left untreated. However, about 10 million Americans annually receive medical attention for their sports-related injuries. That equates to almost 26

per 1,000 people. The highest rate is among children age five to 14 years old (59.3 per 1,000 people). As many as 20% of children who play sports get hurt, and about 25% of their injuries are classified as serious. Boys aged 12 to 17 are the highest risk group. More than 775,000 boys and girls under age 14 are treated in hospital emergency rooms for sports-related injuries.

Injury rates are highest for athletes who participate in contact sports, but the most serious injuries are associated with individual activities. Between one-half and two-thirds of childhood sports injuries occur during practice, or in the course of unorganized athletic activity.

Baseball and softball are the leading causes of sports-related facial trauma in the United States, with 68% of these injuries caused by contact with the ball rather than player-player collision or being hit by a swung bat.

Types of sports injuries

About 95% of sports injuries are minor soft tissue traumas.

The most common sports injury is a bruise (contusion). It is caused when blood collects at the site of an injury and discolors the skin.

Sprains account for one-third of all sports injuries. A sprain is a partial or complete tear of a ligament, a strong band of tissue that connects bones to one another and stabilizes joints.

A strain is a partial or complete tear of:

- muscle (tissue composed of cells that enable the body to move)
- tendon (strong connective tissue that links muscles to bones)

Inflammation of a tendon (**tendinitis**) and inflammation of one of the fluid-filled sacs that allow tendons to move easily over bones (**bursitis**) usually result from minor stresses that repeatedly aggravate the same part of the body. These conditions often occur at the same time.

SKELETAL INJURIES. **Fractures** account for 5–6% of all sports injuries. The bones of the arms and legs are most apt to be broken. Sports activities rarely involve fractures of the spine or skull. The bones of the legs and feet are most susceptible to stress fractures, which occur when muscle strains or contractions make bones bend. Stress fractures are especially common in ballet dancers, long-distance runners, and in people whose bones are thin.

Shin splints are characterized by soreness and slight swelling of the front, inside, and back of the lower leg, and by sharp **pain** that develops while exercising and gradually intensifies. Shin splints are caused by overuse or by stress fractures that result from the repeated foot pounding associated with activities such as aerobics, long-distance running, basketball, and volleyball.

A compartment syndrome is a potentially debilitating condition in which the muscles of the lower leg grow too large to be contained within membranes that enclose them. This condition is characterized by **numbness and tingling**. Untreated compartment syndrome can result in long-term loss of function.

BRAIN INJURIES. Brain injury is the primary cause of fatal sports-related injuries. **Concussion**, which is also called mild traumatic brain injury or MTBI, can result from even minor blows to the head. A concussion can cause loss of consciousness and may affect:

- balance
- comprehension
- coordination
- hearing

- memory
- vision

Causes and symptoms

Common causes of sports injuries include:

- athletic equipment that malfunctions or is used incorrectly
- falls
- forceful high-speed collisions between players
- wear and tear on areas of the body that are continually subjected to stress

Symptoms include:

- instability or obvious dislocation of a joint
- pain
- swelling
- weakness

Diagnosis

Symptoms that persist, intensify, or reduce the athlete's ability to play without pain should be evaluated by an orthopedic surgeon. Prompt diagnosis often can prevent minor injuries from becoming major problems, or causing long-term damage.

An orthopedic surgeon should examine anyone:

- who is prevented from playing by severe pain associated with acute injury
- whose ability to play has declined due to chronic or long-term consequences of an injury
- whose injury has caused visible deformities in an arm or leg.

The physician will perform a **physical examination**, ask how the injury occurred, and what symptoms the patient has experienced. X rays and other imaging studies of bones and soft tissues may be ordered.

Anyone who has suffered a blow to the head should be examined immediately, and at five-minute intervals until normal comprehension has returned. The initial examination measures the athlete's:

- awareness
- concentration
- short-term memory

Subsequent evaluations of concussion assess:

- dizziness
- headache
- nausea
- visual disturbances

Treatment

Treatment for minor soft tissue injuries generally consists of:

- compressing the injured area with an elastic bandage
- elevation
- ice
- rest.

Anti-inflammatories, taken by mouth or injected into the swelling, may be used to treat bursitis. Anti-inflammatory medications and exercises to correct muscle imbalances usually are used to treat tendinitis. If the athlete keeps stressing inflamed tendons, they may rupture, and casting or surgery is sometimes necessary to correct this condition.

Orthopedic surgery may be required to repair serious sprains and strains.

Controlling inflammation as well as restoring normal use and mobility are the goals of treatment for overuse injuries.

Athletes who have been injured are usually advised to limit their activities until their injuries are healed. The physician may suggest special exercises or behavior modifications for athletes who have had several injuries. Athletes who have been severely injured may be advised to stop playing altogether.

Prevention

Every child who plans to participate in organized athletic activity should have a pre-season sports physical. This special examination is performed by a pediatrician or family physician who:

- carefully evaluates the site of any previous injury
- may recommend special stretching and strengthening exercises to help growing athletes create and preserve proper muscle and joint interaction
- pays special attention to the cardiovascular and skeletal systems.

Telling the physician which sport the athlete plays will help that physician determine which parts of the body will be subjected to the most stress. The physician then will be able to suggest to the athlete steps to take to minimize the chance of getting hurt.

Other injury-reducing game plans include:

- being in shape
- knowing and obeying the rules that regulate the activity
- not playing when tired, ill, or in pain

- not using steroids, which can improve athletic performance but cause life-threatening problems
- taking good care of athletic equipment and using it properly
- wearing appropriate protective equipment

On a larger scale, sports injuries are becoming a public health concern in America. Prevention efforts include wearing protective devices (such as bicycle helmets and pads when skating or skateboarding), and educating both children and adults about safety. Other preventive efforts include changes in the rules of the game or sport to minimize injuries. For example, wearing goggles will be mandatory in women's lacrosse in order to reverse the rising rate of eye and other facial injuries in that sport. Research also continues on improving equipment. For example, thick rubber insoles can help prevent against repetitive injuries from running but scientists recently observed that they can add to injuries in sports such as soccer, where athletes need to make quick changes of direction. On the other hand, recent improvements in the design and construction of football helmets have been credited with a significant decline in the frequency and severity of head injuries among football players.

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ORGANIZATIONS

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Sports vision see **Vision training**

Spouse abuse see **Abuse**

Sprains and strains

Definition

Sprain refers to damage or tearing of ligaments or a joint capsule. Strain refers to damage or tearing of a muscle.

Description

When excessive force is applied to a joint, the ligaments that hold the bones together may be torn or damaged. This results in a sprain and its seriousness depends on how badly the ligaments are torn. Any joint can be sprained but the most frequently injured joints are the ankle, knee, and finger.

Strains are tears in the muscle. Sometimes called pulled muscles, they usually occur because of over-exertion or improper lifting techniques. Sprains and strains are common. Anyone can have them.

Children under age eight are less likely to have sprains than are older people. Children's ligaments are tighter and their bones are more apt to break before a ligament tears. People who are active in sports suffer more strains and sprains than less active people. Repeated sprains in the same joint make the joint less stable and more prone to future sprains.

Causes and symptoms

There are three grades of sprains. Grade I sprains are mild injuries where there is no tearing of the

ligament and no joint function is lost, although there may be tenderness and slight swelling.

Grade II sprains are caused by a partial tear in the ligament. These sprains are characterized by obvious swelling, extensive bruising, **pain**, difficulty bearing weight, and reduced function of the joint.

Grade III, or third degree, sprains are caused by complete tearing of the ligament where there is severe pain, loss of joint function, widespread swelling and bruising, and the inability to bear weight. These symptoms are similar to those of bone **fractures**.

Strains can range from mild muscle stiffness to great soreness. Strains result from overuse of muscles, improper use of the muscles, or as the result of injury in another part of the body when the body compensates for pain by altering the way it moves.

Diagnosis

Grade I sprains and mild strains are usually self-diagnosed. Grade II and III sprains are often seen by a physician, who takes x rays of the area to differentiate between a sprain and a fracture.

Treatment

Grade I sprains and mild strains can be treated at home. Basic **first aid** for sprains consists of RICE: Rest, Ice for 48 hours, Compression (wrapping in an elastic bandage), and Elevation of the sprain above the level of the heart. Over-the-counter pain medication such as **acetaminophen** (Tylenol) or ibuprofen (Motrin) can be taken for pain.

In addition to RICE, people with grade II and grade III sprains in the ankle or knee usually need to use crutches until the sprains have healed enough to bear weight. Sometimes, **physical therapy** or home exercises are needed to restore the strength and flexibility of the joint.

Grade III sprains are usually immobilized in a cast for several weeks to see if the sprain heals. Pain medication is prescribed. Surgery may be necessary to relieve pain and restore function. Athletic people under age 40 are the most likely candidates for surgery, especially with grade III knee sprains. For complete healing, physical therapy usually will follow surgery.

Alternative treatment

Alternative practitioners endorse RICE and conventional treatments. In addition, nutritional therapists recommend vitamin C and bioflavonoids to supplement a diet high in whole grains, fresh fruits, and vegetables. Anti-inflammatories, such as bromelain

KEY TERMS

Ligament—Tough, fibrous connective tissue that holds bones together at joints.

(a proteolytic enzyme from pineapples) and turmeric (*Curcuma longa*), may also be helpful. The homeopathic remedy arnica (*Arnica montana*) may be used initially for a few days, followed by ruta (*Ruta graveolens*) for joint-related injuries or *Rhus toxicodendron* for muscle-related injuries. If surgery is needed, alternative practitioners can recommend pre- and post-surgical therapies that will enhance healing.

Prognosis

Moderate sprains heal within two to four weeks but it can take months to recover from severe ligament tears. Until recently, tearing the ligaments of the knee meant the end to an athlete's career. Improved surgical and rehabilitative techniques now offer the possibility of complete recovery. However, once a joint has been sprained, it will never be as strong as it was before.

Prevention

Sprains and strains can be prevented by warming-up before exercising, using proper lifting techniques, wearing properly fitting shoes, and taping or bracing the joint.

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Tish Davidson, A.M.

Sputum culture

Definition

Sputum is material coughed up from the lungs and expectorated (spit out) through the mouth. A sputum culture is done to find and identify the microorganism causing an infection of the lower respiratory tract such as **pneumonia** (an infection of the lung). If a microorganism is found, more testing is done to determine which **antibiotics** will be effective in treating the infection.

Purpose

A person with a **fever** and a continuing **cough** that produces pus-like material and/or blood may have an infection of the lower respiratory tract. Infections of the lungs and bronchial tubes are caused by several types of microorganisms, including bacteria, fungi (molds and yeast), and viruses. A **chest x ray** provides visual evidence of an infection; a culture can grow the microorganism causing the infection. The microorganism is grown in the laboratory so it can be identified and tested for its response to medications, such as antifungals and antibiotics.

Description

Based on the clinical condition of the patient, the physician determines what group of microorganism is likely to be causing the infection, and then orders one or more specific types of cultures: bacterial, viral, or fungal (for yeast and molds). For all culture types, the sputum must be collected into a sterile container. The sputum specimen must be collected carefully so that bacteria that normally live in the mouth and saliva do not contaminate the sputum and complicate the process of identifying the cause of the infectious agent. Once in the laboratory, each culture type is handled differently.

Bacterial culture

A portion of the sputum is smeared on a microscope slide for a Gram stain. Another portion is spread over the surface of several different types of culture plates, and placed in an incubator at body temperature for one to two days.

A Gram stain is done by staining the slide with purple and red stains, then examining it under a microscope. Gram staining checks that the specimen does not contain saliva or material from the mouth. If many epithelial (skin) cells and few white blood cells are seen, the specimen is not pure sputum and is not adequate for culture. Depending on laboratory policy, the specimen may be rejected and a new specimen requested. If many white blood cells and bacteria of one type are seen, this is an early confirmation of infection. The color of stain picked up by the bacteria (purple or red), their shape (such as round or rectangular), and their size provide valuable clues as to their identity and helps the physician predict what antibiotics might work best before the entire test is completed. Bacteria that stain purple are called gram-positive; those that stain red are called gram-negative.

During incubation, bacteria present in the sputum sample multiply and will appear on the plates as visible

colonies. The bacteria are identified by the appearance of their colonies, by the results of biochemical tests, and through a Gram stain of part of a colony.

A sensitivity test, also called antibiotic susceptibility test, is also done. The bacteria are tested against different antibiotics to determine which will treat the infection by killing the bacteria.

The initial result of the Gram stain is available the same day or in less than an hour if requested by the physician. An early report, known as a preliminary report, is usually available after one day. This report will tell if any bacteria have been found yet, and if so, their Gram stain appearance—for example, a gram-negative rod, or a gram-positive cocci. The final report, usually available in one to three days, includes complete identification and an estimate of the quantity of the bacteria and a list of the antibiotics to which they are sensitive.

Fungal culture

To look for mold or yeast, a fungal culture is done. The sputum sample is spread on special culture plates that will encourage the growth of mold and yeast. Different biochemical tests and stains are used to identify molds and yeast. Cultures for fungi may take several weeks.

Viral culture

Viruses are a common cause of pneumonia. For a viral culture, sputum is mixed with commercially-prepared animal cells in a test tube. Characteristic changes to the cells caused by the growing virus help identify the virus. The time to complete a viral culture varies with the type of virus. It may take from several days to several weeks.

Special procedures

Tuberculosis is caused by a slow-growing bacteria called *Mycobacterium tuberculosis*. Because it does not easily grow using routine culture methods, special procedures are used to grow and identify this bacteria. When a sputum sample for tuberculosis first comes into the laboratory, a small portion of the sputum is smeared on a microscope slide and stained with a special stain, called an acid-fast stain. The stained sputum is examined under a microscope for tuberculosis organisms, which pick-up the stain, making them visible. This smear is a rapid screen for the organism and allows the physician to receive a preliminary report within 24 hours.

To culture for tuberculosis, portions of the sputum are spread on and placed into special culture

plates and tubes of broth that promote the growth of the organism. Growth in broth is faster than growth on culture plates. Instruments are available that can detect growth in broth, speeding the process even further. Growth and identification may take two to four weeks.

Other microorganisms that cause various types of lower respiratory tract infections also require special culture procedures to grow and identify. *Mycoplasma pneumoniae* causes a mild to moderate form of pneumonia, commonly called walking pneumonia; *Bordetella pertussis* causes **whooping cough**; *Legionella pneumophila*, Legionnaire's disease; *Chlamydia pneumoniae*, an atypical pneumonia; and *Chlamydia psittaci*, parrot fever.

Pneumocystis carinii causes pneumonia in people with weakened immune systems, such as people with **AIDS**. This organism does not grow in culture. Special stains are done on sputum when pneumonia caused by this organism is suspected. The diagnosis is based on the results of these stains, the patient's symptoms, and medical history.

Sputum culture is also called sputum culture and sensitivity.

It is possible that sputum cultures will eventually be replaced in the diagnosis of tuberculosis by newer molecular techniques. These advanced methods speed the diagnostic process as well as improve its accuracy. As of late 2002, four molecular techniques are increasingly used in laboratories around the world to diagnose TB. They include polymerase chain reaction to detect mycobacterial DNA in patient specimens; nucleic acid probes to identify mycobacteria in culture; restriction fragment length polymorphism analysis to compare different strains of TB for epidemiological studies; and genetic-based susceptibility testing to identify drug-resistant strains of mycobacteria.

Preparation

The specimen for culture should be collected before antibiotics are begun. Antibiotics in the person's system may prevent microorganisms present in the sputum from growing in culture.

The best time to collect a sputum sample is early in the morning, before having anything to eat or drink. The patient should first rinse his or her mouth with water to decrease mouth bacteria and dilute saliva. Through a deep cough, the patient must cough up sputum from within the chest. Taking deep breaths and lowering the head helps bring up the sputum. Sputum must not be held in the mouth but immediately spat into a sterile container. For tuberculosis, the

KEY TERMS

Acid-fast stain—A special stain done to microscopically identify the bacteria that cause tuberculosis.

Culture—A laboratory test done to grow and identify microorganisms causing infection.

Gram stain—Microscopic examination of a portion of a bacterial colony or sample from an infection site after it has been stained by special stains. Certain bacteria pick up and retain the purple stain; these bacteria are called gram-positive. Other bacteria lose the purple stain and retain the red stain; these bacteria are called gram-negative. The color of the bacteria, in addition to their size and shape, provide clues as to the identity of the bacteria.

Normal flora—The mixture of bacteria normally found at specific body sites.

Pneumonia—An infection of the lungs.

Sputum—Material coughed up from the lower respiratory tract and expectorated through the mouth.

Sensitivity test—A test that determines which antibiotics will kill the bacteria that has been isolated from a culture.

physician may want the patient to collect sputum samples on three consecutive mornings.

If coughing up sputum is difficult, a health care worker can have the patient breathe in sterile saline produced by a nebulizer. This nebulized saline coats the respiratory tract, loosening the sputum, and making it easier to cough up. Sputum may also be collected by a physician during a **bronchoscopy** procedure. Bronchoscopy, however, is not regarded as a cost-effective way of obtaining a useful sample.

If tuberculosis is suspected, collection of sputum should be carried out in an **isolation** room, with all attending healthcare workers wearing masks.

In addition to special precautions in collecting sputum when tuberculosis is suspected, workers in hospital laboratories must take extra care to inactivate unstained smear preparations that may contain *M. tuberculosis*. The most effective deactivation technique is the use of a solution of 5% phenol in ethanol.

Normal results

Sputum from a healthy person would have no growth on culture. A mixture of microorganisms,

however, normally found in a person's mouth and saliva often contaminate the culture. If these microorganisms grow in the culture, they may be reported as normal flora contamination.

Abnormal results

The presence of bacteria and white blood cells on the Gram stain and the isolation of a microorganism from culture, other than normal flora contamination, is evidence of a lower respiratory tract infection.

Microorganisms commonly isolated from sputum include: *Streptococcus pneumoniae*, *Haemophilus influenzae*, *Staphylococcus aureus*, *Legionella pneumophila*, *Mycoplasma pneumoniae*, *Klebsiella pneumoniae*, *Pseudomonas aeruginosa*, *Bordetella pertussis*, and *Escherichia coli*.

Resources

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Squamous cell carcinoma of the skin

Definition

A squamous cell carcinoma is a skin **cancer** that originates from squamous keratinocytes in the epidermis, the top layer of the skin. *Squamous* is a term that indicates a surface with a scaly nature.

Description

Squamous keratinocytes are flattened unpigmented skin cells in the middle of the epidermis. When they become cancerous, these cells invade the dermis (the layer of skin just below the epidermis) and spread out into the normal skin. They become visible as a small growth or area of change in the skin's appearance.

Most squamous cell carcinomas appear on areas that have been exposed to the sun: the head and neck, forearms, backs of the hands, upper part of the torso, and lower legs. Many develop in precancerous patches called actinic keratoses. Actinic keratoses are rough, scaly patches on the skin that usually start to show up in middle age. They are associated with a lifetime's exposure to the sun. Estimates of the chance that an actinic keratosis will turn into a squamous cell carcinoma vary from 0.24 to 20%.

Squamous cell carcinomas can also originate in old **scars** and **burns**, long-standing sores, and other areas of chronic skin irritation. These tumors tend to be more dangerous than those that arise in actinic keratoses.



Squamous cell carcinoma on the nose. (Dr P. Marazzi/Photo Researchers, Inc.)

The least dangerous type of squamous cell carcinoma is called Bowen's disease, intraepithelial squamous cell carcinoma, or squamous cell carcinoma *in situ*. Bowen's disease can show up anywhere on the skin but it is especially common on the head and neck. This cancer usually grows slowly but may evolve into a more serious, spreading form if it is not removed.

Other types of squamous cell carcinomas grow fairly quickly and can develop within a few months. These tumors may spread in the skin along the blood vessels, nerves, and muscles. They can also metastasize, or spread to other areas. On the average, 2–6% of squamous cell carcinomas metastasize but the rate varies with the tumor site. At least 95% of the tumors that originate in actinic keratoses remain in the skin; but up to 38% of the cancers from scars are metastatic. Metastasis is also more likely when the cancer originates on the ear, lip, or genitalia, is large or deep, or develops in someone with a severely suppressed immune system. Cancers that regrow after treatment, and tumors that spread along the nerves are particularly dangerous.

Demographics

Squamous cell carcinoma is the second most common type of skin cancer in North America. There are between 80,000 and 100,000 cases diagnosed each year in the United States.

Squamous cell carcinomas are more common in the older adult population rather than the young. Overall, the chance of developing one is about 7–11%. The likelihood increases with exposure to the sun, and is greatest for fair-skinned individuals who tan poorly. Living near the equator, where ultraviolet light is more intense, also increases the risk. A weakened immune system—for instance, from an organ transplant, or AIDS—can also increase the risk of developing a squamous cell carcinoma by a factor of 5 to 250.

Squamous cell carcinomas tend to be most dangerous in individuals with dark skin. The mortality rate for African-Americans with squamous cell carcinomas is 17–24%, much higher than the 2% **death** rate for white males with nonmelanoma skin cancer. One reason for this disparity is that the cancers that develop in dark skin are more likely to come from old scars and burns than from actinic keratoses.

Causes and symptoms

Squamous cell carcinoma is caused by genetic damage to a skin cell. A number of factors can increase the risk that this will happen, but the exact cause is rarely known.

KEY TERMS

Actinic keratosis (plural actinic keratoses)—A rough, dry, scaly patch on the skin associated with sun exposure.

Albinism—A genetic disease characterized by the absence of the normal skin pigment, melanin.

Antioxidant—A substance that can neutralize free radicals. Free radicals are damaging molecules formed from oxygen. Antioxidant vitamins include vitamin E, C, and beta-carotene, a form of vitamin A.

Biopsy—A sample of an organ taken to look for abnormalities. Also, the technique used to take such samples.

Chronic—Long-standing.

Dermis—A layer of skin sandwiched between the epidermis and the fat under the skin. It contains the blood vessels, nerves, sweat glands, and hair follicles.

Epidermis—The thin layer of skin cells at the surface of the skin.

Fluorouracil—A cancer drug.

Interferon alpha—A chemical made naturally by the immune system and also manufactured as a drug.

Local anesthetic—A liquid used to numb a small area of the skin.

Lymph node—A small organ full of immune cells, found in clusters throughout the body. Lymph nodes are where reactions to infections usually begin.

Nonmelanoma skin cancer—A squamous cell carcinoma or basal cell carcinoma.

Nonsteroidal anti-inflammatory drugs (NSAIDS)—A class of drugs that suppresses inflammation. Includes a wide variety of drugs, including aspirin.

Papillomavirus—A member of a group of viruses associated with warts and cervical cancer.

Pathologist—A doctor who specializes in examining cells and other parts of the body for abnormalities.

Precancerous—Abnormal and with a high probability of turning into cancer, but not yet a cancer.

Oncologist—A doctor who specializes in the treatment of cancer.

Retinoids—A class of drugs related to vitamin A.

Selenium—A mineral needed in extremely small quantities by the body. Large amounts can be very toxic.

Xeroderma pigmentosum—A genetic disease characterized by the inability to repair damaged DNA. Individuals with this disease develop an excessive number of skin cancers.

Any of the following changes may be a warning sign that an actinic keratosis is developing into a squamous cell carcinoma:

- pain
- increased redness
- sores or bleeding
- hardening or thickening
- increased size

Most squamous cell carcinomas begin as a small red bump on the skin. More advanced squamous cell carcinomas have the following characteristics:

- a few millimeters to a few centimeters in diameter
- reddish-brown, flesh-colored, pink, or red
- bumpy or flat
- sharp, irregular edges in Bowen's disease; others may have no definite edge
- may be crusted or scaly
- may contain bleeding sores

Diagnosis

Squamous cell carcinomas are usually diagnosed with a **skin biopsy** taken in the doctor's office. This is generally a brief, simple procedure. After numbing the skin with an injection of local anesthetic, the doctor snips out the tumor or a piece of it. This skin sample is sent to a pathologist to be read. It can take up to a week for the biopsy results to come back. Squamous cell carcinomas are graded into categories of one through four. The grading is based on how deeply the tumor penetrates in the skin and how abnormal its cells are. Higher grades are more serious.

Treatment team

Primary care physicians remove some squamous cell carcinomas; other cancers, including larger or more complicated tumors, may be referred to a dermatologist. The services of a plastic surgeon are occasionally necessary. Metastatic tumors are often treated by an oncologist, surgeons, specially trained nurses, and specialists in radiation treatment.

Clinical staging, treatments, and prognosis

Staging

In stage 0 (Bowen's disease), the cancer is very small and has not yet spread from the epidermis to the dermis.

In stage I, the cancer is less than 2 cm (0.8 inches) in diameter. No cancer cells can be found in lymph nodes or other internal organs.

In stage II, the cancer is more than 2 cm (0.8 inches) in diameter. No cancer cells can be found in lymph nodes or other internal organs.

In stage III, cancer cells have been found in nearby lymph nodes or in the bone, muscle, or cartilage beneath the skin.

A stage IV cancer can be any size. In this stage, cancer cells have been discovered in internal organs that are distant from the skin. Squamous cell carcinomas tend to spread to nearby lymph nodes, the liver, and the lungs.

Treatment

The treatment options for a squamous cell carcinoma depend on the size of the tumor, its location, and the likelihood that it will spread aggressively or metastasize. All of the treatments described below generally have cure rates of approximately 90 to 99% for small, localized cancers. The five-year cure rates are highest with Moh's surgery, also called Mohs micrographic surgery.

One option is conventional surgery. The doctor numbs the area with an injection of local anesthetic, then cuts out the tumor and a small margin of normal skin around it. The wound is closed with a few stitches. One advantage of conventional surgery is that the wound usually heals quickly. Another benefit is that the complete cancer can be sent to a pathologist for evaluation. If cancer cells are found in the skin around the tumor, additional treatments can be done.

Laser surgery may be an alternative. A disadvantage to laser surgery is that the **wounds** from some lasers heal more slowly than cuts from a scalpel. The advantage is that bleeding is minimal.

Another option is Moh's micrographic surgery. This technique is a variation of conventional surgery. In this procedure, the surgeon examines each piece of skin under the microscope as it is removed. If any cancer cells remain, another slice is taken from that area and checked. These steps are repeated until the edges of the wound are clear of tumor cells, then the wound is closed. The advantage to this technique is that all of the visible cancer cells are removed but as

much normal skin as possible is spared. Mohs surgery is often used for larger or higher risk tumors and when cosmetic considerations are important. The main disadvantage is that it takes much longer than conventional surgery and requires a specially trained surgeon.

In cryosurgery, liquid nitrogen is used to freeze the tumor and destroy it. This treatment is another type of blind destruction; there is no skin sample to make sure the cancer cells have all been killed. Patients report swelling and **pain** after cryosurgery and a wound appears a few days later where the cells were destroyed. Healing takes about four to six weeks. When the site heals, it has usually lost its normal pigment. There is a risk of nerve damage with this technique. Cryosurgery is generally used only for small cancers in stage 0 and stage I.

In electro dessication and curettage, the physician scoops out the cancer cells with a spoon-shaped instrument called a curette. After most of the tumor is gone, the rest is destroyed with heat from an electrical current. The wound is left open to heal like an abrasion. It leaks fluid, crusts over, and heals during the next two to six weeks. This method is generally used only for the smallest squamous cell carcinomas (stage 0 and stage I). One disadvantage is that there is no skin sample to confirm that the tumor is completely gone. The electrical current used during this surgery can interfere with some **pacemakers**.

Some cases of Bowen's disease can be treated by applying a lotion containing 5-fluorouracil (fluorouracil or 5-FU) for several weeks. This treatment usually gives good cosmetic results. The side effects from 5-fluorouracil include **allergies** to the ingredients, infections, redness, peeling, and crusting, sensitivity to the sun, and changes in skin color. The main disadvantage to this treatment is that the drug cannot penetrate very far and cancer cells in the deeper parts of the tumor may not be destroyed.

Radiation therapy is sometimes used for squamous cell carcinomas, especially when the tumor is at a site where surgery would be difficult or remove a sizeable amount of tissue. This treatment is sometimes combined with surgery for cancers that have metastasized or are likely to. One disadvantage is that tumors returning after radiation tend to grow more quickly than the original cancer. In addition, x rays may promote new skin cancers. The cosmetic results are usually good. In some cases the skin may lose a little pigment, or develop spider veins. Some doctors reserve radiation treatment for those over 60. One drawback of radiation therapy for squamous cell carcinomas in or near the

mouth is that the radiation may cause the tissues inside the mouth to break down.

Chemotherapy is often added to surgery or radiation for stage IV cancers. Retinoids and interferon are experimental treatments that may be helpful.

Prognosis

Because many squamous cell carcinomas are not staged, precise five-year survival rates for each stage are not available. In general, the prognosis is very good for small squamous cell carcinomas that originate in actinic keratoses. However, cancers that were not completely destroyed may regrow. Tumors can redevelop in the scar from the surgery, on the edges of the surgery site, or deep in the skin. Larger or higher-risk tumors, cancers that regrow after treatment, and tumors that have invaded local tissues or metastasized are more difficult to cure. Most metastases show up within the first two years after a skin tumor has been removed. The five-year survival rate for metastatic cancers is 34%.

Alternative and complementary therapies

Alternative treatments for squamous cell carcinoma usually attempt to prevent rather than treat this cancer. Options being tested include antioxidant vitamins, minerals, and green tea extracts.

Coping with cancer treatment

Most squamous cell carcinomas are removed with techniques that cause few, if any, lasting side effects. Patients who have cosmetic concerns may wish to discuss them with their doctors.

Clinical trials

The medical community considers the following treatments to be experimental.

Clinical trials are testing whether interferon alpha, injected into the tumor, can destroy some squamous cell carcinomas. An early report from a combination of interferon alpha and retinoids is promising.

Ongoing trials are also evaluating whether small squamous cell carcinomas can be cured with photodynamic laser therapy. In this technique, a dye activated by laser light destroys the cancer. This dye is spread onto the skin, injected, or drunk. During a waiting period, normal cells clear the dye, then a laser activates the remainder. This technique is only useful for cancers very near the surface of the skin. One side effect after treatment is a period of excessive sun-sensitivity.

Other clinical trials are testing whether retinoids spread onto the skin can prevent or treat squamous cell carcinoma.

Another new experimental approach to squamous cell carcinoma is **gene therapy**. Researchers in Texas reported in 2003 on a Phase III investigation that uses an adenovirus as a vector to carry an altered p53 gene into the cancerous squamous cells. The function of the p53 gene is to maintain the structure of the cell's DNA and to induce the cell to die if its DNA is damaged beyond repair. Phase I and phase II trials have indicated that this approach to treatment has lengthened the survival time in patients with recurrent squamous cell carcinoma.

Prevention

The most important risk factor for squamous cell carcinoma is exposure to the sun (or other source of ultraviolet light) combined with a lighter complexion and inability to tan. Other risk factors include:

- increasing age
- actinic keratoses
- a previous skin cancer
- exposure to arsenic or the chemicals in coal tars
- radiation treatments
- treatment with psoralen and ultraviolet light for psoriasis
- chronic skin damage such as burn scars and ulcers
- infection with some varieties of human papillomavirus
- genetic disorders such as xeroderma pigmentosum and albinism
- a weakened immune system

Most people will receive 80% of their lifetime exposure to the sun before they reach the age of 20. For this reason, prevention should start during childhood and adolescence. Some important steps to prevent squamous cell carcinoma, as well as other skin cancers include:

- Wear protective clothing and a wide-brimmed hat in the sun.
- Stay out of the sun from 10 a.m. to 4 p.m.
- Use a sunscreen that has a sun protection factor (SPF) of at least 15.
- Avoid suntanning booths.

Drugs related to vitamin A (including beta-carotene and retinoids), vitamin E, **nonsteroidal anti-inflammatory drugs** (NSAIDS), and selenium might be able to prevent some skin cancers, but their effectiveness is still in question.

Special concerns

Because many squamous cell carcinomas are found on the face and neck, cosmetic concerns are a priority for many patients. If there is a risk of noticeable scarring or damage, a patient may wish to ask about alternative types of removal or inquire about the services of a plastic surgeon.

After treatment, it is important to return to the doctor periodically to check for regrowth or new skin cancers. Approximately a third to a half of all patients with nonmelanoma skin cancers find a new skin cancer within the next five years. Having a squamous cell carcinoma before the age of 60 may also increase the chance of developing other cancers in internal organs; however, this idea is still very controversial.

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ORGANIZATIONS

- American Skin Association, 150 East 58th Street, 32nd Floor, New York, NY, 10155-0002, (212) 753-8260.
- NIH/National Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse, One AMS Circle, Bethesda, MD, 20892-3675, (301) 495-4484, <http://www.nih.gov/niams>.

The NIAMS conducts and supports basic, clinical, and epidemiologic research and research training and disseminates information on diseases that include many forms of arthritis and diseases of the musculoskeletal system and the skin.

Skin Cancer Foundation, 245 Fifth Avenue, Suite 2402, New York, NY, 10016, (212) 725-5176.

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Rebecca J. Frey, PhD

Squint see **Strabismus**

SSPE see **Subacute sclerosing panencephalitis**

SSRIs see **Selective serotonin reuptake inhibitors**

SSSS see **Staphylococcal scalded skin syndrome**

St. Anthony's fire see **Erysipelas**

St. John's wort

Definition

St. John's wort is a species of the Hypericum genus that is native around the world and was introduced in U.S. The plant has yellow flowers that have been used medicinally for over 2,000 years.

Purpose

St. John's wort has been used to treat mental disorders and nerve **pain**, as a sedative and, as a salve, for **wounds, burns** and topical infections.

Today, it is sometimes used to treat mild depression, **anxiety**, and **sleep disorders**. Some clinical studies support its effectiveness, but two large and well designed ones do not.



St. John's wort flowers. (Photo Researchers, Inc.)

Preparations

The flowering tops are dried, ground and steeped as a tea to be drunk, placed in capsules to be taken by mouth in amounts of 500-1800mg per day, or combined with a salve for topical application.

Precautions

Though used as medicines, herbal products are regulated like dietary supplements in the United States. Thus, manufacturers are responsible only for their production processes. Imported herbals may not meet U.S. manufacturing standards. Approval of herbals is based on traditional use, not demonstrated safety and effectiveness. Before an herbal can be forcefully withdrawn from the market, the FDA must prove that it is unsafe.

Many herbal products vary from stated label potency.

St. John's wort should not be taken by pregnant women or nursing mothers.

St. John's wort may increase sensitivity to sunlight.

Interactions

St. John's wort may interact with, and increase the toxicity of, antidepressants like Prozac.

At higher doses, St. John's wort may reduce the effectiveness of cyclosporine, warfarin, birth control pills, HIV drugs, simvastatin, **digoxin**, and theophylline.

Resources

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James Waun, MD, R Ph

St. Vitus' dance see **Sydenham's chorea**

Stanford-Binet intelligence scales

Definition

The Stanford-Binet intelligence scale is a standarized test that assesses intelligence and cognitive abilities in children and adults aged two to 23.



The Stanford-Binet intelligence scale. (Photo Researchers, Inc.)

Purpose

The Stanford-Binet intelligence scale is used as a tool in school placement, in determining the presence of a learning disability or a developmental delay, and in tracking intellectual development. In addition, it is sometimes included in neuropsychological testing to assess the brain function of individuals with neurological impairments.

Precautions

Although the Stanford-Binet was developed for children as young as two, examiners should be cautious in using the test to screen very young children for developmental delays or disabilities. The test cannot be used to diagnose **mental retardation** in children aged three and under, and the scoring design may not detect developmental problems in preschool-age children.

Intelligence testing requires a clinically trained examiner. The Stanford-Binet intelligence scale should be administered and interpreted by a trained professional, preferably a psychologist.

Description

The Stanford-Binet intelligence scale is a direct descendent of the Binet-Simon scale, the first intelligence scale created in 1905 by psychologist Alfred Binet and Dr. Theophilus Simon. This revised edition, released in 1986, was designed with a larger, more diverse, representative sample to minimize the gender and racial inequities that had been criticized in earlier versions of the test.

The Stanford-Binet scale tests intelligence across four areas: verbal reasoning, quantitative reasoning,

KEY TERMS

Norms—Normative or mean score for a particular age group.

Representative sample—A random sample of people that adequately represents the test-taking population in age, gender, race, and socioeconomic standing.

Standard deviation—A measure of the distribution of scores around the average (mean). In a normal distribution, two standard deviations above and below the mean includes about 95% of all samples.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results. The Stanford-Binet test was standardized on a national representative sample of 5,000 subjects.

abstract/visual reasoning, and short-term memory. The areas are covered by 15 subtests, including vocabulary, comprehension, verbal absurdities, pattern analysis, matrices, paper folding and cutting, copying, quantitative, number series, equation building, memory for sentences, memory for digits, memory for objects, and bead memory.

All test subjects take an initial vocabulary test, which along with the subject's age, determines the number and level of subtests to be administered. Total testing time is 45–90 minutes, depending on the subject's age and the number of subtests given. Raw scores are based on the number of items answered and are converted into a standard age score corresponding to age group, similar to an IQ measure.

The 1997 Medicare reimbursement rate for psychological and neuropsychological testing, including intelligence testing, is \$58.35 an hour. Billing time typically includes test administration, scoring and interpretation, and reporting. Many insurance plans cover all or a portion of diagnostic psychological testing.

Normal results

The Stanford-Binet is a standardized test, meaning that norms were established during the design phase of the test by administering the test to a large, representative sample of the test population. The test has a mean, or average, standard score of 100 and a standard deviation of 16 (subtests have a mean of 50 and a standard deviation of 8). The standard deviation indicates how far above or below the norm the

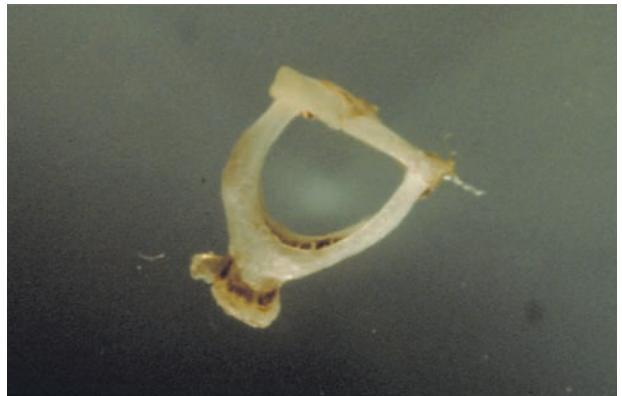
subject's score is. For example, an eight-year-old is assessed with the Stanford-Binet scale and achieves a standard age score of 116. The mean score of 100 is the average level at which all eight-year-olds in the representative sample performed. This child's score would be one standard deviation above that norm.

While standard age scores provide a reference point for evaluation, they represent an average of a variety of skill areas. A trained psychologist will evaluate and interpret an individual's performance on the scale's subtests to discover strengths and weaknesses and offer recommendations based upon these findings.

ORGANIZATIONS

American Psychological Association (APA), 750 First St. NE, Washington, DC, 20002-4242, (202) 336-5500, (800) 374-2721, <http://www.apa.org/>.

Paula Anne Ford-Martin



A human stapes bone (located in middle ear) extracted during a stapedectomy. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)

Next, the surgeon separates the stapes from the incus; freed from the stapes, the incus and malleus bones can now move when pressed. A laser (or other tiny instrument) vaporizes the tendon and arch of the stapes bone, which is then removed from the middle ear.

The surgeon then opens the window that joins the middle ear to the inner ear and acts as the platform for the stapes bone. The surgeon directs the laser's beam at the window to make a tiny opening, and gently clips the prosthesis to the incus bone. A piece of tissue is taken from a small incision behind the ear lobe and used to help seal the hole in the window and around the prosthesis. The eardrum is then gently replaced and repaired, and held there by absorbable packing ointment or a gelatin sponge. The procedure usually takes about an hour and a half.

Good candidates for the surgery are those who have a fixed stapes from otosclerosis, and a conductive hearing loss at least 20 dB. Patients with a severe hearing loss might still benefit from a stapedectomy, if only to improve their hearing to the point where a hearing aid can be of help. The procedure can improve hearing in more than 90% of cases.

Stapedectomy

Definition

Stapedectomy is a surgical procedure in which the innermost bone (stapes) of the three bones (the stapes, the incus, and the malleus) of the middle ear is removed, and replaced with a small plastic tube of stainless-steel wire (a prosthesis) to improve the movement of sound to the inner ear.

Purpose

A stapedectomy is used to treat progressive **hearing loss** caused by **otosclerosis**, a condition in which spongy bone hardens around the base of the stapes. This condition fixes the stapes to the opening of the inner ear so that the stapes no longer vibrates properly; therefore, the transmission of sound to the inner ear is disrupted. Untreated otosclerosis eventually results in total deafness, usually in both ears.

Description

With the patient under local or **general anesthesia**, the surgeon opens the ear canal and folds the eardrum forward. Using an operating microscope, the surgeon is able to see the structures in detail, and evaluates the bones of hearing (ossicles) to confirm the diagnosis of otosclerosis.

Preparation

Prior to admission to the hospital, the patient will be given a hearing test to measure the degree of deafness, and a full ear, nose, and throat exam.

Most surgeons prefer to use general anesthesia; in this case, an injection will be given to the patient before surgery.

KEY TERMS

Cochlea—The hearing part of the inner ear. This snail-shaped structure contains fluid and thousands of microscopic hair cells tuned to various frequencies, in addition to the organ of Corti (the receptor for hearing).

Conductive hearing loss—A type of medically treatable hearing loss in which the inner ear is usually normal but there are specific problems in the middle or outer ears that prevent sound from getting to the inner ear in a normal way.

Incus—The middle of the three bones of the middle ear. It is also known as the “anvil.”

Malleus—One of the three bones of the middle ear. It is also known as the “hammer.”

Ossicles—The three small bones of the middle ear: the malleus (hammer), the incus (anvil) and the stapes (stirrup). These bones help carry sound from the eardrum to the inner ear.

Vertigo—A feeling of dizziness together with a sensation of movement and a feeling of rotating in space.

Aftercare

The patient is usually discharged the morning after surgery. **Antibiotics** are given up to five days after surgery to prevent infection; packing and sutures are removed about a week after surgery.

It is important that the patient not put pressure on the ear for a few days after surgery. Blowing one's nose, lifting heavy objects, swimming underwater, descending rapidly in high-rise elevators, or taking an airplane flight should be avoided.

Right after surgery, the ear is usually quite sensitive, so the patient should avoid loud noises until the ear retrains itself to hear sounds properly.

It is extremely important that the patient avoid getting the ear wet until it has completely healed. Water in the ear could cause an infection; most seriously, water could enter the middle ear and cause an infection within the inner ear, which could then lead to a complete hearing loss. When taking a shower, and washing the hair, the patient should plug the ear with a cotton ball or lamb's wool ball, soaked in Vaseline. The surgeon should give specific instructions about when and how this can be done.

Usually, the patient may return to work and normal activities about a week after leaving the hospital, although if the patient's job involves heavy lifting, three weeks of home rest is recommended. Three days after surgery, the patient may fly in a pressurized aircraft.

Risks

The most serious risk is an increased hearing loss, which occurs in about one percent of patients. Because of this risk, a stapedectomy is usually performed on only one ear at a time.

Less common complications include:

- temporary change in taste (due to nerve damage) or lack of taste
- perforated eardrum
- vertigo that may persist and require surgery
- damage to the chain of three small bones attached to the eardrum
- temporary facial nerve paralysis
- ringing in the ears

Severe **dizziness** or vertigo may be a signal that there has been an incomplete seal between the fluids of the middle and inner ear. If this is the case, the patient needs immediate bed rest, an exam by the ear surgeon, and (rarely) an operation to reopen the eardrum to check the prosthesis.

Normal results

Most patients are slightly dizzy for the first day or two after surgery and may have a slight **headache**. Hearing improves once the swelling subsides, the slight bleeding behind the ear drum dries up, and the packing is absorbed or removed, usually within two weeks. Hearing continues to get better over the next three months.

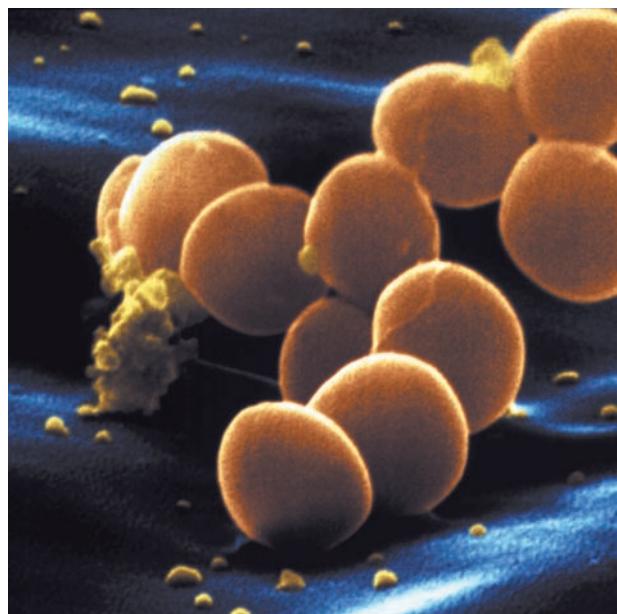
About 90% of patients will have a completely successful surgery, with markedly improved hearing. In 8% of cases, hearing improves, but not quite as patients usually expect. About half the patients who had ringing in the ears (**tinnitus**) before surgery will have significant relief within six weeks after the procedure.

ORGANIZATIONS

American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Road, Alexandria, VA, 22314-2857, (703) 836-4444, <http://www.entnet.org>.

Better Hearing Institute, 1444 I Street, NW, Suite 700,
Washington, DC, 20005, (202) 449-1100, (800) 327-
9355, mail@betterhearing.org, http://www.better
hearing.org/.http://www.betterhearing.org/.

Carol A. Turkington



Staphylococcal infections

Definition

Staphylococcal (staph) infections are communicable infections caused by a staphylococcal bacteria. They are generally characterized by the formation of abscesses. Staphylococcal infections are the leading cause of primary infections originating in hospitals (nosocomial infections) in the United States.

Description

Classified since the early twentieth century as among the deadliest of all disease-causing organisms, staph exists on the skin or inside the nostrils of 20–30% of healthy people. It is sometimes found in breast tissue, the mouth, and the genital, urinary, and upper respiratory tracts.

Although staph bacteria are usually harmless, when injury or a break in the skin enables the bacteria to invade the body and overcome the body's natural defenses, consequences can range from minor discomfort to **death**. Infection is most apt to occur in:

- newborns
- women who are breastfeeding



A close-up of a woman's finger and nail cuticle infected with *Staphylococcus aureus*. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

A micrographic image of *Staphylococcus aureus*. (Oliver Meckes/Photo Researchers, Inc.)

- individuals whose immune systems have been undermined by radiation treatments, chemotherapy, HIV/AIDS, organ transplantation, or medication
- intravenous drug users
- those with surgical incisions, skin disorders, and serious illness like cancer, diabetes, and lung disease
- the elderly, particularly those who live in nursing homes or who are hospitalized.

Types of infections

Staph infections produce pus-filled pockets (abscesses) located just beneath the surface of the skin or deep within the body. Risk of infection is greatest among newborns, the very young, and the very old.

A localized staph infection is confined to a ring of dead and dying white blood cells and bacteria. The skin above it feels warm to the touch. Most of these abscesses eventually burst and pus that leaks onto the skin can cause new infections.

A small fraction of localized staph infections enter the bloodstream and spread through the body. In children, these systemic (affecting the whole body) or disseminated infections frequently affect the ends of the long bones of the arms or legs, causing a bone infection called **osteomyelitis**. When adults develop invasive staph infections, bacteria are most apt to cause abscesses of the brain, heart, kidneys, liver, lungs, or spleen.

Staphylococcus aureus

Named for the golden color of the bacteria grown under laboratory conditions, *S. aureus* is a hardy organism that can survive in extreme temperatures or other inhospitable circumstances. About 70–90% of the population carry this strain of staph in the nostrils at some point in their life. Although present on the skin of only 5–20% of healthy people, as many as 40% carry the bacteria elsewhere, such as in the throat, vagina, or rectum. These people may carry the bacteria for varying periods of time (from hours to years) without developing symptoms or becoming ill.

S. aureus causes a variety of infections. **Boils** and inflammation of the skin surrounding a hair shaft (**folliculitis**) are the most common. Toxic shock (TSS) and scalded skin syndrome (SSSS) are among the most serious.

Methicillin-resistant Staphylococcus aureus infections (MRSA)

S. aureus flourishes in hospitals, where it infects healthcare personnel and patients who have had surgery; who have acute **dermatitis**, insulin-dependent diabetes, or dialysis-dependent **kidney disease**; or who receive frequent allergy-desensitization injections. Staph bacteria can also contaminate bedclothes, catheters, and other objects.

Toxic shock

Toxic shock syndrome is a life-threatening infection characterized by severe **headache**, **sore throat**, **fever** as high as 105°F, and a sunburn-like rash that spreads from the face to the rest of the body. Symptoms appear suddenly; they also include **dehydration** and watery **diarrhea**.

Inadequate blood flow to peripheral parts of the body (shock) and loss of consciousness occur within the first 48 hours. Between the third and seventh day of illness, skin peels from the palms of the hands, soles of the feet, and other parts of the body. Kidney, liver, and muscle damage often occur.

SCALDED SKIN SYNDROME. Rare in adults and most common in newborns and other children under the age of five, scalded skin syndrome originates with a localized skin infection. A mild fever and/or an increase in the number of infection-fighting white blood cells may occur.

A bright red rash spreads from the face to other parts of the body and eventually forms scales. Large, soft blisters develop at the site of infection and elsewhere. When they burst, they expose inflamed skin that looks as if it had been burned.

MISCELLANEOUS INFECTIONS. *S. aureus* can also cause:

- arthritis
- bacteria in the bloodstream (bacteremia)
- pockets of infection and pus under the skin (carbuncles)
- tissue inflammation that spreads below the skin, causing pain and swelling (cellulitis)
- inflammation of the valves and walls of the heart (endocarditis)
- inflammation of tissue that enclosed and protects the spinal cord and brain (meningitis)
- inflammation of bone and bone marrow (osteomyelitis)
- pneumonia

Other strains of staph

***S. epidermidis*.** Capable of clinging to tubing (as in that used for intravenous feeding, etc.), prosthetic devices, and other non-living surfaces, *S. epidermidis* is the organism that most often contaminates devices that provide direct access to the bloodstream.

The primary cause of **bacteremia** in hospital patients, this strain of staph is most likely to infect **cancer** patients, whose immune systems have been compromised, and high-risk newborns receiving intravenous supplements.

S. epidermidis also accounts for two of every five cases of prosthetic valve **endocarditis**. Prosthetic valve endocarditis is endocarditis as a complication of the implantation of an artificial valve in the heart. Although contamination usually occurs during surgery, symptoms of infection may not become evident until a year after the operation. More than half of the patients who develop prosthetic valve endocarditis die.

***STAPHYLOCOCCUS SAPROPHYTICUS*.** Existing within and around the tube-like structure that carries urine from the bladder (urethra) of about 5% of healthy males and females, *S. saprophyticus* is the second most common cause of unobstructed urinary tract infections (UTIs) in sexually active young women. This strain of staph is responsible for 10–20% of infections affecting healthy outpatients.

Causes and symptoms

Staph bacteria can spread through the air, but infection is almost always the result of direct contact with open sores or body fluids contaminated by these organisms.

Staph bacteria often enter the body through inflamed hair follicles or oil glands. Or they penetrate skin damaged by **burns**, cuts and scrapes, infection, insect **bites**, or **wounds**.

Multiplying beneath the skin, bacteria infect and destroy tissue in the area where they entered the body. Staph infection of the blood (staphylococcal bacteremia) develops when bacteria from a local infection infiltrate the lymph glands and bloodstream. These infections, which can usually be traced to contaminated catheters or intravenous devices, usually cause persistent high fever. They may cause shock. They also can cause death within a short time.

Warning signs

Common symptoms of staph infection include:

- pain or swelling around a cut, or an area of skin that has been scraped
- boils or other skin abscesses
- blistering, peeling, or scaling of the skin; this is most common in infants and young children
- enlarged lymph nodes in the neck, armpits, or groin

A family physician should be notified whenever:

- Lymph nodes in the neck, armpits, or groin become swollen or tender.
- An area of skin that has been cut or scraped becomes painful or swollen, feels hot, or produces pus. These symptoms may mean the infection has spread to the bloodstream.
- A boil or carbuncle appears on any part of the face or spine. Staph infections affecting these areas can spread to the brain or spinal cord.
- A boil becomes very sore. Usually a sign that infection has spread, this condition may be accompanied by fever, chills, and red streaks radiating from the site of the original infection.
- Boils that develop repeatedly. This type of recurrent infection could be a symptom of diabetes.

Diagnosis

Blood tests that show unusually high concentrations of white blood cells can suggest staph infection, but diagnosis is based on laboratory analysis of material removed from pus-filled sores, and on analysis of normally uninfected body fluids, such as, blood and urine. Also, x rays can enable doctors to locate internal abscesses and estimate the severity of infection. Needle biopsy (removing tissue with a needle, then examining it under a microscope) may be used to assess bone involvement.

Treatment

Superficial staph infections can generally be cured by keeping the area clean, using soaps that leave a germ-killing film on the skin, and applying warm,

moist compresses to the affected area for 20–30 minutes three or four times a day.

Severe or recurrent infections may require a 7 to 10 day course of treatment with penicillin or other oral antibiotics. The location of the infection and the identity of the causal bacteria determines which of several effective medications should be prescribed.

In case of a more serious infection, antibiotics may be administered intravenously for as long as six weeks. Intravenous antibiotics are also used to treat staph infections around the eyes or on other parts of the face.

Surgery may be required to drain or remove abscesses that form on internal organs, or on shunts or other devices implanted inside the body.

Alternative treatment

Alternative therapies for staph infection are meant to strengthen the immune system and prevent recurrences. Among the therapies believed to be helpful for the person with a staph infection are **yoga** (to stimulate the immune system and promote relaxation), **acupuncture** (to draw heat away from the infection), and herbal remedies. Herbs that may help the body overcome, or withstand, staph infection include:

- Garlic (*Allium sativum*). This herb is believed to have antibacterial properties. Herbalists recommend consuming three garlic cloves or three garlic oil capsules a day, starting when symptoms of infection first appear.
- Cleavers (*Galium aparine*). This anti-inflammatory herb is believed to support the lymphatic system. It may be taken internally to help heal staph abscesses and reduce swelling of the lymph nodes. A cleavers compress can also be applied directly to a skin infection.
- Goldenseal (*Hydrastis canadensis*). Another herb believed to fight infection and reduce inflammation, goldenseal may be taken internally when symptoms of infection first appear. Skin infections can be treated by making a paste of water and powdered goldenseal root and applying it directly to the affected area. The preparation should be covered with a clean bandage and left in place overnight.
- Echinacea (*Echinacea spp.*). Taken internally, this herb is believed to have antibiotic properties and is also thought to strengthen the immune system.
- Thyme (*Thymus vulgaris*), lavender (*Lavandula officinalis*), or bergamot (*Citrus bergamot*) oils. These oils are believed to have antibacterial properties and may help to prevent the scarring that may result from skin infections. A few drops of these oils are added to water and then a compress soaked in the water is applied to the affected area.

KEY TERMS

Abscess—A cavity containing pus surrounded by inflamed tissue.

Endocarditis—Inflammation of the lining of the heart, and/or the heart valves, caused by infection.

Nosocomial infections—Infections that were not present before the patient came to a hospital but were acquired by a patient while in the hospital.

- Tea tree oil (*Melaleuca* spp.). Another infection-fighting herb, this oil can be applied directly to a boil or other skin infection.

Prognosis

Most healthy people who develop staph infections recover fully within a short time. Others develop repeated infections. Some become seriously ill, requiring long-term therapy or emergency care. A small percentage die.

Prevention

Healthcare providers and patients should always wash their hands thoroughly with warm water and soap after treating a staph infection or touching an open wound or the pus it produces. Pus that oozes onto the skin from the site of an infection should be removed immediately. This affected area should then be cleansed with antiseptic or with antibacterial soap.

To prevent infection from spreading from one part of the body to another, it is important to shower rather than bathe during the healing process. Because staph infection is easily transmitted from one member of a household to others, towels, washcloths, and bed linens used by someone with a staph infection should not be used by anyone else. They should be changed daily and laundered separately in hot water with bleach until symptoms disappear.

Children should frequently be reminded not to share:

- brushes, combs, or hair accessories
- caps
- clothing
- sleeping bags
- sports equipment
- other personal items

A diet rich in green, yellow, and orange vegetables can bolster natural immunity. A doctor or nutritionist may recommend **vitamins** or mineral supplements to

compensate for specific dietary deficiencies. Drinking 8 to 10 glasses of water a day can help flush disease-causing organisms from the body.

Because some strains of staph bacteria are known to contaminate artificial limbs, prosthetic devices implanted within the body, and tubes used to administer medication or drain fluids from the body, catheters and other devices should be removed on a regular basis, if possible, and examined for microscopic signs of staph. Symptoms may not become evident until many months after contamination has occurred so this practice should be followed even with patients who show no sign of infection.

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Staphylococcal scalded skin syndrome

Definition

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by a type of bacteria in which large sheets of skin may peel away.

Description

SSSS primarily strikes children under the age of five, particularly infants. Clusters of SSSS cases (epidemics) can occur in newborn nurseries when staff in nurseries accidentally pass the causative bacteria between patients. It can also strike other age groups who have weakened immune systems. Such immunocompromised patients include those with **kidney**

disease, people undergoing cancer chemotherapy, organ transplant patients, and individuals with acquired **immunodeficiency** syndrome (**AIDS**).

Causes and symptoms

SSSS is caused by a type of bacteria called *Staphylococcus aureus*. This bacteria produces a chemical called an epidermolytic toxin ("epiderm," deriving from the Greek words *epi*, meaning on, and *derma*, meaning skin, refers to the top layer of skin; "-lytic," deriving from the Greek word *lysis*, which literally denotes the act of undoing, means breaking or destroying; a toxin is a poison). While the bacteria itself is not spread throughout the body, it affects all of the skin by sending this toxin through the bloodstream.

SSSS begins with a small area of infection. In newborn babies, this may appear as a crusted area around the umbilicus, or in the diaper area. In children between the ages of one and six, a small, red, crusty bump appears near the nose or ear. The child may have no energy and may have a **fever**. The skin becomes sensitive and uncomfortable even before the rash is fully visible. The rash starts out as bright red patches around the original area of crusting. Blisters may appear and the skin may look wrinkled. When the blisters pop, they leave pitted areas. Even gently touching these red patches of skin may cause them to peel away in jagged sheets. The skin below is shiny, moist, and bright pink. Within a day or two, the top layer of skin all over the body is peeling off in large sheets.

The dangers of this illness include the chance that a different kind of bacteria will invade through the open areas in the skin and cause a serious systemic infection (**sepsis**). A lot of body fluid is lost as the skin peels away and the layer underneath dries. **Dehydration** is a danger at this point.

Diagnosis

Although good patient care includes taking specimens of blister fluid and smears from the nose or throat, no bacteria are usually demonstrated. SSSS is usually diagnosed on the basis of the typical progression of symptoms in a child of this age, prone to this disorder. A sample of skin (**skin biopsy**) should be taken, prepared, and examined under a microscope. If the patient's disease is truly SSSS, the biopsy will show a characteristic appearance. There will be no accumulation of those cells usually present in the case of a bacterial infection. Instead, there will be evidence of disruption of only the top layer of skin (epidermis).

KEY TERMS

Epidermis—The top layer of skin.

Epidermolytic—Damaging to the top layer of skin.

Sepsis—An overwhelming infection affecting all the systems of the body.

Treatment

Treatment involves careful attention to avoid the development of dehydration. A variety of lotions and creams are available to apply to areas where the epidermis has peeled away. This both soothes the sensitive areas and protects against drying and further moisture loss.

Prognosis

Most patients heal from SSSS within 10–14 days. Healing occurs without scarring in the majority of patients. **Death** may occur if severe dehydration or sepsis complicate the illness. About 3% of children die of these complications; about 50% of immunocompromised adults die of these complications.

Prevention

As always, good hygiene can prevent the passage of the causative bacteria between people. In the event of an outbreak in a newborn nursery, members of the staff should have nasal smears taken to identify an adult who may be unknowingly carrying the bacteria and passing it on to the babies.

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Staphylococcal food poisoning see **Food poisoning**

Starvation

Definition

Starvation is the result of a severe or total lack of nutrients needed for the maintenance of life.

Description

Adequate **nutrition** has two components, necessary nutrients and energy in the form of calories. It is possible to ingest enough energy without a well-balanced selection of individual nutrients and produce diseases that are noticeably different from those resulting from an overall insufficiency of nutrients and energy. Although all foods are a source of energy for the human body, it is possible to consume a seemingly adequate amount of food without getting the required minimum of energy (calories). For example, marasmus is the result of a diet that is deficient mainly in energy. Children who get enough calories, but not enough protein have kwashiorkor. This is typical in cultures with a limited variety of foods that eat mostly a single staple carbohydrate like maize or rice. These conditions overlap and are associated with multiple vitamin and mineral deficits, most of which have specific names and set of problems associated with them.

- Marasmus produces a very skinny child with stunted growth.
- Children with kwashiorkor have body fat, an enlarged liver, and edema—swelling from excess water in the tissues. They also have growth retardation.
- Niacin deficiency produces pellagra characterized by diarrhea, skin rashes, brain dysfunction, tongue, mouth and vaginal irritation, and trouble swallowing.
- Thiamine (Vitamin B₁) deficiency causes beriberi, which can appear as heart failure and edema, a brain and nerve disease, or both.
- Riboflavin deficiency causes a sore mouth and throat, a skin rash, and anemia.
- Lack of vitamin C (ascorbic acid)—scurvy—causes hair damage, bleeding under the skin, in muscles and joints, gum disease, poor wound healing, and in severe cases convulsions, fever, loss of blood pressure, and death.
- Vitamin B₁₂ is needed to keep the nervous system working properly. It and pyridoxine (vitamin B₆) are both necessary for blood formation.
- Vitamin A deficiency causes at first loss of night vision and eventually blindness from destruction of the cornea, a disease called keratomalacia.
- Vitamin K is necessary for blood clotting.

- Vitamin D regulates calcium balance. Without it, children get rickets and adults get osteomalacia.

Causes and symptoms

Starvation may result from a number of factors. They include:

- anorexia nervosa, which is an eating disorder characterized by extreme calorie restriction
- intentional fasting
- coma
- stroke
- inability to obtain food (famine; child abuse; aftermath of war or other disaster; being lost in wilderness or desert areas)
- severe gastrointestinal disease

Since the body will combat **malnutrition** by breaking down its own fat and eventually its own tissue, a whole host of symptoms can appear. The body's structure, as well as its functions, are affected. Starved adults may lose as much as 50% of their normal body weight.

Characteristic symptoms of starvation include:

- shrinkage of such vital organs as the heart, lungs, ovaries, or testes, and gradual loss of their functions
- chronic diarrhea
- anemia
- reduction in muscle mass and consequent weakness
- lowered body temperature combined with extreme sensitivity to cold
- decreased ability to digest food because of lack of digestive acid production
- irritability and difficulty with mental concentration
- immune deficiency
- swelling from fluid under the skin
- decreased sex drive

Complete starvation in adults leads to **death** within 8 to 12 weeks. In the final stages of starvation, adult humans experience a variety of neurological and psychiatric symptoms, including **hallucinations** and convulsions as well as severe muscle **pain** and disturbances in heart rhythm.

In children, chronic malnutrition is marked by growth retardation. Anemia is the first sign to appear in an adult. Swelling of the legs is next, due to a drop in the protein content of the blood. Loss of resistance to infection follows next, along with poor wound healing. There is also progressive weakness and difficulty swallowing, which may lead to inhaling food. At the same time, the signs of specific nutrient deficiencies may appear.

KEY TERMS

Anemia—Not enough red blood cells in the blood.

Anorexia nervosa—Eating disorder marked by malnutrition and weight loss commonly occurring in young women.

Cornea—The clear part of the front of the eye that admits light.

Kwashiorkor—Severe malnutrition in children caused mainly by a protein-poor diet, characterized by growth retardation.

Marasmus—Severe malnutrition in children caused by a diet lacking mainly in calories. Can also be caused by disease and parasitic infection.

Treatment

If the degree of malnutrition is severe, the intestines may not tolerate a fully balanced diet. They may, in fact, not be able to absorb adequate nutrition at all. Carefully prepared elemental diets or intravenous feeding must begin the treatment. A formula consisting of 42% dried skim milk, 32% edible oil, and 25% sucrose plus electrolyte, mineral, and vitamin supplements is recommended for the first phase of refeeding. The treatment back to health is long and first begins with liquids. Gradually, solid foods are introduced and a daily diet providing 5,000 calories or more is instituted.

Prognosis

People can recover from severe degrees of starvation to a normal stature and function. Children, however, may suffer from permanent mental retardation or growth defects if their deprivation was long and extreme.

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Stasis dermatitis see **Dermatitis**

Static encephalopathy see **Cerebral palsy**

STDs see **Sexually transmitted diseases**

Steatosis see **Fatty liver**

Steele-Richardson-Olszewski syndrome see **Progressive supranuclear palsy**

Stein-Leventhal syndrome see **Polycystic ovary syndrome**

Steinert's disease see **Myotonic dystrophy**

Stem cell therapy see **Bone marrow transplantation**

Stem cell transplantation

Definition

Stem cells are basic human cells that reproduce (replicate) easily, providing a continuous source of new, sometimes different types of cells. A stem cell transplant is a procedure that replaces unhealthy stem cells with healthy ones. Stem cells can be harvested from bone marrow, from peripheral blood and from umbilical cord blood.

Purpose

Physicians use stem cell transplants to treat many diseases that damage or destroy bone marrow, found in the soft fatty tissue inside the bones. Examples of these diseases are leukemia and **multiple myeloma**. Some patients develop bone marrow disorders because of aggressive **cancer** treatments or as result of diseases such as **aplastic anemia**, which causes abnormal blood cell production.

Recent advances in stem cell research have made it a treatment possibility for patients with certain types of lymphomas, genetic disorders, hereditary metabolic disorders and **autoimmune disorders** as well. Researchers are hoping eventually to harvest stem cells to treat diseases such as Parkinson's disease, type 1 diabetes,

Alzheimer's disease, liver disease, arthritis, and spinal cord injuries.

Demographics

The number of stem cell transplants performed continues to increase. It is estimated that between 30,000 to 40,000 transplantations are performed on an annual basis worldwide and this number is increasing by up to 20% yearly.

Description

Stem cell transplants sometimes are called hematopoietic stem cell transplants, bone marrow transplants, or cord blood transplants. Nearly 100 years ago, physicians tried to give patients with leukemia and anemia bone marrow by mouth. These treatments were not successful, but led to experiments showing healthy bone marrow transfused into the blood stream could restore damaged bone marrow.

Today, two types of stem cell transplants are performed most often. When a patient's own stem cells are collected (harvested) then returned to the same patient's body, it is called an autologous transplant. Using stem cells from another person, or a donor, is called allogenic transplant. A third, less common type of transplant, is called a syngeneic transplant in which the donor is an identical twin. In many cases, donor cells come from a close relative, such as a brother or sister. However, the likelihood that a sibling will match the patient is only about 25%. Stem cells may need to come from a person not related to the recipient.

To find out if a patient can receive stem cells from a donor, physicians developed human leukocyte antigen (HLA) testing to match tissue types. The next challenge became finding donors. Throughout the 1980s and 1990s, private individuals, hospitals, foundations, and states worked to set up a nationwide registry of bone marrow donors. The National Marrow Donor Program (NMDP) now has the largest stem cell donor registry in the world. At present, there are more than three million donors registered through the NMDP. However, ethnic minorities represent only a small percentage of the donors to the NMDP making it often more difficult to provide a donor to a member of an ethnic minority requiring a stem cell transplant.

Stem cell transplants normally take place at specialized centers. Procurement of stem cells from the donor can be accomplished in several ways: from the bone marrow, the peripheral blood, and less commonly, from umbilical cord blood.

Donor cells harvested directly from a donor's bone marrow is done in an operating room while the patient (the donor) is under regional or **general anesthesia**. Bone marrow normally is harvested from the top of the hip bone. The marrow usually is filtered, treated, and either transplanted immediately or frozen for later use.

Stem cells harvested from a donor's peripheral blood are collected during apheresis once a process called stem cell mobilization has occurred. Hematopoietic stem cells are typically found in low concentrations in the circulating peripheral blood. Stem cells can be mobilized to enter the peripheral blood by administering the hematopoietic growth factor, granulocyte colony-stimulating factor (G-CSF) (filgrastim or lenograstim) to the donor. After about four days, enough stem cells are available in the circulating peripheral blood to be harvested. During a three to four hour apheresis collection process more stem cells can typically be collected than during a bone marrow harvest without the need for general anesthesia and an operating room setting.

Bone marrow transplantation is considered superior to peripheral blood stem cell transplant (PBSCT) for most nonmalignant conditions. Peripheral blood stem cell transplant is considered superior to bone marrow transplant when rapid engraftment of stem cells is needed. PBSCT is also associated with early hospital discharges, decreased relapse rates, and decreased mortality rates. However, PBSCT is associated with increased incidence of graft versus host disease (GVHD), a post-transplant complication.

Stem cells are transfused through an intravenous (IV) catheter that physicians insert in the patient's neck or chest. The procedure is usually done in the patient's hospital room. This part of the transplant process is referred to as the "rescue process." The stem cells replace malignant or defective cells. Transplanted donor cells travel to the bone cavities and begin replacing old bone marrow.

Precautions

The transplant team will weigh many factors when determining if a patient is a candidate for stem cell transplantation, including overall health and function of many vital organ systems. Stem cell transplantation is an aggressive treatment and may not be recommended for some patients, including those with heart, kidney, or lung disorders. If the patient has an aggressive cancer that has spread throughout the body, he or she may not be considered a candidate for a stem cell transplant. It once was thought that stem cell transplants were not safe in patients over age 60, but research

KEY TERMS

Catheter—A medical device shaped like a tube that physicians can insert into vessels, canals, or passageways to more easily inject or withdraw fluids.

Embryo—A developing human from the time of conception to the end of the eighth week after conception.

Engraftment—The process of transplanted stem cells reproducing new cells.

shows that some elderly patients can safely receive stem cells from donors.

Many ethical and legal factors are impacting the research and development into stem cell transplantation. Much debate surrounds scientific advances. For example, human embryos, fetal tissues and umbilical cords are sources of stem cells that may be transplanted or used for disease research. Some people have ethical problems with the use of embryos in fertility clinics for stem cell research or transplantation. Some link stem cell transplantation for disease with cloning and want to stop funding for stem cell research over fear of human cloning. A study released in 2005 stated that 63% of Americans back embryonic stem cell research and 70% support federal legislation to promote more research. Meanwhile, scientists continue to develop new and exciting possibilities for transplanting stem cells into the human body that may one day lead to new treatments for previously incurable diseases. Many do so with private funding.

Preparation

Standard preparation involves eliminating diseased and damaged cells. The exact process depends on the patient. In many cases when transplantation is done to treat cancer, the patient will receive **chemotherapy**, often in extremely high doses. Some also receive **radiation therapy**. Another goal of preparation is to suppress the immune system. This makes it less likely that the patient's body will reject the donated stem cells. This step is called the conditioning regimen and is considered a crucial element in stem cell transplantation. New advances have been made that allow some of the patient's diseased cells to remain and mix with the new cells. Immediately before transplantation, the treating physician and staff will give the patient special instructions and precautions,

depending on his or her disease and exact procedure. Many serious side effects are associated with the preparative regimens including **nausea**, **vomiting**, hair loss (**alopecia**), **diarrhea**, skin **rashes**, mouth sores and ulcers, as well as lung, liver, and neurological toxicities. Another serious result of the conditioning regimen is **infertility**. Sperm banking may be an option for some men. Preservation of female fertility by banking of oocytes (eggs) has not been as successful.

Aftercare

Stem cells take up to three weeks or longer to begin producing new cells or bone marrow, a process called engraftment. Until engraftment is complete, patients may bleed easily and are at high risk for the development of life-threatening infections. To reduce the risk for infection patients are hospitalized in high-efficiency particulate air (HEPA) filtered rooms that are sealed using positive air pressure. All individuals entering the room should practice strict hand hygiene to minimize the potential for spread of infection. Patients may be required to stay in the hospital for at least one week following transplantation until blood cell counts reach a safe level. Patients who received autologous transplants can often be managed on an outpatient basis. Once home, patients usually must be closely monitored, be careful not to risk infection, may be anemic, and may be extremely fatigued. Most patients will receive prophylactic antibiotic and antifungal therapy for 75–100 days after the transplant and will be monitored very closely for the occurrence of graft-versus-host disease and other transplant-related side effects.

Risks

In addition to the risk of a life-threatening infection following a stem cell transplant, patients receiving stem cells from donors risk serious complications from graft-versus-host disease (GVHD). GVHD is caused when the donor's cells react against the patient's (recipient's) tissue. Sometimes, the patient's body simply rejects the new cells. Researchers continue to explore ways to lessen risks of complications following stem cell transplants.

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ORGANIZATIONS

- International Myeloma Foundation (IMF), 12650 Riverside Dr., Suite 206, North Hollywood, CA, 91607–3421, (800) 452-CURE, <http://www.myeloma.org>.
- National Marrow Donor Program (NMDP), 3001 Broadway St. NE, Suite 100, Minneapolis, MN, 55413–1753, (800) 627-7692, <http://www.marrow.org>.

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Stent see **Coronary stent**

Stereotactic surgery see **Gamma knife surgery**

Sterilization see **Tubal ligation; Vasectomy**

- progestins
- androgens

However, the term "steroid" commonly refers to anabolic-androgenic steroids (AASs), which are synthetic derivatives of the male sex hormone testosterone.

Purpose

Anabolic-androgenic steroids are growth inducers. They were first developed in Europe in the 1930s to treat malnourishment and to promote healing following surgery. They are most commonly prescribed for hormone-replacement therapy; however they are also prescribed for a wide variety of other conditions including:

- cachexia (weight loss and muscle wasting) from severe conditions such as HIV/AIDS, chronic obstructive pulmonary disease (COPD), liver or kidney failure, some cancers, infection, surgery, injury, or unexplained inability to maintain adequate weight
- various types of anemia
- certain types of breast cancer in women
- hereditary angioedema
- acute and chronic wounds
- malnutrition with weight loss
- severe burns
- short stature
- osteoporosis
- primary or secondary hypogonadism
- side effects of long-term corticosteroid use

In addition to their legitimate medical uses, AASs are widely abused for building muscle, increasing strength, losing fat and flab, enhancing athletic performance, and otherwise engaging in high-risk behavior. Their anabolic effects build muscle and their androgenic effects develop male sexual characteristics in both males and females. At medically prescribed dosages the growth effects of anabolic steroids are slow or unnoticeable. However at high abusive doses muscles increase rapidly in size and strength and AAS may increase energy and libido.

Competitive weightlifters began using AASs in the 1950s and their use gradually spread to other sports. Most major athletic competitions, including the Olympic Games, the Wimbledon tennis tournament, the Tour de France bicycle race, and major-league professional sports teams, now routinely screen athletes for steroid use. Some steroids are easily detectable with a urine screen since the products of steroid metabolism can be identified in the urine for six months or longer after the drugs are discontinued. However the technology for detecting steroids in blood or urine—and distinguishing the use of AASs from the products of

Steroids

Definition

Steroids constitute a large class of naturally occurring and synthetic chemicals including sterols, such as cholesterol, and various corticosteroid and sex hormones that have wide-ranging effects. There are five classes of steroid hormones:

- mineralocorticoids
- glucocorticoids
- estrogens

normal metabolism or legitimate medications—is often one step behind the design of new steroid drugs.

Description

Steroids are chemicals containing a “steroid nucleus” consisting of four rings of 17 carbon atoms. They are fat-soluble compounds found in both plants and animals that function as hormones—chemical messengers. In humans steroids are produced from cholesterol in the endocrine glands, such as the adrenal cortex and the gonads (ovary and testis). Glands secrete the hormones directly into the bloodstream where they are transported to distant parts of the body and mediate various vital physiologic functions ranging from suppressing inflammation to regulating events during **pregnancy**.

Anabolic-androgenic steroids have three mechanisms of action:

- They help metabolize proteins.
- They speed up the synthesis of skeletal muscle tissue.
- They provide a “rush” that enables an athlete to train harder and longer by temporarily masking fatigue.

Common AASs preparations include:

- testosterone esters: testosterone propionate, cypionate, and enanthate
- testosterone derivatives: methyltestosterone (which is rarely used), methandrostenolone, and fluoxymesterone (which is rarely used clinically but is abused)
- nandrolone derivatives: nandrolone decanoate, ethylestrenol, and trenbolone
- dihydrotestosterone (DHT) derivatives: oxandrolone, stanozolol, and oxymetholone

AASs are taken orally as tablets or capsules, by injection into muscles, or through the skin via ointments or transdermal patches. They are sometimes combined with creatine, protein powders, and/or various antioxidant formulations. These are considered to be ergogenic aids—substances that enhance the body’s production, use, or recovery of energy and provide athletes with a competitive advantage.

U.S. brand names

AASs that are prescribed for medical purposes in the United States include:

- Depo-Testosterone (testosterone cypionate) for low testosterone levels
- Android (methyltestosterone) for treating hot flashes in postmenopausal women
- Nandrolone decanoate, an injectable preparation available only as a generic and relatively safe at

clinical doses for treating osteoporosis, but one of the most widely abused AASs

- Durabolin (nandrolone phenpropionate), a parenteral injection
- Oxandrin (oxandrolone), oral tablets prescribed for HIV/AIDS-related conditions, bone pain from osteoporosis, to prevent certain side effects of corticosteroids, and sometimes abused by female athletes
- Winstrol (stanozolol), oral tablets for treating hereditary angioedema
- Anadrol-50 (oxymetholone), oral tablets that are abused worldwide and are considered to be carcinogenic

In the past steroid supplements such as androstenedione (“Andro”) were commercially available in the United States. They were declared illegal in 2004 in an amendment to the Controlled Substances Act. The only remaining legal steroid supplement in the United States is dehydroepiandrosterone (DHEA), which may or may not be converted to testosterone in the body.

Many athletes and other people obtain and use AASs illegally. Estimates of steroid **abuse** in the general adult population of the United States range from 0.5–5%. Although most anabolic steroid users are adult males, misuse is increasing among women and adolescents.

Street names for veterinary steroids that are abused include:

- Abolic
- Dianabol
- Equipoise
- Finajet/Finaject
- Parabolin
- Trenbolone
- Winstrol V

Street names for oral steroids that are abused include:

- Anadrol
- Anavar
- Maxibolin
- Methyltestosterone
- Parabolin
- Primobolin
- Primobolin
- Proviron
- Winstrol

Street names for abused injectable steroids include:

- Anastrofin
- Bolasterone
- Deca-Duabolin

KEY TERMS

Anabolic—Causing muscle and bone growth and a shift from fat to muscle in the body.

Androgenic—Causing testosterone-like, masculinizing effects.

Angioedema—Patches of swelling of the skin, subcutaneous layers, mucus membranes, and sometimes internal organs.

Cachexia—Physical wasting and malnutrition, usually from chronic disease.

Cholesterol—A steroid alcohol in cells and body fluids that serves as a precursor for hormones and other steroids.

Corticosteroid—A steroid, such as cortisone, produced by the adrenal cortex.

Creatine—A nitrogen-containing substance found in muscle.

Estrogen—The primary female sex hormone.

Hormones—Chemical messengers that are carried by the bloodstream to various organs where they effect functioning, often by stimulatory action.

Sex hormones—Hormones that are responsible for sexual characteristics and reproductive functioning.

Steroids—A class of hormones and drugs that includes sex and stress hormones and anti-inflammatory medications, contraceptives, and growth-promoting substances.

Sterols—Steroid alcohols, such as cholesterol, that are widely distributed in the body.

Testosterone—The primary male sex hormone.

- Delatestryl
 - Dep-testosterone
 - Dihydrolone
 - Durabolin
 - Dymethzine
 - Enoltestovis
 - Methatriol
 - Primobolin
 - Primobolin
 - Quinolone
 - Sustanon 250
 - Therobolin
 - Trophobolene
- “Designer” steroids include:
- THG (tetrahydrogestrinone)
 - Madol (desoxymethyltestosterone)
 - Genabol (norbolethone)
 - Equipoise and others (boldenone undecylenate)

Canadian brand names

Canadian AASs include Deca-Durabolin (nandrolone decanoate) and Anapolon 50 (oxymetholone).

International brand names

There are many thousands of international brand names for AASs and in some countries they are available over the counter. Methandrostenolone (Dianabol, Danabol, DBOL, Reforvit-b) and ethylestrenol (Maxibolin) have been discontinued in the United States but are still

manufactured internationally. Trenbolone, a veterinary drug in the United States, is a widely abused European prescription drug. Winstrol (stanozolol) is marketed internationally as well as in the United States, comes in both oral and injectable forms, and is widely abused.

Recommended dosage

Prescribed dosages of AASs depend on the specific drug and the condition being treated. Examples of dosages include:

- for malnutrition and cachexia from COPD: an initial 250-milligram (mg) injection of testosterone followed by 12 mg per day of stanozolol
- for renal-failure patients on hemodialysis: 100 mg per week of nandrolone
- for quadriplegic patients: 20 mg per day of oxandrolone
- for weight loss from alcoholic hepatitis: 80 mg per day of oxandrolone
- for decreasing the frequency and severity of angioedema attacks: 2 mg of stanozolol three times daily

Dosages used by AAS abusers are often 50–100 times higher than those for treating medical conditions. In one survey of 500 AAS users, more than half reported taking 1,000 mg or more every week. In addition 13% acknowledged using such unsafe injection practices such as sharing needles, reusing needles, or sharing multi-dose vials of steroids.

In addition to higher doses, AAS abusers may practice:

- stacking: taking two or more AASs together, using more than one route of administration, or mixing AASs with other drugs such as stimulants or painkillers
- cycling: alternating periods of steroid use with periods of abstinence
- pyramiding: cycling of increasing doses over several weeks followed by decreasing doses Some users believe that cycling and pyramiding maximize the desirable effects of steroids while reducing the undesirable effects, although there is no scientific evidence to support this and cycling may build tolerance, requiring higher dosages.

Precautions

Although AASs are considered to be relatively safe at the usual prescribed dosages, at abusive dosages they can result in a wide range of serious health problems, including:

- abnormal lipid profiles
- early heart attacks
- strokes
- kidney failure
- severe liver problems including jaundice, tumors, and cancer
- depression
- severe psychiatric disturbances

Unsafe injection of AASs can result in HIV and **hepatitis B** and C infections. It also has been suggested that steroids may be gateway drugs for **narcotics** abuse and it is possible to become psychologically addicted to them.

Withdrawal from high doses of AASs may require:

- medication to relieve withdrawal symptoms
- antidepressants
- hormones to restore normal hormonal function

Pediatric

AASs should be used with extreme caution in children. They can cause early **puberty** and lead to premature cessation of bone growth, resulting in permanent short stature.

A 2008 survey found that 1.4% of eighth graders, 1.4% of tenth graders, and 2.2% of twelfth graders had used AASs. Some were athletes attempting to increase their strength and size; others were simply attempting to speed up their growth to keep pace with their peers. Many teenagers also are attracted by the psychological rush that comes with steroid abuse. Prevention programs often target school- or community-sponsored

athletic teams, coaches, and team leaders. It has been recommended that educational prevention programs start with middle-school athletes.

Geriatric

Elderly patients can be more sensitive to the effects of AASs. They should not be used in elderly people with prostate problems, fluid buildup, or abnormal liver function.

Pregnant or breastfeeding

Pregnant women or women who are planning to become pregnant should never take AASs because they are known to cause **birth defects**. Although it is not known whether most AASs pass into breast milk, lactating women should avoid these drugs.

Other conditions and allergies

People with the following conditions should never take an AAS:

- high blood calcium levels (hypercalcemia)
- prostate cancer
- breast cancer in males
- breast cancer with high calcium levels in females
- severe kidney damage

The following conditions may be contraindications for AASs:

- heart or blood vessel disease
- previous heart attack
- high blood cholesterol
- bleeding or clotting problems
- diabetes, since AASs can affect blood sugar levels
- liver or kidney problems

Side effects

AASs usually have few side effects at medical dosages. However at the much higher doses used to improve body image or athletic performance, side effects can be serious and sometimes irreversible. Excessive use can cause harmful imbalances in hormones and body chemistry.

Common side effects of AASs include:

- excitability
- changes in sex drive
- swelling of the feet and ankles
- baldness
- breast swelling
- change in skin color

Steroids

- diarrhea
- nausea
- vomiting
- insomnia
- bladder irritation
- fertility problems
- testicular problems
- difficulty achieving an erection

Rare but more severe side effects of medically prescribed dosages of AASs include:

- severe allergic reactions
- acne, especially in women
- darkened urine
- deepening of the voice in women
- increased facial and body hair in women
- menstrual changes
- frequent painful erections
- behavioral or emotional changes
- jaundice

Side effects of AAS abuse can include:

- shrinking testicles, falling sperm counts, gynecomastia (swelling and enlargement of the breasts), increased urination, and enlarged prostate glands in males
- virilization in females, including hirsutism (growth of body and facial hair), male-pattern baldness, cessation of menstruation, decreased breast size, deepening of the voice, and abnormal enlargement of the clitoris
- an increase in “bad” cholesterol (LDL) and a decrease in “good” cholesterol (HDL)
- water retention leading to high blood pressure and stroke
- heart attacks
- liver and kidney tumors
- insomnia
- drastic mood swings ranging from mania to depression
- aggression, paranoia, irritability, delusions, hostility, psychosis

Pediatric

Side effects of AASs in young people can include:

- acne
- early puberty
- an initial growth spurt
- premature cessation of growth due to closure of growth plates

Other conditions and allergies

AAS preparations can contain various ingredients that may cause allergic reactions in some people.

Interactions

AASs may interact with:

- anticoagulants such as warfarin (Coumadin)
- carbamazepine
- insulin or oral diabetes medications
- corticosteroids
- eucalyptus
- kava (*Piper methysticum*)

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American College of Sports Medicine, PO Box 1440, Indianapolis, IN, 46202-1440, (317) 637-9200, (317) 634-7817, <http://www.acsm.org>.

National Institute on Drug Abuse (NIDA), 6001 Executive Boulevard, Room 5213, Bethesda, MD, 20892-9561, (301) 443-1124, information@nida.nih.gov, http://www.drugabuse.gov/NIDAHome.html.

Jill U. Adams
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Stillbirth

Definition

A stillbirth is defined as the **death** of a fetus at any time after the twentieth week of **pregnancy**. Stillbirth is also referred to as intrauterine fetal death (IUFD).

Description

It is important to distinguish between a stillbirth and other words that describe the unintentional end of a pregnancy. A pregnancy that ends before the twentieth week is called a **miscarriage** rather than a stillbirth, even though the death of the fetus is a common cause of miscarriage. After the twentieth week, the unintended end of a pregnancy is called a stillbirth if the infant is dead at birth and premature delivery if it is born alive.

Factors that increase a mother's risk of stillbirth include: age over 35, **malnutrition**, inadequate prenatal care, **smoking**, and alcohol or drug **abuse**.

Causes and symptoms

Causes

A number of different disorders can cause stillbirth. They include:

- Pre-eclampsia and eclampsia. These are disorders of late pregnancy characterized by high blood pressure, fluid retention, and protein in the urine.
- Diabetes in the mother.
- Hemorrhage.
- Abnormalities in the fetus caused by infectious diseases, including syphilis, toxoplasmosis, German measles (rubella), and influenza.
- Severe birth defects, including spina bifida. Birth defects are responsible for about 20% of stillbirths.
- Postmaturity. Postmaturity is a condition in which the pregnancy has lasted 41 weeks or longer.
- Unknown causes. These account for about one-third of stillbirths.

KEY TERMS

Alpha-fetoprotein analysis—A blood test that can be done after the sixteenth week of pregnancy to evaluate the possibility of spina bifida and other birth defects in the fetus.

Electronic fetal nonstress test—A test in which electronic monitors attached to the mother's abdomen to detect contractions of the uterus as well as the baby's heartbeat and movements.

Miscarriage—The spontaneous end of a pregnancy before the twentieth week. The death of the fetus is a common cause of miscarriage.

Oxytocin—A drug that is given to induce labor in some cases of stillbirth.

Pre-eclampsia and eclampsia—Disorders of late pregnancy associated with high blood pressure, fluid retention, and protein in the urine. They can cause stillbirth.

Premature delivery—The birth of a live baby when a pregnancy ends spontaneously after the twentieth week.

Symptoms

In most cases the only symptom of stillbirth is that the mother notices that the baby has stopped moving. In some cases, the first sign of fetal death is **premature labor**. Premature labor is marked by a rush of fluid from the vagina, caused by the tearing of the membrane around the baby; and by abdominal cramps or contractions.

Diagnosis

When the mother notices that fetal movement has stopped, the doctor can use several techniques to evaluate whether the baby has died. The doctor can listen for the fetal heartbeat with a stethoscope, use Doppler ultrasound to detect the heartbeat, or give the mother an electronic fetal nonstress test. In this test, the mother lies on her back with electronic monitors attached to her abdomen. The monitors record the baby's heart rate, movements, and contractions of the uterus.

Treatment

Medical

In most cases of intrauterine death, the mother will go into labor within two weeks of the baby's death. If the mother does not go into labor, the doctor will bring

on (induce) labor in order to prevent the risk of hemorrhage. Labor is usually induced by giving the mother a drug (oxytocin) that cause the uterus to contract.

Follow-up therapy

Emotional support from family and friends, self-help groups, and counseling by a mental health professional can help bereaved parents cope with their loss.

Prognosis

With the exception of women with diabetes, women who have a stillbirth have as good a chance of carrying a future pregnancy to term as women who are pregnant for the first time.

Prevention

The risk of stillbirth can be lowered to some extent by good prenatal care and the mother's avoidance of exposure to infectious diseases, smoking, alcohol abuse, or drug consumption. Tests before delivery (**antepartum testing**), such as ultrasound, the alpha-fetoprotein blood test, and the electronic fetal non-stress test, can be used to evaluate the health of the fetus before there is a stillbirth.

Resources

BOOKS

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ORGANIZATIONS

Compassionate Friends, P.O. Box 3696, Oak Brook, IL, 60522, (630) 990-0010, (630) 990-0246, (877) 969-0010, <http://www.compassionatefriends.org>.

GriefNet, GriefNet, Ann Arbor, MI, 48106-3272, cendra@griefnet.org, <http://www.griefnet.org>.

Hannah's Prayer, PO Box 15053, Long Beach, CA, 90815, (562) 335-4130, <http://www.hannah.org>.

M.E.N.D. (Mommies Enduring Neonatal Death), P.O. Box 1007 , Coppell, TX, 75019, (972) 506-9000, rebekah@mend.org, <http://www.mend.org>.

Pregnancy and Infant Loss Support (SHARE), 402 Jackson Street, St. Charles, MO, 63301, (636) 947-6164, (800) 821-6819.

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Stings see **Bites and stings**

Stockholm syndrome

Definition

Stockholm syndrome refers to a group of psychological symptoms that occur in some persons in a captive or hostage situation. It has received considerable media publicity in recent years because it has been used to explain the behavior of such well-known kidnapping victims as Patty Hearst (1974) and Elizabeth Smart (2002). The term takes its name from a bank robbery in Stockholm, Sweden, in August 1973. The robber took four employees of the bank (three women and one man) into the vault with him and kept them hostage for 131 hours. After the employees were finally released, they appeared to have formed a paradoxical emotional bond with their captor; they told reporters that they saw the police as their enemy rather than the bank robber and that they had positive feelings toward the criminal. The syndrome was first named by Nils Bejerot (1921–1988), a medical professor who specialized in **addiction** research and served as a psychiatric consultant to the Swedish police during the standoff at the bank. Stockholm syndrome is also known as Survival Identification Syndrome.

Description

Stockholm syndrome is considered a complex reaction to a frightening situation and experts do not agree completely on all of its characteristic features or on the factors that make some people more susceptible than others to developing it. One reason for the disagreement is that it would be unethical to test theories about the syndrome by experimenting on human beings. The data for understanding the syndrome are derived from actual hostage situations since 1973 that differ considerably from one another in terms of location, number of people involved, and time frame. Another source of disagreement concerns the extent to which the syndrome can be used to explain other historical phenomena or more commonplace types of abusive relationships. Many researchers believe that Stockholm syndrome helps to explain certain behaviors of survivors of World War II concentration camps; members of religious cults; battered wives; incest survivors; and physically or emotionally abused children as well as persons taken hostage by criminals or terrorists.

Most experts, however, agree that Stockholm syndrome has three central characteristics:

- The hostages have negative feelings about the police or other authorities.
- The hostages have positive feelings toward their captor(s).

- The captors develop positive feelings toward the hostages.

Causes & symptoms

Stockholm syndrome does not affect all hostages (or persons in comparable situations); in fact, a Federal Bureau of Investigation (FBI) study of over 1200 hostage-taking incidents found that 92% of the hostages did *not* develop Stockholm syndrome. FBI researchers then interviewed flight attendants who had been taken hostage during airplane hijackings, and concluded that three factors are necessary for the syndrome to develop:

- The crisis situation lasts for several days or longer.
- The hostage takers remain in contact with the hostages; that is, the hostages are not placed in a separate room.
- The hostage takers show some kindness toward the hostages or at least refrain from harming them. Hostages abused by captors typically feel anger toward them and do not usually develop the syndrome.

In addition, people who often feel helpless in other stressful life situations or are willing to do anything in order to survive seem to be more susceptible to developing Stockholm syndrome if they are taken hostage.

People with Stockholm syndrome report the same symptoms as those diagnosed with **post-traumatic stress disorder** (PTSD): **insomnia**, nightmares, general irritability, difficulty concentrating, being easily startled, feelings of unreality or confusion, inability to enjoy previously pleasurable experiences, increased distrust of others, and flashbacks.

Diagnosis

Stockholm syndrome is a descriptive term for a pattern of coping with a traumatic situation rather than a diagnostic category. Most psychiatrists would use the diagnostic criteria for **acute stress disorder** or posttraumatic stress disorder when evaluating a person with Stockholm syndrome.

Treatment

Treatment of Stockholm syndrome is the same as for PTSD, most commonly a combination of medications for short-term sleep disturbances and **psychotherapy** for the longer-term symptoms.

Prognosis

The prognosis for recovery from Stockholm syndrome is generally good, but the length of treatment needed depends on several variables. These include the

KEY TERMS

Coping—In psychology, a term that refers to a person's patterns of response to stress. Some patterns of coping may lower a person's risk of developing Stockholm syndrome in a hostage situation.

Flashback—The re-emergence of a traumatic memory as a vivid recollection of sounds, images, and sensations associated with the trauma. The person having the flashback typically feels as if they are reliving the event. Flashbacks were first described by doctors treating combat veterans of World War I (1914–1918).

Identification with an aggressor—In psychology, an unconscious process in which a person adopts the perspective or behavior patterns of a captor or abuser. Some researchers consider it a partial explanation of Stockholm syndrome.

Regression—In psychology, a return to earlier, usually childish or infantile, patterns of thought or behavior.

Syndrome—A set of symptoms that occur together.

nature of the hostage situation; the length of time the crisis lasted, and the individual patient's general coping style and previous experience(s) of trauma.

Prevention

Prevention of Stockholm syndrome at the level of the larger society includes further development of crisis intervention skills on the part of law enforcement as well as strategies to prevent kidnapping or hostage-taking incidents in the first place. Prevention at the individual level is difficult as of the early 2000s because researchers have not been able to identify all the factors that may place some persons at greater risk than others; in addition, they disagree on the specific psychological mechanisms involved in Stockholm syndrome. Some regard the syndrome as a form of regression (return to childish patterns of thought or action) while others explain it in terms of emotional **paralysis** ("frozen fright") or identification with the aggressor.

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- American Psychiatric Association (APA), 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209, (888) 357-7924, apa@psych.org, <http://www.psych.org>.
- Federal Bureau of Investigation (FBI), 935 Pennsylvania Avenue, NW, Washington, DC, 20535-0001, (202) 324-3000, <http://www.fbi.gov>.

Rebecca Frey, PhD

Stomach acid determination see **Gastric acid determination**



An excised section of a human stomach showing a cancerous tumor (center, triangular shape). (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Stomach cancer

Definition

Stomach **cancer** (also known as gastric cancer) is a disease in which the cells forming the inner lining of the stomach become abnormal and start to divide uncontrollably, forming a mass called a tumor.

Description

The stomach is a J-shaped organ that lies in the left and central portion of the abdomen. The stomach produces many digestive juices and acids that mix with food and aid in the process of digestion. There are five regions of the stomach that doctors refer to when determining the origin of stomach cancer. These are:

- the cardia, area surrounding the cardiac sphincter which controls movement of food from the esophagus into the stomach,
- the fundus, upper expanded area adjacent to the cardiac region,

- the antrum, lower region of the stomach where it begins to narrow,
- the prepyloric, region just before or nearest the pylorus,
- and the pylorus, the terminal region where the stomach joins the small intestine

Cancer can develop in any of the five sections of the stomach. Symptoms and outcomes of the disease will vary depending on the location of the cancer.

Based on previous data from the National Cancer Institute and the United States Census, the American Cancer Society estimates that 21,700 Americans will be diagnosed with stomach cancer during 2001 and approximately 13,000 deaths will result from the disease. In most areas, men are affected by stomach cancer nearly twice as often as women. Most cases of stomach cancer are diagnosed between the ages of 50 and 70, but in families with a hereditary risk of stomach cancer, younger cases are more frequently seen.

Stomach cancer is one of the leading causes of cancer deaths in several areas of the world, most notably Japan and other Asian countries. In Japan it appears almost ten times as frequently as in the United States. The number of new stomach cancer cases is decreasing in some areas, however, especially in developed countries. In the United States, incidence rates have dropped from 30 individuals per 100,000 in the 1930s, to only 8 in 100,000 individuals developing stomach cancer by the 1980s. The use of refrigerated foods and increased consumption of fresh fruits and vegetables, instead of preserved foods with high salt content, may be a reason for the decline.

Causes and symptoms

While the exact cause for stomach cancer has not been identified, several potential factors have lead to increased numbers of individuals developing the disease and therefore, significant risk has been associated. Diet, work environment, exposure to the bacterium *Helicobacter pylori*, and a history of stomach disorders, such as ulcers or polyps, are some of these believed causes.

Studies have shown that eating foods with high quantities of salt and nitrates increases the risk of stomach cancer. The diet in a specific region can have a great impact on its residents. Making changes to the types of foods consumed has been shown to decrease likelihood of disease, even for individuals from countries with higher risk. For example, Japanese people who move to the United States or Europe and change the types of foods they eat have a far lower chance of developing the disease than do Japanese people who remain in Japan and do not change their dietary habits. Eating recommended amounts of fruit and vegetables may lower a person's chances of developing this cancer.

A high risk for developing stomach cancers has been linked to certain industries as well. The best proven association is between stomach cancer and persons who work in coal mining and those who work processing timber, nickel, and rubber. An unusually large number of these workers have been diagnosed with this form of cancer.

Several studies have identified a bacterium (*Helicobacter pylori*) that causes stomach ulcers (inflammation in the inner lining of the stomach). Chronic (long-term) infection of the stomach with these bacteria may lead to a particular type of cancer (lymphomas or mucosa-associated lymphoid tissue [MALT]) in the stomach.

Another risk factor is the development of polyps, benign growths in the lining of the stomach. Although polyps are not cancerous, some may have the potential to turn cancerous. People in blood group A are also at

elevated risk for this cancer for unknown reasons. Other speculative causes of stomach cancer include previous stomach surgery for ulcers or other conditions, or a form of anemia known as pernicious anemia.

Stomach cancer is a slow-growing cancer. It may be years before the tumor grows very large and produces distinct symptoms. In the early stages of the disease, the patient may only have mild discomfort, **indigestion**, **heartburn**, a bloated feeling after eating, and mild **nausea**. In the advanced stages, a patient will have loss of appetite and resultant weight loss, stomach pains, **vomiting**, difficulty in swallowing, and blood in the stool. Stomach cancer often spreads (metastasizes) to adjoining organs such as the esophagus, adjacent lymph nodes, liver, or colon.

Diagnosis

Unfortunately, many patients diagnosed with stomach cancer experience **pain** for two or three years before informing a doctor of their symptoms. When a doctor suspects stomach cancer from the symptoms described by the patient, a complete medical history will be taken to check for any risk factors. A thorough **physical examination** will be conducted to assess all the symptoms. Laboratory tests may be ordered to check for blood in the stool (**fecal occult blood test**) and anemia (low red blood cell count), which often accompany gastric cancer.

In some countries, such as Japan, it is appropriate for patients to be given routine screening examinations for stomach cancer, as the risk of developing cancer in that society is very high. Such screening might be useful for all high-risk populations. Due to the low prevalence of stomach cancer in the United States, routine screening is usually not recommended unless a family history of the disease exists.

Whether as a screening test or because a doctor suspects a patient may have symptoms of stomach cancer, **endoscopy** or barium x rays are used in diagnosing stomach cancer. For a barium x ray of the upper gastrointestinal tract, the patient is given a chalky, white solution of barium sulfate to drink. This solution coats the esophagus, the stomach, and the small intestine. Air may be pumped into the stomach after the barium solution in order to get a clearer picture. Multiple x rays are then taken. The barium coating helps to identify any abnormalities in the lining of the stomach.

In another more frequently used test, known as upper gastrointestinal endoscopy, a thin, flexible, lighted tube (endoscope) is passed down the patient's throat and into the stomach. The doctor can view the lining of the esophagus and the stomach through the

tube. Sometimes, a small ultrasound probe is attached at the end of the endoscope. This probe sends high frequency sound waves that bounce off the stomach wall. A computer creates an image of the stomach wall by translating the pattern of echoes generated by the reflected sound waves. This procedure is known as an endoscopic ultrasound or EUS.

Endoscopy has several advantages, in that the physician is able to see any abnormalities directly. In addition, if any suspicious-looking patches are seen, biopsy forceps can be passed painlessly through the tube to collect some tissue for microscopic examination. This is known as a biopsy. EUS is beneficial because it can provide valuable information on depth of tumor invasion.

After stomach cancer has been diagnosed and before treatment starts, another type of x-ray scan is taken. Computed tomography (CT) is an imaging procedure that produces a three-dimensional picture of organs or structures inside the body. CT scans are used to obtain additional information in regard to how large the tumor is and what parts of the stomach it borders; whether the cancer has spread to the lymph nodes; and whether it has spread to distant parts of the body (metastasized), such as the liver, lung, or bone. A CT scan of the chest, abdomen, and pelvis is taken. If the tumor has gone through the wall of the stomach and extends to the liver, pancreas, or spleen, the CT will often show this. Although a CT scan is an effective way of evaluating whether cancer has spread to some of the lymph nodes, it is less effective than EUS in evaluating whether the nodes closest to the stomach are free of cancer. However, CT scans, like barium x rays, have the advantage of being less invasive than upper endoscopy.

Laparoscopy is another procedure used to stage some patients with stomach cancer. This involves a medical device similar to an endoscope. A laparoscopy is a minimally invasive surgery technique with one or a few small incisions, which can be performed on an outpatient basis, followed by rapid recovery. Patients who may receive **radiation therapy** or **chemotherapy** before surgery may undergo a laparoscopic procedure to determine the precise stage of cancer. The patient with bone pain or with certain laboratory results should be given a **bone scan**.

Benign gastric neoplasms are tumors of the stomach that cause no major harm. One of the most common is called a submucosal leiomyoma. If a leiomyoma starts to bleed, surgery should be performed to remove it. However, many leiomyomas require no treatment. Diagnosis of stomach cancers should be conducted

carefully so that if the tumor does not require treatment the patient is not subjected to a surgical operation.

Treatment

More than 95% of stomach cancers are caused by adenocarcinomas, malignant cancers that originate in glandular tissues. The remaining 5% of stomach cancers include lymphomas and other types of cancers. It is important that gastric lymphomas be accurately diagnosed because these cancers have a much better prognosis than stomach adenocarcinomas. Approximately half of the people with gastric lymphomas survive five years after diagnosis. Treatment for gastric lymphoma involves surgery combined with chemotherapy and radiation therapy.

Staging of stomach cancer is based on how deep the growth has penetrated the stomach lining; to what extent (if any) it has invaded surrounding lymph nodes; and to what extent (if any) it has spread to distant parts of the body (metastasized). The more confined the cancer, the better the chance for a cure.

One important factor in the staging of adenocarcinoma of the stomach is whether or not the tumor has invaded the surrounding tissue and, if it has, how deep it has penetrated. If invasion is limited, prognosis is favorable. Disease tissue that is more localized improves the outcome of surgical procedures performed to remove the diseased area of the stomach. This is called a resection of the stomach.

Several distinct ways of classifying stomach cancer according to cell type have been proposed. The Lauren classification is encountered most frequently. According to this classification system, gastric adenocarcinomas are either called intestinal or diffuse. Intestinal cancers are much like a type of intestinal cancer called intestinal carcinoma. Intestinal tumors are more frequently found in males and in older patients. The prognosis for these tumors is better than that for diffuse tumors. Diffuse tumors are more likely to infiltrate, that is, to move into another organ of the body.

Because symptoms of stomach cancer are so mild, treatment often does not commence until the disease is well advanced. The three standard modes of treatment for stomach cancer include surgery, radiation therapy, and chemotherapy. While deciding on the patient's treatment plan, the doctor takes into account many factors. The location of the cancer and its stage are important considerations. In addition, the patient's age, general health status, and personal preferences are also taken into account.

Surgery

In the early stages of stomach cancer, surgery may be used to remove the cancer. Surgical removal of adenocarcinoma is the only treatment capable of eliminating the disease. Laparoscopy is often used before surgery to investigate whether or not the tumor can be removed surgically. If the cancer is widespread and cannot be removed with surgery, an attempt will be made to remove blockage and control symptoms such as pain or bleeding. Depending on the location of the cancer, a portion of the stomach may be removed, a procedure called a partial **gastrectomy**. In a surgical procedure known as total gastrectomy, the entire stomach may be removed. However, doctors prefer to leave at least part of the stomach if possible. Patients who have been given a partial gastrectomy achieve a better quality of life than those having a total gastrectomy and typically lead normal lives. Even when the entire stomach is removed, the patients quickly adjust to a different eating schedule. This involves eating small quantities of food more frequently. High-protein foods are generally recommended.

Partial or total gastrectomy is often accompanied by other surgical procedures. Lymph nodes are frequently removed and nearby organs, or parts of these organs, may be removed if cancer has spread to them. Such organs may include the pancreas, colon, or spleen.

Preliminary studies suggest that patients who have tumors that cannot be removed by surgery at the start of therapy may become candidates for surgery later. Combinations of chemotherapy and radiation therapy are sometimes able to reduce disease for which surgery is not initially appropriate. Preliminary studies are being performed to determine if some of these patients can become candidates for surgical procedures after such therapies are applied.

Chemotherapy

Whether or not patients undergoing surgery for stomach cancer should receive chemotherapy is a controversial issue. Chemotherapy involves administering anti-cancer drugs either intravenously (through a vein in the arm) or orally (in the form of pills). This can either be used as the primary mode of treatment or after surgery to destroy any cancerous cells that may have migrated to distant sites. Most cancers of the gastrointestinal tract do not respond well to chemotherapy, however, adenocarcinoma of the stomach and advanced stages of cancer are exceptions.

Chemotherapy medicines such as doxorubicin, mitomycin C, and 5-fluorouracil, used alone, provide benefit to at least one in five patients. Combinations of agents may provide even more benefit, although it is

not certain that this includes longer survival. For example, some doctors use what is called the FAM regimen, which combines 5-fluorouracil, doxorubicin, and mitomycin. Some doctors prefer using 5-fluorouracil alone to FAM since side effects are more moderate. Another combination some doctors are using involve high doses of the medications methotrexate, 5-fluorouracil, and doxorubicin. Other combinations that have shown benefit include the ELF regimen, a combination of leucovorin, 5-fluorouracil, and etoposide. The EAP regimen, a combination of etoposide, doxorubicin, and cisplatin is also used.

Although chemotherapy using a single medicine is sometimes used, the best response rates are often achieved with combinations of medicines. Therefore, in addition to studies exploring the effectiveness of new medicines there are other studies attempting to evaluate how to best combine existing forms of chemotherapy to bring the greatest degree of help to patients.

Radiation therapy

Radiation therapy is often used after surgery to destroy the cancer cells that may not have been completely removed during surgery. To treat stomach cancer, external beam radiation therapy is generally used. In this procedure, high-energy rays from a machine that is outside of the body are concentrated on the area of the tumor. In the advanced stages of gastric cancer, radiation therapy is used to ease symptoms such as pain and bleeding. However, studies of radiation treatment for stomach cancer have shown that the way it has been used it has been ineffective for many patients.

Researchers are actively assessing the role of chemotherapy and radiation therapy used before a surgical procedure is conducted. They are searching for ways to use both chemotherapy and radiation therapy so that they increase the length of survival of patients more effectively than current methods are able to do.

Prognosis

Overall, approximately 20% of patients with stomach cancer live at least five years following diagnosis. Patients diagnosed with stomach cancer in its early stages have a far better prognosis than those for whom it is in the later stages. In the early stages, the tumor is small, lymph nodes are unaffected, and the cancer has not migrated to the lungs or the liver. Unfortunately, only about 20% of patients with stomach cancer are diagnosed before the cancer had spread to the lymph nodes or formed a distant metastasis.

It is important to remember that statistics on prognosis may be misleading. Newer therapies are being

KEY TERMS

Adenocarcinoma—Malignant cancers that originate in the tissues of glands or that form glandular structures.

Anemia—A condition in which iron levels in the blood are low.

Barium x ray (upper GI)—An x-ray test of the upper part of the gastrointestinal (GI) tract (including the esophagus, stomach, and a small portion of the small intestine) after the patient is given a white, chalky barium sulfate solution to drink. This substance coats the upper GI and the x rays reveal any abnormality in the lining of the stomach and the upper GI.

Biopsy—Removal of a tissue sample for examination under the microscope to check for cancer cells.

Chemotherapy—Treatment of cancer with synthetic drugs that destroy the tumor either by inhibiting the growth of the cancerous cells or by killing the cancer cells.

Endoscopic ultrasound (EUS)—A medical procedure in which sound waves are sent to the stomach wall by an ultrasound probe attached to the end of an endoscope. The pattern of echoes generated by the

reflected sound waves are translated into an image of the stomach wall by a computer.

External radiation therapy—Radiation therapy that focuses high-energy rays from a machine on the area of the tumor.

Infiltrate—A tumor that moves into another organ of the body.

Polyp—An abnormal growth that develops on the inside of a hollow organ such as the colon, stomach, or nose.

Radiation therapy—Treatment using high-energy radiation from x-ray machines, cobalt, radium, or other sources.

Total gastrectomy—Surgical removal (excision) of the entire stomach.

Upper endoscopy—A medical procedure in which a thin, lighted, flexible tube (endoscope) is inserted down the patient's throat. Through this tube the doctor can view the lining of the esophagus, stomach, and the upper part of the small intestine.

developed rapidly and five-year survival has not yet been measured with these. Also, the largest group of people diagnosed with stomach cancer are between 60 and 70 years of age, suggesting that some of these patients die not from cancer but from other age-related diseases. As a result, some patients with stomach cancer may be expected to have longer survival than did patients just ten years ago.

Prevention

Avoiding many of the risk factors associated with stomach cancer may prevent its development. Excessive amounts of salted, smoked, and pickled foods should be avoided, as should foods high in nitrates. A diet that includes recommended amounts of fruits and vegetables is believed to lower the risk of several cancers, including stomach cancer. The American Cancer Society recommends eating at least five servings of fruits and vegetables daily and choosing six servings of food from other plant sources, such as grains, pasta, beans, cereals, and whole grain bread.

Abstaining from tobacco and excessive amounts of alcohol will reduce the risk for many cancers. In countries where stomach cancer is common, such as Japan, early detection is important for successful treatment.

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National Coalition for Cancer Survivorship, 1010 Wayne Ave., Suite 770, Silver Spring, MD, 20910, (301) 650-9127, (301) 565-9670, (888) 650-9127, info@canceradvocacy.org, <http://www.canceradvocacy.org>.

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Bob Kirsch

Stomach flu see **Gastroenteritis**

Stomach flushing

Definition

Stomach flushing is the repeated introduction of fluids into the stomach through a nasogastric tube, and their subsequent withdrawal by **nasogastric suction**.

Purpose

Stomach flushing is performed to aid in controlling gastrointestinal bleeding or to cleanse the stomach of poisons.

Controlling stomach bleeding

Bleeding from the esophagus due to ruptured veins or bleeding from the stomach due to ulcers is a medical emergency. In an attempt to stop the bleeding, the stomach is flushed with large quantities of body-temperature saline solution or ice water. This procedure is called stomach flushing or gastric lavage.

Stomach flushing to control bleeding is not uniformly accepted, and some experts believe it is of little benefit and exposes the patient to unnecessary risks. It is usually done in conjunction with the administration of drugs to constrict the blood vessels.

Stomach flushing to remove poisons

At one time, stomach flushing was common practice to remove certain poisons. Recent thinking by the American Academy of Clinical Toxicology is that stomach flushing should not be used routinely with poisoned patients. It is useful only if the patient has swallowed a life-threatening quantity of poison, and when the flushing can be done within 60 minutes of having swallowed the poison.

Precautions

In **poisoning** cases, stomach flushing should not be used if the poison is a strong corrosive acid (hydrochloric acid, sulfuric acid), alkali (lye, ammonia), or a volatile hydrocarbon such as gasoline. Stomach flushing should also not be done on patients who are having convulsions. Patients who are losing or have lost consciousness must have their airways intubated before a nasogastric tube is inserted.

Description

Stomach flushing is performed in a hospital emergency room or intensive care unit by an

KEY TERMS

Electrolytes—Salts and minerals that ionize in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost any major biochemical reaction in the body.

Saline—A salt water solution that mimics the concentration of electrolytes in the blood.

emergency room physician or gastroenterologist. A nasogastric tube is inserted, and small amounts of saline or ice water are introduced into the stomach and withdrawn. The procedure is repeated until the withdrawn fluid is clear.

Preparation

Little preparation is necessary for this procedure other than educating the patient as to what will happen. The patient should remove dental appliances before the nasogastric tube is inserted.

Aftercare

After stomach flushing, the patient's vital signs will be monitored. Checks will be made for fluid and electrolyte imbalances. If necessary, additional treatment to prevent gastrointestinal bleeding or poisoning will be done.

Risks

In poisoning cases, stomach flushing delays the administration of **activated charcoal**, which may be more beneficial to treating the patient than flushing the stomach. In addition, stomach flushing may stimulate bleeding from the esophagus or stomach. The patient may inhale some of the stomach contents, causing aspiration, **pneumonia**, or infection in the lungs. Fluid and electrolyte imbalances are more likely to occur in older, sicker patients. Mechanical damage to the throat is more likely in patients who are uncooperative.

Normal results

Stomach flushing is usually tolerated by patients and is a temporary treatment, performed in conjunction with other therapies.

Resources

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Tish Davidson, A.M.

Stomach removal see *Gastrectomy*

Stomach resection see *Gastrectomy*

Stomachache

Definition

A stomachache is **pain** in the abdomen, between the bottom of the ribcage and the crease of the groin. Stomachaches are common in children and can have many causes—physical, psychological, and emotional.

Demographics

All children have occasional stomachaches. Stomachaches account for many visits to doctors and emergency rooms, as well as many missed school days. Approximately one third of children have been seen by a physician for abdominal pain by the age of 15, but only a small proportion of these represent serious problems. Complaints of abdominal pain are more common in children under age 11 than in older children and teens.

Chronic or recurrent abdominal pain (RAP) affects as many as 15% of children, especially between the ages of 4 and 12. In addition, as many as 2% of all children may experience abdominal migraines. Females are more affected by these than males.

Description

Although parents may be frightened or frustrated by their child’s stomachaches—especially when the cause is not obvious—abdominal pain that lasts less than three hours is rarely serious. There are different types of stomachaches:

- A localized stomachache is pain that is limited to one part of the abdomen.
- A generalized stomachache is pain involving at least half of the abdomen. It can occur with many different illnesses and conditions and usually resolves without medical treatment.
- A cramping-type stomachache is often relieved by passing gas or stool.

- Chronic abdominal pain or RAP, also called functional abdominal pain, is defined as occurring at least once a month for at least three months and is severe enough to interfere with a child’s activities. “Functional” means that the pain is real, but is not caused by a disease or other medical condition. It may be related to diet, stress, psychological or emotional problems, or nervous system immaturity or increased sensitivity.
- Abdominal migraines are stomachaches or cramping, usually near the navel or midline, that typically occur in reaction to the same triggers that cause migraine headaches.

Risk factors

Risk factors for stomachaches depend on the underlying physical, psychological, or emotional causes. Depression and **anxiety disorders** are risk factors for RAP in children and adolescents. Recurrent stomachaches in preschoolers have been shown to correlate with maternal depression. Abdominal migraines usually occur in children with a family history of migraine.

Causes and symptoms

Stomachaches in children are rarely serious and often have no identifiable cause. Mild stomachaches are often caused by overeating, gas pains, or **indigestion**. Foods that are too spicy or greasy can give children stomachaches. Changes in eating habits or bowel movements are also common causes, particularly in children under age 12. Constipation—the inability to pass a stool—is one of the most common causes of lower abdominal cramping.

Toddlers do not necessarily distinguish between physical pain and physical or emotional needs. Thus, a stomachache in a young child can indicate **fatigue**, hunger, or the need for a bowel movement.

Strep throat, caused by the *Streptococcus pyogenes* bacterium, accounts for up to 10% of acute stomachaches in children. Other infections that can cause stomachaches include:

- gastroenteritis or stomach flu—a viral infection that causes stomach cramps, diarrhea, and/or vomiting
- food poisoning—often caused by bacteria from undercooked or spoiled food—that can also cause severe diarrhea and/or vomiting lasting less than 12 hours
- a cough
- an ear infection
- chicken pox
- urinary tract infection
- pneumonia
- parasitic infections

Other physical causes of stomachaches in children include:

- swallowing air (aerophagia), which can result from anxiety or fear or from chewing gum or drinking carbonated beverages
- acid reflux or gastroesophageal reflux disease (GERD), in which stomach contents back up or reflux into the esophagus
- menstruation in girls
- lactose intolerance or difficulty digesting lactose—the sugar in milk products
- other food intolerances
- food allergies, such as a peanut allergy
- celiac disease—an inherited immune system response to gluten in flour
- latex allergy
- excessive caffeine
- irritable bowel syndrome
- inflammatory bowel disease
- poisoning from a plant, mushroom, drug, or chemical
- various medications, including drugs that have constipation as a side effect
- a swallowed foreign object
- injury to the abdomen

Pain that starts near the navel and progresses to severe pain in the lower right side of the abdomen can suggest **appendicitis**. Other symptoms of appendicitis include **fever**, loss of appetite, **vomiting**, and increasingly severe pain. Other rare but serious causes of stomachaches include:

- lead poisoning
- kidney infection
- a blocked intestine
- intussusception—slippage of a length of intestine into an adjacent portion, usually resulting in intestinal obstruction
- intestinal malrotation or twisting of a portion of the intestine

Stomachaches—especially RAP—in children aged three and older are very often stress-related. Stress- or anxiety-related causes of abdominal pain can include:

- separation anxiety, as when starting school
- social anxiety, such as attending a birthday party
- performance anxiety, such as a school exam
- over-excitement
- worry
- fear

- bullying
- abuse
- neglect
- depression
- anxiety disorders such as post-traumatic stress syndrome (PTSD)

Stomachaches can come on suddenly (acute) or persist for weeks or months (chronic). Stomachaches that last less than five minutes, even if they recur for many days, are unlikely to be serious. Stomach pain may be dull, sharp, or recurring cramps. The specific symptoms of a stomachache may suggest the cause:

- Indigestion or stomach flu can cause generalized abdominal pain.
- Pain and nausea or vomiting from a minor abdominal injury often subsides in just a few minutes.
- Stomachache, chest pain, and coughing are the most common symptoms of GERD in children.
- The closer the pain is to the navel, the more likely it is to be functional abdominal pain.
- When a stomachache is the only symptom, it is most often functional (the “Rule of Ones”).
- A mild generalized pain or cramps that become more severe over a period of hours may suggest an intestinal blockage.
- Localized pain that occurs suddenly and worsens can indicate a serious problem, such as inflammation of an abdominal organ, appendicitis, gallbladder disease, or peptic ulcer disease.
- Severe sudden pain or pain that increases with coughing or movement is occasionally a symptom of a serious medical problem.
- Symptoms of abdominal migraines can include sudden severe pain across the midline that may last for an hour or up to three days, as well as nausea, vomiting, paleness, and an inability to eat.

Although an older child will complain of a stomachache and can point to the area of pain and describe its severity, children younger than five or six may not be able to describe their stomachache accurately. Young children may simply hold their belly or point to it. Babies may fuss and cry, draw their legs up toward their bellies, or refuse to eat.

A physician should be consulted if a child has severe sudden abdominal pain, new mild abdominal pain that becomes more severe over a period of hours or days, or chronic or frequent stomachaches that interfere with normal activities such as school, play, eating, or sleeping through the night. A stomachache accompanied by any of the following symptoms also requires medical consultation:

KEY TERMS

Abdominal migraine—A variant form of a migraine headache; moderate to severe midline abdominal pain, usually occurring in children with a family history of migraine.

Appendicitis—Inflammation of the appendix—a narrow blind tube in the lower right abdomen.

Cognitive-behavioral therapy (CBT)—Treatment that identifies negative thoughts and behaviors and helps develop more positive approaches.

Constipation—The delayed or infrequent passage of dry, hard feces.

Functional pain—Pain that does not have a structural or organic cause.

Gastroenteritis—Stomach flu; inflammation of the lining of the stomach and intestines.

Gastroesophageal reflux disease (GERD)—Recurrent heartburn or acid indigestion caused by leakage of the stomach contents into the esophagus.

Recurrent abdominal pain (RAP)—Functional abdominal pain; stomachaches in children that recur at least once a month and that are not caused by an underlying medical condition.

- severe vomiting
- chronic severe diarrhea
- gastrointestinal bleeding
- unexplained fever
- persistent pain in the right side of the abdomen
- weight loss
- slow growth
- a family history of inflammatory bowel disease

A stomachache may be a medical emergency if the child:

- appears to be very ill
- becomes still or cannot stand or walk
- may have been poisoned by a plant, mushroom, medication, or chemical
- has had recent abdominal injury
- has a fever above 104°F (40°C) that is not reduced with fever medication
- is severely dehydrated
- is having difficulty breathing

Diagnosis

Examination

The healthcare provider will take a medical history and perform a **physical examination**, looking for signs of swelling and pressing on the child's belly to identify painful points. The physician will ask for information about the stomachache including:

- its intensity—mild, moderate, or severe
- whether the pain is crampy, a steady ache, sharp, or burning
- localization
- constancy

- duration
- recurrence
- foods, activities, or other factors that improve or worsen the pain
- any accompanying symptoms
- any other factors such as injury, recent travel, or drinking untreated water

Tests

Stomachaches do not usually require diagnostic tests. If appendicitis or another serious condition is suspected, then blood tests may be performed.

Procedures

Depending on the suspected cause of the stomachache, ultrasound imaging may be performed. X rays also may be taken.

Treatment

Traditional

Treatment of stomachaches depends on the suspected cause. They often do not require treatment. A special diet or specific eating instructions may be suggested. If **stress, anxiety**, or other psychological or emotional causes are suspected, the child may be referred to a psychologist or other mental health professional. Appendicitis requires an **appendectomy**, which is surgical removal of the appendix.

Drugs

Medications, especially **aspirin** and other **nonsteroidal anti-inflammatory drugs** (NSAIDs) such as ibuprofen and naproxen, can irritate the stomach lining and worsen stomachaches. If the stomachache is

accompanied by a fever above 102°F (39°C), a child may be given an appropriate dose of **acetaminophen** (Tylenol). Children should not be given a laxative for stomach cramps.

Children with GERD may be treated with drugs called **proton pump inhibitors** that suppress stomach acid. In 2008 the U.S. Food and Drug Administration (FDA) approved esomeprazole (Nexium) for the short-term treatment of GERD in children aged 1 to 11.

Alternative

Peppermint tea, peppermint oil capsules, or ginger can help relieve mild stomachaches. Behavioral interventions have had the most success in reducing or eliminating RAP. These therapies include:

- cognitive-behavioral therapy (CBT)
- guided imagery
- distraction therapy
- relaxation techniques, including breathing and progressive muscle relaxation
- biofeedback
- self-hypnosis
- coping skills for children and parents

Home remedies

Stomachaches are most often treated at home. Mild stomachaches may be treated by:

- bed rest
- consuming plenty of clear fluids (water, broth, tea, or fruit juice diluted with water) to prevent dehydration
- taking frequent, small sips of fluids
- eating several small meals instead of two or three large meals
- eating mild foods, such as rice, dry toast, crackers, gelatin, or applesauce
- avoiding high-fat and spicy foods, most fruits, and caffeinated and carbonated drinks for at least 48 hours after symptoms have passed
- having a child sit on the toilet and try to pass a stool, which can relieve stomach pain due to constipation or diarrhea
- sitting in warm water to relax the anus and help release stool
- preparing for a child to vomit by having a pan ready, since young children may refer to nausea as a tummy ache

Dietary changes have not been shown to be effective in treating frequent stomachaches in children.

However, increasing dietary fiber is a simple and inexpensive way to help some children.

Prognosis

Most mild stomachaches clear up within 30 minutes to 2 hours. Stomach cramps from **gastroenteritis** often precede each bout of **vomiting** and **diarrhea** and may last for several days. Stomachaches from **food poisoning** usually last less than 12 hours. With stomachaches that have serious causes, such as appendicitis, the pain continues to worsen and becomes constant.

Young children with RAP are more likely than other children to have behavioral problems and they are at greater risk for developing anxiety disorders as young adults. Children with abdominal migraines usually develop migraine headaches as they become older.

Prevention

Stomachaches in children are often preventable. Parents should ensure that their children:

- get plenty of sleep
- develop regular eating habits
- wash their hands before eating
- eat slowly
- avoid overeating
- avoid eating before bed
- have regular bowel movements and develop regular bowel habits
- eat fiber-rich foods that encourage regular bowel movements
- have only limited chewing gum and carbonated beverages to prevent swallowing air
- always correctly use car child-safety seats or seat belts to prevent abdominal injuries

Although children require less dietary fiber than adults, fiber may help prevent stomachaches in some children. The American Dietetic Association's formula for daily fiber intake, in grams, for children aged 3 to 18, is the child's age plus five grams. Fruits, vegetables, and whole grains are good sources of fiber. Dietary fiber should be increased gradually. Children should drink extra water or milk when increasing dietary fiber.

Resources

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ORGANIZATIONS

- American Academy of Family Physicians, 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2680, (913) 906-6000, (800) 274-6000, (913) 906-6075, <http://www.aafp.org>.
- American Academy of Pediatrics, 141 Northwest Point Blvd., Elk Grove Village, IL, 60007-1098, (874) 434-4000, (874) 434-8000, kidsdocs@aap.org, <http://www.aap.org>.
- American Dietetic Association, 120 South Riverside Plaza, Suite 2000, Chicago, IL, 60606-6995, (800) 877-1600, <http://www.eatright.org>.
- International Foundation for Functional Gastrointestinal Disorders, P.O. Box 170864, Milwaukee, WI, 53217-8076, (414) 964-1799, (888) 964-2001, (414) 964-7176, iffgd@iffgd.org, <http://aboutkidsgi.org>.

Margaret Alic, PhD

Stomatitis

Definition

Inflammation of the mucous lining of any of the structures in the mouth, which may involve the cheeks, gums, tongue, lips, and roof or floor of the mouth. The word “stomatitis” literally means inflammation of the



This patient is afflicted with stomatitis, a common inflammatory disease of the mouth. (*Custom Medical Stock Photo, Inc. Reproduced by permission.*)

mouth. The inflammation can be caused by conditions in the mouth itself, such as poor **oral hygiene**, poorly fitted dentures, or from mouth **burns** from hot food or drinks, or by conditions that affect the entire body, such as medications, allergic reactions, or infections.

Description

Stomatitis is an inflammation of the lining of any of the soft-tissue structures of the mouth. Stomatitis is usually a painful condition, associated with redness, swelling, and occasional bleeding from the affected area. **Bad breath** (halitosis) may also accompany the condition. Stomatitis affects all age groups, from the infant to the elderly.

Causes and symptoms

A number of factors can cause stomatitis; it is a fairly common problem in the general adult population in North America. Poorly fitted oral appliances, cheek biting, or jagged teeth can persistently irritate the oral structures. Chronic mouth breathing due to plugged nasal airways can cause dryness of the mouth tissues, which in turn leads to irritation. Drinking beverages that are too hot can burn the mouth, leading to irritation and **pain**. Diseases, such as herpetic infections (the **common cold** sore), **gonorrhea**, **measles**, leukemia, **AIDS**, and lack of vitamin C can present with oral signs. Other systemic diseases associated with stomatitis include inflammatory bowel disease (IBD) and Behcet’s syndrome, an inflammatory multi-system disorder of unknown cause.

Aphthous stomatitis, also known as recurrent aphthous ulcers (RAU) or **canker sores**, is a specific type of stomatitis that presents with shallow, painful ulcers that are usually located on the lips, cheeks, gums, or roof or

floor of the mouth. These ulcers can range from pinpoint size to up to 1 in (2.5 cm) or more in diameter. Though the causes of canker sores are unknown, nutritional deficiencies, especially of vitamin B₁₂, folate, or iron is suspected. Generalized or contact stomatitis can result from excessive use of alcohol, spices, hot food, or tobacco products. Sensitivity to mouthwashes, toothpastes, and lipstick can irritate the lining of the mouth. Exposure to heavy metals, such as mercury, lead, or bismuth can cause stomatitis. Thrush, a fungal infection, is a type of stomatitis.

Diagnosis

Diagnosis of stomatitis can be difficult. A patient's history may disclose a dietary deficiency, a systemic disease, or contact with materials causing an allergic reaction. A **physical examination** is done to evaluate the oral lesions and other skin problems. Blood tests may be done to determine if any infection is present. Scrapings of the lining of the mouth may be sent to the laboratory for microscopic evaluation, or cultures of the mouth may be done to determine if an infectious agent may be the cause of the problem.

Treatment

The treatment of stomatitis is based on the problem causing it. Local cleansing and good oral hygiene are fundamental. Sharp-edged foods such as peanuts, tacos, and potato chips should be avoided. A soft-bristled toothbrush should be used, and the teeth and gums should be brushed carefully; the patient should avoid banging the toothbrush into the gums. Local factors, such as ill-fitting dental appliances or sharp teeth, can be corrected by a dentist. An infectious cause can usually be treated with medication. Systemic problems, such as AIDS, leukemia, and anemia are treated by the appropriate medical specialist. Minor mouth burns from hot beverages or hot foods will usually resolve on their own in a week or so. Chronic problems with aphthous stomatitis are treated by first correcting any vitamin B₁₂, iron, or folate deficiencies. If those therapies are unsuccessful, medication can be prescribed which can be applied to each aphthous ulcer with a cotton-tipped applicator. This therapy is successful with a limited number of patients. More recently, low-power treatment with a carbon dioxide laser has been found to relieve the discomfort of recurrent aphthae. Major outbreaks of aphthous stomatitis can be treated with tetracycline **antibiotics** or **corticosteroids**. Valacyclovir has been shown to be effective in treating stomatitis caused by herpesviruses.

KEY TERMS

Aphthous stomatitis—A specific type of stomatitis presenting with shallow, painful ulcers. Also known as *canker sores*.

Stomatitis—Inflammation of the lining of the mouth, gums, or tongue.

Thrush—A form of stomatitis caused by *Candida* fungi and characterized by cream-colored or bluish patches on the tongue, mouth, or pharynx.

Patients may also be given topical anesthetics (usually a 2% lidocaine gel) to relieve pain and a protective paste (Orabase) or a coating agent like Kaopectate to protect eroded areas from further irritation from dentures, braces, or teeth.

Alternative treatment

Alternate treatment of stomatitis mainly involves prevention of the problem. Patients with such dental appliances as dentures should visit their dentist on a regular basis. Patients with systemic diseases or chronic medical problems need to ask their health care provider what types of oral problems they can expect from their particular disease. These patients must also contact their medical clinic at the first sign of problems. Common sense needs to be exercised when consuming hot foods or drinks. Use of tobacco products should be discouraged. Alcohol should be used in moderation. Mouthwashes and toothpastes known to the patient to cause problems should be avoided.

Botanical medicine can assist in resolving stomatitis. One herb, calendula (*Calendula officinalis*), in tincture form (an alcohol-based herbal extract) and diluted for a mouth rinse, can be quite effective in treating aphthous stomatitis and other manifestations of stomatitis.

More recently, a group of researchers in Brazil have reported that an extract made from the leaves of *Trichilia glabra*, a plant found in South America, is effective in killing several viruses that cause stomatitis.

Prognosis

The prognosis for the resolution of stomatitis is based on the cause of the problem. Many local factors can be modified, treated, or avoided. Infectious causes of stomatitis can usually be managed with medication, or, if the problem is being caused by a certain drug, by changing the offending agent.

Prevention

Stomatitis caused by local irritants can be prevented by good oral hygiene, regular dental checkups, and good dietary habits. Problems with stomatitis caused by systemic disease can be minimized by good oral hygiene and closely following the medical therapy prescribed by the patient's health care provider.

Resources

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ORGANIZATIONS

- American Dental Association, 211 E. Chicago Ave., Chicago, IL, 60611-2678, (312) 440-2500, <http://www.ada.org>.
- American Medical Association, 515 N. State St., Chicago, IL, 60654, (800) 621-8335, <http://www.ama-assn.org/>.

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Stone removal see **Gallstone removal**

Stool culture

Definition

Stool culture is a test to identify bacteria in patients with a suspected infection of the digestive tract. A sample of the patient's feces is placed in a special medium where bacteria is then grown. The bacteria that grow in

the culture are identified using a microscope and biochemical tests.

Purpose

Stool culture is performed to identify bacteria or other organisms in persons with symptoms of gastrointestinal infection, most commonly **diarrhea**. Identification of the organism is necessary to determine the treatment of the patient's infection or to trace the cause of an outbreak or epidemic of certain types of diarrhea.

According to the Centers for Disease Control and Prevention (CDC), doctors are most likely to order a stool culture for patients with any of the following characteristics: **AIDS**, bloody stools, diarrhea lasting longer than three days, high **fever**, history of recent travel abroad, or severe **dehydration**.

Precautions

Stool culture is performed only if an infection of the digestive tract is suspected. The test has no harmful effects.

Description

Stool culture may also be called fecal culture. To obtain a specimen for culture, the patient is asked to collect a stool sample into a special sterile container. In some cases, the container may contain a transport solution. Specimens may need to be collected on three consecutive days. It is important to return the specimen to the doctor's office or the laboratory in the time specified by the physician or nurse. Laboratories do not accept stool specimens contaminated with water, urine, or other materials.

The culture test involves placing a sample of the stool on a special substance, called a medium, that provides nutrients for certain organisms to grow and reproduce. The medium is usually a thick gel-like substance. The culture is done in a test tube—or on a flat round culture plate—which is incubated at the proper temperature for growth of the bacteria. After a colony of bacteria grows in the medium, the type of bacteria is identified by observing the colony's growth, its physical characteristics, and its microscopic features. The bacteria may be dyed with special stains that make it easier to identify features specific to particular bacteria.

The length of time needed to perform a stool culture depends on the laboratory where it is done and the culture methods used. Stool culture usually takes 72 hours or longer to complete. Some organisms may take several weeks to grow in a culture.

An antibiotic sensitivity test may be done after a specific bacterium is identified. This test shows which **antibiotics** will be most effective for treating the infection.

Although most intestinal infections are caused by bacteria, in some cases a fungal or viral culture may be necessary. The most common bacterial infections of the digestive tract are caused by *Shigella*, *Salmonella*, *Campylobacter*, and *Yersinia*. Patients taking certain antibiotics may be susceptible to infection with *Clostridium difficile*. In some cases, as with *Clostridium difficile*, the stool culture is used to detect the toxin (poison or harmful chemical) produced by the bacteria.

Patients with AIDS, or other immune system diseases, may also have gastrointestinal infections caused by such fungi as *Candida*, or viral organisms including cytomegalovirus.

Several intestinal parasites may cause gastrointestinal infection and diarrhea. Parasites are not cultured, but are identified microscopically in a test called "Stool Ova and Parasites."

Insurance coverage for stool culture may vary among different insurance plans. This common test usually is covered if ordered by a physician approved by the patient's insurance plan, and if it is done at an approved laboratory.

Alternative methods

Newer methods of testing stool samples for specific disease organisms include various forms of polymerase chain reaction (PCR) assays. One type that has been used to test for several different types of intestinal viruses at the same time is the RT-PCR, which stands for reverse transcriptase polymerase chain reaction. This assay measures changes in an organism's messenger RNA. RT-PCR assays have several advantages over standard stool cultures: they require only very small samples of material; they can be performed much more rapidly; and they can be used to test environmental water for virus contamination as well as human stool samples.

Preparation

The physician or other healthcare provider will ask the patient for a complete medical history and perform a **physical examination** to determine possible causes of the gastrointestinal problem. Information about the patient's diet, any medications taken, and recent travel may provide clues to the identity of possible infectious organisms.

KEY TERMS

Bismuth—A substance used in medicines to treat diarrhea, nausea, and indigestion.

Enteric—Pertaining to the intestine.

Enterotoxigenic—Refers to an organism that produces toxins in the gastrointestinal tract that cause such things as vomiting, diarrhea, and other symptoms of food poisoning.

Feces—Material excreted by the intestines.

Flora—Refers to normal bacteria found in a healthy person.

Gastrointestinal—Referring to the digestive tract; the stomach and intestines.

Psyllium hydrophilic mucilloid—A plant material contained in some laxatives.

Sterile—Free of microorganisms.

Toxin—A poison; usually refers to protein poisons produced by bacteria, animals, and some plants.

Stool culture normally does not require any special preparation. Patients do not need to change their diet before collecting the specimen. Intake of some substances can contaminate the stool specimen and should not be taken the day before collection. These substances include castor oil, bismuth, and laxative preparations containing psyllium hydrophilic mucilloid.

Normal results

Bacteria that are normally found in the intestines include *Pseudomonas* and *Escherichia coli*. These enteric bacteria (bacteria of the gastrointestinal system) are considered normal flora and usually do not cause infection in the digestive tract.

Abnormal results

Bacteria that do not normally inhabit the digestive tract, and that are known to cause gastrointestinal infection include *Shigella*, *Salmonella*, *Campylobacter*, and *Yersinia*. *Clostridium difficile* produces a toxin that can cause severe diarrhea. Other bacteria that produce toxins are *Staphylococcus aureus*, *Bacillus cereus*, and enterotoxigenic (producing disease in the digestive system) *Escherichia coli*. Although *Escherichia coli* is a normal bacteria found in the intestines, the enterotoxigenic type of this bacteria can be acquired from eating contaminated meat, juice, or

fruits. It produces a toxin that causes severe inflammation and bleeding of the colon.

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ORGANIZATIONS

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

Toni Rizzo
Rebecca J. Frey, PhD

Stool fat test

Definition

Stool fats, also known as fecal fats, or fecal lipids, are fats that are excreted in the feces. When secretions from the pancreas and liver are adequate, emulsified dietary fats are almost completely absorbed in the small intestine. When a malabsorption disorder or other cause disrupts this process, excretion of fat in the stool increases.

Purpose

This test evaluates digestion of fats by determining excessive excretion of lipids in patients exhibiting signs of malabsorption, such as weight loss, abdominal distention, and scaly skin.

Precautions

Drugs that may increase fecal fat levels include **enemas** and **laxatives**, especially mineral oil. Drugs that may decrease fecal fat include Metamucil and barium. Other substances that can affect test results include alcohol, potassium chloride, **calcium** carbonate, neomycin, kanamycin, and other broad-spectrum **antibiotics**.

Description

Excessive excretion of fecal fat is called steatorrhea, a condition that is suspected when the patient has large, "greasy," and foul-smelling stools. Both digestive and absorptive disorders can cause steatorrhea. Digestive disorders affect the production and release of the enzyme lipase from the pancreas, or bile from the liver, which are substances that aid digestion of fats; absorptive disorders disturb the absorptive and enzyme functions of the intestine. Any condition that causes malabsorption or maldigestion is also associated with increased fecal fat. As an example, children with **cystic fibrosis** have mucous plugs that block the pancreatic ducts. The absence or significant decrease of the pancreatic enzymes, amylase, lipase, trypsin, and chymotrypsin limits fat protein and carbohydrate digestion, resulting in steatorrhea due to fat malabsorption.

Both qualitative and quantitative tests are used to identify excessive fecal fat. The qualitative test involves staining a specimen of stool with a special dye, then examining it microscopically for evidence of malabsorption, such as undigested muscle fiber and various fats. The quantitative test involves drying and weighing a 72-hour stool specimen, then using an extraction technique to separate the fats, which are subsequently evaporated and weighed. This measurement of the total output of fecal fat per 24 hours in a three-day specimen is the most reliable test for steatorrhea.

Preparation

This test requires a 72-hour stool collection. The patient should abstain from alcohol during this time and maintain a high-fat diet (100 g/day) for three days before the test, and during the collection period. The patient should call the laboratory for instructions on how to collect the specimen.

Normal results

Reference values vary from laboratory to laboratory but are generally found within the range of 5–7 g/24 hr.

It should be noted that children, especially infants, cannot ingest the 100 g/day of fat that is suggested for the test. Therefore, a fat retention coefficient is determined by measuring the difference between ingested fat and fecal fat, and expressing that difference as a percentage. The figure, called the fat retention coefficient, is 95% or greater in healthy children and adults. A low value is indicative of steatorrhea.

Abnormal results

Increased fecal fat levels are found in cystic fibrosis, malabsorption secondary to other conditions like Whipple's disease or **Crohn's disease**, maldigestion secondary to pancreatic or bile duct obstruction, and "short-gut" syndrome secondary to surgical resection, bypass, or congenital anomaly.

Resources

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Janis O. Flores

Precautions

Stool O & P is performed if an infection of the digestive tract is suspected. The test has no harmful effects.

Description

Examination of the stool for ova and parasites is done to diagnose parasitic infection of the intestines. The test may be done in the doctor's office or a laboratory. The patient collects a stool sample in one or more sterile containers containing special chemical fixatives. The feces should be collected directly into the container. It must not be contaminated with urine, water, or other materials. Three specimens are often needed—collected every other day, or every third day. However, as many as six specimens may be needed to diagnose the amoeba *Entamoeba histolytica*. The specimen does not need to be refrigerated. It should be delivered to the doctor's office or laboratory within 12 hours.

In the laboratory, the stool sample is observed for signs of parasites and their eggs. Some parasites are large enough to be seen without a microscope. For others, microscope slides are prepared with fresh unstained stool, and with stool dyed with special stains. These preparations are observed with a microscope for the presence of parasites or their eggs.

An unstained stool examination for ova and parasites normally only takes a few minutes. If specimen staining and other preparation is done, the test may take longer. When the specimen is sent to a laboratory, the results may take eight to 24 hours to be reported.

The most common intestinal parasites in North America that cause infections are:

- roundworms: *Ascaris lumbricoides*
- hookworms: *Necator americanus*
- pinworms: *Enterobius follicularis*
- tapeworms: *Diphyllobothrium latum*, *Taenia saginata*, and *Taenia solium*
- protozoa: *Entamoeba histolytica* (an amoeba), and *Giardia lamblia* (a flagellate)

Numerous other parasites are found in other parts of the world. These may be contracted by travelers to other countries. Patients with acquired immune deficiency syndrome (**AIDS**) or other immune system disorders are commonly infected with the parasites in the *Microsporidia* family, *Cryptosporidium*, and *Isospora belli*.

Insurance coverage for stool ova and parasites may vary among different insurance plans. This test usually is covered if ordered by a physician approved by the

Stool O & P test

Definition

The stool O & P test is the stool ova and parasites test. In this test, a stool sample is examined for the presence of intestinal parasites and their eggs, which are called ova.

Purpose

The ova and parasites test is performed to look for and identify intestinal parasites and their eggs in persons with symptoms of gastrointestinal infection. Patients may have no symptoms, or experience **diarrhea**, blood in the stools, and other gastrointestinal distress. Identification of a particular parasite indicates the cause of the patient's disease and determines the medication needed to treat it.

KEY TERMS

Amoeba—A type of protozoa (one-celled animal) that can move or change its shape by extending projections of its cytoplasm.

Bismuth—A substance used in medicines to treat diarrhea, nausea, and indigestion.

Cryptosporidium—A type of parasitic protozoa.

Feces—Material excreted by the intestines.

Flagellate—A microorganism that uses flagella (hair-like projections) to move.

Gastrointestinal—Referring to the digestive tract; the stomach and intestines.

Isospora belli—A type of parasitic protozoa.

Microsporida—A type of parasitic protozoa.

Ova—Eggs.

Parasite—An organism that lives on or inside another living organism (host), causing damage to the host.

Pathogenic—Disease-causing.

Protozoa—One-celled eukaryotic organisms belonging to the kingdom Protista.

Sterile—Free of microorganisms.

patient's insurance plan, and if it is done at an approved laboratory.

Preparation

The physician, or other healthcare provider, will ask the patient for a complete medical history, and perform a **physical examination** to determine possible causes of the gastrointestinal symptoms. Information about the patient's diet, any medications taken, and recent travel may provide clues to the identity of possible infectious parasites.

Collecting a stool sample for ova and parasite detection normally does not require any special preparation. Patients do not need to change their diet before collecting the specimen. Patients should avoid taking any medications or treatments containing mineral oil, castor oil, or bismuth, magnesium or other antidiarrheal medicines, or **antibiotics** for 7 to 10 days before collecting the specimen.

Normal results

Normally, parasites and eggs should not be found in stools. Some parasites are not pathogenic, which means they do not cause disease. If these are found, no treatment is necessary.

Abnormal results

The presence of any pathogenic parasite indicates an intestinal parasitic infection. Depending on the parasite identified, other tests may need to be performed to determine if the parasite has invaded other parts of the body. Some parasites travel from the intestines to other parts of the body and may already have caused damage to other tissues by the time a diagnosis is made. For

example, the roundworm, *Ascaris* penetrates the intestinal wall and can cause inflammation in the abdomen. It can also migrate to the lungs and cause **pneumonia**. This kind of injury can occur weeks before the roundworm eggs show up in the stool.

Other types of damage caused by intestinal parasites include anemia due to hemorrhage caused by hookworms, and anemia caused by depletion of vitamin B₁₂ through the action of tapeworms.

When a parasite is identified, the patient can be treated with the appropriate medications to eliminate the parasite.

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Toni Rizzo

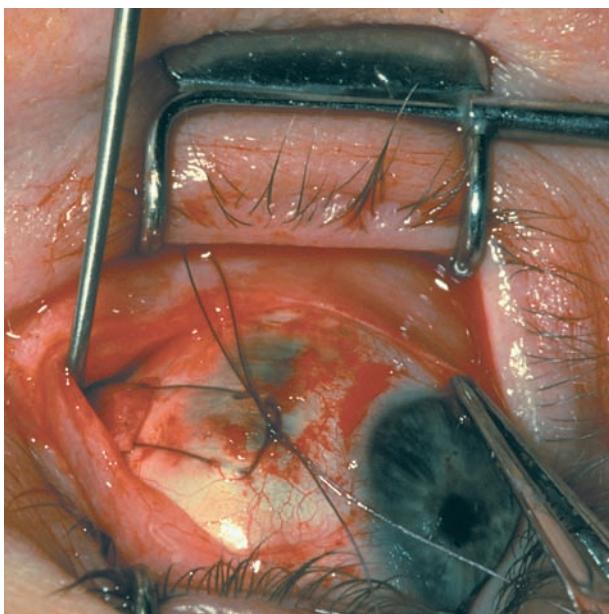
Stool occult blood test see **Fecal occult blood test**

Stool ova and parasites test see **Stool O & P test**

Strabismus

Definition

Strabismus is a condition in which the eyes do not point in the same direction. It can also be referred to as a tropia or squint.



A close-up of ophthalmic surgery being performed to correct strabismus. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Description

Strabismus occurs in 2–5% of all children. About half are born with the condition, which causes one or both eyes to turn:

- inward (esotropia or “crossed eyes”)
- outward (exotropia or “wall eyes”)
- upward (hypertropia)
- downward (hypotropia)

Strabismus is equally common in boys and girls. It sometimes runs in families.

Types of strabismus

Esotropia is the most common type of strabismus in infants. Accommodative esotropia develops in children under age two who cross their eyes when focusing on objects nearby. This usually occurs in children who are moderately to highly farsighted (hyperopic).

Another common form of strabismus, exotropia, may only be noticeable when a child looks at far-away objects, daydreams, or is tired or sick.

Sometimes the eye turn is always in the same eye; however sometimes the turn alternates from one eye to the other’.

Most children with strabismus have comitant strabismus. No matter where they look, the degree of deviation does not change. In incomitant strabismus,

the amount of misalignment depends upon which direction the eyes are pointed.

False strabismus (pseudostrabismus)

A child may appear to have a turned eye, however this appearance may actually be due to:

- extra skin that covers the inner corner of the eye
- a broad, flat nose
- eyes set unusually close together or far apart

This condition, false strabismus, usually disappears as the child’s face grows. An eye doctor needs to determine whether the eyeturn is true or pseudostrabismus.

With normal vision, both eyes send the brain the same message. This binocular fixation (both eyes looking directly at the same object) is necessary to see three-dimensionally and to aid in depth perception. When an eye is misaligned, the brain receives two different images. Young children learn to ignore distorted messages from a misaligned eye, but adults with strabismus often develop double vision (diplopia).

A baby’s eyes should be straight and parallel by three or four months of age. A child who develops strabismus after the age of eight or nine years is said to have adult-onset strabismus.

Causes and symptoms

Strabismus can be caused by a defect in muscles or the part of the brain that controls eye movement. It is especially common in children who have:

- brain tumors
- cerebral palsy
- Down syndrome
- hydrocephalus
- other disorders that affect the brain

Diseases that cause partial or total blindness can cause strabismus. So can extreme farsightedness, **cataracts**, eye injury, or having much better vision in one eye than the other.

In adults, strabismus is usually caused by:

- diabetes
- head trauma
- stroke
- brain tumor
- other diseases affecting nerves that control eye muscles

The most obvious symptom of strabismus is an eye that isn’t always straight. The deviation can vary

from day to day or during the day. People who have strabismus often squint in bright sunlight or tilt their heads to focus their eyes.

Diagnosis

Every baby's eyes should be examined by the age of six months. A baby whose eyes have not straightened by the age of four months should be examined to rule out serious disease.

A pediatrician, family doctor, ophthalmologist, or optometrist licensed to use diagnostic drugs uses drops that dilate the pupils and temporarily paralyze eye-focusing muscles to evaluate visual status and ocular health. Early diagnosis is important. Some eye turns may be a result of a tumor. Untreated strabismus can damage vision in the unused eye and possibly result in lazy eye (**amblyopia**).

Treatment

Preserving or restoring vision and improving appearance may involve one or more of the following:

- glasses to aid in focusing and straighten the eye(s)
- patching to force infants and young children to use and straighten the weaker eye
- eye drops or ointments as a substitute for patching or glasses, or to make glasses more effective
- surgery to tighten, relax, or reposition eye muscles
- medication injected into an overactive eye muscle to allow the opposite muscle to straighten the eye
- vision training (also called eye exercises)

Prognosis

Early consistent treatment usually improves vision and appearance. The most satisfactory results are achieved if the condition is corrected before the age of seven years old.

ORGANIZATIONS

American Academy of Ophthalmology (AAO), P. O. Box 7424, San Francisco, CA, 94120-7424, (415) 561-8500, (415) 561-8500, <http://www.aao.org>.

American Academy of Pediatric Ophthalmology and Strabismus (AAPOS), PO Box 193832, San Francisco, CA, 94119-3832, (415) 561-8505, (415) 561-8531, aapos@aoa.org, <http://www.aapos.org>.

American Optometric Association, 243 North Lindbergh Blvd., St. Louis, MO, 63141, (314) 991-4100, (314) 991-4101, (800) 365-2219, <http://www.aoa.org/>.

Maureen Haggerty

Strawberry marks see **Birthmarks**

Strengthening exercises see **Exercise**

Strep culture see **Throat culture**

Strep test see **Streptococcal antibody tests**

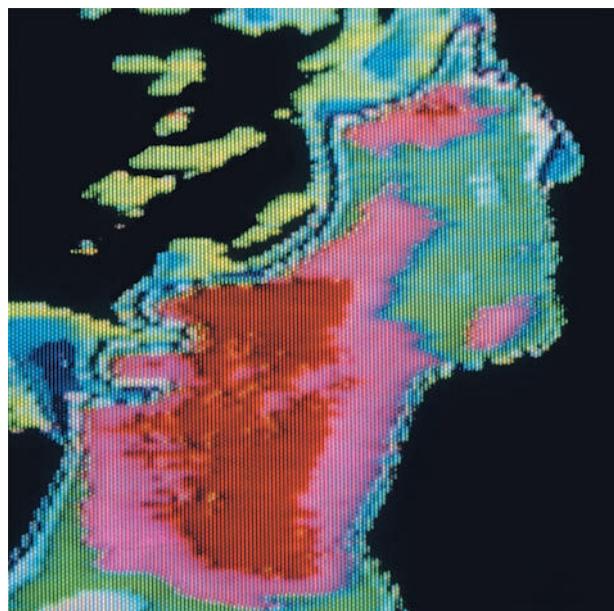
Strep throat

Definition

Streptococcal **sore throat**, or strep throat as it is more commonly called, is an infection of the mucous membranes lining the pharynx (throat). Sometimes the tonsils are also infected (**tonsillitis**). The disease is caused by group A *Streptococcus* bacteria. Untreated strep throat may develop into **rheumatic fever** or other serious conditions.

Description

Strep throat accounts for between 5–10% of all sore throats. Although anyone can get strep throat, it is most common in school-age children. People who smoke, who are fatigued, run down, or who live in damp, crowded conditions are also more likely to become infected. Children under age two and adults who are not around children are less likely to get the



A thermographic image showing a streptococcal sore throat, or strep throat. (© Howard Sochurek/Corbis.)

disease; their sore throats are usually caused by viruses, not strep bacteria.

Strep throat occurs most frequently from November to April. The disease passes directly from person to person by coughing, sneezing, and close contact. Very occasionally the disease is passed through food, when a food handler infected with strep bacteria accidentally contaminates food by coughing or sneezing. Statistically, if someone in the household is infected, one out of every four other household members is likely to get strep throat within two to seven days.

Causes and symptoms

A person with strep throat suddenly develops a painful sore throat one to five days after being exposed to streptococcus bacteria. The **pain** is indistinguishable from sore throats caused by other types of bacteria or viruses.

The infected person usually feels tired and has a **fever**, sometimes accompanied by chills, **headache**, muscle aches, swollen lymph glands, and **nausea**. Young children may complain of abdominal pain. The tonsils look swollen and are bright red, with white or yellow patches of pus on them. Sometimes the roof of the mouth is red or has small red spots. Often a person with strep throat has **bad breath**.

Despite these common symptoms, strep throat can be deceptive. It is possible to have the disease and not show any of these symptoms. Many young children complain only of a headache and stomach ache, without the characteristic sore throat symptoms.

Occasionally, within a few days of developing the sore throat, an individual may develop a fine, rough, sunburn-like rash over the face and upper body and have a fever of 101–104°F (38.3–40°C). The tongue becomes bright red, with a flecked, strawberry-like appearance. When a rash develops, this form of strep throat is called **scarlet fever**. The rash is a reaction to toxins (poisons) released by streptococcus bacteria. Scarlet fever is no more dangerous than strep throat, and it is treated the same way. The rash disappears in about five days. One to three weeks later, patches of skin may peel off, as might occur with a **sunburn**, especially on the fingers and toes.

Untreated strep throat can cause rheumatic fever and rheumatic heart disease (damage to the heart caused by rheumatic fever). This is a serious illness, although it is uncommon in the United States. The most recent outbreak in the U.S. occurred in the mid-1980s. Worldwide, however, 90,000 people are estimated to die from rheumatic heart disease each year, with the highest rates occurring in developing

countries. Rheumatic fever occurs most often in children between the ages of 5 and 15, and may have a genetic component since it seems to run in families. Although the strep throat that causes rheumatic fever is contagious, rheumatic fever itself is not.

Rheumatic fever begins one to six weeks after an untreated streptococcal infection. The joints, especially the wrists, elbows, knees, and ankles become red, sore, and swollen. The infected person develops a high fever, and possibly a rapid heartbeat when lying down, paleness, **shortness of breath**, and fluid retention. A red rash over the trunk may come and go for weeks or months. An acute attack of rheumatic fever lasts about three months.

Rheumatic fever can cause permanent damage to the heart and heart valves. It can be prevented by promptly treating **streptococcal infections** with **antibiotics**. It does not occur if all the streptococcus bacteria are killed within the first 10–12 days after infection.

In the 1990s, outbreaks of a virulent strain of group A *Streptococcus* were reported to cause a toxic-shock-like illness and a severe invasive infection called necrotizing fasciitis, which destroys skin and muscle tissue. Although these diseases are caused by group A *Streptococci*, they rarely begin with strep throat. Usually the streptococcus bacteria enter the body through a skin wound. These complications are rare. However, since the **death** rate in necrotizing fasciitis is 30–50%, it is wise to seek prompt treatment for any streptococcal infection.

Diagnosis

Diagnosis of a strep throat by a doctor begins with a **physical examination** of the throat and chest. The doctor will also look for signs of other illness, such as a sinus infection or **bronchitis**, and seek information about whether the patient has been around other people with strep throat. If it appears that the patient may have strep throat, the doctor will do laboratory tests.

There are two types of tests to determine if a person has strep throat. A rapid strep test can determine only the presence of streptococcal bacteria in material collected on a sterile swab from the throat. It will not tell the doctor whether the sore throat is caused by another kind of bacteria or if it is caused by a virus. The results of a rapid strep test are available in about 20 minutes. The advantage of this test is the speed with which a diagnosis can be made.

The rapid strep test has a false negative rate of about 25%. In other words, in about one out of every four cases where no strep is detected by the rapid strep test, the patient actually does have strep throat.

Because of this, when a rapid strep test is negative, the doctor often does a **throat culture**.

For a rapid strep test or a throat culture, a nurse will use a sterile swab to reach down into the throat and obtain a sample of material from the sore area. The procedure takes only a few seconds, but may cause gagging.

For a throat culture, a sample of swabbed material is cultured, or grown, in the laboratory on a medium that allows technicians to determine what kind of bacteria are present. Results take 24–48 hours. The test is very accurate and will show the presence of other kinds of bacteria besides *Streptococci*. It is important not to take any leftover antibiotics before visiting the doctor and having a throat culture. Even small amounts of antibiotics can suppress the bacteria and mask its presence in the throat culture, thus preventing the patient from getting proper treatment.

In the event that rheumatic fever is suspected, the doctor does a blood test. This test, called an antistreptolysin-O test, will tell the doctor whether the person has recently been infected with strep bacteria. This helps the doctor distinguish between rheumatic fever and **rheumatoid arthritis**.

Treatment

Strep throat is treated with antibiotics. Penicillin is the preferred medication. Oral penicillin must be taken for 10 days. Patients need to take the entire amount of antibiotic prescribed and not discontinue taking the medication when they feel better. Stopping the antibiotic early can lead to a return of the strep infection. Occasionally, a single injection of long-acting penicillin (Bicillin) is given instead of 10 days of oral treatment.

About 10% of the time, penicillin is not effective against the strep bacteria. When this happens a doctor may prescribe other antibiotics such as cefuroxime (Ceftin), cefixime (Suprax), cefpodoxime proxetil (Vantin), loracarbef (Lorabid), cefditoren (Spectracef), azithromycin (Zithromax), clindamycin (Cleocin), or a cephalosporin (Keflex, Durocef, Ceclor). Erythromycin (Eryzole, Pedazole, Ilosone), another inexpensive antibiotic, can be given to people who are allergic to penicillin. Scarlet fever is treated with the same antibiotics as strep throat.

Without treatment, the symptoms of strep throat begin subsiding in four or five days. However, because of the possibility of developing rheumatic fever, it is important to treat strep throat promptly with antibiotics. If rheumatic fever does occur, it is also treated with antibiotics. Anti-inflammatory drugs, such as **steroids**, are

used to treat joint swelling. **Diuretics** are used to reduce water retention. Once the rheumatic fever becomes inactive, children may continue on low doses of antibiotics to prevent a reoccurrence. Necrotizing fasciitis is treated with hospitalization and intravenous antibiotics.

Home care for strep throat

There are home care steps that people can take to ease the discomfort of their strep symptoms.

- Take acetaminophen or ibuprofen for pain. Aspirin should not be given to children because of its association with an increase in Reye's Syndrome, a serious disease.
- Gargle with warm double strength tea or warm salt water, made by adding one teaspoon of salt to eight ounces of water, to relieve sore throat pain.
- Drink plenty of fluids, but avoid acidic juices like orange juice because they irritate the throat.
- Eat soft, nutritious foods like noodle soup. Avoid spicy foods.
- Avoid smoke and smoking.
- Rest until the fever is gone, then resume strenuous activities gradually.
- Use a room humidifier, as it may make sore throat sufferers more comfortable.
- Be aware that antiseptic lozenges and sprays may aggravate the sore throat rather than improve it.

Alternative treatment

Alternative treatment focuses on easing the symptoms of strep throat through herbs and botanical medicines. Some practitioners suggest using these treatments in addition to antibiotics, since they primarily address the comfort of the patient and not the underlying infection. Many practitioners recommend *Lactobacillus acidophilus* to offset the suppressive effects of antibiotics on the beneficial bacteria of the intestines.

Some suggested treatments include:

- Inhaling fragrances of the essential oils of lavender (*Lavandula officinalis*), thyme (*Thymus vulgaris*), eucalyptus (*Eucalyptus globulus*), sage (*Salvia officinalis*), and sandalwood (Aromatherapy).
- Gargling with a mixture of water, salt, and turmeric (*Curcuma longa*) powder or astringents, such as alum, sumac, sage, and bayberry (Ayurvedic medicine).
- Taking osha root (*Ligusticum porteri*) internally for infection or drinking tea made of sage, echinacea (*Echinacea spp.*) and cleavers (*Gallium aparine*). Osha root has an unpleasant taste many children will not accept (Botanical medicine).

KEY TERMS

Lactobacillus acidophilus—A bacteria found in yogurt that changes the balance of the bacteria in the intestine in a beneficial way.

Prognosis

Patients with strep throat begin feeling better about 24 hours after starting antibiotics. Symptoms rarely last longer than five days. People remain contagious until after they have been taking antibiotics for 24 hours. Children should not return to school or childcare until they are no longer contagious. Food handlers should not work for the first 24 hours after antibiotic treatment because strep infections are occasionally passed through contaminated food. People who are not treated with antibiotics can continue to spread strep bacteria for several months.

About 10% of strep throat cases do not respond to penicillin. People who have even a mild sore throat after a 10 day treatment with antibiotic should return to their doctor. An explanation for this may be that the person is a carrier of strep but that something other than a strep bacterium is causing the sore throat.

Taking antibiotics within the first week of a strep infection will prevent rheumatic fever and other complications. If rheumatic fever does occur, the outcomes vary considerably. Some cases may be cured. In others there may be permanent damage to the heart and heart valves. In rare cases, rheumatic fever can be fatal.

Necrotizing fasciitis has a death rate of 30–50%. Patients who survive often suffer a great deal of tissue and muscle loss. Fortunately, this complication of a streptococcus infection is very rare.

Prevention

There is no way to prevent getting a strep throat. However, the risk of getting one or passing one on to another person can be minimized by:

- washing hands well and frequently, especially after nose blowing or sneezing and before food handling
- disposing of used tissues properly
- avoiding close contact with someone who has a strep throat
- not sharing food and eating utensils with anyone
- not smoking

Resources

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Tish Davidson, A.M.

Streptobacillary rat-bite fever see **Rat-bite fever**

Streptococcal antibody tests

Definition

Streptococcal infections are caused by a microorganism called *Streptococcus*. Three streptococcal antibody tests are available: the antistreptolysin O titer (ASO), the antideoxyribonuclease-B titer (anti-Dnase-B, or ADB), and the streptozyme test.

Purpose

The antistreptolysin O titer, or ASO, is ordered primarily to determine whether a previous group A *Streptococcus* infection has caused a poststreptococcal disease, such as **scarlet fever**, **rheumatic fever**, or a **kidney disease** called **glomerulonephritis**.

The anti-DNase-B (ADB) test is performed to determine a previous infection of a specific type of *Streptococcus*, group A beta-hemolytic *Streptococcus*. Identification of infections of this type are particularly important in suspected cases of acute rheumatic **fever** (ARF) or acute glomerulonephritis.

Streptozyme is a screening test used to detect antibodies to several streptococcal antigens. An antigen is a substance that can trigger an immune response, resulting in production of an antibody as part of the body’s defense against infection and disease.

Precautions

For the ASO test, increased levels of fats, called beta lipoproteins, in the blood can neutralize streptolysin O and cause a false-positive ASO titer. **Antibiotics**, which reduce the number of streptococci and thereby suppress ASO production, may decrease ASO levels. **Steroids**, which suppress the immune system, consequently may also suppress ASO production. Also Group A streptococcal infections of the skin may not produce an ASO response. Antibiotics also may decrease anti-DNase-B (ADB) levels.

Description

Streptococcal infections are caused by bacteria known as *Streptococcus*. There are several disease-causing strains of streptococci (groups A, B, C, D, and G), which are identified by their behavior, chemistry, and appearance. Each group causes specific types of infections and symptoms. These antibody tests are useful for group A streptococci. Group A streptococci are the most virulent species for humans and are the cause of **strep throat**, **tonsillitis**, wound and skin infections, blood infections (septicemia), scarlet fever, **pneumonia**, rheumatic fever, **Sydenham's chorea** (formerly called St. Vitus' dance), and glomerulonephritis.

Although symptoms may suggest a streptococcal infection, the diagnosis must be confirmed by tests. The best procedure, and one that is used for an acute infection, is to take a sample from the infected area for culture, a means of growing bacteria artificially in the laboratory. However, cultures are useless about two to three weeks after initial infection, so the ASO, anti-DNase-B, and streptozyme tests are used to determine if a streptococcal infection is present.

Antistreptolysin O titer (ASO)

The ASO titer is used to demonstrate the body's reaction to an infection caused by group A beta-hemolytic streptococci. Group A streptococci produce the enzyme streptolysin O, which can destroy (lyse) red blood cells. Because streptolysin O is antigenic (contains a protein foreign to the body), the body reacts by producing antistreptolysin O (ASO), which is a neutralizing antibody. ASO appears in the blood serum one week to one month after the onset of a strep infection. A high titer (high levels of ASO) is not specific for any type of poststreptococcal disease, but it does indicate if a streptococcal infection is or has been present.

Serial (several given in a row) ASO testing is often performed to determine the difference between an acute or convalescent blood sample. The diagnosis of

a previous strep infection is confirmed when serial titers of ASO rise over a period of weeks, then fall slowly. ASO titers peak during the third week after the onset of acute symptoms of a streptococcal disease; at six months after onset, approximately 30% of patients exhibit abnormal titers.

Antideoxyribonuclease-B titer (anti-DNase B, or ADB)

Anti-DNase-B, or ADB, also detects antigens produced by group A strep, and is elevated in most patients with rheumatic fever and poststreptococcal glomerulonephritis. This test is often done concurrently with the ASO titer, and subsequent testing is usually performed to detect differences in the acute and convalescent blood samples. When ASO and ADB are performed concurrently, 95% of previous strep infections are detected. If both are repeatedly negative, the likelihood is that the patient's symptoms are not caused by a poststreptococcal disease.

When evaluating patients with acute rheumatic fever, the American Heart Association recommends the ASO titer rather than ADB. Even though the ADB is more sensitive than ASO, its results are too variable. It also should be noted that, while ASO is the recommended test, when ASO and ADB are done together, the combination is better than either ASO or ADB alone.

Streptozyme

The streptozyme test is often used as a screening test for antibodies to the streptococcal antigens NADase, DNase, streptokinase, streptolysin O, and hyaluronidase. This test is most useful in evaluating suspected poststreptococcal disease following *Streptococcus pyogenes* infection, such as rheumatic fever.

Streptozyme has certain advantages over ASO and ADB. It can detect several antibodies in a single assay, it is technically quick and easy, and it is unaffected by factors that can produce false-positives in the ASO test. The disadvantages are that, while it detects different antibodies, it does not determine which one has been detected, and it is not as sensitive in children as in adults. In fact, borderline antibody elevations, which could be significant in children, may not be detected at all. As with the ASO and ADB, a serially rising titer is more significant than a single determination.

Preparation

These tests are performed on blood specimens drawn from the patient's vein. The patient does not need to fast before these tests.

KEY TERMS

Antibody—A protein manufactured by a type of white blood cells called lymphocytes, in response to the presence of an antigen, or foreign protein, in the body. Because bacteria, viruses, and other organisms commonly contain many antigens, antibodies are formed against these foreign proteins to neutralize or destroy the invaders.

Antigen—A substance that can trigger a defensive response in the body, resulting in production of an antibody as part of the body's defense against infection and disease. Many antigens are foreign proteins not found naturally in the body, and include bacteria, viruses, toxins, and tissues from another person used in organ transplantation.

Glomerulonephritis—An inflammation of the glomeruli, the filtering units of the kidney. Damage to these structures hampers removal of waste products, salt, and water from the bloodstream, which may

cause serious complications. This disorder can be mild and cause no symptoms, or so severe enough to cause kidney failure.

Rheumatic fever—A disease that causes inflammation in various body tissues. Rare in most developed countries, but reported to be on the increase again in parts of the United States. Joint inflammation occurs, but more serious is the frequency with which the disease permanently damages the heart. The nervous system may also be affected, causing Sydenham's chorea.

Sydenham's chorea—A childhood disorder of the central nervous system. Once called St. Vitus' dance, the condition is characterized by involuntary, jerky movements that usually follow an attack of rheumatic fever. Rare in the United States today, but a common disorder in developing countries. Usually resolves in two to three months with no long-term adverse effects.

Risks

The risks associated with these tests are minimal but may include slight bleeding from the blood-drawing site, **fainting** or feeling lightheaded after the blood is drawn, or blood accumulating under the puncture site (hematoma).

Normal results

Antistreptolysin O titer:

- adult: 160 Todd units/mL
- child: 6 months to 2 years: 50 Todd units/mL; 2 to 4 years: 160 Todd units/mL; 5 to 12 years: 170–330 Todd units/mL
- newborn: similar to the mother's value

Antideoxyribonuclease-B titer:

- adult: 85 units
- child (preschool): 60 units
- child (school age): 170 units

Streptozyme: less than 100 streptozyme units.

Abnormal results

Antistreptolysin O titer: Increased levels are seen after the second week of an untreated infection in acute streptococcal infection, and are increased with acute rheumatic fever, acute glomerulonephritis (66%

of patients will not have high ASO titers), and scarlet fever.

Antideoxyribonuclease-B titer: Increased titers are seen in cases of acute rheumatic fever and post-streptococcal glomerulonephritis.

Streptozyme: As this is a screening test for antibodies to streptococcal antigens, increased levels require more definitive tests to confirm diagnosis.

Resources

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Streptococcal gangrene see **Gangrene**

Streptococcal infections

Definition

Streptococcal (strep) infections are communicable diseases that develop when bacteria of the genus *Streptococcus* invade parts of the body and overwhelm the



The scarlet fever rash on this person's arm was caused by a streptococcal infection. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

body's immune system. Not every streptococcal infection causes detectable symptoms.

Description

Streptococcal bacteria produce symptoms that vary widely in location and severity—everything from skin infections to sore throats and **scarlet fever** to rare, but frequently fatal, necrotizing fasciitis and streptococcal toxic **shock**. Many people have some form of streptococcus bacteria in their body at some point in their life without necessarily showing any symptoms of infection. Nevertheless, a person who hosts bacteria without showing signs of infection is a carrier and can pass the infection on to others.

Types of infection

Primary strep infections invade healthy tissue. **Strep throat**, more formally called streptococcal pharyngitis, is the most common type of primary strep infection. It accounts for between 5–10% of all sore throats, and is especially common among school-aged children. Secondary strep infections invade tissue already weakened by injury or illness. Secondary strep infections frequently affect the bones, ears, eyes, joints, or intestines. Both primary and secondary strep infections can travel from affected tissues to lymph glands or enter the bloodstream and spread throughout the body (systemic infection). Many different strains of streptococcal bacteria have been identified since the 1930s. Types A, B, C, D, and G are the strains most likely to make people ill.

GROUP A STREPTOCOCCUS. All Group A strep (GAS) is the form of streptococcal bacteria most likely to be associated with serious illness. GAS is found worldwide. The incidence of respiratory strep A

infections (strep throat, for example) is highest in cold climates and among young children. The incidence of GAS skin infections is highest in the tropics.

Two of the most severe invasive GAS infections are necrotizing fasciitis or flesh-eating bacteria disease, which causes the destruction of muscle tissue and fat, and **toxic shock syndrome**, a rapidly progressive disorder that causes shock and damages internal organs. In the mid-2000s, particularly virulent (strong and causing serious illness) strains of GAS bacteria appeared to be increasing. GAS is also the type of strep responsible for strep throat and **scarlet fever**. Strep throat is very common and is usually not serious. If untreated, however, strep throat can lead to **rheumatic fever**, which can permanently damage the heart and other organs.

GROUP B STREPTOCOCCUS. Group B strep (GBS) most often affects pregnant women, infants, the elderly, and chronically ill adults such as those with HIV/AIDS. Streptococcal infection occurs when bacteria contaminate cuts or open sores or otherwise penetrate the body's natural defenses. GBS exists in the reproductive tract of between 5 and 40% of all women. Most of these women are carriers who do not develop symptoms of infection. Nevertheless, they can transmit the bacterium to their newborns during **childbirth**. In the United States in 2006, two to three of every 1,000 live-born babies had a GBS infection. However, the number of babies dying of neonatal GBS infection has been declining steadily since the 1980s, most likely as the result of prevention programs initiated by the United States Centers for Disease Control (CDC).

About 75% of infected infants develop early-onset infection. Sometimes evident within a few hours of birth and always apparent within the first week of life, this condition causes inflammation of the membranes covering the brain and spinal cord (**meningitis**), **pneumonia**, blood infection (**sepsis**) and other problems.

Late-onset GBS develops between the ages of seven days and three months. It often causes meningitis. About half of all cases of this rare condition can be traced to mothers who are GBS carriers. The cause of the others is unknown.

Elderly individuals, especially those with other health problems, are also at higher risk of contracting a serious GBS infection that can spread to the entire body.

GROUP C STREPTOCOCCUS. Group C streptococcus (GCS) is a common source of infection in animals. It rarely causes human illness.

GROUP D STREPTOCOCCUS. Group D streptococcus (GDS) is a common cause of wound infections in hospital patients. GDS is also associated with:

- abnormal growth of tissue in the gastrointestinal tract
- urinary tract infection (UTI)
- uterine infections in women who have just given birth

GROUP G STREPTOCOCCUS. Normally present on the skin, in the mouth and throat, and in the intestines and genital tract, Group G strep (GGS) is most likely to lead to infection in alcoholics and in people who have **cancer**, **diabetes mellitus**, **rheumatoid arthritis**, and other conditions that suppress immune-system activity.

GGS can cause a variety of serious infections, including:

- bacteria in the bloodstream (bacteremia)
- inflammation of the connective tissue structure surrounding a joint (bursitis)
- endocarditis (a condition that affects the lining of the heart chambers and the heart valves)
- meningitis
- inflammation of bone and bone marrow (osteomyelitis)
- inflammation of the lining of the abdomen (peritonitis)

Causes and symptoms

Streptococcal infection occurs when bacteria contaminate cuts or open sores or otherwise penetrate the body's natural defenses.

GAS

GAS is transmitted by direct contact with saliva, nasal discharge, or open **wounds** of someone who has the infection. Chronic illness, **kidney disease** treated by dialysis, and steroid use increase vulnerability to infection.

About one of five people with GAS infection develops a sore, inflamed throat (strep throat), and pus on the tonsils. The majority of those infected by GAS either have no symptoms or develop enlarged lymph nodes, fever, **headache**, **nausea**, **vomiting**, weakness, and a rapid heartbeat.

Necrotizing fasciitis, also called flesh-eating bacteria disease, can be caused by a virulent strain of GAS. In this rare disease (only 500 cases have been reported since 1883), tissues become gangrenous and rapidly decompose from the interior outward to the skin, resulting in muscle and skin loss. The **death** rate is as high as 75%. Toxic shock syndrome is characterized by severe headache, **sore throat**, fever as high as 105°F (40.6°C), **dehydration**, watery **diarrhea**, and a

sunburn-like rash that spreads from the face to the rest of the body. Symptoms develop suddenly and can be fatal.

GBS

A pregnant woman who has GBS infection can develop infections of the bladder, blood, and urinary tract, and deliver a baby who is infected or stillborn. The risk of transmitting GBS infection during birth is highest in a woman whose labor begins before the 37th week of **pregnancy** or lasts more than 18 hours or who:

- becomes a GBS carrier during the final stages of pregnancy
- has a GBS urinary-tract infection
- has already given birth to a baby infected with GBS
- develops a fever during labor

Among men, and in women who are not pregnant, the average age of infection with GBS is 64. African Americans appear to be significantly more susceptible to infection by this strain of strep than any other racial group. The most common consequences of GBS infection are pneumonia and infections of blood, skin, and soft tissue.

Miscellaneous symptoms

Other symptoms associated with strep infection include:

- anemia
- elevated white blood cell counts
- inflammation of the epiglottis (epiglottitis)
- heart murmur
- high blood pressure
- infection of the heart muscle
- kidney inflammation (nephritis)
- swelling of the face and ankles

Diagnosis

Strep bacteria can be obtained by swabbing the back of the throat, vagina, rectum, or the infected area with a sterile cotton swab. There are two types of tests to determine if a person has a strep infection. A rapid strep test uses material collected on a sterile swab from the throat or other area where strep is suspected. This test can be done in a doctor's office and can determine only the presence of streptococcal bacteria. It is most often used to determine if a person has strep throat. The results of a rapid strep test are available in about 20 minutes. The advantage of this test is the speed with which a diagnosis can be made.

The rapid strep test has a false negative rate of about 25%. In other words, in about one out of every four cases where no strep is detected by the rapid strep test, the patient actually does have a strep infection. Because of this, when a rapid strep test is negative, the doctor often does a culture test.

For a culture, a sample of swabbed material is grown in the laboratory on a medium that allows technicians to determine what kind of bacteria are present. Results take 24–48 hours. The test is very accurate and will show the presence of other kinds of bacteria besides *Streptococci*.

Treatment

Penicillin is often the antibiotic of choice to treat strep infections. Oral penicillin is usually taken for 10 days for infections such as strep throat and longer for systemic infections. Patients need to take the entire amount of antibiotic prescribed and not discontinue taking the medication when they feel better. Stopping the antibiotic early can lead to a return of the strep infection. Occasionally, a single injection of long-acting penicillin (Bicillin) is given instead of 10 days of oral treatment. It takes less than 24 hours for **antibiotics** to eliminate an infected person's ability to transmit strep bacteria.

About 10% of the time, penicillin is not effective against the strep bacteria. When this happens a doctor may prescribe other antibiotics such as cefuroxime (Ceftin), cefixime (Suprax), cefpodoxime proxetil (Vantin), loracarbef (Lorabid), cefditoren (Spectracef), azithromycin (Zithromax), clindamycin (Cleocin), or a cephalosporin (Keflex, Durocef, Ceclor). Erythromycin (Eryzole, Pedazole, Ilosone), another inexpensive antibiotic, can be given to people who are allergic to penicillin. Scarlet fever is treated with the same antibiotics as strep throat.

Without treatment, the symptoms of untreated strep throat begin subsiding in four or five days. However it is important to treat strep infections promptly with antibiotics because of the possibility of developing secondary disorders or infections. For example, rheumatic fever and rheumatic heart disease may develop from untreated strep throat.

Guidelines developed by the American Academy of Obstetrics and Gynecology (AAOG), the American Academy of Pediatrics (AAP), and the Centers for Disease Control and Prevention (CDC) recommend administering intravenous antibiotics to a woman at high risk of passing GBS infection on to her child, and offering the medication to any pregnant woman who wants it.

Initiating antibiotic therapy at least four hours before birth allows medication to become concentrated enough to protect the baby during its passage through the birth canal.

Newborns infected with GBS during or shortly after birth may die. Those who survive can require lengthy hospital stays and may develop vision or **hearing loss** and other permanent disabilities.

Alternative treatment

Conventional medicine is very successful in treating strep infections. However, several alternative therapies, including homeopathy and botanical medicine, may help relieve symptoms or support the person with a strep infection. For example, several herbs, including garlic (*Allium sativum*), **echinacea** (*Echinacea spp.*), and goldenseal (*Hydrastis canadensis*), are believed to strengthen the immune system, thus helping the body fight a current infection, as well as helping prevent future infections.

Prognosis

Most people who develop strep infections are treated with antibiotics and recover promptly without complications. Strep throat, for example, is almost never fatal. However, GAS is that results in systemic (involving the whole body) infection has a death rate of 25–40%. Streptococcal toxic shock and necrotizing fasciitis also have high death rates. GBS infection can be fatal in newborns and the elderly.

Prevention

Exposure to infected people should be avoided. However, the risk of getting one or passing one on to another person can be minimized by:

- washing hands well and frequently, especially after nose blowing or sneezing and before food handling
- disposing of used tissues properly
- avoiding close contact with someone who has a strep throat
- not sharing food and eating utensils with anyone
- not smoking

Resources

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Streptococcal sore throat see **Strep throat**

Streptococcal toxic shock syndrome see
Toxic shock syndrome

Streptomycin see **Aminoglycosides**

Streptozyme test see **Streptococcal antibody tests**

Stressful life events

- Death of spouse, family member, or close friend
- Divorce
- Marital separation
- Jail term/sentencing of close family member or friend
- Personal injury or illness
- Marriage
- Loss of job due to termination
- Retirement
- Pregnancy
- Change in financial state

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Risk factors

Risk factors for stress-related illnesses are a mix of personal, interpersonal, and social variables. These factors include lack or loss of control over one's physical environment and lack or loss of social support networks. People who are dependent on others (e.g. children or the elderly) or who are socially disadvantaged (because of race, gender, educational level, or similar factors) are at greater risk of developing stress-related illnesses. Other risk factors include feelings of helplessness, hopelessness, extreme fear or anger, and cynicism or distrust of others.

Recent research indicates that some vulnerability to stress is genetic. Scientists at the University of Wisconsin and King's College London discovered that people who inherited a short, or stress-sensitive, version of the serotonin transporter gene were almost three times as likely to experience depression following a stressful event as people with the long version of the gene. Further research is likely to identify other genes that affect susceptibility to stress.

Demographics

Nearly everyone experiences stress in their lives at some time. One study found that about 75% of those surveyed reported experiencing at least some stress in the previous two weeks. Occasional stress is an expected part of life for most people, and while often unpleasant, does not lead to long-term negative outcomes. In some cases however, severe or prolonged stress can lead to illness.

Causes and symptoms

Causes

The causes of stress can include any event or occurrence that a person considers a threat to his or

Stress

Definition

Stress is defined as an organism's total response to environmental demands or pressures. When stress was first studied in the 1950s, the term was used to denote both the causes and the experienced effects of these pressures. More recently, however, the word stressor has been used for the stimulus that provokes a stress response.

Description

Stress in humans results from interactions between persons and their environment that are perceived as straining or exceeding their adaptive capacities and threatening their well being. The element of perception indicates that human stress responses reflect differences in personality as well as differences in physical strength or general health. One recurrent disagreement among researchers concerns the definition of stress in humans. Is it primarily an external response that can be measured by changes in glandular secretions, skin reactions, and other physical functions, or is it an internal interpretation of, or reaction to, a stressor; or is it both?

her coping strategies or resources. Researchers generally agree that a certain degree of stress is a normal part of a living organism's response to the inevitable changes in its physical or social environment, and that positive, as well as negative, events can generate stress. Stress-related disease, however, results from excessive and prolonged demands on an individual's coping resources.

Stress can come from many different situations or events, and can affect different people in different ways. Things that cause stress in one individual may not cause a negative response in another individual. A specific upcoming event can cause significant stress, such as an upcoming job interview, presentation, or final exam. Even upcoming events that are considered positive or exciting can cause stress, such as a wedding or the birth of a baby. When an upcoming event is the source of stress, the individual may experience a constant, nagging worry about the event. He or she may visualize the event repeatedly, and may worry about things that could go wrong. It is often hard to stop thinking about the event. This type of stress can take a significant toll when it persists over long periods of time.

Stress also may be caused by things going on in the world. Events such as the terrorist attacks of September 11th, 2001 are believed to have increased stress in people not only in the United States, but also around the world. Events like terrorist attacks can lead to worry about the safety of family members and loved ones, worries about when another attack may happen, or concerns about many other things. These kind of worries, when they occur over a long period of time, can lead to stress and related negative outcomes.

A traumatic event, such as a car accident, earthquake, **rape**, or even witnessing a traumatic event, can cause severe stress. In these cases an individual may mentally relive the event repeatedly. This is a normal response to a traumatic event but if it persists for more than a short time or does not seem to be getting better, the individual may have **post-traumatic stress disorder** (PTSD), a serious psychiatric illness that occurs in individuals who have participated in or witnessed traumatic events. PTSD is particularly common as among warfighters who are constantly under stress while engaged in military campaigns.

Stress also can be caused, not just by a single event, but also by the presence of issues and problems that are upsetting or frustrating in day-to-day life. These can include worries about money, pressure at work, and problems with a relationship. Events such

as divorce or the **death** of a loved one can also cause stress for many months or years.

Symptoms

The symptoms of stress can be either physical or psychological or both. Physical symptoms may include problems sleeping, **indigestion**, stomach pains, chest pains, **fatigue**, **headache**, back or neck **pain**, and many others. Psychological signs of stress include **anxiety**, frustration, irritability, and even depression. These symptoms may not be problematic if they occur for a short period of time but if they are ongoing an individual should consider seeing a doctor.

Stress-related physical illnesses, such as **irritable bowel syndrome**, heart attacks, arthritis, and chronic headaches, can result from long-term overstimulation of a part of the nervous system that regulates the heart rate, blood pressure, and digestive system. Stress-related emotional illness results from inadequate or inappropriate responses to major changes in one's life situation, such as marriage, completing one's education, becoming a parent, losing a job, or retirement. Psychiatrists sometimes use the term "adjustment disorder" to describe this type of illness. In the workplace, stress-related illness often takes the form of burnout—a loss of interest in or ability to perform one's job due to long-term high stress levels. For example, **palliative care** nurses are at high risk of burnout due to their inability to prevent their patients from dying or even to relieve their physical suffering in some circumstances.

Diagnosis

When the doctor suspects that a patient's illness is connected to stress, he or she will take a careful history that includes stressors in the patient's life (e.g. family or employment problems, other illnesses, recent major life changes). Many physicians will evaluate the patient's personality as well, in order to assess his or her coping resources and emotional response patterns. There are several personality inventories and **psychological tests** that doctors can use to help diagnose the amount of stress that the patient experiences and the coping strategies that he or she uses to deal with them. A variation on this is to identify what the patient perceives as threatening as well as stressful. Stress-related illness can be diagnosed by primary care doctors, as well as by those who specialize in psychiatry. The doctor will need to distinguish between **adjustment disorders** and anxiety or **mood disorders**, and between psychiatric disorders and physical illnesses (e.g., abnormal thyroid activity) that have psychological side effects.

KEY TERMS

Adjustment disorder—A psychiatric disorder marked by inappropriate or inadequate responses to a change in life circumstances. Depression following retirement from work is an example of adjustment disorder.

Biofeedback—A technique in which patients learn to modify certain body functions, such as temperature or pulse rate, with the help of a monitoring machine.

Burnout—An emotional condition, marked by tiredness, loss of interest, or frustration, that interferes with job performance. Burnout is usually regarded as the result of prolonged stress.

Stress hardiness—A personality characteristic that enables persons to stay healthy in stressful circumstances. It includes belief in one's ability to influence the situation; being committed to or fully engaged in one's activities; and having a positive view of change.

Stress management—A category of popularized programs and techniques intended to help people deal more effectively with stress.

Stressor—A stimulus, or event, that provokes a stress response in an organism. Stressors can be categorized as acute or chronic, and as external or internal to the organism.

Treatment

Recent advances in the understanding of the many complex connections between the human mind and body have produced a variety of mainstream approaches to stress-related illness. Present treatment regimens may include one or more of the following:

- Medications. These may include drugs to control blood pressure or other physical symptoms of stress, as well as drugs that affect the patient's mood (tranquilizers or antidepressants).
- Stress management programs. These may be either individual or group treatments, and usually involve analysis of the stressors in the patient's life. They often focus on job or workplace-related stress.
- Behavioral approaches. These strategies include relaxation techniques, breathing exercises, and physical exercise programs including walking.
- Massage. Therapeutic massage relieves stress by relaxing the large groups of muscles in the back, neck, arms, and legs.
- Cognitive therapy. These approaches teach patients to reframe or mentally reinterpret the stressors in their lives in order to modify the body's physical reactions.
- Meditation and associated spiritual or religious practices. Recent studies have found positive correlations between these practices and stress hardiness.

Alternative treatment

Treatment of stress is one area in which the boundaries between traditional and alternative therapies have changed in recent years, in part because some forms of physical exercise (**yoga**, **tai chi**, aikido) that were once

associated with the counterculture have become widely accepted as useful parts of mainstream **stress reduction** programs. Other alternative therapies for stress that are occasionally recommended by mainstream medicine include **aromatherapy**, dance therapy, **biofeedback**, nutrition-based treatments (including dietary guidelines and **nutritional supplements**), **acupuncture**, homeopathy, and herbal medicine.

Prognosis

The prognosis for recovery from a stress-related illness is dependent on a wide variety of factors in a person's life, many of which are genetically determined (e.g. race, sex, illnesses that run in families) or beyond the individual's control (e.g. economic trends, cultural stereotypes and prejudices, death of a loved one). It is possible, however, for an individual to learn new responses to stress and, thus, change their experiences of it. A person's ability to remain healthy in stressful situations is sometimes referred to as stress hardiness. Stress-hardy people have a cluster of personality traits that strengthen their ability to cope. These traits include believing in the importance of what they are doing; believing that they have some power to influence their situation; and viewing life's changes as positive opportunities rather than as threats.

Prevention

Complete prevention of stress is neither possible nor desirable because stress is an important stimulus of human growth and creativity, as well as an inevitable part of life. In addition, specific strategies for stress prevention vary widely from person to person, depending on the nature and number of the stressors

in an individual's life, and the amount of control he or she has over these factors. In general, however, a combination of attitudinal and behavioral changes works well for most patients. An important form of prevention may be parental modeling of healthy attitudes and behaviors within the family.

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ORGANIZATIONS

- The American Institute of Stress, 124 Park Avenue, Yonkers, NY, 10703, (914) 963-1200, (914) 965-6267, Stress125@optonline.net, <http://www.stress.org>.
- National Institute of Mental Health, 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD, 20892-9663, (301) 443-4513, TTY (301) 443-8431, (866) 615-6464, TTY (866) 415-8051, (301) 443-4279, nimhinfo@nimh.gov, <http://www.nih.nih.gov>.
- United States Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (404) 639-3534, (800)-CDC-INFO (800)-232-4636, TTY: (888) 232-6348, inquiry@cdc.gov, <http://www.cdc.gov>.

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Stress reduction

Definition

Stress is the body's normal response to anything that disturbs its natural physical, emotional, or mental balance. Stress reduction refers to various strategies that counteract this response and produce a sense of relaxation and tranquility.

Purpose

Although stress is a natural phenomenon of living, stress that is not controlled and that continues for a long period of time can seriously compromise health.

For this reason, stress must be understood, managed, and appropriately reduced. Several very different strategies and therapies are available that can help with relaxation and stress management.

Precautions

Stress reduction can only present a problem if an individual attributes an actual, serious condition or disease to being simply a stress-related response and avoids consulting a physician.

People who have undergone a severe trauma (criminal assault, combat, natural disaster, car accident, etc.) may experience symptoms of **post-traumatic stress disorder** (PTSD) or **acute stress disorder** (ASD). These disorders are defined by their temporal connection to a traumatic event in the patient's life, and are characterized by a cluster of **anxiety** and dissociative symptoms. They interfere with the patient's normal level of functioning, and require some form of supportive therapy. People who experience a sense of detachment or unreality, emotional numbing, a general feeling of being dazed, **amnesia** for part of the traumatic event, or similar symptoms should consult a medical doctor in addition to using other approaches to stress reduction.

Description

Everyone encounters stress every day. Although most people think of stress as negative, it is neither good nor bad but is neutral or nonspecific. Stress may be internal (from within ourselves) or external (such as noise from the environment) and does not always result from something unpleasant. Stress can also be caused by events that are positive, like an upcoming wedding or the birth of a child. A certain amount of stress is actually essential to being sufficiently stimulated to meet the challenges of everyday life, but when stress is constant and acute, it can have dangerous consequences.

The specific and immediate cause of stress is called the **stressor**. A stressor can be something dramatic or terrible, such as a violent experience or the **death** of a loved one, or it can be a positive and rewarding event, like marriage or a promotion. The stressor can be internal, such as feelings of guilt or anger felt in a relationship, or it can be external, such as a natural disaster or the ordinary rigors and frustrations of commuting. It can also have a physical source, like simple **exercise** or hard work, or it can be strictly mental, like worry. Our bodies react the same way physiologically no matter what the source of the stress.

From a physical standpoint, the body reacts to stress in a standard and predictable manner. When

stress occurs, the brain immediately receives nerve impulses. These impulses initiate an automatic sequence carried out by the body's sympathetic nervous system: it begins with stimulation of the brain's hypothalamus, which sends nerve impulses to both the adrenal and the pituitary glands. Also called the "fight or flight" response, this automatic physiological process is known to have evolved in humans and animals to enable them to cope with sudden, life-threatening emergencies. When faced with a major stressor, the body's biochemistry instantly hurtles into a ready mode that marshals all the possible resources necessary to either escape or do battle. Thus, the adrenal glands located on top of the kidneys provide an instant surge of adrenaline, quickening the heart rate and blood flow and providing every cell with extra oxygen. They also release cortisol or hydrocortisone, causing an increase in both amino acids (the building blocks of proteins) and blood sugar. These are required for tissue repair to take place. Finally, the pituitary gland at the base of the brain releases a variety of hormones, including endorphins, that act as natural painkillers and permit the body to do things it ordinarily could not do. Thus, very shortly after a stressor is recognized by the body, the heart and breathing rate spike, the pupils dilate to improve vision, perspiration increases, digestion slows, and the body is aroused, energized, and temporarily has an increased threshold for **pain**. This sequence of events allows individuals to do whatever is required to save themselves, whether it is to flee from a predator or engage in combat and fend off an attack.

While these automatic physiological responses served early humans well and were essential to survival of the species, today's men and women rarely must literally fight for their lives or dodge and elude a predator. Yet the body's automatic response to stress has remained largely unchanged in a radically changed world. Whether caveman or corporate executive, when the fight-or-flight response kicks in, a three-stage process begins. Stage one is the alarm stage in which the body releases hormones and prepares for extreme physical action. Resistance is stage two in which the body attempts to resist yet adapt to the stress and to repair any damage done. The final stage, exhaustion, occurs if the stress remains constant. It is especially dangerous since stage one's physical response may begin all over again. The persistence of stress and stage three's exhaustion is the point at which disease can occur. The body may then experience severe debilitating conditions like migraine, heart irregularities, and mental illness. Some bodily functions may even shut down altogether.

Although different individuals may have different levels of stress tolerance, chronic stress will eventually

wear down even the strongest of people. Prolonged stress can cause biochemical imbalances that weaken the immune system and invite serious illness. Overall, stress that persists is known to interfere with digestion and, more seriously, alter brain chemistry, create hormonal imbalances, increase heart rate, raise blood pressure, and negatively affect both metabolic and immune function. It is also important to recognize that although stress itself is not a disease, it can worsen any number of already serious physical conditions. While long-term stress can seriously affect one's quality of life and lead to major, sometimes fatal, diseases, prolonged stress also results in the everyday miseries of **headache** and allergy, digestive disorders and **fatigue**, irritable bladder and **impotence, insomnia**, anxiety, depression, and chronic aches and pains. Researchers exploring the connection between stress and susceptibility to colds exposed stressed individuals (who had experienced a death in the family, became divorced, or had recently moved) to cold viruses and then tested for antibodies a month later. Results indicated that severely stressed individuals were four times more likely to become infected.

Stress can cause or contribute to illness but reducing stress may have the opposite effect and may strengthen the body and even encourage healing. The most important step toward reducing the stress may be to understand the nature of stress and to learn how to gain some control over it. Being able to recognize that he or she is stressed is the first step in reducing stress. Of the many signs and symptoms that indicate the presence of stress, some are obvious and require only common sense to recognize. Short-term noticeable effects of stress include sweaty palms and other types of perspiration, dilated pupils, and difficulty in swallowing ("a lump in the throat"). Tightness in the chest is another stress signal as are stomach problems and some skin conditions. Stress that is the result of prolonged anxiety (a sense of apprehension) often results in feelings of panic or actual trembling, fatigue, insomnia, and **shortness of breath**, heart **palpitations** and dizziness, and irritability. Although none of these symptoms is pleasant, they are relatively minor compared to the silent but much more serious internal effects that can lead to serious health problems.

Fortunately, stress and the negative effects it creates can be reduced by a wide variety of therapeutic approaches. When successfully applied, many of these therapies and strategies can both reduce stress and begin reverse its damage. Before selecting a particular therapy, it is important to be able to distinguish unhealthy stress from stress that is not negative. Researchers have found that the most important variable among

types of stress is an individual's sense of control over the situation. The least harmful stress scenario is one in which an individual feels that he or she has a sufficient degree of control or that the situation is highly predictable. Put simply, predictable pain is less stressful because individuals know when to relax (gaining relief from pain as well as protecting themselves from its damaging effects). But when individuals have no warning of pain, they are in a state of constant stress. An example from daily life might be the difference between the stress experienced during a move by parents, who are in control and making decisions, and a child who does not have a say in the matter. The former can pick and choose when to enter or engage a stressful situation or problem but the latter has no control nor any ability to predict when such a situation will arise and are constantly on alert or in a state of anxiety.

For those with little control over situations that make them anxious, there are two ways to deal with the stress. One is to remove or reduce the stressor, and the other is to increase resistance to it. Although there are many strategies to achieve each of these, all of them can be reduced to some variation of a single simple concept—relaxation. While there is no one single technique or therapy that will help everyone, the combination of life-style change, diet, exercise, and relaxation will allow nearly all types of individuals to better manage the stress in their lives. Although relaxation is at the core of most stress reduction methods, it is not something that everyone can fully achieve without assistance and guidance.

There are a number of relaxation therapies that enable an individual to achieve deep, beneficial relaxation. They can be grouped into the following general categories: mind-body therapies, body work and movement therapies, and herbal-based **diets** and natural regimens. Many of the specific techniques in these categories can be part of a self-help or self-care approach, although some require the help of an experienced practitioner.

Therapies that focus on a connection between the mind and body are based on the fact that thinking patterns and emotions can have physical effects on the body. These techniques encourage the individual to take control and learn techniques for coping with stressors rather than trying to eliminate them. Such therapies range from individual counseling and **meditation** or involvement with a support group to **guided imagery** and **biofeedback**. They all have the common goal of evoking the physiological relaxation response in which a person can achieve beneficial internal results such as lowering blood pressure and decreasing gastric acid secretion.

Body work and movement therapies include techniques ranging from dance therapy and massage to **reflexology** and **rolfing**. Body work is based partly on the therapeutic power of human touch and can also include manipulation, realignment, and posture correction. Movement therapies are a particular form of physical exercise, although they attempt to do much more than simply get a person into shape. Most usually emphasize the mind/body connection and strive to put people in better touch with both their bodies and their feelings. Body work and movement therapies can be as vigorous as deep tissue manipulation or as simple and minimal as the Alexander technique's light posture corrections.

Herbal remedies for stress are usually part of a larger system of natural, **holistic medicine**. Whether Chinese traditional medicine, its counterpart from India, or the homeopathy of the West, all these systems of natural medicine have a holistic focus and emphasize the need for inner balance. All demonstrate how the individual's physical, emotional, mental, and spiritual states are connected and use natural substances as part of the treatment for reducing stress. Such therapies range from the occasional purging (cleansing) of **Ayurvedic medicine** to the sleep-inducing properties of chamomile tea. They also can include the use of cayenne to relieve pain, fragrant essential oils from flowers to evoke a pleasing response and relieve tension, or aloe vera to soothe burned skin.

Some of the more common therapies and techniques available for reducing stress include:

- Acupuncture. Insertion of needles at certain spots under the skin for the purpose of attaining balance by either releasing blocked energy or draining off excess energy.
- Alexander technique. Improving the alignment of head, neck, and back to achieve efficient posture and movement.
- Aromatherapy. Massage with essential oils from flowers to affect mood and produce a sense of well-being.
- Art therapy. Creating something allows free expression and results in feelings of achievement and mood change.
- Autogenic training therapy. A form of deep meditation or self-hypnosis.
- Autosuggestion therapy. A form of verbal therapy involving repetition of a positive idea.
- Ayurvedic medicine. A complete system of daily living based on awareness of one's particular constitution.

- Behavioral therapy. A variety of psychotherapies that are based on changing behavior through retraining.
- Bach flower therapy. Herbal remedies that are prepared from flowers acting energetically to soothe the mind and body.
- Bioenergetics. A practice that encourages sudden release of tensions by crying or kicking.
- Biofeedback. Monitoring rates of body functions and using data to influence and gain control over autonomic functions.
- Breathing for relaxation. Stylized breathing technique to control and lower body functions.
- Counseling. Work with a therapist trained in talking-based therapy.
- Dance movement therapy. Freedom of expression through movement.
- Feldenkrais method. Slow, light movements alter habits and reeducate neuromuscular system.
- Flotation therapy. Floating in a soundproof tank with no external stimulation.
- Guided imagery. Creating a mental picture of what is desired. Also called Creative imagery or Visualization.
- Herbal medicine. Uses substances derived from plants as treatment instead of synthetic drugs.
- Homeopathy. Uses minute doses of plant, animal, and mineral substances to stimulate the body's natural healing.
- Hydrotherapy. Use of water internally and externally for healing purposes.
- Hypnotherapy. Hypnosis in order to identify and release patterns that keep an individual from a personal balance point.
- Kinesiology. Uses muscle testing to correct imbalances in the body's "energy system." Also called Touch for Health.
- Massage. Use of touch and manipulation to soothe. Can also employ vigorous deep tissue manipulation.
- Meditation. Deep, relaxed, receptive, and focused concentration on a single object, sound, or word.
- Music therapy. Playing or listening to music to create an emotional reaction.
- Naturopathy. A complete health care system that uses a variety of natural healing therapies.
- Psychotherapy. A talking-based therapy with a mental health professional to get at the root of a conflict, modify behavior or change disruptive thought patterns.
- Reflexology. Manipulation of zones of the feet that relate to the major organs, glands, and areas of the body.
- Rolfing. Vigorous manipulation of the body's connective tissue to restore "balance."
- Shiatsu. Traditional Japanese finger pressure massage therapy.
- Sound therapy. Uses sound waves to slow the body's autonomic system.
- Tai chi chuan. System of slow, continuous exercises based on rhythm and equilibrium.
- Yoga. System of exercises that combines certain positions with deep breathing and meditation.

These and many other techniques, systems, and therapies are available to the person seeking to reduce and manage stress. Some methods are very simple and can be easily learned, while others are high-tech and often involve a practitioner. A search for common elements among most of these stress-reducing systems reveals several strategies that many people may be able to employ on their own. However, it is important to know and recognize the signals of stress. Further, it is easier to resist the negative effects of stress by eating properly and getting sufficient sleep and exercise.

Nearly all stress-reducing systems are geared to evoking some degree of beneficial mind/body relaxation and most include some version of the following:

- mental time out
- deep breathing
- meditation and singular focus
- gentle, repetitive exercise

The best stress reduction system is the one that works for the individual. Whether stress can be relieved by laughter, mellow music, repetition of a single word, self-massage, vigorous activity, or simply by doing everyday chores in a mindful state of heightened awareness, it is important that stress be recognized and managed every day. Studies have shown that regular relaxation eventually makes the body less responsive to its stress hormones and acts as a sort of natural tranquilizer. It is as if people can build their own defense against the stress response.

Many companies have introduced workplace stress management programs to improve their employees' health. These programs typically include instruction on emotional refocusing or restructuring, and have been shown to be beneficial in reducing the participants' blood pressure, heart rate, and other signs of emotional upset. In addition, stress management programs designed for persons in specific high-stress occupations (medicine, law enforcement, emergency response, etc.) have proved to be effective in reducing burnout and helping members of these professions cope with the specific stresses of their respective jobs.

KEY TERMS

Adrenal gland—A pair of glands that rest on the top of each kidney that produce steroids, such as sex hormones and those concerned with metabolic functions.

Amino acid—Organic acids that are the main components of proteins and are synthesized by living cells.

Antibody—A type of protein produced in the blood in response to a foreign substance that destroys the intruding substance; it is responsible for immunity.

Burnout—An emotional condition marked by tiredness, loss of interest, or frustration that interferes with job performance. Burnout is usually regarded as the result of prolonged stress.

Chronic—Long-term or frequently recurring.

Debilitating—Weakening, or reducing the strength of.

Dilate—To enlarge, open wide, or distend.

Endorphins—A group of proteins with powerful pain-killing properties that originate naturally in the brain.

Holistic—That which pertains to the entire person, involving the body, mind, and spirit.

Hydrocortisone—A steroid hormone produced by the adrenal glands that provides resistance to stress.

Hypothalamus—A part of the brain that controls some of the body's automatic regulatory functions.

Immune function—The state in which the body recognizes foreign materials and is able to neutralize them before they can do any harm.

Impotence—The inability of the male to engage in sexual intercourse because of insufficient erection.

Insomnia—Inability to sleep under normal conditions.

Metabolic function—Those processes necessary for the maintenance of a living organism.

Neuromuscular—Relating to nerve and muscle or their interaction.

Physiological—Dealing with the functions and processes of the body.

Pituitary gland—A gland at the base of the brain responsible for growth, maturation, and reproduction.

Sympathetic nervous system—That part of the autonomic nervous system that affects contraction of muscles and blood vessels. Stimulation of this system by a stressor triggers the production of hormones that prepare the body for fight or flight

Therapeutic—Curative or healing.

An additional general strategy for handling stress in family life or the workplace is the cultivation of a group of character traits that has been termed “psychological hardiness.” These traits include believing in the importance of what one is doing, believing that one has some power to influence the immediate situation, and viewing life’s changes as positive opportunities rather than as threats. These qualities are sometimes referred to as the “3 Cs,” which stand for commitment, control and challenge. Approaches to stress reduction that enhance these qualities are especially beneficial.

Newer trends in stress reduction

One trend in stress reduction in the 2000s is the development of stress management programs or stress reduction strategies tailored to specific categories of people, often defined by their occupation or by a chronic health condition. For example, journalists who cover traumatic events are increasingly recognized as susceptible to developing posttraumatic stress disorder. In July of 2007 a study presented at the annual meeting of the

American Society for Clinical Oncology showed men taught stress management techniques before surgery for **prostate cancer** had improved outcomes after both 6 and 12 months.

Another new trend in stress reduction is the development of programs designed for communities as well as individuals. After the events of September 11, 2001, many mental health professionals recognized that acts of terrorism or mass violence affect large groups of people and that psychiatric interventions need to address stress as a group experience as well as an individual one.

Risks

All relaxation-based therapies to reduce stress are generally considered free of serious risk.

Normal results

Learning how to manage stress has the short-term benefits of giving people some sense of control in their lives, providing them with positive coping strategies,

and making them more relaxed and healthier. The long-term benefits can be a stronger immune system, proper hormonal balance, and reduced susceptibility to disease.

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Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

National Child Traumatic Stress Network, University of California, Los Angeles. 11150 W. Olympic Blvd., Suite 650, Los Angeles, CA, 90064, (310) 235-2633, (310) 235-2612, <http://www.nctsnet.org/nccts>.

National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Bethesda, MD, 20892, (301) 443-4513, (301) 443-4279, (866) 615-6464, nimhinfo@nih.gov, <http://www.nimh.nih.gov>.

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Stress test

Definition

A stress test is primarily used to identify **coronary artery disease**. It requires patients to **exercise** on a treadmill or exercise bicycle while their heart rate, blood pressure, electrocardiogram (ECG), and symptoms are monitored.

Purpose

The body requires more oxygen during exercise than when it is at rest. To deliver more oxygen during exercise, the heart has to pump more oxygen-rich blood. Because of the increased stress on the heart, exercise can reveal coronary problems that are not apparent when the body is at rest. This is why the stress test, though not perfect, remains the best initial, noninvasive, practical coronary test.

The stress test is particularly useful for detecting **ischemia** (inadequate supply of blood to the heart muscle) caused by blocked coronary arteries. Less commonly, it is used to determine safe levels of exercise in people with existing coronary artery disease.

Description

A technician affixes electrodes to the patient's chest, using adhesive patches with a special gel that conducts electrical impulses. Typically, electrodes are placed under each collarbone and each bottom rib, and six electrodes are placed across the chest in a rough outline of the heart. Wires from the electrodes are connected to an ECG, which records the electrical activity picked up by the electrodes.

The technician runs resting ECG tests while the patient is lying down, then standing up, and then breathing heavily for half a minute. These baseline tests can later be compared with the ECG tests performed while the patient is exercising. The patient's blood pressure is taken and the blood pressure cuff is left in place so that blood pressure can be measured periodically throughout the test.

KEY TERMS

Angina—Chest pain from a poor blood supply to the heart muscle due to stenosis (narrowing) of the coronary arteries.

Cardiac arrhythmia—An irregular heart rate (frequency of heartbeats) or rhythm (the pattern of heartbeats).

Defibrillator—A device that delivers an electric shock to the heart muscle through the chest wall in order to restore a normal heart rate.

False negative—Test results showing no problem when one exists.

False positive—Test results showing a problem when one does not exist.

Hypertrophy—The overgrowth of muscle.

Ischemia—Diminished supply of oxygen-rich blood to an organ or area of the body.

The patient begins riding a stationary bicycle or walking on a treadmill. Gradually the intensity of the exercise is increased. For example, if the patient is walking on a treadmill, then the speed of the treadmill increases and the treadmill is tilted upward to simulate an incline. If the patient is on an exercise bicycle, then the resistance or “drag” is gradually increased. The patient continues exercising at increasing intensity until reaching the target heart rate (generally set at a minimum of 85% of the maximal predicted heart rate based on the patient’s age) or experiences severe **fatigue**, **dizziness**, or chest **pain**. During the test, the patient’s heart rate, ECG, and blood pressure are monitored.

Sometimes other tests, such as **echocardiography** or thallium scanning, are used in conjunction with the exercise stress test. For instance, studies suggest that women have a high rate of false negatives (results showing no problem when one exists) and false positives (results showing a problem when one does not exist) with the stress test. They may benefit from another test, such as exercise echocardiography. People who are unable to exercise may be injected with drugs, such as adenosine, which mimic the effects of exercise on the heart, and then given a thallium scan. The thallium scan or echocardiogram are particularly useful when the patient’s resting ECG is abnormal. In such cases, interpretation of exercise-induced ECG abnormalities is difficult.

Preparation

Patients are usually instructed not to eat or smoke for several hours before the test. They should be advised to inform the physician about any medications they are taking, and to wear comfortable sneakers and exercise clothing.

Aftercare

After the test, the patient should rest until blood pressure and heart rate return to normal. If all goes well, and there are no signs of distress, the patient may return to his or her normal daily activities.

Risks

There is a very slight risk of myocardial infarction (a **heart attack**) from the exercise, as well as cardiac arrhythmia (irregular heart beats), **angina**, or cardiac arrest (about 1 in 100,000). The exercise stress test carries a very slight risk (1 in 100,000) of causing a heart attack. For this reason, exercise stress tests should be attended by health care professionals with immediate access to defibrillators and other emergency equipment.

Patients are cautioned to stop the test should they develop any of the following symptoms:

- unsteady gait
- confusion
- skin that is grayish or cold and clammy
- dizziness or fainting
- a drop in blood pressure
- angina (chest pain)
- cardiac arrhythmias (irregular heartbeat)

Normal results

A normal result of an exercise stress test shows normal electrocardiogram tracings and heart rate, blood pressure within the normal range, and no angina, unusual dizziness, or **shortness of breath**.

A number of abnormalities may appear on an exercise stress test. Examples of exercise-induced ECG abnormalities are ST segment depression or heart rhythm disturbances. These ECG abnormalities may indicate deprivation of blood to the heart muscle (ischemia) caused by narrowed or blocked coronary arteries. Stress test abnormalities generally require further diagnostic evaluation and therapy.

Patient education

Patients must be well prepared for a stress test. They should not only know the purpose of the test but also signs and symptoms that indicate the test should be stopped. Physicians, nurses, and ECG technicians can ensure patient safety by encouraging them to immediately communicate discomfort at any time during the stress test.

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- National Heart, Lung, and Blood Institute, Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301) 592-8573, <http://www.nhlbi.nih.gov>.

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Stridor

Definition

Stridor is a term used to describe noisy breathing in general and to refer specifically to a high-pitched crowing sound associated with **croup**, respiratory infection, and airway obstruction.

Description

Stridor occurs when erratic air currents attempt to force their way through breathing passages narrowed by:

- illness
- infection
- the presence of foreign objects
- throat abnormalities

Stridor can usually be heard from a distance but is sometimes audible only during deep breathing. Someone who has stridor may crow and wheeze when:

- inhaling
- exhaling
- inhaling and exhaling

Most common in young children, whose naturally small airways are easily obstructed, stridor can be a symptom of a life-threatening respiratory emergency.

Causes and symptoms

During childhood, stridor is usually caused by infection of the cartilage flap (epiglottis) that covers the opening of the windpipe to prevent **choking** during swallowing. It can also be caused by a toy or other tiny object the child has tried to swallow.

Laryngomalacia is a common cause of a rapid, low-pitched form of stridor that may be heard when a baby inhales. This harmless condition does not require medical attention. It usually disappears by the time the child is 18 months old.

The most common causes of stridor in adults are:

- abscess or swelling of the upper airway
- paralysis or malfunction of the vocal cords
- tumor

Other common causes of stridor include:

- enlargement of the thyroid gland (goiter)
- swelling of the voice box (laryngeal edema)
- narrowing of the windpipe (tracheal stenosis)

When stridor is caused by a condition that slowly narrows the airway, crowing and **wheezing** may not develop until the obstruction has become severe.

Diagnosis

When stridor is present in a newborn, pediatricians and neonatologists look for evidence of:

- heart defects inherent at birth (congenital)
- neurological disorders
- general toxicity

If examinations do not reveal the reasons for the baby's noisy breathing, the air passages are assumed to be the cause of the problem.

Listening to an older child or adult breathe usually enables pediatricians, family physicians, and pulmonary specialists to estimate where an airway

obstruction is located. The extent of the obstruction can be calculated by assessing the patient's:

- complexion
- chest movements
- breathing rate
- level of consciousness

X rays and direct examination of the voice box (larynx) and breathing passages indicate the exact location of the obstruction or inflammation. Flow-volume loops and pulse oximetry are diagnostic tools used to measure how much air flows through the breathing passages, and how much oxygen those passages contain.

Pulmonary function tests may also be performed.

Treatment

The cause of this condition determines the way it is treated.

Life-threatening emergencies may require:

- the insertion of a breathing tube through the mouth and nose (tracheal intubation)
- the insertion of a breathing tube directly into the windpipe (tracheostomy)

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Maureen Haggerty

Stroke

Definition

Stroke is a life-threatening condition that occurs when the blood supply to a part of the brain is suddenly cut off or when brain tissue is damaged by bleeding into the brain. There are two main types of stroke. Ischemic stroke occurs when a clot blocks an artery to the brain; this type accounts for about 80% of strokes. The other type, hemorrhagic stroke, occurs when a blood vessel in the brain bursts, allowing blood to spill out into brain

tissue. The blood upsets the chemical balance that the nerve cells in the brain need to function.

Demographics

According to the Centers for Disease Control and Prevention (CDC), stroke is the third leading cause of death in the United States as of 2009, being responsible for about 160,000 deaths each year. About 795,000 Americans have strokes each year, 550,000 for the first time and 245,000 having a second or third stroke. Of these cases, approximately 625,000 are ischemic strokes. The total cost of stroke to the American economy per year as of 2009 is approximately \$68.9 billion. By the year 2025, the annual number of strokes is expected to reach 1 million. As of 2009, more than 4.4 million people in the United States are stroke survivors. Worldwide, the World Health Organization estimates that 15 million people suffer a stroke each year, resulting in 5 million deaths and 5 million permanently disabled survivors.

About 50,000 Americans have a **transient ischemic attack** (TIA) in an average year; of this group, 35% will have a severe stroke at some point in the future.

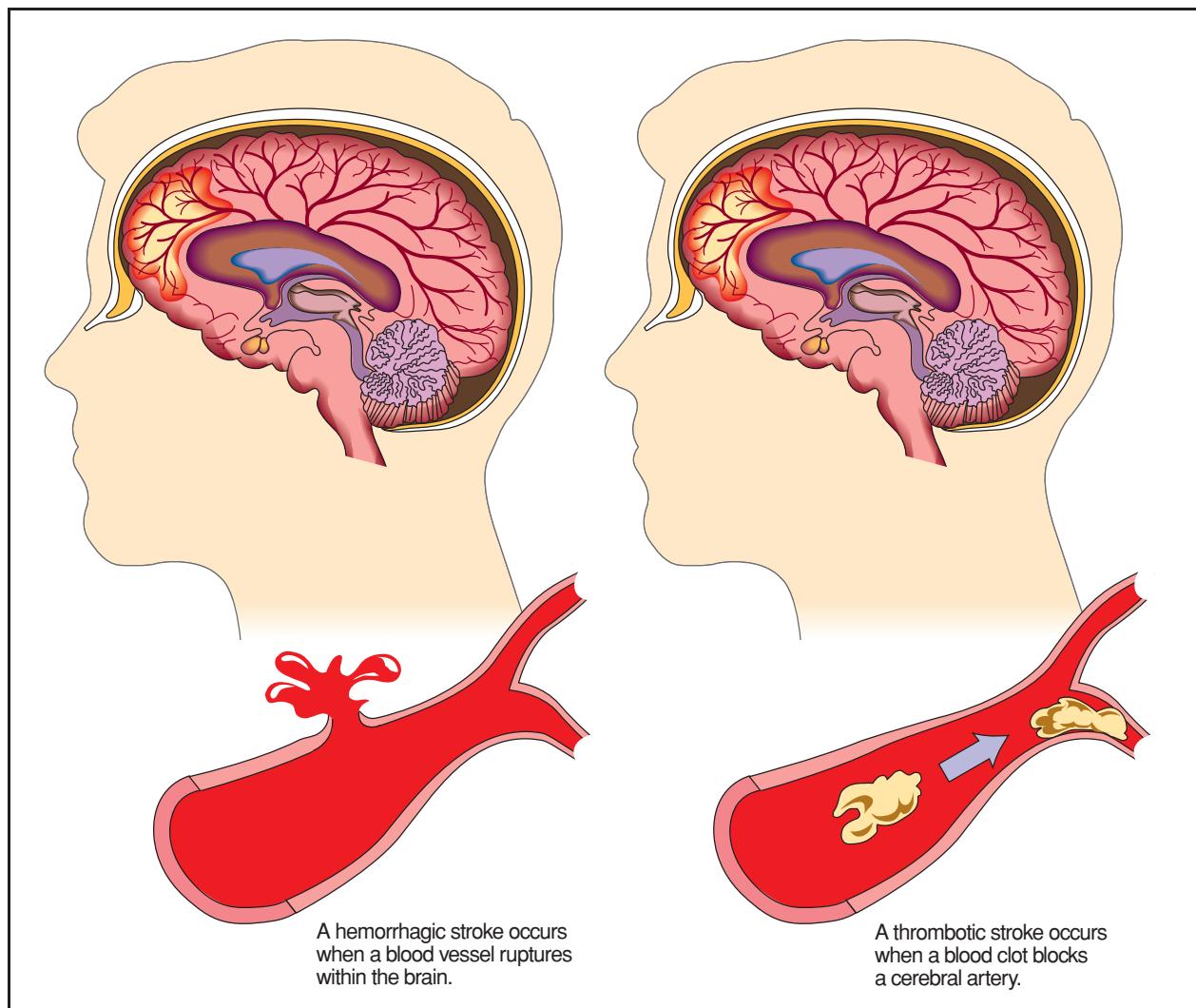
Strokes can affect people in any age group; however, the risk increases sharply in people over 55 years of age. 75% percent of all strokes in Canada and the United States occur in people over 64. Men are 1.25 times more likely to have strokes than women; however, women are more likely to die of stroke because they are usually older when they have their first stroke.

Strokes in children are rare—about six cases per 100,000 children per year in North America. About a third of these cases are in newborns.

African Americans have an increased risk of stroke compared to other racial and ethnic groups in the United States and they are also more likely to suffer a stroke at younger ages. Hispanics are at lesser risk of stroke than African Americans but they also tend to have strokes at relatively young ages. African Americans between the ages of 45 and 55 die from stroke 4–5 times more often than Caucasians in the same age group.

Description

Stroke is usually a sudden occurrence. A stroke occurs when blood flow is interrupted to part of the brain. Without blood to supply oxygen and nutrients and to remove waste products, brain cells quickly begin to die. Depending on the region of the brain affected, a stroke may cause **paralysis**, speech impairment, loss of memory and reasoning ability, **coma**, or death. A stroke also is sometimes called a brain attack or a cerebrovascular accident (CVA).



A hemorrhagic stroke (left) compared to a thrombotic stroke (right). (Illustration by Hans & Cassady, Inc. Reproduced by permission of Gale, a part of Cengage Learning.)

Some people have a warning event called a transient ischemic attack (TIA) or mini-stroke. A TIA has the same symptoms as a full-blown stroke but goes away in a few minutes or hours, leaving no permanent effects. It is, however, an indication that the person is at risk of a major stroke and should see their doctor right away. A TIA offers the person an opportunity to take preventive action.

Stroke is a medical emergency requiring immediate treatment. Prompt treatment improves the chances of survival and increases the degree of recovery that may be expected. A person who may have suffered a stroke should be seen in a hospital emergency room without delay. Treatment to break up a blood clot, the major cause of stroke, must begin within three hours of the stroke to be effective. Improved medical

treatment of all types of stroke has resulted in a dramatic decline in death rates in recent decades. In 1950, nine in ten stroke patients died, compared to slightly less than one in three in the twenty-first century. However, about two-thirds of stroke survivors will have disabilities ranging from moderate to severe.

Risk factors

Risk factors for stroke in adults include:

- Hypertension (high blood pressure). This is the most important single risk factor for stroke.
- High blood cholesterol levels.
- Age over 55.
- A family history of stroke, TIA, or heart attack.

- Diabetes.
- Smoking. Smoking doubles a person's risk of ischemic stroke.
- Personal history of previous stroke or TIA.
- Obesity.
- Heavy use of cocaine.
- Irregular heart rhythm.
- Heavy drinking. Alcohol consumption raises a person's blood pressure.
- Use of birth control pills or hormone replacement therapy.

Risk factors for stroke in children include:

- Congenital (present at birth) malformations of blood vessels and other structures in the brain.
- Infections of the brain like encephalitis and meningitis.
- Head trauma.
- Blood disorders, particularly sickle cell disease.

Causes and symptoms

Causes

Stroke is caused by a loss of blood supply to the brain resulting either from a clot blocking an artery or from bleeding into or around the brain. Ischemic stroke can result from two types of clots. The first is an embolus, which is a free-floating clot produced in the heart or somewhere else in the body that travels to a blood vessel in the brain. The second type of clot is called a thrombus. It is formed within an artery in the head or neck and grows there until it is large enough to block the artery. **Atherosclerosis**, a disease of the blood vessels in which fatty deposits build up along the walls of the vessels, is a common cause of this type of clot.

ISCHEMIC STROKE. A cerebral **embolism** occurs when a blood clot from elsewhere in the circulatory system breaks free. If it becomes lodged in an artery supplying the brain, either in the brain or in the neck, it can cause a stroke. The most common cause of cerebral embolism is atrial fibrillation, a disorder of the heart beat. In atrial fibrillation, the upper chambers (atria) of the heart beat weakly and rapidly instead of slowly and steadily. Blood within the atria is not completely emptied. This stagnant blood may form clots within the atria, which can then break off and enter the circulation. Atrial fibrillation is a factor in about 15% of all strokes. The risk of a stroke from atrial fibrillation can be dramatically reduced with daily use of anticoagulant medication.

Cerebral thrombosis occurs when a blood clot, or thrombus, forms within the brain itself, blocking the flow of blood through the affected vessel. Clots most

often form due to "hardening" (atherosclerosis) of brain arteries. Cerebral thrombosis occurs most often at night or early in the morning. Cerebral thrombosis is often preceded by a transient ischemic attack, or TIA, sometimes called a "mini-stroke." In a TIA, blood flow is temporarily interrupted, causing short-lived stroke-like symptoms. Recognizing the occurrence of a TIA, and seeking immediate treatment, is an important step in stroke prevention.

HEMORRHAGIC STROKE. Hemorrhagic stroke can occur when an aneurysm—a weak spot in the wall of an artery—suddenly bursts. High blood pressure is the most common cause of this type of hemorrhagic stroke. Hemorrhagic stroke can also occur when the walls of an artery become thin and brittle; they can then break and leak blood into the brain. Hemorrhagic stroke can take one of two forms: the blood can leak directly into brain tissue from an artery in the brain, or it can leak from an artery near the surface of the brain into the space between the skull and the membranes covering the brain.

The vessels most likely to break are those with preexisting defects such as an aneurysm. An aneurysm is a bulge or pouch in a blood vessel caused by weakening of the arterial wall. Brain aneurysms are surprisingly common; according to **autopsy** studies, about 6% of all Americans have them. Aneurysms rarely cause symptoms until they burst, however. Aneurysms are most likely to burst when blood pressure is highest, and controlling blood pressure is an important preventive strategy.

Intracerebral hemorrhage affects vessels within the brain itself, while **subarachnoid hemorrhage** affects arteries at the brain's surface, just below the protective arachnoid membrane. Intracerebral hemorrhages represent about 10% of all strokes, while subarachnoid hemorrhages account for about 7%.

In addition to depriving affected tissues of blood supply, the accumulation of fluid within the inflexible skull creates excess pressure on brain tissue, which can quickly lead to death. Nonetheless, recovery may be more complete for a person who survives hemorrhage than for one who survives a clot because the effects of blood deprivation usually are not as severe.

The death of brain cells triggers a chain reaction in which toxic chemicals created by cell death affect other nearby cells. This is one reason why prompt treatment can have such a dramatic effect on final recovery.

Symptoms

Stroke has five major signs or symptoms. The American Stroke Association has a quick symptom checklist called "Give Me 5" that can be used by a friend,

KEY TERMS

Aneurysm—A pouchlike bulging of a blood vessel. Aneurysms can rupture, leading to stroke.

Atrial fibrillation—A disorder of the heart beat associated with a higher risk of stroke. In this disorder, the upper chambers (atria) of the heart do not completely empty when the heart beats, which can allow blood clots to form.

Cerebral embolism—A blockage of blood flow through a vessel in the brain by a blood clot that formed elsewhere in the body and traveled to the brain.

Cerebral thrombosis—A blockage of blood flow through a vessel in the brain by a blood clot that formed in the brain itself.

Comorbid—Referring to the presence of one or more diseases or disorders in addition to the patient's primary disorder.

Deficit—In medicine, the loss or impairment of a function or ability.

Dysphagia—The medical term for difficulty in swallowing.

Embolus—The medical term for a clot that forms in the heart and travels through the circulatory system to another part of the body.

Intracerebral hemorrhage—A cause of some strokes in which vessels within the brain begin bleeding.

Ischemia—Loss of blood supply to a tissue or organ resulting from the blockage of a blood vessel.

Platelets—Small irregularly shaped blood cells involved in the formation of blood clots.

Statins—A group of medications given to lower blood cholesterol levels that work by inhibiting an enzyme involved in cholesterol formation. Statins are also known as HMG-CoA reductase inhibitors.

Subarachnoid hemorrhage—A cause of some strokes in which arteries on the surface of the brain begin bleeding.

Thrombus—A blood clot that forms inside an intact blood vessel and remains there.

Tissue plasminogen activator (tPA)—A substance that is sometimes given to patients within three hours of a stroke to dissolve blood clots within the brain.

Transient ischemic attack (TIA)—A brief stroke lasting from a few minutes to 24 hours. TIAs are sometimes called mini-strokes.

relative, coworker, or caregiver as well as by a person who thinks they may be having a stroke:

- Walk: Is the person having trouble with balance or coordination?
- Talk: Is speech difficult or slurred? Is the person's face drooping?
- Reach: Is one side of the body weak or numb?
- See: Is vision partly or entirely lost?
- Feel: Does the person have a sudden severe headache with no obvious cause?

Other symptoms of stroke that some patients experience include drooling, uncontrollable eye movements, personality or mood changes, drowsiness, loss of memory, or loss of consciousness.

A person with stroke can have more than one of these symptoms at the same time. The important feature to keep in mind is that the symptoms of an embolic ischemic stroke come on suddenly, which helps in distinguishing stroke from other causes of **dizziness**, vision problems, or **headache**. The symptoms of a thrombotic stroke come on more gradually.

A child having a stroke may lose bladder control, have a seizure, or have **nausea and vomiting** as well as the symptoms associated with stroke in adults.

Diagnosis

The diagnosis of stroke includes taking the patient's history and obtaining an account of the patient's present symptoms. In younger patients, the doctor will ask about recent drug use, head trauma, use of **oral contraceptives**, or bleeding disorders. In middle-aged and older patients, the doctor will ask about such risk factors as **hypertension**, **diabetes mellitus**, tobacco use, high cholesterol, and a history of **coronary artery disease**, coronary artery bypass surgery, or atrial fibrillation.

An important part of the history-taking is finding out when the symptoms began and when the patient was last seen normal. Information from family, bystanders, or emergency personnel is often critical to prompt diagnosis and treatment, particularly when tissue plasminogen activator (tPA) therapy is an option. If the patient has awakened with the symptoms of stroke, then the

time of onset is defined as the time the patient was last seen without symptoms.

Examination

The next step is a complete physical and neurological examination to rule out the possibility that the patient's symptoms are being caused by a **brain tumor**. The examination has several purposes: checking the patient's airway, breathing, and circulation; identifying any neurological deficits; identifying the potential cause(s) of the stroke; and identifying any comorbid conditions the patient may have. The neurologist may use the National Institutes of Health Stroke Scale (NIHSS), which is a checklist that allows the doctor to record the patient's level of consciousness; visual function; ability to move; ability to feel sensations; ability to move the facial muscles; and ability to talk.

Tests

Other tests used to diagnose stroke include:

- Blood tests. These can reveal the existence of blood disorders that increase a person's risk of stroke.
- Computed tomography (CT) scan. This type of imaging test is one of the first tests given to a patient suspected of having a stroke. It helps the doctor determine the cause of the stroke and the extent of brain injury.
- Magnetic resonance imaging (MRI). This imaging test is useful in pinpointing the location of small or deep brain injuries.
- Electroencephalogram (EEG). This test measures the brain's electrical activity.
- Blood flow tests. These are done to detect the location and size of any blockages in the blood vessels. One type of blood flow test uses ultrasound to produce an image of the arteries in the neck leading into the brain. Another type of blood flow test, called angiography, uses a special dye injected into blood vessels that will show up on an x ray.
- Echocardiography. This type of test uses ultrasound to produce an image of the heart. It can be useful in determining whether an embolus from the heart caused the patient's stroke.

Treatment

Traditional

Treatment of stroke depends on whether it is ischemic or hemorrhagic. Ischemic stroke is treated first with blood thinners, often **aspirin** or another drug known as warfarin. If the patient is seen by a specialized stroke team within 3 hours of the attack, he

or she may be treated with a drug called tissue plasminogen activator or tPA, described more fully in the next section. It is critical, however, to be sure that the patient has an ischemic rather than a hemorrhagic stroke, as blood-thinning drugs can make a hemorrhagic stroke worse.

Ischemic stroke can also be treated by surgery. The two procedures most commonly used are **endarterectomy**, a procedure in which the surgeon removes the fatty deposits caused by atherosclerosis from the inside of one of the main arteries to the brain; and placing a tube made of metallic mesh called a stent inside the artery to prevent recurrent narrowing of the artery.

Hemorrhagic stroke is treated by removing pooled blood from the brain and repairing damaged blood vessels. To prevent another hemorrhagic stroke, the surgeon may use a procedure called aneurysm clipping. In this procedure, the surgeon clamps the weak spot in the artery away from the rest of the blood vessel, which reduces the chances that it will burst and bleed. Endovascular treatment may be used for aneurysms that are difficult to reach surgically. In this procedure, a catheter is guided from a larger artery up into the brain to reach the aneurysm. Small coils of wire are discharged into the aneurysm, which plug it and block off blood flow from the main artery.

Drugs

Emergency treatment of stroke from a blood clot is aimed at dissolving the clot. This "thrombolytic therapy" currently is performed most often with tissue plasminogen activator, or tPA. tPA must be administered within three hours of the stroke event. Therefore, patients who awaken with stroke symptoms are ineligible for tPA therapy, as the time of onset cannot be accurately determined. tPA therapy has been shown to improve recovery and decrease long-term disability in selected patients. tPA therapy carries a 6.4% risk of inducing a cerebral hemorrhage, however, and is not appropriate for patients with bleeding disorders, very high blood pressure, known aneurysms, any evidence of intracranial hemorrhage, or incidence of stroke, head trauma, or intracranial surgery within the past three months. Patients with clot-related (thrombotic or embolic) stroke who are ineligible for tPA treatment may be treated with heparin or other blood thinners, or with aspirin or other anti-clotting agents in some cases.

Emergency treatment of hemorrhagic stroke is aimed at controlling intracranial pressure. Intravenous

urea or mannitol plus hyperventilation is the most common treatment. **Corticosteroids** also may be used. Patients with reversible bleeding disorders, such as those due to anticoagulant treatment, should have these bleeding disorders reversed, if possible.

Rehabilitation

Rehabilitation refers to a comprehensive program designed to regain function as much as possible and compensate for permanent losses. Approximately 10% of stroke survivors recover without any significant disability and able to function independently. Another 10% are so severely affected that they must remain institutionalized for severe disability. The remaining 80% can return home with appropriate therapy, training, support, and care services.

Rehabilitation is coordinated by a team of medical professionals and may include the services of a neurologist, a physician who specializes in rehabilitation medicine (physiatrist), a physical therapist, an occupational therapist, a speech-language pathologist, a nutritionist, a mental health professional, and a social worker. Rehabilitation services may be provided in an acute care hospital, rehabilitation hospital, long-term care facility, outpatient clinic, or at home.

The rehabilitation program is based on the patient's individual deficits and strengths. Strokes on the left side of the brain primarily affect the right half of the body, and vice versa. In addition, in left brain dominant people, who constitute a significant majority of the population left brain strokes usually lead to speech and language deficits, while right brain strokes may affect spatial perception. Patients with right brain strokes also may deny their illness, neglect the affected side of their body, and behave impulsively.

Rehabilitation may be complicated by cognitive losses, including diminished ability to understand and follow directions. Poor results are more likely in patients with significant or prolonged cognitive changes, sensory losses, language deficits, or incontinence.

PREVENTION OF COMPLICATIONS. Rehabilitation begins with prevention of stroke recurrence and other medical complications. The risk of stroke recurrence may be reduced with many of the same measures used to prevent stroke, including quitting **smoking** and controlling blood pressure.

One of the most common medical complications following stroke is deep venous thrombosis, in which a clot forms within a limb immobilized by paralysis. Clots that break free often become lodged in an artery feeding the lungs. This type of **pulmonary embolism** is a common cause of death in the weeks following a

stroke. Resuming activity within a day or two after the stroke is an important preventive measure, along with use of elastic stockings on the lower limbs. Drugs that prevent clotting may be given, including intravenous heparin and oral warfarin.

Weakness and loss of coordination of the swallowing muscles may impair swallowing (dysphagia) and allow food to enter the lower airway. This may lead to aspiration **pneumonia**, another common cause of death shortly after a stroke. Dysphagia may be treated with retraining exercises and temporary use of pureed foods.

Depression occurs in 30–60% of stroke patients. Antidepressants and **psychotherapy** may be used in combination. Other medical complications include urinary tract infections, pressure ulcers, falls, and seizures.

TYPES OF REHABILITATIVE THERAPY. Brain tissue that dies in a stroke cannot regenerate. In some cases, the functions of that tissue may be performed by other brain regions after a training period. In other cases, compensatory actions may be developed to replace lost abilities.

Physical therapy is used to maintain and restore range of motion and strength in affected limbs, and to maximize mobility in walking, wheelchair use, and transferring (from wheelchair to toilet or from standing to sitting, for instance). The physical therapist advises on mobility aids such as wheelchairs, braces, and canes. In the recovery period, a stroke patient may develop muscle spasticity and **contractures**, or abnormal contractions. Contractures may be treated with a combination of stretching and splinting.

Occupational therapy improves such self-care skills as feeding, bathing, and dressing, and helps develop effective compensatory strategies and devices for activities of daily living. A speech-language pathologist focuses on communication and swallowing skills. When dysphagia is a problem, a nutritionist can advise alternative meals that provide adequate **nutrition**.

Mental health professionals may be involved in the treatment of depression or loss of thinking (cognitive) skills. A social worker may help coordinate services and ease the transition out of the hospital back into the home. Both social workers and mental health professionals may help counsel the patient and family during the difficult rehabilitation period. Caring for a person affected with stroke requires learning a new set of skills and adapting to new demands and limitations. Home caregivers may develop **stress**, **anxiety**, and depression. Caring for the caregiver is an important part of the overall stroke treatment program.

Support groups can provide an important source of information, advice, and comfort for stroke patients and for caregivers. Joining a support group can be one of the most important steps in the rehabilitation process.

First aid

It is useful for friends, coworkers, or bystanders to know the basics of **first aid** for stroke victims. If someone appears to be having a stroke, the most important first step is to call for emergency help *at once*. Stroke is a medical emergency; the sooner the person is evaluated and treated, the better their chances of recovery. The drug presently considered most useful in treating stroke must be given within 3–4 hours of the attack to be effective.

Additional measures that can be taken to help the affected person while waiting for the emergency response team:

- If the person stops breathing, give them mouth-to-mouth resuscitation.
- If they are vomiting, tilt their head to one side to prevent them from swallowing the material.
- Do *not* give them anything to eat or drink.

Prognosis

The prognosis of stroke depends on the person's age, the type and location of the stroke, and the amount of time elapsed between diagnosis and treatment. In general, patients with ischemic stroke have a better prognosis than those with hemorrhagic stroke. In one study in the Boston area, 19% of patients with ischemic stroke died within the first 30 days of the attack compared to 35% with hemorrhagic stroke.

Stroke is fatal for about 27% of white males, 52% of black males, 23% of white females, and 40% of black females. Stroke survivors may be left with significant deficits. Emergency treatment and comprehensive rehabilitation can significantly improve both survival and recovery. One recent study found that treating stroke survivors with certain antidepressant medications, even if they were not depressed, could increase their chances of living longer. People who received the treatment were less likely to die from cardiovascular events than those who did not receive **antidepressant drugs**.

About 10% of stroke patients recover enough function to live independently without help; another 50% can remain at home with outside assistance. The remaining 40% require long-term care in a nursing home.

Stroke in children can be devastating. Between 20% and 35% of newborns who survive a stroke will go on to have a second stroke. More than 66% of older children who suffer strokes will have cognitive deficits, seizures, behavioral problems, changes in personality, or physical disabilities. Unlike adult survivors, children who survive strokes may develop **mental retardation, epilepsy, or cerebral palsy**.

Prevention

Many strokes are preventable with proper self-care. People cannot change some risk factors for stroke, such as race, age, sex, or family history, but they can control several other risk factors:

- They can quit smoking, drinking heavily, or using cocaine.
- They can keep their weight at a healthy level.
- They can exercise regularly, eat a healthy diet, and take medications for high blood pressure if they are diagnosed with it.
- They can take steps to lower their risk of diabetes or high blood cholesterol levels.
- They can lower the level of emotional stress in their life or learn to manage stress more effectively.
- They can get regular checkups for abnormal heart rhythms if they have been diagnosed with such problems.
- They can see their doctor at once if they have a TIA.

People with no previous history of stroke may be given certain drugs as preventive measures. These drugs include statins (drugs that lower blood cholesterol levels) and platelet antiaggregants (medications intended to prevent platelets in the blood from forming clumps that may lead to clots).

Damage from stroke may be significantly reduced through emergency treatment. Knowing the symptoms of stroke is as important as knowing those of a **heart attack**. Patients with stroke symptoms should seek emergency treatment without delay, which may mean dialing 911 rather than their family physician.

Treatment of atrial fibrillation may significantly reduce the risk of stroke. Preventive anticoagulant therapy may benefit those with untreated atrial fibrillation. Warfarin (Coumadin) has proven to be more effective than aspirin for patients at higher risk of stroke. Warfarin is, however, complicated to use because it interacts with a large number of other drugs and requires frequent monitoring by the patient's physician. A new drug called ximelagatran (Exanta) with fewer side effects was introduced in Europe but rejected by the U.S. Food and Drug Administration in 2004 because of

indications of liver damage in 5–6% of subjects in clinical trials. The drug was withdrawn from the European market in 2006.

A recent innovation is the use of computer technology to allow stroke experts in one hospital to evaluate and diagnose a patient in another hospital that might not have a specialist available. Called TeleStroke, the network allows a patient to be evaluated for ischemic stroke within the three-hour time limit for the effective use of tPA. More recently, stroke specialists have proposed an updated version of TeleStroke called TeleStroke 2.0, which would be a Web-based system that could be accessed from any desktop or laptop computer, not just those connected to videoconferencing equipment.

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ORGANIZATIONS

American Academy of Neurology (AAN), 1080 Montreal Avenue, Saint Paul, MN, 55116, (651)-695-2717, (800) 879-1960, (651) 695-2791, <http://www.aan.com/>.

American Stroke Association (ASA), 7272 Greenville Avenue, Dallas, TX, 75231, (888) 4-STROKE, (214) 706-5231, strokeassociation@heart.org, <http://www.strokeassociation.org/presenter.jhtml?identifier=1200037>.

Brain Aneurysm Foundation (BAF), 269 Hanover Street, Building 3, Hanover, MA, 02339, (781) 826-5556, (888) 272-4602, office@bafound.org, <http://www.bafound.org/>.

- National Heart, Lung, and Blood Institute (NHLBI), Health Information Center, P.O. Box 30105, Bethesda, MD, 20824-0105, (301)-592-8573, (240)-629-3246, nhlbiinfo@nhlbi.nih.gov, <http://www.nhlbi.nih.gov/>.
- National Institute of Neurological Disorders and Stroke (NINDS), P.O. Box 5801, Bethesda, MD, 20824, (800)-352-9424, (301)-496-5751, <http://www.ninds.nih.gov/index.htm>.
- National Stroke Association (NSA), 9707 E. Easter Lane, Centennial, CO, 80112, (800)-STROKES, (303)-649-1328, info@stroke.org, <http://www.stroke.org/site/PageNavigator/HOME>.

Richard Robinson
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Strongyloidiasis see **Threadworm infection**

Structural integration see **Rolfing**

Stupor see **Coma**

Stuttering

Definition

There is no standard definition of stuttering (sometimes called stammering) but most attempt to define stuttering as blockages, coordination, or fragmentations of the forward flow of speech (fluency). These stoppages, referred to as disfluencies, are often excessive and characterized by specific types of disfluency. These types of disfluencies include repetitions of sounds and syllables, prolongation of sounds, and blockages of airflow. Individuals who stutter are often aware of their stuttering and feel a loss of control when they are disfluent. Both children and adult stutterers expend an excessive amount of physical and mental energy when speaking. Older children and adults who stutter show myriad negative reactive behaviors, feelings, and attitudes. These behaviors, referred to as secondary behaviors, make the disorder more severe and difficult.

Demographics

Stuttering is a relatively low-prevalence disorder. According to the National Institute on Deafness and Other Communication Disorders (NIDCD), about 3 million people in the United States stutter as of 2010. Across all cultures, roughly 1% of people currently has a stuttering disorder. This figure differs from incidence, or number of individuals who have been diagnosed with stuttering at some point in their

lives. Research suggests that roughly 5% of the population has been diagnosed with a stuttering disorder at some point across the life span. This difference suggests that a significant number of individuals who stutter develop through or outgrow the problem.

Stuttering is most common in children between the ages of two and five, the period when they are learning to speak. The majority of these children will eventually stop stuttering. In adults, approximately three times as many men stutter as women. The sex ratio seems to be lower in early childhood, with a similar number of girls and boys stuttering. The ratio of boys to girls appears to get larger as children become older. This phenomenon suggests that males are more likely to continue to stutter than females.

Description

Stuttering is a confusing and often misunderstood developmental speech and language disorder. Before discussing stuttering, it is important to understand the concepts of speech fluency and disfluency. Fluency is generally described as the forward flow of speech. For most speakers, fluent speech is easy and effortless. Fluent speech is free of any interruptions, blockages, or fragmentations. Disfluency is defined as a breakdown or blockage in the forward flow of speech, or fluency. For all speakers, some occurrence of disfluency is normal. For example, people may insert short sounds or words, referred to as interjections or fillers when speaking; examples of such are “um,” “like,” or “uh.” Also, speakers might repeat phrases, revise words or phrases, or sometimes repeat whole words for the purpose of clarification. For young children, disfluency is a part of the normal development of speech and language, especially during the preschool years (between the ages of two and five years).

The occurrence of disfluency is not the same as stuttering, though stuttered speech is characterized by an excessive amount of disfluency. The disfluencies produced by people who stutter will often be similar to those in the speech of individuals who do not stutter; however, certain types of disfluent behavior are likely to appear only in the speech of people who stutter. These disfluencies are sound and syllable repetitions (i.e. ca-ca-ca-cat), sound prolongations (“sssss–salad,” “fffffish”), and complete blockages of airflow. These behaviors, often referred to as stuttering type disfluencies, distinguish stuttered speech from nonstuttered speech.

Unlike speakers who do not stutter, most people who stutter react negatively to their disfluencies. A person may develop a number of physical reactions, including tension of the muscles involved in speech (tongue, jaw, lips, or chest, for example) and tension

in muscles not related to speech (such as shoulders, limbs, and forehead). In addition to these physiological reactions, people who stutter often have negative emotional reactions to the disorder. Among the emotions that people who stutter report are embarrassment, guilt, and frustration.

Finally, many people who stutter develop a number of negative attitudes and beliefs regarding themselves and speaking—because of their stuttering. These may be negative attitudes and beliefs in certain speaking situations, with people with whom they interact, and in their own abilities. These physiological, emotional, and attitudinal (cognitive) reactions to stuttering, described as secondary stuttering behaviors, are often very disruptive to the communication process and the person's life.

Stuttering behaviors can develop and vary throughout the life span. Sometimes, children will experience periods when stuttering appears to go away for a time, only to return in a more severe pattern. Many children, (estimates range between 50 and 80%) will develop normal fluency after periods of stuttering. For those who continue to stutter during late childhood, adolescence, and into adulthood, stuttering can become a chronic problem. Lifelong efforts will be needed to cope successfully with the behavior.

Due to the effect that stuttering has on communication, the person who stutters may experience certain difficulties in various parts of his or her life. These problems might be secondary to factors inside the person (symptoms of stuttering) and outside the person (society's attitudes toward stuttering and other barriers). For example, many people who stutter report difficulties in social settings. Children who stutter often experience teasing and other social penalties. Adolescents and adults also report a variety of social problems. Academic settings may be difficult for children who stutter because of the emphasis most schools place on verbal performance in the classroom.

There is some evidence that people who stutter confront barriers in employment, at least for jobs that require interacting with the public. These barriers may take the form of inability to do certain tasks easily (talking on the phone, for example), limitations in job choices, and discrimination in the hiring and promotion processes.

Risk factors

According to the Stuttering Foundation of America (SFA), the following are known risk factors for stuttering:

- Family history of stuttering.

- Sex. Boys are at greater risk of stuttering than girls.
- Age when stuttering begins. Children who begin to stutter before age three are more likely to outgrow it than those who begin to stutter later.
- Time elapsed since onset of stuttering. Children who have been stuttering longer than six months are less likely to outgrow the condition.

As far as is known as of 2010, race, ethnicity, or native language are not risk factors for stuttering. The rate of stuttering and sex ratio are thought to be similar throughout the world, although some researchers think that rates of stuttering are higher in Africa than elsewhere. What does seem to vary most widely from country to country is cultural attitudes toward stuttering.

Causes and symptoms

Although research has not identified a single cause of stuttering as of 2010, there appear to be several factors that are viewed as being important to the onset and development of stuttering. Therefore, stuttering is often described as multifactorial and having possibly multiple causes. First, there is a genetic predisposition to stutter, as evidenced by studies of families and twins. In 2010, researchers at NIDCD identified three specific genes—one on chromosome 12 and two on chromosome 16—that are linked to stuttering. The genes are known as GNPTAB, GNPTG, and NAGPA.

A second important factor in stuttering the onset of stuttering is the physiological makeup of people who stutter. Research suggests that the brains of people who stutter may function abnormally during speech production. These differences in functioning may lead to breakdowns in speech production and to the development of disfluent speech.

Third, there is some evidence that speech and language development is an important issue in understanding the development of stuttering. Studies have found some evidence that children show stuttering type behaviors may also have other difficulties with speech-language. Additionally, children with speech-language delays often show stuttering-type behaviors. Finally, environmental issues have a significant impact on the development of stuttering behaviors. An environment that is overly stressful or demanding may cause children to have difficulties developing fluent speech. Although the environment, in particular parental behaviors, does not cause stuttering, it is an important factor that might adversely affect a child who is operating at a reduced capacity for developing fluent speech.

There is no evidence that stuttering is secondary to a psychological disturbance. It is reasonable to assume

that stuttering might have some effect on psychological adjustment and a person's ability to cope with speaking situations. People who stutter might experience a lower self-esteem, and some might report feeling depressed. These feelings and difficulties with coping are most likely the result and not the cause of stuttering. In addition, several research studies have reported that many people who stutter report high levels of **anxiety** and **stress** when they are talking and stuttering. These feelings, psychological states, and difficulties with coping are most likely the result and not the cause of stuttering.

Generally, children begin to stutter between the ages of two and five years. This type of stuttering is categorized as developmental stuttering. Another major category of stuttering is called neurogenic. Neurogenic stuttering results from brain damage, such as a **stroke**, a traumatic injury to the brain, or a degenerative neurological disease. In other cases, stuttering may be secondary to a psychological conversion disorder due to a psychologically traumatic event. When stuttering has abrupt onset secondary to a psychological trauma, it is described as psychogenic stuttering. As of 2010, psychogenic stuttering is considered the least common type.

As stated earlier, the primary symptoms of stuttering include excessive disfluency, both stuttering and normal types (core behaviors), as well as physical, emotional, and cognitive reactions to the problem. These behaviors vary in severity across people who stutter from very mild to very severe. Additionally, behaviors will vary considerably across different speaking situations. There are specific situations when people tend to experience more stuttering (such as talking on the phone or with an authority figure) or less stuttering (speaking with a pet or to themselves, for example). It is likely that this variability might even extend to people having periods (days and even weeks) when they can maintain normally fluent or nonstuttered speech.

Other symptoms that may be associated with stuttering in children include eye blinking, tremor in the lips or jaw, and tension or movement of the face or upper body.

Diagnosis

Examination

Speech-language pathologists (SLPs) are responsible for making the diagnosis and managing the treatment of adults and children who stutter. Preferably, a board-certified speech-language pathologist should be sought for direct intervention or consulting. Diagnosis of stuttering, or identifying children at risk for stuttering, is difficult because most children show excessive

KEY TERMS

Developmental—Referring to a speech problem or other disorder that arises during a specific stage in human development.

Disfluency—Any difficulty in fluent speech, including stuttering.

Neurogenic—Referring to a disorder associated with damage to the central nervous system.

Psychogenic—Referring to a disorder associated with mental or emotional conflict. At one time most stuttering was considered psychogenic, but recent research indicates that psychogenic stuttering is the least common form.

disfluencies in their speech. With children, diagnostic procedures include the collection and analysis of speech and disfluent behaviors in a variety of situations. In addition, the child's general speech-language abilities will be evaluated.

Finally, the speech-language pathologist will interview parents and teachers regarding the child's general developmental, speech-language development, and their perceptions of the child's stuttering behaviors. For adults and older children, diagnostic procedures will also include gathering and analyzing speech samples from a variety of settings. In addition, the speech-language pathologist will conduct a lengthy interview with the person about their stuttering and history of their stuttering problem. Finally, the person who stutters might be asked to report his or her attitudes and feelings related to stuttering, either while being interviewed or by completing a series of questionnaires.

Tests

In some cases, the child may be referred to an otolaryngologist (specialist in ear, nose, and throat disorders) or a neurologist to rule out the possibility that abnormalities in the structure of the child's tongue or mouth, or a brain disorder, are related to the stuttering.

Treatment

Traditional

GENERAL CONSIDERATIONS. Experts generally accept the view that conducting interventions with children and families early in childhood (preschool) is the most effective means of total recovery from stuttering. The chances for a person to recover fully from stuttering by obtaining near-normal fluency are

reduced as the person ages. This is the reason that early intervention is critical. For older children and adults for which stuttering has become a chronic disorder, the focus of therapy is on developing positive coping mechanisms for dealing with the problem. This therapy varies in success based on the individual.

TREATMENT OPTIONS FOR YOUNG CHILDREN.

Treatment of young children generally follows one of two basic approaches. These approaches may also be combined into a single treatment program. The first type of approach, often referred to as indirect therapy, focuses on altering the environment to allow the child opportunities to develop fluent speech. With this approach, counseling parents regarding the alteration of behaviors that affect fluency is the focus. For example, parents may be taught to reduce the amount of household stress or in the level of speech-language demands being placed on the child. In addition, parents may be advised to change characteristics of their speech, such as their speech rate and turn-taking style; this is done to help their children develop more fluent speech.

The other basic approach in treatment with young children targets the development of fluent speech. This type of approach, often referred to as direct therapy, teaches children to use skills that will help them improve fluency, and they are sometimes given verbal rewards for producing fluent speech.

TREATMENT OPTIONS FOR OLDER CHILDREN AND ADULTS.

Treatment approaches for older children and adults usually take one of two forms. These approaches target either helping the person to modify his or her stuttering or fluency. Approaches that focus on modifying stuttering usually teach individuals to reduce the severity of their stuttering behaviors by identifying and eliminating all of the secondary or reactive behaviors. Individuals also work to reduce the amount of emotional reaction toward stuttering.

Finally, the speech-language pathologist will help the individual to learn techniques that allow them to stutter in an easier manner. Therapy does not focus on helping the individual to speak fluently, although most individuals will attain higher levels of fluency if this approach is successful. The other groups of approaches focus on assisting adults and children who stutter to speak more fluently. This type of therapy, which focuses less on changing secondary and emotional reactions, helps the person to modify their speech movements in a specific manner that allows for fluent sounding speech. These procedures require the individual to focus on developing new speech patterns. This often requires a significant amount of practice and skill. The successful outcome of these approaches is nonstuttered, fluent sounding speech. Many therapists integrate

stuttering modification and fluency shaping approaches into more complete treatment programs. In addition, psychological counseling may be used to supplement traditional **speech therapy**.

Drugs

There is no drug that has been approved by the Food and Drug Administration (FDA) just for the treatment of stuttering. Although various medications used to treat **epilepsy**, depression, and **anxiety disorders** have been tried as therapy for stuttering, all of these agents have problematic side effects, and none has been particularly successful in treating stuttering. There are, however, isolated case studies of successful treatment of neurogenic stuttering using **antipsychotic drugs**. As of 2010, NIDCD does not recommend the use of drugs to treat stuttering.

Another method of therapy for stuttering is the use of electronic devices to improve fluency. One type of device fits into the ear like a hearing aid and digitally replays the child's voice so that it sounds as if the child is speaking in unison with someone else. Another type of device, called delayed auditory feedback, forces the child to speak more slowly so that his or her voice will not sound distorted through the machine. NIDCD researchers maintain that the evidence about the success of these devices is mixed as of 2010: "Questions remain about how long such effects may last and whether people are able to easily use these devices in real-world situations."

Prognosis

The prognosis for developmental stuttering is generally good. About 65% of preschool children who stutter outgrow the condition and 74% recover by their early teens. In general, girls have a better prognosis than boys.

According to the American Speech-Language-Hearing Association (ASHA), no single factor can be used to estimate a given child's prognosis for full recovery from stuttering. Complete recovery from severe stuttering is most likely when children and their families receive treatment close to the time of onset. Thus, early identification and treatment of stuttering is critical. For older children and adults, stuttering becomes a chronic problem that requires a lifetime of formal and self-directed therapy. For individuals who show this more chronic form of the disorder, internal motivation for change and support from significant others is going to be an important part of recovery.

Friends, an association for young people who stutter, was founded in 1997 by John Ahlbach, an adult

who stutters, and Lee Caggiano, a speech-language therapist who specializes in stuttering and is the mother of a son who stutters. Friends offers online support to young people who stutter, their parents, and speech-language professionals who work with them. The group also publishes books and a bi-monthly newsletter as well as holding an annual convention.

Prevention

With the exception of the small number of cases in which stuttering is caused by a traumatic brain injury, the condition is not preventable as of 2010.

Resources

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National Institute on Deafness and Other Communication Disorders (NIDCD). "Stuttering." (Accessed September 23, 2010) <http://www.nidcd.nih.gov/health/voice/stutter.html>.

ORGANIZATIONS

American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Rd., Alexandria, VA, 22314-2857, (703) 836-4444, <http://www.entnet.org/>.

American Speech-Language-Hearing Association (ASHA), 2200 Research Blvd., Rockville, MD, 20850-3289, (301) 296-5700, <http://www.asha.org/default.htm>.

Friends: The National Association of Young People Who Stutter, [c/o Lee Caggiano] 38 South Oyster Bay Rd., Syosset, NY, 11791, (866) 866-8335, LCAGGIA-NO@aol.com, <http://www.friendswhostutter.org/>.

International Stuttering Association (ISA), [c/o Joseph Lukong Tardzenyuy, Secretary] PO Box 9598, Douala, Cameroon, admin@stutterisa.org, <http://www.stutterisa.org/index.html>.

National Institute on Deafness and Other Communication Disorders (NIDCD), 31 Center Dr., MSC 2320, Bethesda, MD, 20892-2320, (800) 241-1044, (301) 770-8977, nidcdinfo@nidcd.nih.gov, <http://www.nidcd.nih.gov/index.asp>.

Stuttering Foundation of America (SFA), 3100 Walnut Grove Rd., Suite 603, Memphis, TN, 38111-0749, (901) 452-7343, (800) 992-9392, (901) 452-3931, info@stutteringhelp.org, <http://www.stutteringhelp.org/Default.aspx?tabid=4>.

Rodney Gabel, PhD
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Stye see **Eyelid disorders**

Subacute sclerosing panencephalitis

Definition

Subacute sclerosing panencephalitis is a rare, progressive brain disorder caused by an abnormal immune response to the **measles** virus.

KEY TERMS

Measles encephalitis—A serious complication of measles occurring in about one out of every 1,000 cases, causing headache, drowsiness, and vomiting seven to ten days after the rash appears. Seizures and coma can follow, which may lead to retardation and death.

Description

This fatal condition is a complication of measles, and affects children and young adults before the age of 20. It usually occurs in boys more often than in girls but is extremely rare, appearing in only one out of a million cases of measles.

Causes and symptoms

Experts believe this condition is a form of measles **encephalitis** (swelling of the brain), caused by an improper response by the immune system to the measles virus.

The condition begins with behavioral changes, **memory loss**, irritability, and problems with school work. As the neurological damage increases, the child experiences seizures, involuntary movements, and further neurological deterioration. Eventually, the child starts suffering from progressive **dementia**. The optic nerve begins to shrink and weaken (atrophy) and subsequently the child becomes blind.

Diagnosis

Blood tests and spinal fluid reveal high levels of antibodies to measles virus, and there is a characteristically abnormal electroencephalogram (EEG), or brain wave test. Typically, there is a history of measles infection two to ten years before symptoms begin.

Treatment

There is no standard treatment and a number of **antiviral drugs** have been tested with little success. Treatment of symptoms, including the use of **anticonvulsant drugs**, can be helpful.

Prognosis

While there may be periodic remissions during the course of this disease, it is usually fatal (often from **pneumonia**) within one to three years after onset.

ORGANIZATIONS

National Institute of Allergies and Infectious Diseases, 6610 Rockledge Drive, MSC 6612, Bethesda, MD, 20892-6612, (301) 496-5717, (301) 402-3573, (866) 284-4107, ocpostoffice@niaid.nih.gov, <http://www.niaid.nih.gov>.
National Organization for Rare Disorders, P.O. Box 8923, New Fairfield, CT, 06812-8923, (800) 999-6673, <http://www.rarediseases.org>.

Carol A. Turkington

Subacute spongiform encephalopathy see
Creutzfeldt-Jakob disease

Subacute thyroiditis see **Thyroiditis**

Subarachnoid hemorrhage

Definition

A subarachnoid hemorrhage is an abnormal and very dangerous condition in which blood collects beneath the arachnoid mater, a membrane that covers the brain. This area, called the subarachnoid space, normally contains cerebrospinal fluid. The accumulation of blood in the subarachnoid space can lead to **stroke**, seizures, and other complications. Additionally, subarachnoid hemorrhages may cause permanent brain damage and a number of harmful biochemical events in the brain. A subarachnoid hemorrhage and the related problems are frequently fatal.

Description

Subarachnoid hemorrhages are classified into two general categories: traumatic and spontaneous. Traumatic refers to brain injury that might be sustained in an accident or a fall. Spontaneous subarachnoid hemorrhages occur with little or no warning and are frequently caused by ruptured aneurysms or blood vessel abnormalities in the brain.

Traumatic brain injury is a critical problem in the United States. According to annual figures compiled by the Brain Injury Association, approximately 373,000 people are hospitalized, more than 56,000 people die, and 99,000 survive with permanent disabilities due to traumatic brain injuries. The leading causes of injury are bicycle, motorcycle, and automobile accidents, with a significant minority due to accidental falls, and sports and recreation mishaps.

Exact statistics are not available on traumatic subarachnoid hemorrhages, but several large clinical studies have found an incidence of 23–39% in relation to

severe **head injury**. Furthermore, subarachnoid hemorrhages have been described in the medical literature as the most common brain injury found during **autopsy** investigations of head trauma.

Spontaneous subarachnoid hemorrhages are often due to an aneurysm (a bulge or sac-like projection from a blood vessel) which bursts. **Arteriovenous malformations** (AVMs), which are abnormal interfaces between arteries and veins, may also rupture and release blood into the subarachnoid space. Both aneurysms and AVMs are associated with weak spots in the walls of blood vessels and account for approximately 60% of all spontaneous subarachnoid hemorrhages. The rest may be attributed to other causes, such as **cancer** or infection, or are of unknown origin.

In industrialized countries, it is estimated that there are 6.5–26.4 cases of spontaneous subarachnoid hemorrhage per 100,000 people annually. Certain factors raise the risk of suffering a hemorrhage. Aneurysms are acquired over a person's lifetime and are rarely a factor in subarachnoid hemorrhage before age 20. Conversely, AVMs are present at birth. In some cases, there may be a genetic predisposition for aneurysms or AVMs. Other factors that have been implicated, but not definitively linked to spontaneous subarachnoid hemorrhages, include **atherosclerosis**, cigarette use, extreme alcohol consumption, and the use of illegal drugs, such as **cocaine**. The exact role of high blood pressure is somewhat unclear, but since it does seem linked to the formation of aneurysms, it may be considered an indirect risk factor.

The immediate danger due to subarachnoid hemorrhage, whether traumatic or spontaneous, is **ischemia**. Ischemia refers to tissue damage caused by restricted or blocked blood flow. The areas of the brain that do not receive adequate blood and oxygen can suffer irreparable injury, leading to permanent brain damage or **death**. An individual who survives the initial hemorrhage is susceptible to a number of complications in the following hours, days, and weeks.

The most common complications are intracranial **hypertension**, vasospasm, and **hydrocephalus**. Intracranial hypertension, or high pressure within the brain, can lead to further bleeding from damaged blood vessels; a complication associated with a 70% fatality rate. Vasospasm, or blood vessel constriction, is a principal cause of secondary ischemia. The blood vessels in the brain constrict in reaction to chemicals released by blood breaking down within the subarachnoid space. As the blood vessels become narrower, blood flow in the brain becomes increasingly restricted. Approximately one third of spontaneous subarachnoid hemorrhages and 30–60% of

traumatic bleeds are followed by vasospasm. Hydrocephalus, an accumulation of fluid in the chambers of the brain (ventricles) due to restricted circulation of cerebrospinal fluid, follows approximately 15% of subarachnoid hemorrhages. Because cerebrospinal fluid cannot drain properly, pressure accumulates on the brain, possibly prompting further ischemic complications.

Causes and symptoms

Whether through trauma or disease, subarachnoid hemorrhages are caused by blood being released by a damaged blood vessel and accumulating in the subarachnoid space. Symptoms associated with traumatic subarachnoid hemorrhage may or may not resemble those associated with spontaneous hemorrhage, as trauma can involve multiple injuries with overlapping symptoms.

Typically, a spontaneous subarachnoid hemorrhage is indicated by a sudden, severe **headache**. **Nausea**, **vomiting**, and **dizziness** frequently accompany the **pain**. Loss of consciousness occurs in about half the cases of spontaneous hemorrhage. A **coma**, usually brief, may occur. A stiff neck, **fever**, and aversion to light may appear following the hemorrhage. Neurologic symptoms may include partial **paralysis**, loss of vision, seizures, and speech difficulties.

Spontaneous subarachnoid hemorrhages may be preceded by warning signs prior to the initial bleed. Sentinel, or warning, headaches may be present in the days or weeks before an aneurysm or AVM ruptures. These headaches can be accompanied by dizziness, nausea, and **vomiting**, and possibly neurologic symptoms. Approximately 50% of AVMs are discovered before they bleed significantly; however, most aneurysms are not diagnosed before they rupture.

Diagnosis

To make a diagnosis, a health-care provider takes a detailed history of the symptoms and does a **physical examination**. The symptoms may mimic other disorders and diagnosis can be complicated, especially if the individual is unconscious. The sudden, severe headache can fuel suspicion of a subarachnoid hemorrhage or similar event, and a computed tomography scan (CT scan) or **magnetic resonance imaging** (MRI) scan is considered essential to a quick diagnosis. The MRI is less sensitive than the CT in detecting acute subarachnoid bleeding, but more sensitive in diagnosing AVM or aneurysm.

A CT scan reveals blood that has escaped into the subarachnoid space. For the best results, the scan should be done within 12 hours of the hemorrhage. If

KEY TERMS

Aneurysm—A weak point in a blood vessel where the pressure of the blood causes the vessel wall to bulge outwards. An aneurysm may also appear as a sac-like projection from the blood vessel wall.

Arachnoid mater—One of three membranes that encase the brain and spinal cord. The arachnoid mater is the middle membrane.

Arteriovenous malformation—An abnormal tangle of arteries and veins in which the arteries feed directly into the veins without a normal intervening capillary bed.

Atherosclerosis—An abnormal condition in which lipids, or fats, form deposits on the inside walls of blood vessels.

Cerebral angiography—A medical test in which an x-ray visible dye is injected into blood vessels to allow them to be imaged on an x ray.

Cerebrospinal fluid—The clear, normally colorless fluid found within the subarachnoid space.

Computerized tomography (CT) scan—Cross-sectional x rays of the body compiled to create a three-dimensional image of the body's internal structures.

Hemorrhage—The escape of blood from blood vessels.

Hydrocephalus—Englargement of the chambers in the brain (ventricles) caused by an accumulation of cerebrospinal fluid.

Intracranial hypertension—Abnormally high pressure within the brain.

Ischemia—A condition in which blood flow is cut off or restricted from a particular area. The tissue becomes starved of oxygen and nutrients, resulting in tissue death.

Ischemic—Referring to ischemia.

Lumbar puncture—A diagnostic procedure in which a needle is inserted into the lower spine to withdraw a small amount of cerebrospinal fluid. This fluid is examined to assess trauma to the brain.

Subarachnoid—Referring to the space underneath the arachnoid mater.

Vasospasm—The constriction or narrowing of blood vessels. In cases of hemorrhage, the constriction is prompted by chemical signals from the escaped blood as it breaks down.

if this is not possible, **lumbar puncture** and examination of the cerebrospinal fluid is advised. Lumbar puncture is also done in cases in which the CT scan doesn't reveal a hemorrhage, but there is a high suspicion that one has occurred. In subarachnoid hemorrhage, cerebrospinal fluid shows red blood cells and/or xanthochromia, a yellowish tinge caused by blood breakdown products. Xanthochromia first appears six to 12 hours after subarachnoid hemorrhage, making it advisable to delay lumbar puncture until at least 12 hours after the onset of symptoms for a more definite diagnosis.

Once a hemorrhage, AVM, or aneurysm has been diagnosed, further tests are done to pinpoint the damage. The CT scan may be useful in giving the general location, but cerebral **angiography** maps out the exact details. This procedure involves injecting a special dye into the blood stream. This dye makes blood vessels visible in x rays of the area.

Treatment

The initial course of treatment focuses on stabilizing the hemorrhage victim. Depending on the individual's condition, this may involve intubation and mechanical ventilation, supplemental oxygen, intravenous fluids,

and close monitoring of vital signs. If the person suffers seizures, an anticonvulsant, such as phenytoin (Dilantin), is administered. Nimodipine, a **calcium** channel blocker, may be given to prevent vasospasm and its complications. Sedatives and medications for pain, nausea, and vomiting are administered as needed.

Once the individual is stabilized, cerebral angiography is done to locate the damaged blood vessel. This information and the individual's condition are considered before attempting surgical treatment. Surgery is necessary to remove the damaged area of the blood vessel and prevent a second hemorrhage. The specific neurosurgical procedures depend on the location and type of blood vessel damage. Typically, clip ligation is the preferred means of treating an aneurysm, and surgical excision, radiosurgery, or endovascular embolization are used to manage an AVM.

Prognosis

Individuals who are conscious and demonstrate few neurologic symptoms when they reach medical help have the best prognosis. However, the overall prospects for subarachnoid hemorrhage patients are generally not good. Of the individuals who suffer an aneurysmal

hemorrhage, approximately 15% do not live long enough to get medical treatment. Another 20–40% will not survive the complications caused by the hemorrhage, and approximately 12% of the survivors will experience permanent neurologic disability. Neurologic disabilities may include partial paralysis, weakened or numbed areas of the body, cognitive or speech difficulties, and vision problems. Individuals whose subarachnoid hemorrhages occur as a result of AVMs have a slightly better prognosis, although the risk of death is approximately 10–15% for each hemorrhage.

Subarachnoid hemorrhage associated with traumatic brain injury has a poor prognosis. In clinical studies, 46–78% of head injury cases involving subarachnoid hemorrhage resulted in severe disability, vegetative survival, or death. Furthermore, it is possible that traumatic subarachnoid hemorrhages are accompanied by additional injuries, which would further diminish survival and recovery rates.

Prevention

Traumatic brain injury is the leading cause of subarachnoid hemorrhages, so it follows that efforts to prevent head injury would prevent these hemorrhages. Since accidents cannot always be prevented, measures to minimize potential damage are always advisable. Use of activity-appropriate protective gear, such as bicycle helmets, motorcycle helmets, and sports head gear, is strongly encouraged and promoted by medical associations, consumer organizations, advocacy groups, and health-care professionals. These same groups also advise using seat belts in automobiles.

Spontaneous subarachnoid hemorrhages are more difficult to prevent. Since there may be a genetic component to aneurysms and AVMs, close relatives to individuals with these conditions may consider being screened to assess their own status. Quitting **smoking** and keeping blood pressure within normal limits may also reduce the risk of suffering a spontaneous subarachnoid hemorrhage.

ORGANIZATIONS

Brain Injury Association of America, 1608 Spring Hill Road, Suite 110, Vienna, VA, 22182, (703) 761-0750, (703) 761-0755, <http://www.biausa.org>.

National Stroke Association, 9707 E Easter Lane Building B, Centennial, CO, 80112, (303) 649-1328, (800) 787-6537, Info@stroke.org, <http://www.stroke.org>.

Julia Barrett

Subdural empyema see **Central nervous system infections**

Subdural hematoma

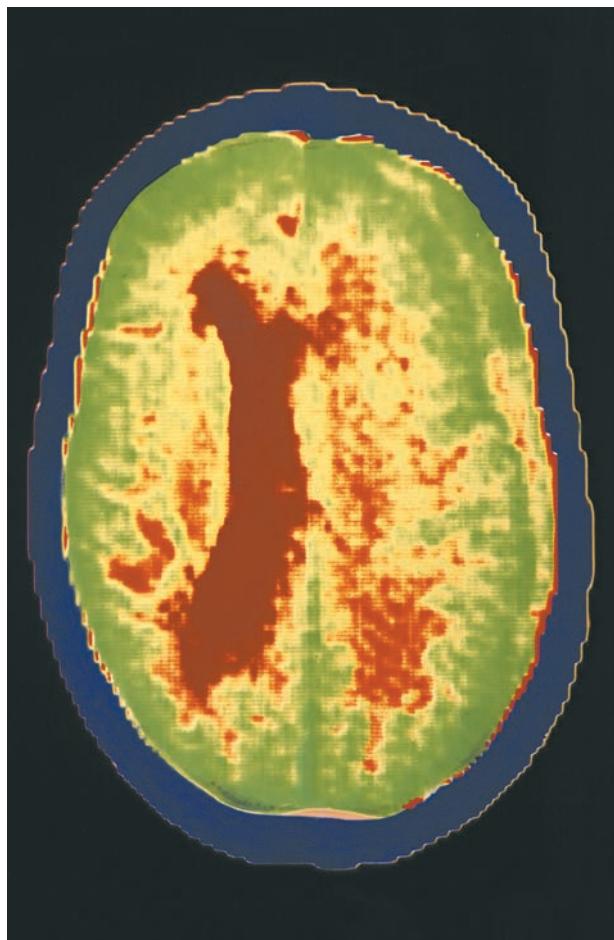
Definition

A subdural hematoma is a collection of blood in the space between the outer layer (dura) and middle layers of the covering of the brain (the meninges). It is most often caused by torn, bleeding veins on the inside of the dura as a result of a blow to the head.

Description

Subdural hematomas most often affect people who are prone to falling. Only a slight hit on the head or even a fall to the ground without hitting the head may be enough to tear veins in the brain, often without fracturing the skull. There may be no external evidence of the bruising on the brain's surface.

Small subdural hematomas may not be very serious, and the blood can be slowly absorbed over several



CT scan indicating subdural hematoma highlighted as a red mass on the center left of the brain. (Photo Researchers, Inc.)



Subdural hematoma present on autopsied body. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

weeks. Larger hematomas, however, can gradually enlarge over several weeks, even though the bleeding has stopped. This enlargement can compress the brain itself, possibly leading to **death** if the blood is not drained.

The time between the injury and the appearance of symptoms can vary from less than 48 hours to several weeks, or more. Symptoms appearing in less than 48 hours are due to an acute subdural hematoma. This type of bleeding is often fatal, and results from tearing of the venous sinus. If more than two weeks have passed before symptoms appear, the condition is called a chronic subdural hematoma, resulting from tearing of the smaller vein. The young and the old are most likely to experience a chronic condition. This chronic form is less risky, as pressure of the veins against the skull lessens the bleeding. Prompt medical care can reduce the probability of permanent brain damage.

Causes and symptoms

A subdural hematoma is caused by an injury to the head that tears blood vessels. In childhood, hematomas are a common complication of falls. A subdural hematoma also may be an indication of **child abuse**, as evidenced by **shaken baby syndrome**.

Symptoms tend to fluctuate, and include:

- headache
- episodes of confusion and drowsiness
- one-sided weakness or paralysis
- lethargy
- enlarged or asymmetric pupils
- convulsions or loss of consciousness after head injury
- coma

KEY TERMS

Corticosteroids—A group of drugs similar to natural corticosteroid hormones produced by the adrenal glands. The drugs have a wide variety of applications, including use for inflammatory disorders and swelling.

Diuretics—A group of drugs that helps remove excess water from the body by increasing the amount lost by urination.

Fontanelle—One of the two soft areas on a baby's scalp; a membrane-covered gap between the bones of the skull.

A doctor should be contacted immediately if symptoms appear. Because these symptoms mimic the signs of a **stroke**, the patient should tell the doctor about any **head injury** within the previous few months.

In an infant, symptoms may include increased pressure within the skull, growing head size, bulging fontanelle (one of two soft spots on a infant's skull), **vomiting**, irritability, lethargy, and seizures. In cases of child abuse, there may be **fractures** of the skull or other bones.

Diagnosis

A chronic subdural hematoma can be difficult to diagnose but a slow loss of consciousness after a head injury is assumed to be a hematoma unless proven otherwise. The hematoma can be confirmed with **magnetic resonance imaging** (MRI), which is the preferred type of scan; a hematoma can be hard to detect on a computed tomography scan (CT scan), depending on how long after the hemorrhage the CT is done.

Treatment

Small hematomas that do not cause symptoms may not need to be treated. Otherwise, the hematoma should be surgically removed. Liquid blood can be drained from burr holes drilled into the skull. The surgeon may have to open a section of skull to remove a large hematoma or to tie off the bleeding vein.

Corticosteroids and **diuretics** can control brain swelling. After surgery, **anticonvulsant drugs** (such as phenytoin) may help control or prevent seizures, which can begin as late as two years after the head injury.

Prognosis

If treatment is provided soon enough, recovery is usually complete. **Headache**, **amnesia**, attention problems, **anxiety**, and giddiness may continue for some time after surgery. Most symptoms in adults usually disappear within six months, with further improvement over several years. Children tend to recover much faster.

Prevention

Because a subdural hematoma usually follows a head injury, preventing head injury can prevent a hematoma.

ORGANIZATIONS

American Academy of Neurology, 1080 Montreal Ave., St. Paul, MN, 55116, (651) 695-2717, (651) 695-2791, (800) 879-1960, memberservices@aan.com, <http://www.aan.com>.

Brain Injury Association of America, 1608 Spring Hill Road, Suite 110, Vienna, VA, 22182, (703) 761-0750, (703) 761-0755, <http://www.biausa.org>.

Head Injury Hotline, P.O. Box 84151, Seattle, WA, 98124, (206) 621-8558, <http://www.headinjury.com>.

Head Trauma Support Project, P.O. Box 215666, Sacramento, CA, 95821, (916) 568-6660.

Carol A. Turkington

Subdural hemorrhage see **Subdural hematoma**

Subluxations see **Dislocations and subluxations**

Substance abuse and dependence

Definition

Substance abuse and dependence refer to any continued pathological use of a medication, non-medically indicated drug (called drugs of abuse), or toxin. They normally are distinguished as follows.

Substance abuse is any pattern of substance use that results in repeated adverse social consequences related to drug-taking—for example, interpersonal conflicts, failure to meet work, family, or school obligations, or legal problems. Substance dependence, commonly known as **addiction**, is characterized by the physiological and behavioral symptoms related to substance use. These symptoms include the need for increasing amounts of the substance to maintain desired effects,

withdrawal if drug-taking ceases, and a great deal of time spent in activities related to substance use.

Substance abuse is more likely to be diagnosed among those who have just begun taking drugs and is often an early symptom of substance dependence. However, substance dependence can appear without substance abuse, and substance abuse can persist for extended periods of time without a transition to substance dependence.

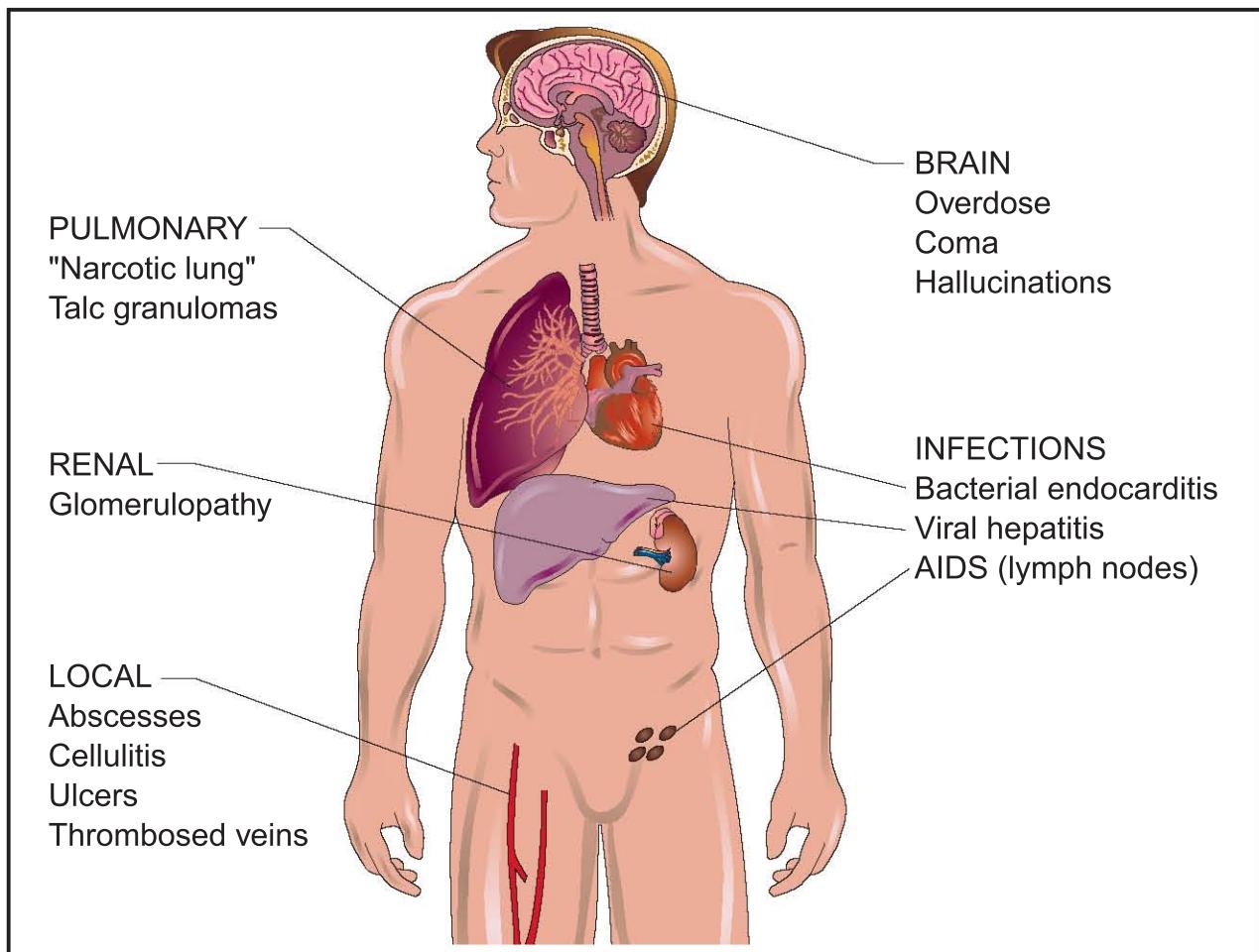
Description

Substance abuse and dependence are disorders that affect all population groups although specific patterns of abuse and dependence vary with age, gender, culture, and socioeconomic status. According to data from the National Longitudinal Alcohol Epidemiologic Survey, 13.3% of a survey group of Americans exhibited symptoms of alcohol dependence during their lifetime, and 4.4% exhibited symptoms of alcohol dependence during the past 12 months. According to the United States Department of Health and Human Services' National Survey on Drug Use and Health, in 2005 9.1% of the population age 12 or older (about 22.2 million people) were classified as having substance abuse or dependence within the last year. About 7.7% (18.7 about million people) were classified as having alcohol abuse or dependence.

Although substance dependence can begin at any age, people aged 18 to 25 have been found to have higher substance abuse and dependency rates than other age groups. Individuals who first used drugs or alcohol at a young age are more likely to have drug abuse and dependence problems later in life than those who first used drugs or alcohol at an older age. Gender proportions vary according to the class of drugs, but substance abuse and dependence is about twice as likely to occur in men than in women.

In addition to being an individual health disorder, substance abuse and dependence may be viewed as a public health problem with far-ranging health, economic, and social implications. Substance-related disorders are associated with teen **pregnancy** and the transmission of **sexually transmitted diseases** (STDs), as well as failure in school, unemployment, domestic violence, homelessness, and crimes such as **rape and sexual assault**, aggravated assault, robbery, burglary, and larceny. Many different estimates have been made for the economic cost of substance abuse and dependence, and most estimate it at tens or hundreds of billions of dollars.

The term substance, when discussed in the context of substance abuse and dependence, refers to



Substance abuse often causes a variety of medical abnormalities and conditions throughout the body, as shown in the illustration above. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

medications, drugs of abuse, and toxins. These substances have an intoxicating effect, desired by the user, which can have either stimulating (speeding up) or depressive/sedating (slowing down) effects on the body. Substance dependence and/or abuse can involve any of the following 10 classes of substances:

- alcohol
- amphetamines (including “crystal meth,” some medications used in the treatment of attention deficit disorder [ADD], and amphetamine-like substances found in appetite suppressants)
- cannabis (including marijuana and hashish)
- cocaine (including “crack”)
- hallucinogens (including LSD, mescaline, and MDMA [“ecstasy”])
- inhalants (including compounds found in gasoline, glue, and paint thinners)

- nicotine (including that found in cigarettes and smokeless tobacco)
- opioids (including morphine, heroin, codeine, methadone, oxycodone [Oxycontin (TM)])
- phencyclidine (including PCP, angel dust, ketamine)
- sedative, hypnotic, and anxiolytic (anti-anxiety) substances (including benzodiazepines such as valium, barbiturates, prescription sleeping medications, and most prescription anti-anxiety medications)

Caffeine has been identified as a substance in this context, but as yet there is insufficient evidence to establish whether caffeine-related symptoms fall under substance abuse and dependence.

Substances of abuse may thus be illicit drugs, readily available substances such as alcohol or glue, over-the-counter drugs, or prescription medications. In many cases, a prescription medication that becomes a substance of abuse may have been a legal, medically

Percentage of population abusing illicit drugs or alcohol, by gender and age

Gender	Illicit drugs	Alcohol
Female	2.2	5.1
Male	3.4	9.7
Age	Illicit drugs	Alcohol
12–17	4.6	4.9
18–25	7.8	17.2
26 or older	1.7	6.0

SOURCE: Substance Abuse and Mental Health Services Administration (SAMHSA), Office of Applied Studies, 2008 *National Survey on Drug Use and Health* (September 2009). Available online at: <http://oas.samhsa.gov/NSDUH/latest.htm> (accessed June 10, 2010).

(Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.)

indicated prescription for the user, but the pattern of use diverges from the use prescribed by the physician.

Causes and symptoms

Causes

The causes of substance dependence are not well established, but three factors are believed to contribute to substance-related disorders: genetic factors, psychopathology, and social learning. In genetic epidemiological studies of **alcoholism**, the probability of identical twins both exhibiting alcohol dependence was significantly greater than with fraternal twins, thus suggesting a genetic component in alcoholism. It is unclear, however, whether the genetic factor is related to alcoholism directly, or whether it is linked to other psychiatric disorders that are known to be associated with substance abuse. For example, there is evidence that alcoholic males from families with **depressive disorders** tend to have more severe courses of substance dependence than alcoholic men from families without such family histories.

These and other findings suggest substance abuse may be a way to relieve the symptoms of a psychological disorder. In this model, unless the underlying pathology is treated, attempts to permanently stop substance dependence are ineffective. Psychopathologies that are associated with substance dependence include antisocial personality disorder, **bipolar disorder**, depression, **anxiety disorder**, and **schizophrenia**.

A third factor related to substance dependence is social environment. In this model, drug-taking is essentially a socially learned behavior. Local social norms

determine the likelihood that a person is exposed to the substance and whether continued use is reinforced. For example, individuals may, by observing family or peer role models, learn that substance use is a normal way to relieve daily stresses. External penalties, such as legal or social sanctions, may reduce the likelihood of substance abuse.

At the level of neurobiology, it is believed that substances of abuse operate through similar pathways in the brain. The chemical changes induced by the stimulation of these pathways by initial use of the substance lead to the desire to continue substance use, and eventually to substance dependence.

Symptoms

The DSM-IV-TR identifies seven criteria (symptoms), at least three of which must be met during a given 12-month period, for the diagnosis of substance dependence:

- Tolerance, as defined either by the need for increasing amounts of the substance to obtain the desired effect or by experiencing less effect with extended use of the same amount of the substance.
- Withdrawal, as exhibited either by experiencing unpleasant mental, physiological, and emotional changes when drug-taking ceases or by using the substance as a way to relieve or prevent withdrawal symptoms.
- Longer duration of taking substance or use in greater quantities than was originally intended.
- Persistent desire or repeated unsuccessful efforts to stop or lessen substance use.
- A relatively large amount of time spent in securing and using the substance, or in recovering from the effects of the substance.
- Important work and social activities reduced because of substance use.
- Continued substance use despite negative physical and psychological effects of use.

Although not explicitly listed in the DSM-IV-TR criteria, “craving,” or the overwhelming desire to use the substance regardless of countervailing forces, is a universally-reported symptom of substance dependence.

Symptoms of substance abuse, as specified by DSM-IV-TR, include one or more of the following occurring during a given 12-month period:

- Substance use resulting in a recurrent failure to fulfill work, school, or home obligations (such as work absences, substance-related school suspensions, or neglect of children).

- Substance use in physically hazardous situations such as driving or operating machinery.
- Substance use resulting in legal problems such as drug-related arrests.
- Continued substance use despite negative social and relationship consequences of use.

In addition to the general symptoms, there are other physical signs and symptoms of substance abuse that are related to specific drug classes:

- Signs and symptoms of alcohol intoxication include such physical signs as slurred speech, lack of coordination, unsteady gait, memory impairment, and stupor, as well as behavior changes shortly after alcohol ingestion, including inappropriate aggressive behavior, mood volatility, and impaired functioning.
- Amphetamine users may exhibit rapid heartbeat, elevated or depressed blood pressure, dilated (enlarged) pupils, weight loss, excessively high energy levels, inability to sleep, confusion, and occasional paranoid psychotic behavior.
- Cannabis users may exhibit red eyes with dilated pupils, increased appetite, dry mouth, and rapid pulse. They may also be sluggish and slow to react.
- Cocaine users may exhibit rapid heart rate, elevated or depressed blood pressure, dilated pupils, and weight loss, in addition to wide variations in their energy level, severe mood disturbances, psychosis, and paranoia.
- Users of hallucinogens may exhibit anxiety or depression, paranoia, and unusual behavior in response to hallucinations (imagined sights, voices, sounds, or smells that appear real). Signs include dilated pupils, rapid heart rate, tremors, lack of coordination, and sweating. Flashbacks, or the re-experiencing of a hallucination long after stopping substance use, are also a symptom of hallucinogen use.
- Users of inhalants experience dizziness, spastic eye movements, lack of coordination, slurred speech, and slowed reflexes. Associated behaviors may include belligerence, tendency toward violence, apathy, and impaired judgment.
- Opioid drug users exhibit slurred speech, drowsiness, impaired memory, and constricted (small) pupils. They may appear slowed in their physical movements.
- Phencyclidine users exhibit spastic eye movements, rapid heartbeat, decreased sensitivity to pain, and lack of muscular coordination. They may show belligerence, predisposition to violence, impulsiveness, and agitation.
- Users of sedative, hypnotic, or anxiolytic drugs show slurred speech, unsteady gait, inattentiveness, and

impaired memory. They may also display inappropriate behavior, mood volatility, and impaired functioning.

Other signs are related to the form in which the substance is used. For example, heroin, certain other opioid drugs, and certain forms of **cocaine** may be injected. A person using an injectable substance may have “track marks” (outwardly visible signs of the site of an injection, with possible redness and swelling of the vein in which the substance was injected). Furthermore, poor judgment brought on by substance use can result in the injections being made under dangerously unhygienic conditions. These unsanitary conditions and the use of shared needles are risk factors for major infections of the heart, as well as infection with HIV (the virus that causes **AIDS**), certain forms of hepatitis (a liver infection), and **tuberculosis**.

Cocaine is often taken as a powdery substance, which is “snorted” through the nose. This can result in frequent nosebleeds, sores in the nose, and even erosion (an eating away) of the nasal septum (the structure that separates the two nostrils).

Overdosing on a substance is a frequent complication of substance abuse. **Drug overdose** can be purposeful (with **suicide** as a goal), or due to carelessness, the unpredictable strength of substances purchased from street dealers, the mixing of more than one type of substance, or as a result of the increasing doses that a person must take to experience a similar level of effect. Substance overdose can be a life-threatening emergency, with the specific symptoms depending on the type of substance used. Substances with depressive effects may dangerously slow the breathing and heart rate, drop the body temperature, and result in general unresponsiveness. Substances with stimulatory effects may dangerously increase the heart rate and blood pressure, produce abnormal heart rhythms, increase body temperature, induce seizures, and cause erratic behavior.

Diagnosis

Tools used in the diagnosis of substance dependence include screening questionnaires and patient histories, **physical examination**, and laboratory tests. A simple and popular screening tool is the CAGE questionnaire. CAGE refers to the first letters of each word that forms the basis of each of the four questions of the screening exam:

- Have you ever tried to Cut down on your substance use?
- Have you ever been Annoyed by people trying to talk to you about your substance use?
- Do you ever feel Guilty about your substance use?

- Do you ever need an Eye opener (use of the substance first thing in the morning) in order to start your day?

A “yes” answer to two or more of these questions is an indication that the individual should be referred for a more thorough work-up for substance dependency or abuse.

In addition to CAGE, other screening questionnaires are available. Some are designed for particular population groups such as pregnant women, and others are designed to more thoroughly assess the severity of substance dependence. These questionnaires, known by their acronyms, include AUDIT, HSS, HSQ, PRIME-MD, ACE, TWEAK, s-MAST, and SADD. There is some variability among questionnaires in terms of how accurately and comprehensively they can identify individuals as substance dependent.

Patient history, as taken through the direct interview, is important for identifying physical symptoms and psychiatric factors related to substance use. Family history of alcohol or other substance dependency is also useful for diagnosis.

A physical examination may reveal signs of substance abuse. These signs are specific to the substances used, and may include needle marks, tracks, or nasal erosion.

With the individual’s permission, substance use can be detected through laboratory testing of his or her blood, urine, or hair. Laboratory testing, however, may be limited by the sensitivity and specificity of the testing method, and by the time elapsed since the person last used the drug.

One of the most difficult aspects of diagnosis involves overcoming the patient’s denial. Denial is a psychological state during which a person is unable to acknowledge the (usually negative) circumstances of a situation. In this case, denial leads a person to underestimate the degree of his or her substance use and of the problems associated with the substance use.

Treatment

According to the American Psychiatric Association, there are three goals for the treatment of people with substance use disorders: (1) the patient abstains from or reduces the use and effects of the substance; (2) the patient reduces the frequency and severity of relapses; and (3) the patient develops the psychological and emotional skills necessary to restore and maintain personal, occupational, and social functioning.

In general, before treatment can begin, many treatment centers require that the patient undergo **detoxification**. Detoxification is the process of weaning the

patient from his or her regular substance use. Detoxification can be accomplished “cold turkey,” by complete and immediate cessation of all substance use, or by slowly decreasing (tapering) the dose the individual is taking, to minimize the side effects of withdrawal. Some substances must be tapered because “cold turkey” methods of detoxification are potentially life threatening. In some cases, medications may be used to combat the physical and psychological symptoms of withdrawal. For example, **methadone** is used to help patients adjust to the tapering off of heroin use.

Treatment itself consists of three parts: (1) assessment; (2) formulation of a treatment plan; (3) psychiatric management. The first step in treatment is a comprehensive medical and psychiatric evaluation of the patient. This evaluation includes:

- a history of the patient’s past and current substance use, and its cognitive, psychological, physiological, and behavioral effects
- a medical and psychiatric history and examination
- a history of psychiatric treatments and outcomes
- a family and social history
- screening of blood, breath, or urine for substances
- other laboratory tests to determine the presence of other conditions commonly found with substance use disorders

After the assessment is made, a treatment plan is formulated. Treatment plans vary according to the needs of the specific patient and can change for the same patient as it is seen how he or she responds to different elements of treatment. Plans typically involve the following elements: (1) a strategy for the psychiatric management of the patient; (2) a strategy for reducing effects or use of substances, or for abstinence; (3) efforts to ensure compliance with the treatment program and to prevent relapse; (4) treatments for other conditions associated with substance use. Initial therapy and treatment setting (hospital, residential treatment, partial hospitalization, or outpatient) decisions are made as part of the treatment plan, but because substance use disorders are considered a chronic condition requiring long-term care, these plans can and do change through the course of treatment.

The third step, psychiatric management of the patient, is the implementation of the treatment plan. Psychiatric management of the patient includes establishing a trusting relationship between clinician and patient; monitoring the patient’s progress; managing the patient’s relapses and withdrawal; diagnosing and treating associated psychiatric disorders; and helping the patient adhere to the treatment plan through

therapy and the development of skills and social interactions that reinforce a drug-free lifestyle.

As part of the treatment process, patients typically undergo psychosocial therapy and, in some cases, pharmacologic treatment. Psychosocial therapeutic modalities include **cognitive-behavioral therapy**, behavioral therapy, individual psychodynamic or interpersonal therapy, **group therapy**, **family therapy**, and self-help groups. Pharmacologic treatment may include medications that ease withdrawal symptoms, reduce cravings, interact negatively with substances of abuse to discourage drug-taking, or treat associated psychiatric disorders.

Alternative treatment

The efficacy of alternative treatments for substance use disorders remains for the most part ambiguous. Some studies suggest that **acupuncture** can be used to help treat cocaine addiction. However, a 2007 meta-analysis (summary analysis of studies) found that there was no reproducible scientific data indicating that acupuncture was helpful. A similar meta-analysis reported that acupuncture also had no statistically significant effect on **smoking** cessation.

There has been movement toward examining some touted treatments in more rigorous clinical trials. In particular, there has been some interest in *Pueraria lobata*, or kudzu, an herb that has reputedly been used in Chinese medicine to treat alcoholism. Preclinical trials of an herbal formula with kudzu have shown that increased consumption of the herbal formula is associated with decreased consumption of alcohol. Toxicity studies show few ill effects of the formula, and human trials are currently being undertaken to more fully evaluate the efficacy of this treatment.

The effectiveness of electroacupuncture (the practice of acupuncture accompanied by the application of low levels of electrical current at acupuncture points) in alleviating opiate withdrawal symptoms is also being examined. Preclinical trials suggest that electroacupuncture treatment given prior to the administration of naxolone seems to alleviate the withdrawal effects of naxolone.

Prognosis

Recovery from substance use is notoriously difficult, even with exceptional treatment resources. Although relapse rates are difficult to accurately obtain, the NIAAA cites evidence that 90% of alcohol dependent users experience at least one relapse within the 4 years after treatment. Relapse rates for heroin and nicotine users are believed to be similar. Certain pharmacological

KEY TERMS

Addiction—The state of being both physically and psychologically dependent on a substance.

Dependence—A state in which a person requires a steady concentration of a particular substance to avoid experiencing withdrawal symptoms.

Detoxification—A process whereby an addict is withdrawn from a substance.

Intoxication—The mental, physical, or emotional state produced by a substance.

Street drug—A substance purchased from a drug dealer; may be a legal substance, sold illicitly (without a prescription, and not for medical use), or it may be a substance that is illegal to possess.

Tolerance—A phenomenon whereby a drug user becomes physically accustomed to a particular dose of a substance, and requires increasing dosages in order to obtain the same effects.

Withdrawal—Those side effects experienced by a person who has become physically dependent on a substance, upon decreasing the substance's dosage or discontinuing its use.

treatments, however, have been shown to reduce relapse rates.

Relapses are most likely to occur within the first 12 months of having discontinued substance use. Triggers for relapses can include any number of life stresses (problems on the job or in the marriage, loss of a relationship, **death** of a loved one, financial stresses), in addition to seemingly mundane exposure to a place, situation, or acquaintance associated with previous substance use.

The development of adaptive life skills and ongoing drug-free social support are believed to be two important factors in avoiding relapse. The effect of the support group Alcoholics Anonymous has been intensively studied, and many studies have found that long-term sobriety appears to be positively related to Alcoholics Anonymous attendance and involvement. Support for family members in addition to support for the individual in recovery is also important. Because substance dependence has a serious impact on family functioning, and because family members may inadvertently maintain behaviors that initially led to the substance dependence, ongoing therapy and support for family members should not be neglected.

Prevention

Prevention is best aimed at teenagers and young adults aged 18–24 who are at very high risk for substance experimentation. Prevention programs should include an education component that outlines the risks and consequences of substance use and a training component that gives advice on how to resist peer pressure to use drugs.

Furthermore, prevention programs should work to identify and target children who are at relatively higher risk for substance abuse. This group includes victims of physical or **sexual abuse**, children of parents who have a history of substance abuse, and children with poor school performance and/or attention deficit disorder. These children may require more intensive intervention.

Resources

BOOKS

- Newton, David E. *Substance Abuse: A Reference Handbook*. Santa Barbara, CA: ABC-CLIO, 2010.
Walcott, Terri A., ed. *Drug and Alcohol Abuse Research Focus*. Hauppauge, NY: Nova Science Publishers, 2007.

PERIODICALS

- “Inhalant Abuse Becomes Focus of SAMHSA Guidelines, Prevention Efforts.” *Alcoholism & Drug Abuse Weekly* March 22, 2004: 1–4.

ORGANIZATIONS

- National Institute on Drug Abuse, 6001 Executive Blvd., Room 5213, Bethesda, MD, (301) 443-1124, information@nida.nih.gov, <http://drugabuse.gov>.

Genevieve Pham-Kanter
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Substance dependence see **Substance abuse and dependence**

Sucralfate see **Antiulcer drugs**

Sucrose intolerance see **Carbohydrate intolerance**

Sudden cardiac death

Definition

Sudden cardiac **death** (SCD) is an unexpected death due to heart problems, which occurs within one hour from the start of any cardiac-related symptoms. SCD is sometimes called cardiac arrest.

KEY TERMS

Defibrillator—A device which delivers a controlled electric shock to the heart to return it to normal beating rhythm.

Ventricular fibrillation—When the lower chamber of the heart quivers instead of pumping in an organized way.

Ventricular tachycardia—A rapid heartbeat, usually over 100 beats per minute.

Description

When the heart suddenly stops beating effectively and breathing ceases, a person is said to have experienced sudden cardiac death.

SCD is not the same as actual death. In actual death, the brain also dies. The important difference is that sudden cardiac death is potentially reversible. If it is reversed quickly enough, the brain will not die.

Sudden cardiac death is also not the same as a **heart attack**. A heart attack (myocardial infarction) is the result of a blockage in an artery which feeds the heart, so the heart becomes starved for oxygen. The part that has been starved is damaged beyond repair, but the heart can still beat effectively.

Causes and symptoms

Sudden cardiac death is usually caused by **ventricular fibrillation** (the lower chamber of the heart quivers instead of pumping in an organized rhythm). Ventricular fibrillation almost never returns to normal by itself, so the condition requires immediate intervention. **Ventricular tachycardia** can also lead to sudden cardiac death. The risk for SCD is higher for anyone with heart disease.

When the heart stops beating effectively and the brain is being deprived of oxygenated blood, a medical emergency exists.

Diagnosis

Diagnosis of sudden cardiac death is made when there is a sudden loss of consciousness, breathing stops, and there is no effective heart beat.

Treatment

When sudden cardiac death occurs, the first priority is to establish the flow of oxygenated blood to the brain.

The next priority is to restore normal rhythm to the heart. Forcing air into the mouth will get oxygen into the lungs. Compressing the chest simulates a pumping heart and will get some blood flow to the lungs, brain, and coronary arteries. This method is called **cardiopulmonary resuscitation** (CPR). When trained help arrives, they will attempt to establish a normal heart beat by using a device called a defibrillator.

If sudden cardiac death occurs outside the hospital setting, cardiopulmonary resuscitation (CPR) must begin within four to six minutes and advanced **life support** measures must begin within eight minutes, to avoid brain death. CPR requires no special medical skills and training is available for the ordinary person nationwide.

Prognosis

Sudden cardiac death is reversible in most people if treatment is begun quickly. However, of the people who are resuscitated, 40% will have another SCD within two years if they do not receive appropriate treatment for the underlying cause of the episode.

Prevention

In order to prevent sudden cardiac death, underlying heart conditions must be addressed. Medications and implantable cardioverter-defibrillators may be used.

ORGANIZATIONS

American Heart Association National Center, 7272 Greenville Avenue, Dallas, TX, 75231, (800) 242-8721, Review.personal.info@heart.org.

Dorothy Elinor Stonely

Sudden infant death syndrome

Definition

Sudden infant **death** syndrome (SIDS) is the unexplained death without warning of an apparently healthy infant, usually during sleep. It is also known as crib death and cot death. According to the Centers for Disease Control and Prevention (CDC), a SIDS death is one that cannot be explained after a thorough investigation is conducted, including a complete **autopsy**, examination of the death scene, and review of the baby's clinical history. The American Academy of Pediatrics (AAP) issued a similar definition in its position paper on SIDS in 2005: SIDS is "the sudden

Ten leading causes of infant death in the United States

- Congenital malformations
- Pre-term/low birth weight
- Sudden infant death syndrome (SIDS)
- Maternal complications
- Accidents and unintentional injuries
- Cord and placental complications
- Bacterial sepsis of newborn
- Respiratory distress
- Diseases of the circulatory system
- Neonatal hemorrhage

SOURCE: U.S. Department of Health and Human Services, National Center for Health Statistics, "Deaths: Final Data for 2007," *National Vital Statistics Reports* 58, no. 19 (May 2010). Available online at: <http://www.cdc.gov/nchs/nvss.htm> (accessed June 10, 2010).

Sudden infant death syndrome is the third leading cause of death among infants in the United States. (*Table by PreMediaGlobal. Reproduced by permission of Gale, a part of Cengage Learning.*)

death of an infant under 1 year of age, which remains unexplained after a thorough case investigation, including performance of a complete autopsy, examination of the death scene, and review of the clinical history."

Demographics

According to the CDC, SIDS is the leading cause of death among American infants between the ages of 1 and 12 months, and is the third leading cause overall of infant mortality in the United States. SIDS is responsible for about 1 death per 2,000 live births as of the early 2000s; however, this figure is more than 50% lower than the figures for 1990, largely as a result of the "Back to Sleep" campaign. The CDC reports that as of 2010, more than 4500 infants die each year in the United States "of no obvious cause." About half these deaths are attributed to SIDS, with the remainder classified as Sudden Unexplained Infant Death or SUID.

Most SIDS deaths occur in babies between 2 and 4 months of age; only 1 percent occur in newborns. About 80% of all SIDS deaths involve infants younger than 5 months. Boys are more likely than girls to die of SIDS; 60–70% of SIDS cases involve boys. Most SIDS deaths in North America occur during the winter and early spring, which are the peak times for respiratory infections. A recent epidemic of a viral disease in the surrounding community is associated with an increased number of SIDS cases in that area.

According to the National Institute for Child Health and Human Development (NICHD), African American babies are twice as likely as Caucasian babies to die from SIDS, and Native American and Alaska Native babies are three times as likely. The same high rates of SIDS cases that occur among Native Americans in the United States are found among First Nations tribes in Canada and aboriginal groups in Australia and New Zealand. The reason for these differences is not yet known as of 2010 but may be related to other risk factors listed below.

Description

In the typical SIDS case, the parents or caregivers put the baby to bed after feeding him or her. Checking on the baby shortly after bedtime indicates that everything is normal; however, the baby is later found dead, usually in the position in which he or she had been placed at bedtime or naptime.

In most cases of SIDS, the parents state that the child was apparently healthy; however, some parents of infants who died of SIDS report that their babies "were not themselves" in the hours before death. In a number of cases, the parents report that the baby had **diarrhea** and **vomiting** at some point in the two weeks prior to death. As of 2010, doctors do not know whether these digestive problems are related to SIDS in some way or are only coincidental.

Risk factors

Studies indicate that some mothers are at increased risk of having their child die of SIDS:

- Those who smoke during pregnancy and after childbirth.
- Those who abuse drugs or alcohol.
- Those who are underweight or suffer from malnutrition.
- Those who have children less than one year apart.
- Teenage mothers. The more children the mother has while still in her teens, the greater the risk of SIDS.
- Those who are obese.

Apart from sleeping position, some babies are at increased risk of SIDS:

- Babies who are born prematurely.
- Babies who weigh 4 pounds or less at birth.
- Babies who are not breastfed.
- Babies born during the fall or winter.
- Babies who had a sibling who died of SIDS.
- Babies who are part of a set of twins, triplets, or quadruplets.

- Babies who are exposed to tobacco smoke.
- Babies put to sleep in an overheated room.
- Babies whose parents practice co-sleeping (the baby shares the parents' bed at night).
- Babies who are overdressed for sleep or covered with too many blankets.

Causes and symptoms

The cause of SIDS is not yet known for certain, although at least 70 different theories have been proposed as of 2010. As of 2010 there are 20 clinical trials under way evaluating apnea, preterm birth, the use of pacifiers, sleeping position, and secondhand smoke in the home as risk factors for SIDS. It is likely that some cases of SIDS are the result of a combination of factors. Doctors have proposed several different theories for SIDS:

- Bacterial infections. A British study published in May 2008 reported that some cases of SIDS appear to result from previously undetected bacterial infections.
- Abnormalities in the part of the brain stem that controls breathing. A study published in the *Journal of the American Medical Association* in the fall of 2007 is one of the strongest pieces of evidence so far that innate differences in brain structure may put some babies at increased risk of SIDS.
- Smothering caused by sleeping on the stomach. This theory holds that babies put to sleep lying on the stomach may breathe in their own exhaled carbon dioxide because they do not have the same ability as older children to move their heads during sleep to get more oxygen.
- Episodes of apnea (sudden cessation of breathing). Babies sometimes stop breathing periodically for reasons that are still not completely understood.
- Abnormalities in heart rhythm. About 10% of babies who die of SIDS have been found to have sudden episodes of extremely rapid heartbeat.
- Triple-risk theory. This theory proposes to explain SIDS as the end result of three factors: a biological vulnerability (such as a weakened heart or abnormal brain stem), an environmental problem (such as sleeping on the stomach), and being too young to regulate breathing and other vital functions as effectively as older children.
- Genetic factors. There is evidence as of late 2009 that 5–10% of SIDS cases are associated with genetic mutations that affect potassium channels in the heart tissue. This abnormality increases the risk of irregular heart rhythms and sudden cardiac death. Some researchers also think that the relatively high

KEY TERMS

Apnea—Temporary cessation of breathing. It may be intentional (holding one's breath) or involuntary, resulting from criminal assault (choking), a neurological disorder, an upper respiratory infection, or accidental trauma.

Autopsy—The examination of a body after death to determine the cause of death.

Brain stem—The lower part of the brain directly connected to the spinal cord. It controls breathing and other vital functions.

Co-sleeping—Allowing a baby to sleep in the same bed as its parents. It is also called bed sharing.

Congenital—Existing or present at the time of birth.

Crib death—Another name for SIDS. It is often called cot death in the United Kingdom, Australia, and New Zealand.

Infanticide—Intentional killing of a child within the first year of life.

Postmortem—Referring to the period following death.

rate of SIDS among African Americans compared to other racial groups may be related to a genetic predisposition to irregular heart rhythms. Research in this area is ongoing as of 2010.

Still other theories about the cause of death in SIDS include immune system disorders that cause changes in the baby's heart rate and breathing patterns during sleep, or a metabolic disorder that causes a buildup of fatty acids in the baby's system.

Theories that are no longer accepted include the notion that SIDS is caused by vaccinations, by dust mites or other insects in the crib mattress, or by toxic gases released by materials used in the manufacture of crib mattresses.

Diagnosis

The diagnosis of SIDS is primarily a diagnosis of exclusion. This means that it is given only after other possible causes of the baby's death have been ruled out. Known risk factors aid in the diagnosis. Unlike the pattern in other diseases, however, the diagnosis of SIDS can only be given postmortem. It is recommended that all infants who die in their sleep receive an autopsy to determine the cause. Autopsies indicate a definite explanation in about 20% of cases of sudden infant death. In addition, an autopsy can often put to rest any doubts the parents may have. Investigation of the location of the death is also useful in determining the child's sleeping position, bedding, room temperature, and similar factors.

The American Academy of Pediatrics (AAP) estimates that between 1% and 5% of cases of SIDS actually involve infanticide. As a result, the AAP has drawn up a list of criteria that must be met in order to distinguish a case of SIDS from child abuse:

- There has been a complete autopsy of the baby performed by a licensed medical examiner, and the autopsy findings are consistent with a diagnosis of SIDS.
- There is no evidence of head trauma or significant disease.
- There is no evidence of trauma to the baby's bones.
- Such other possible causes of death as pneumonia, metabolic disorders, dehydration, severe birth defects, massive infection, trauma to the abdomen, or carbon monoxide poisoning have been ruled out.
- There is no evidence that the baby was given alcohol, drugs, or other toxic substances.
- There is no evidence of foul play when the death scene is investigated.
- The baby's medical history does not indicate previous health problems.

Treatment

There is nothing that can be done to treat the infant when SIDS occurs. Treatment of the parents includes support and understanding; however, the doctor and other health professionals must at the same time conduct a thorough investigation into the circumstances surrounding the baby's death. There are some differences among the states as to the way in which the postmortem (after death) investigation is carried out, but all states require an investigation before the death can be defined as SIDS as of 2010. It is understandably difficult for parents to accept the need for an autopsy and an evaluation of the bed and room in which the baby died when they are grieving; unfortunately, ruling out the possibility of abuse or intentional suffocation of the child is a legal necessity.

Circumstances that concern doctors as well as law enforcement when a baby dies suddenly include:

- The child was 7 months of age or older. SIDS is unusual in this age group.
- The pregnancy was unwanted.
- There have been previous unexplained infant deaths in the family.
- Family members have a history of arrests for violent or intoxicated behavior.

Prevention

The NICHD and CDC recommend the following precautions to reduce the risk of SIDS:

- Infants should always be placed on their backs to sleep when they are left alone; they should be placed on their stomachs *only* when they are awake and supervised by someone responsible.
- If the baby sleeps in a crib, the crib's mattress should be firm and fit snugly into the crib frame. Such other firm sleeping surfaces as bassinets or cradles are also fine.
- The baby should be dressed in a sleeper or pajama to keep it warm rather than being covered by a blanket.
- Parents who co-sleep with a baby should *never* smoke, drink alcohol, or use drugs when sleeping with the baby. It is better to have the baby sleep in a crib or bassinet next to the parents' bed rather than sharing the bed.
- Parents should *never* put a baby on a couch, waterbed, or pillow for a nap.
- Parents should *never* smoke in the same room as the baby or allow anyone else to do so.
- Caregivers should *never* place the baby to sleep or nap with any pillows, stuffed toys, bumper pads, comforters, quilts, or sheepskins.

Resources

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ORGANIZATIONS

American Academy of Pediatrics (AAP), 141 Northwest Point Boulevard, Elk Grove Village, IL, 60007, (847) 434-4000, (847) 434-8000, <http://www.aap.org/>.

American SIDS Institute, 509 Augusta Drive, Marietta, GA, 30067, (770) 426-8746, (800) 232-SIDS, (770) 4261369, <http://sids.org/index.htm>.

Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) 232-4636, cdcinfo@cdc.gov, <http://www.cdc.gov>.

National Association of Medical Examiners (NAME), 430 Pryor Street SW, Atlanta, GA, 30312, (404) 730-4781, <http://thename.org>.

National Institute of Child Health and Human Development (NICHD), Bldg 31, Room 2A32, MSC 2425, 31 Center Drive, Bethesda, MD, 20892, (800) 370-2943, (866) 760-5947, NICHDInformationResourceCenter@mail.nih.gov, <http://www.nichd.nih.gov>.

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Sugar diabetes see **Diabetes mellitus**

Sugar intolerance see **Carbohydrate intolerance**

Suicide

Definition

Suicide is defined as the intentional taking of one's own life. In some European languages, for example German, the word for suicide translates into English as "self-murder." Until the end of the twentieth century approximately, suicide was considered a criminal act; legal terminology used the Latin phrase *felo-de-se*, which means "a crime against the self." Much of the social stigma that is still associated with suicide derives from its former connection with legal judgment as well as with religious condemnation.

In the social climate of 2009, however, suicidal behavior is most commonly regarded—and responded to—as a psychiatric or medical emergency. Law enforcement personnel may be involved in preventing an attempted suicide or taking suicidal individuals to a hospital emergency department but not in arresting these persons for breaking the law.

Demographics

In the United States, the rate of suicide has continued to rise since the 1950s. More people in the general population die from suicide than homicide in North America. There are almost 11 suicide deaths each year for every 100,000 people living in the United States, and for every suicide, there are between 8 and 25 attempts. There are over 30,000 suicides each year in the United States, or about 82 each day; and each day about 1,500 people attempt suicide.

The demographics of suicide in the United States vary considerably from state to state, with rates higher than the national average in the West and lower in the Midwest and Northeast. Some states, like Alaska, have suicide rates that are almost twice the national average; others, such as Massachusetts, have notably lower rates.

These variations from state to state result in part from differences in age and ethnic distributions and gender ratios among the states. In 2008, suicide was the eleventh leading cause of death in the United States, according to the National Institute of Mental Health (NIMH); it was the eighth leading cause of death among males and sixteenth leading cause of death among females. Males are four times more likely than females to succeed in their suicide attempts, but females report attempting suicide at some point in their lives three times as often as men. Among ethnic groups, suicide rates are highest among white males, followed closely by American Indian and Native Alaskan males. The increase in the overall suicide rate in the United States between 1999 and 2005 was due primarily to an increase in suicides among whites aged 40–64, with white middle-aged women experiencing the largest annual increases. As of 2009, the average age of people who completed suicide in the United States is 40 years.

In terms of age, the highest number of suicides are committed by people under age 40, but suicide rates (percentages in a given group) increase with age. People over age 65 have high suicide rates, with men outnumbering women who commit suicide nearly four to one. The ratio of attempted suicides to completed suicides among people over 65 is thought to be as low as 4:1. By contrast, according to the National Strategy for Suicide Prevention (NSSP), seniors are more likely than younger persons to use highly lethal means of suicide. According to a Canadian study published in 2008, seniors are most likely to use firearms to commit suicide, followed by hanging, self-poisoning, and leaping from heights.

The incidence of suicide and attempted suicide among seniors is widely perceived as a growing public health problem in the United States; as of 2009, older adults represent about 13% of the U.S. population but account for 20% of suicides. According to the NIMH, the highest suicide rate in the nation is for Caucasian men ages 85 and older: 65.3 deaths per 100,000 persons, about six times the national U.S. rate of 10.8 per 100,000.

The overall rate of suicide among young people has declined slowly since 1992, but it still remains the third leading cause of death in age groups spanning children 10 years old to young adults up to age 24. Suicidal behavior is rare in prepubertal children, probably because of their

relative inability to plan and execute a suicide attempt. Children as young as 5, however, have succeeded in killing themselves by leaping out of windows or shooting themselves—as happened with one five-year-old who witnessed his mother kill herself with a gun and imitated her behavior several months later. Suicides among young people ages 15 to 24 show an extreme male bias, with the exception of Hispanics: Four times as many males as females aged 15 to 19 and six times as many males age 20 to 24 in the general population committed suicide in 2004. Over half the suicides in this group were firearm-related, and males in general are far more likely to use firearms. According to the NIMH, the rates of suicide for American youth in 2004 (the most recent data available) were as follows:

- Children between the ages of 10 and 14: 1.3 suicides per 100,000.
- Teenagers between the ages of 15 and 19: 8.2 per 100,000.
- Young adults between the ages of 20 and 24: 12.5 per 100,000.

Race or ethnicity is a factor in suicide rates among young people just as it is among older adults. Suicide rates among American Indian and Alaskan natives between 15 and 34 years are almost twice the national average for this age range. Young Hispanic females make significantly more suicide attempts than their male or non-Hispanic counterparts.

Suicide has become a major social and medical problem around the world, not just in North America. Worldwide suicide rates have increased by 60% since 1960. The World Health Organization (WHO) reported that over one million people worldwide died from suicide in the year 2005 (the most recent year for which data are available), more than were murdered or killed in war. That is a global mortality rate of 16:100,000—or one death by suicide every 40 seconds. According to WHO, more suicides occur in Asia than in any other region of the world—with China, Japan, and India accounting for 40% of the world's suicides. China is also the only country in the world where more women than men take their own lives, with female suicides representing 58% of the total. Rates among young people have risen even faster, to the point where they are now the age group at highest risk in 35% of the world's countries.

Description

Historical background

Attitudes toward suicide have varied throughout history. The ancient Greeks considered it an offense against the state, which was deprived of contributions by potentially useful citizens. The Romans, by

comparison, thought that suicide could be a noble form of death, although they legislated against persons taking their own lives before an impending criminal conviction in order to insure their families' financial inheritance. Early Christianity, which downplayed the importance of life on earth, was not critical of suicide until the fourth century, when St. Augustine condemned it as a sin because it violated the sixth commandment ("Thou shalt not kill"). Eventually, the Roman Catholic Church excommunicated and even denied funeral rites to people who killed themselves. The medieval theologian St. Thomas Aquinas condemned suicide because it usurped God's power over life and death. In the *Divine Comedy*, the great writer Dante placed suicides in one of the lowest circles of Hell. The view of suicide as a sin prevailed in Western societies for hundreds of years, and many people are still influenced by it, either consciously or unconsciously. Suicide was a felony and attempted suicide a misdemeanor in England until 1961.

One of the greatest influences on twentieth-century notions about suicide has been the French sociologist Emile Durkheim's 1897 work *Le suicide*. Analyzing French statistics on suicide, Durkheim concluded that suicide is primarily a function of the strength or weakness of a person's ties to family, religion, and community. Persons with weak social ties and those for whom such ties have been disrupted (such as divorced or widowed people) are the most vulnerable to suicide. Durkheim also categorized suicide into four types. Altruistic suicide is actually approved by society, as in the case of a soldier who throws himself on a hand grenade to protect his comrades. In egoistic suicide, individuals kill themselves because they lack the social ties that could motivate them to go on living. Anomic suicide occurs following the loss of a spouse, child, job, or other significant connection to the community, and fatalistic suicides are committed by people driven to despair by dire external circumstances from which there appears to be no escape.

Twenty years after the publication of Durkheim's work, Sigmund Freud provided the first theory that addressed suicide in terms of one's inner mental and emotional state. In *Mourning and Melancholia* (1917), he proposed that suicide was the result of turning hostility toward a loved one back on oneself. In *Man against Himself* (1936), Karl Menninger extended Freud's contribution to the psychodynamic study of suicide, relating it to such other forms of self-destructive behavior as **alcoholism** or drug **abuse**. Some people still refer to such behavior as "slow-motion suicide."

Types of suicidal behavior

Some mental health professionals distinguish five levels of suicidal behavior: completed suicide; suicide

attempts, which are potentially fatal; suicide gestures, which involve acting-out behavior that is not necessarily lethal; suicide gambles; and suicidal ideation, or thinking about suicide. An example of a suicide gesture would be cutting one's wrist just deeply enough to draw blood from the skin but not deeply enough to sever veins and arteries. The suicide gamble is a type of suicidal behavior in which the person takes the risk that he or she will be discovered in time and that the discoverer will save them. The poet Sylvia Plath's suicide in 1963 is considered an example of a suicide gamble. Plath gassed herself in the kitchen by turning on her oven without lighting it, but left a note on the door for her children's new nanny, had opened the windows in the children's bedroom to protect them, and had sealed the door to the kitchen with dish towels.

Suicidal ideation, or thinking about suicide, is even more common than suicide gestures or attempted suicide. Suicidal ideation spans a continuum from nonspecific thoughts such as "life is not worth living" to specific ideation. Community surveys indicate that between 12 and 25% of primary and high school children have some form of suicidal ideation, whereas 5 to 10% combine suicidal ideation with a plan or intent to make a suicide attempt. Not surprisingly, specific ideation is more closely associated with risk for attempted suicide, and frequently occurs in combination with other risk factors.

Risk factors

Some factors increase a person's risk of suicide:

- Male sex.
- Age over 75.
- A family history of suicide.
- A history of suicide attempts.
- Caucasian race.
- A history of abuse in childhood.
- Traumatic experiences after childhood.
- Recent stressful events, such as separation or divorce, job loss, or death of spouse.
- Chronic medical illness. Patients with AIDS have a rate of suicide 20 times that of the general population.
- Chronic, severe, or intractable pain.
- Loss of mobility or independence.
- Access to a firearm. Death by firearms now accounts for the majority of suicides in the United States.
- Alcohol or substance abuse. While mood-altering substances do not cause a person to kill himself or herself, they weaken impulse control.
- High blood cholesterol levels.

• Presence of a psychiatric illness. Over 90% of Americans who commit suicide have a mental illness. Major depression accounts for 60% of suicides, followed by schizophrenia, alcoholism, substance abuse, borderline personality disorder, Huntington's disease, and epilepsy. The lifetime mortality due to suicide in psychiatric patients is 15% for major depression; 20% for bipolar disorder; 18% for alcoholism; 10% for schizophrenia; and 5–10% for borderline and certain other personality disorders.

In children and adolescents, the most common triggers of suicidal behavior involve interpersonal conflict or loss, most frequently with parents or romantic attachment figures. Family discord, physical or **sexual abuse**, and an upcoming legal or disciplinary crisis are also commonly associated with completed and attempted suicide. The most serious suicide attempts leave suicide notes, show evidence of planning, and use an irreversible method. Most adolescent suicide attempts, though, are of relatively low intent and lethality, and only a minority actually want to die. Usually, children and adolescents who attempt suicide want to escape psychological **pain** or unbearable circumstances, gain attention, influence others, or communicate such strong feelings as rage or love.

Factors that lower the risk of suicide in adults include:

- A significant friendship network outside the workplace.
- Religious faith and practice, especially those that discourage suicide and value life.
- A stable marriage.
- A close-knit extended family.
- A strong interest in or commitment to a project or cause that brings people together: community service, environmental concerns, neighborhood associations, animal rescue groups, etc.

Causes and symptoms

Suicide is an act that represents the end result of a combination of factors in any individual. One model that has been used by clinicians to explain why people suffering under the same life stresses respond differently is known as the stress/diathesis model. Diathesis is a medical term for a predisposition that makes some people more vulnerable to thoughts of suicide. In addition to factors at the individual level, factors in the wider society have been identified as contributing to the rising rate of suicide in the United States:

- Stresses on the nuclear family, including more frequent divorce and economic hardship.

KEY TERMS

Assisted suicide—A form of self-inflicted death in which individuals voluntarily bring about their own death with the help of another, usually a physician, relative, or friend. Assisted suicide is sometimes called physician-assisted death (PAD).

Cortisol—A hormone released by the cortex (outer portion) of the adrenal gland when a person is under stress. Cortisol levels are now considered a biological marker of suicide risk.

Dexamethasone test—A test that serves as a marker of suicide risk by reflecting signaling activity between the brain and the adrenal gland.

Diathesis—The medical term for predisposition. The stress/diathesis model is a diagram that is used to explain why some people are at greater risk of suicidal behavior than others.

Euthanasia—The act of putting individuals or animals to death painlessly or allowing them to die by withholding medical services, usually because of an incurable disease; also called mercy killing.

Frontal cortex—The part of the human brain associated with aggressiveness and impulse control. Abnormalities in the frontal cortex are associated with an increased risk of suicide.

Serotonin—A chemical that occurs in the blood and nervous tissue and functions to transmit signals across the gaps between neurons in the central nervous system. Abnormally low levels of serotonin are associated with depression and an increased risk of suicide.

Self-deliverance—Another term for assisted suicide, more commonly used in Great Britain than in the United States.

Suicide gesture—Attempted suicide characterized by a low-lethality method, low level of intent or planning, and little physical damage; sometimes called pseudocide.

Suicide magnet—A bridge, tall building, or geographic location that acquires a reputation for attracting people who want to commit suicide and attempt it.

- The loss of a set of moral values held in common by the entire society.
- The weakening of churches, synagogues, neighborhood associations, and other mid-range social groups outside the family. In the past, these institutions often provided a sense of belonging for people from troubled or emotionally distant families.
- Frequent geographical moves, which makes it hard for people to make and keep long-term friendships outside their immediate family.
- Sensationalized treatment of suicide in the mass media. A number of research studies have shown that there is a definite risk of “contagion” or copycat suicides from irresponsible reporting, particularly among impressionable adolescents. One group of researchers has estimated that as many as 6% of all suicides in the developed countries are copycat suicides.
- The development over the past century of medications that allow relatively painless suicide. For most of human history, the available means of suicide were uncertain, painful, or both.
- The easy availability of lethal methods of suicide, most notably firearms, and so-called suicide magnets such as bridges or tall buildings that do not have suicide barriers and are easy to reach even for

teenagers too young to drive or seniors who have given up driving. The Golden Gate Bridge in San Francisco is the most notorious suicide magnet in the United States; others include the Aurora Bridge in Seattle, the Sunshine Skyway Bridge in Florida, and the Duke Ellington Bridge in Washington, D.C. Suicide magnets elsewhere in the world include the Aokigahara Forest at the base of Mount Fuji in Japan, and Beachy Head in the United Kingdom.

The role of the Internet in the rate of adolescent suicide has been debated. On the one hand, there are websites and chat rooms that foster preoccupation with suicide and offer detailed descriptions of suicide methods. There are even instances of adolescents recruiting other adolescents over the Internet to join them in a suicide pact, as happened in Japan in October 2004. Seven young people who had met via the Internet committed group suicide by inhaling carbon monoxide from a charcoal burner inside a locked van. Other websites attack psychiatry and mental health professionals, which may steer some vulnerable young people away from seeking help. On the other hand, there are many supportive websites for teens that offer resources (including peer counseling) and contact information for getting help if they are considering suicide.

Diagnosis

The diagnosis of a suicide attempt is often made when the patient either goes to the emergency room of a hospital to seek help or is taken there by family members or first responders. In many cases the patient will have written a suicide note, talked about his or her intention, or begun to carry out a plan to kill themselves. If the patient is not conscious, the doctor will obtain as much information as possible from family members or first responders.

Treatment of attempted suicide

Suicide attempts can be broadly categorized along a continuum that ranges from planned attempts involving highly lethal methods that fail by good fortune to impulsive or poorly planned attempts using less lethal methods.

Adults

An adult suicide attempt of any kind, however, is treated as a psychiatric emergency by police or other rescue personnel. Treatment in a hospital emergency room includes a complete psychiatric evaluation, a **mental status examination**, and a detailed assessment of the circumstances surrounding the attempt. The physician will interview the person's relatives or anyone else who accompanied the patient in order to obtain as much information as possible. Some questions that the physician will ask include whether the patient had a detailed plan for suicide; whether he or she had the means of suicide at hand; what the patient hoped to gain by killing themselves (freedom from pain, reunion with a dead loved one, solution to financial problems, etc.); and whether the patient had any tendencies toward homicide. As a rule, suicide attempts requiring advance planning and the use of violent or highly lethal methods are regarded as the most serious. The patient will be kept under observation while decisions are made about the need for hospitalization.

People who have attempted suicide and who are considered a serious danger to themselves or to others can be legally hospitalized against their will. The doctor bases the decision on the severity of the patient's depression or agitation; the presence of other suicide risk factors, including a history of previous suicide attempts, **substance abuse**, recent stressful events, and symptoms of **psychosis**; and the availability of friends, relatives, or other social support. If the attempt is judged to be a nonlethal suicide gesture, and the patient has adequate support outside the hospital, then he or she may be released after the psychiatric assessment is completed.

Adolescent

The first step in the care of a suicidal teenager is to determine the degree of suicidal risk and the appropriate level of care. It is critical to obtain a no-suicide contract with the patient and family, in which the patient promises to refrain from self-destructive behavior and to notify the professional or caregiver if he or she does feel suicidal again. Treatment of the suicidal youngster should proceed on four levels: (1) removal of firearms and dangerous medications from the home; (2) treatment of the underlying psychiatric disorders; (3) remediation of social and problem-solving skills; (4) evaluation of the patient's home and school environment; and (5) family education about psychiatric problems and suicidal risk.

Another important aspect of aftercare is continuity of treatment; the growing complexity and specialization of the healthcare system means that suicidal children and adolescents are frequently shuffled from one clinic or facility to another. Lack of continuity of care places these young people at an increased risk of additional suicide attempts.

Ethical issues related to suicide

Several ethical issues related to suicide have emerged as public policy matters in the early twenty-first century. The most controversial of these are the notion of a "right to suicide" and the question of assisted suicide.

Right to suicide

The idea that suicide is a right among the elderly or those with terminal illnesses surfaced with the 1991 publication of Derek Humphry's *Final Exit*, a controversial book described by its author as a how-to manual for suicide and assisted suicide. Humphry is the founder of the Euthanasia Research and Guidance Organization (ERGO), known until 2003 as the Hemlock Society. Humphry maintains that people have a right to choose the time, place, and method of their death and that rational suicide is a legitimate and even reasonable choice.

People who are often overlooked in discussions of the right to commit suicide, however, are the relatives and friends who are bereaved by the suicide. It is estimated that each person who commits suicide leaves six survivors to deal with the aftermath. On the basis of this figure, there are at least 4.5 million survivors of suicide in the United States. In addition to the grief that ordinarily accompanies death, survivors of suicide often struggle with feelings of guilt and shame as well. Some people have blamed Humphry and his book for their loved one's decision to commit suicide.

Assisted suicide

Questions pertaining to the legalization of assisted suicide for persons suffering from a terminal illness are connected in part to increases in the average lifespan. Physician-assisted suicide (also known as physician-assisted death or PAD) was legalized in the Netherlands in April 2001 and in the states of Oregon, Washington, and Montana. As of 2009 it was also legal in Belgium and is practiced openly in Switzerland. It is important to distinguish between physician-assisted suicide and euthanasia, or mercy killing. Assisted suicide, which is called “self-deliverance” in Britain, refers to individuals bringing about their own death with the help of another person. Because the other person is often a physician, the act is often called doctor-assisted suicide.

Euthanasia strictly speaking means that the physician or other person is the one who performs the last act that causes death. For example, if a physician injects a patient with a lethal dose of a pain-killing medication, the physician is performing euthanasia. If the physician leaves the patient with a loaded syringe and the patient injects himself or herself with it, the act is an assisted suicide. As of 2009, assisted suicide is illegal everywhere in the United States except Oregon, Washington, and Montana; and euthanasia is illegal in all fifty states. The *Merck Manual of Geriatrics* states: “Physicians can provide treatment intended to minimize [a patient’s] physical and emotional suffering, even if a secondary result is the shortening of life, but they cannot specifically intend to hasten death.”

Media treatment of suicide

In 1989, the Centers for Disease Control and Prevention (CDC) sponsored a national workshop to address the connection between sensationalized media treatments of suicide and the rising rate of suicide among American youth. The CDC and the American Association of Suicidology subsequently adopted a set of guidelines for media coverage of suicide intended to reduce the risk of copycat suicides.

The CDC guidelines point out that the following types of reporting may increase the risk of copycat suicides:

- Presenting oversimplified explanations of suicide, when in fact many factors usually contribute to it. One example concerns the suicide of the widow of a man who was killed in the collapse of the World Trade Center on September 11, 2001. Most newspapers that covered the story described her death as due solely to the act of terrorism, even though she had a history of depressive illness.

- Excessive, ongoing, or repetitive coverage of the suicide.
- Sensationalizing the suicide by inclusion of morbid details or dramatic photographs.
- Giving “how-to” descriptions of the method of suicide.
- Referring to suicide as an effective coping strategy or as a way to achieve temporary fame or other goals.
- Glorifying the act of suicide or the person who commits suicide.
- Focusing on the person’s positive traits without mentioning his or her problems.

Traditional

People who survive a suicide attempt are usually treated with a combination of antidepressant medications and **psychotherapy**.

Drugs

In 2003 the Food and Drug Administration (FDA) approved the use of clozapine (Clozaril), an antipsychotic medication, for the treatment of patients with **schizophrenia** who have attempted suicide.

TREATMENT OF SUICIDE SURVIVORS. In addition to the grief that ordinarily accompanies death, survivors of a friend or relative’s suicide often struggle with feelings of guilt and shame as well. In spite of a general liberalization of social attitudes since World War II, suicide is still stigmatized in many parts of Europe and the United States. Survivors often benefit from group or individual psychotherapy in order to work through such issues as wondering whether they could have prevented the suicide or whether they are at increased risk of committing suicide themselves. Increasing numbers of clergy as well as mental health professionals are taking advanced training in counseling survivors of suicide.

Prognosis

The prognosis for a person who has attempted suicide is generally favorable, although further research needs to be done. Many different studies have followed individuals who attempted suicide to determine how likely individuals who attempt suicide once are likely to die by suicide. These studies have generally found that the likelihood is less than 10%. Some have found the likelihood of eventual death by suicide to be 6% or lower. A doctor who studied 515 people who attempted suicide between 1937 and 1971 found that 94% were still alive at the time of his study or had died of natural causes. In general individuals who attempt suicide and rate highly on intent to commit suicide and hopelessness

may be more likely to commit suicide at a later time. These findings may have been taken to indicate that suicidal behavior is more likely to be a passing response to an acute crisis than a reflection of a permanent state of mind.

Prevention

One reason that suicide is such a tragedy is that most self-inflicted deaths are potentially preventable. Many suicidal people change their minds if they can be helped through their immediate crisis; Dr. Richard Seiden, a specialist in treating survivors of suicide attempts, puts the high-risk period at 90 days after the crisis. Some potential suicides change their minds during the actual attempt; for example, a number of people who survived jumping off the Golden Gate Bridge told interviewers afterward that they regretted their action even as they were falling and that they were grateful they survived.

Brain research is an important means of suicide prevention. Known biological markers for an increased risk of suicide can now be correlated with personality profiles linked to suicidal behavior under **stress** to help identify individuals at risk. One new clinical parameter that may be considered with personality profiles is the dexamethasone suppression test, which serves as an indicator of hyperactivity of a neuroendocrine hormonal pathway between the brain and the adrenal gland. Another clinical parameter that may be combined with psychological assessment is an assessment of serotonin function based on cholesterol levels, with high levels indicating an increased risk of suicide. In addition, brain imaging studies using **positron emission tomography (PET)** are being used to detect abnormal patterns of serotonin uptake in specific regions of the brain. Genetic studies are also yielding new information about inherited predispositions to suicide.

In the late 2000s research was ongoing to discover better methods of treating depression and other disorders that may influence a person's decision to commit suicide. In addition, primary care physicians are continually learning how to better identify and intervene when treating suicidal patients. An estimated 67% of all adults, and 80% of seniors who complete suicide, have seen a physician within a month of their death. Thus primary care physicians are in a good position to evaluate their patients for signs of depression. The good news is that depression in adults in any age group is highly treatable, particularly when antidepressant medications are combined with psychotherapy.

Warning signs of suicidal thinking have been identified:

- Reading a lot of books or articles on death and suicide
- Talking a lot about death or suicide or expressing feelings of hopelessness
- Stockpiling medications
- Refusing to take care of oneself
- Sudden interest in guns
- Giving away cherished possessions, writing long letters, or making other elaborate farewells
- Disrupted sleep patterns
- Hurriedly revising a will
- Increased intake of alcohol or prescription drugs

People who are concerned about a friend or relative at risk of self-harm should take the following steps:

- Become educated about warning signs and risk factors.
- Identify physicians and other healthcare professionals who know the person and can provide help; and keep their telephone numbers readily available.
- Talk openly with the person about his or her feelings. Although many people are afraid to ask whether someone is thinking about suicide for fear of angering them or giving them an idea, in many cases honest concern is welcomed by the individual.
- Call the local hospital emergency department or 911 if the person seems to be at immediate risk of suicide.

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ORGANIZATIONS

- American Academy of Child and Adolescent Psychiatry (AACAP), 3615 Wisconsin Avenue, N.W., Washington, DC, 20016-3007, (202) 966-7300, (202) 966-2891, <http://www.aacap.org/>.
- American Association of Suicidology (AAS), 5221 Wisconsin Avenue, NW, Washington, DC, 20015, (202) 237-2280, (202) 237-2282, <http://www.suicidology.org/web/guest/home>.
- American Foundation for Suicide Prevention (AFSP), 120 Wall Street, 22nd Floor, New York, NY, 10005, (212) 363-3500, (888) 333-AFSP, (212) 363-6237, inquiry@afsp.org, <http://www.afsp.org/>.
- American Psychiatric Association, 1000 Wilson Boulevard, Suite 1825, Arlington, VA, 22209-3901, (703) 907-7300, apa@psych.org, <http://www.psych.org/>.
- National Institute of Mental Health (NIMH), 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD, 20892-9663, (301) 443-4513, (866) 615-6464, (301) 443-4279, nimhinfo@nih.gov, <http://www.nimh.nih.gov/index.shtml>.
- National Suicide Prevention Lifeline, (800) 273-TALK

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Sulfacetamide see **Antibiotics, ophthalmic**

Sulfamethoxazole and trimethoprim see
Sulfonamides

Sulfinpyrazone see **Gout drugs**

Sulfisoxazole see **Sulfonamides**

Sulfonamides

Definition

Sulfonamides are medicines that prevent the growth of bacteria in the body.

Purpose

Sulfonamides are used to treat many kinds of infections caused by bacteria and certain other microorganisms. Physicians may prescribe these drugs to treat urinary tract infections, ear infections, frequent or long-lasting **bronchitis**, bacterial **meningitis**, certain eye infections, *Pneumocystis carinii*pneumonia, **traveler's diarrhea**, and a number of other kinds of infections. These drugs will *not* work for colds, flu, and other infections caused by viruses.

Description

Sulfonamides, also called sulfa medicines, are available only with a physician's prescription. They are sold in tablet and liquid forms. Some commonly used sulfonamides are sulfisoxazole (Gantrisin) and the combination drug sulfamethoxazole and trimethoprim (Bactrim, Cotrim).

Recommended dosage

The recommended dosage depends on the type of sulfonamide, the strength of the medicine, and the medical problem for which it is being taken. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage.

Always take sulfonamides exactly as directed. To make sure the infection clears up completely, take the medicine for as long as it has been prescribed. Do not stop taking the drug just because symptoms begin to improve. Symptoms may return if the drug is stopped too soon.

Sulfonamides work best when they are at constant levels in the blood. To help keep levels constant, take the medicine in doses spaced evenly through the day and night. Do not miss any doses. For best results, take the medicine with a full glass of water and drink several more glasses of water every day. This will help prevent some of the medicine's side effects.

Precautions

Symptoms should begin to improve within a few days of beginning to take this medicine. If they do not, or if they get worse, check with the physician who prescribed the medicine.

Although such side effects are rare, some people have had severe and life-threatening reactions to sulfonamides. These include sudden, severe liver damage, serious blood problems, breakdown of the outer layer of the skin, and a condition called Stevens-Johnson syndrome, in which people get blisters around the mouth,

eyes, or anus. Call a physician immediately if any of these signs of a dangerous reaction occur:

- skin rash or reddish or purplish spots on the skin
- other skin problems, such as blistering or peeling
- fever
- sore throat
- cough
- shortness of breath
- joint pain
- pale skin
- yellow skin or eyes

This medicine may cause **dizziness**. Anyone who takes sulfonamides should not drive, use machines or do anything else that might be dangerous until they have found out how the drugs affect them.

Sulfonamides may cause blood problems that can interfere with healing and lead to additional infections. Avoid injuries while taking this medicine. Be especially careful not to injure the mouth when brushing or flossing the teeth or using a toothpick. Do not have dental work done until the blood is back to normal.

This medicine may increase sensitivity to sunlight. Even brief exposure to sun can cause a severe **sunburn** or a rash. While being treated with this medicine, avoid being in direct sunlight, especially between 10 a.m. and 3 p.m.; wear a hat and tightly woven clothing that covers the arms and legs; use a sunscreen with a skin protection factor (SPF) of at least 15; protect the lips with a sun block lipstick; and do not use **tanning** beds, tanning booths, or sunlamps.

Babies under 2 months should not be given sulfonamides unless their physician has ordered the medicine.

Older people may be especially sensitive to the effects of sulfonamides, increasing the chance of unwanted side effects, such as severe skin problems and blood problems. Patients who are taking water pills (**diuretics**) at the same time as sulfonamides may also be more likely to have these problems.

Special conditions

People with certain medical conditions or who are taking certain other medicines can have problems if they take sulfonamides. Before taking these drugs, be sure to let the physician know about any of these conditions:

ALLERGIES Anyone who has had unusual reactions to sulfonamides, water pills (diuretics), diabetes medicines, or glaucoma medicine in the past should let his or her physician know before taking sulfonamides. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

PREGNANCY In studies of laboratory animals, some sulfonamides cause **birth defects**. The drugs' effects on human fetuses have not been studied. However, pregnant women are advised not to use this medicine around the time of labor and delivery, because it can cause side effects in the baby. Women who are pregnant or who may become pregnant should check with their physicians about the safety of using sulfonamides during pregnancy.

BREASTFEEDING Sulfonamides pass into breast milk and may cause liver problems, anemia, and other problems in nursing babies whose mothers take the medicine. Because of those problems, women should not breastfeed when they are under treatment with this drug. Women who are **breastfeeding** and who need to take this medicine should check with their physicians to find out how long they need to stop breastfeeding.

OTHER MEDICAL CONDITIONS Before using sulfonamides, people with any of these medical problems should make sure their physicians are aware of their conditions:

- anemia or other blood problems
- kidney disease
- liver disease
- asthma or severe allergies
- alcohol abuse
- poor nutrition
- abnormal intestinal absorption
- porphyria
- folic acid deficiency
- deficiency of the enzyme glucose-6-phosphate dehydrogenase (G6PD)

USE OF CERTAIN MEDICINES Taking sulfonamides with certain other drugs may affect the way the drugs work or may increase the chance of side effects.

Side effects

The most common side effects are mild **diarrhea**, **nausea**, **vomiting**, dizziness, **headache**, loss of appetite, and tiredness. These problems usually go away as the body adjusts to the drug and do not require medical treatment.

More serious side effects are not common, but may occur. If any of the following side effects occur, check with a physician immediately:

- itching or skin rash
- reddish or purplish spots on the skin
- other skin problems, such as redness, blistering, peeling

KEY TERMS

Anemia—A lack of hemoglobin—the compound in blood that carries oxygen from the lungs throughout the body and brings waste carbon dioxide from the cells to the lungs, where it is released.

Bronchitis—Inflammation of the air passages of the lungs.

Fetus—A developing baby inside the womb.

Inflammation—Pain, redness, swelling, and heat that usually develop in response to injury or illness.

Meningitis—Inflammation of tissues that surround the brain and spinal cord.

***Pneumocystis carinii* pneumonia**—A lung infection that affects people with weakened immune systems, such as people with AIDS or people taking medicines that weaken the immune system.

Porphyria—A disorder in which porphyrins build up in the blood and urine.

Porphyrin—A type of pigment found in living things.

Urinary tract—The passage through which urine flows from the kidneys out of the body.

- severe, watery or bloody diarrhea
- muscle or joint aches
- fever
- sore throat
- cough
- shortness of breath
- unusual tiredness or weakness
- unusual bleeding or bruising
- pale skin
- yellow eyes or skin
- swallowing problems

Other rare side effects may occur. Anyone who has unusual symptoms while taking sulfonamides should get in touch with his or her physician.

Interactions

Sulfonamides may interact with a large number of other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. Anyone who takes sulfonamides should let the physician know all other medicines he or she is taking. Among the drugs that may interact with sulfonamides are:

- acetaminophen (Tylenol)
- medicine for overactive thyroid
- male hormones (androgens)
- female hormones (estrogens)
- other medicines used to treat infections
- birth control pills
- medicines for diabetes such as glyburide (Micronase)
- anticoagulants such as warfarin (Coumadin)
- disulfiram (Antabuse), used to treat alcohol abuse
- amantadine (Symmetrel), used to treat flu and also Parkinson's disease
- water pills (diuretics) such as hydrochlorothiazide (HCTZ, HydroDIURIL)
- the anticancer drug methotrexate (Rheumatrex)
- antiseizure medicines such as valproic acid (Depakote, Depakene)

The list above does not include every drug that may interact with sulfonamides. Be sure to check with a physician or pharmacist before combining sulfonamides with any other prescription or nonprescription (over-the-counter) medicine.

Nancy Ross-Flanigan

Sumatriptan see **Antimigraine drugs**

Sunburn

Definition

Inflammation of the skin caused by overexposure to the sun.

Description

Sunburn is caused by exposure to the ultraviolet (UV) rays of the sun. There are two types of ultraviolet rays, UVA and UVB. UVA rays penetrate the skin more deeply and can cause melanoma in susceptible people. UVB rays, which do not penetrate as deeply, cause sunburn and wrinkling. Most UVB rays are absorbed by **sunscreens**, but only about half the UVA rays are absorbed.

Skin **cancer** from sun overexposure is a serious health problem in the United States, affecting more than a million Americans each year. One out of 87 will develop **malignant melanoma**, the most serious type of skin cancer, and 8,100 of them will die each year.



Sunburn

This person has a second-degree sunburn on the back of the neck. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

Fair-skinned people are most susceptible to sunburn because their skin produces only small amounts of the protective pigment called melanin. People trying to get a tan too quickly in strong sunlight are also more vulnerable to sunburn. While they have a lower risk, even the darkest-skinned people can get skin cancer.

Repeated sun overexposure and burning can prematurely age the skin, causing yellowish, wrinkled skin. Overexposure can increase the risk of skin cancer, especially a serious burn in childhood.

Causes and symptoms

The ultraviolet rays in sunlight destroy cells in the outer layer of the skin, damaging tiny blood vessels underneath. When the skin is burned, the blood vessels dilate and leak fluid. Cells stop making protein. Their DNA is damaged by the ultraviolet rays. Repeated DNA damage can lead to cancer.

When the sun burns the skin, it triggers immune defenses, which identify the burned skin as foreign. At the same time, the sun transforms a substance on the skin which interferes with this immune response. While this substance keeps the immune system from attacking a person's own skin, it also means that any malignant cells in the skin will be able to grow freely.

Sunburn causes skin to turn red and blister. Several days later, the dead skin cells peel off. In severe cases, the burn may occur with sunstroke (**vomiting**, **fever** and collapse).

Diagnosis

Visual inspection and a history of exposure to the sun.

KEY TERMS

Malignant melanoma—The most deadly of the three types of skin cancer.

Sunscreen—Products which block the damaging rays of the sun. Good sunscreens contain either para-aminobenzoic acid (PABA) or benzophenone, or both. Sunscreen protection factors range from 2–45.

Treatment

Aspirin can ease **pain** and inflammation. Tender skin should be protected against the sun until it has healed. In addition, apply:

- calamine lotion
- sunburn cream or spray
- cool tap water compress
- colloidal oatmeal (Aveeno) baths
- dusting powder to reduce chafing

People who are severely sunburned should see a doctor, who may prescribe corticosteroid cream to speed healing.

Alternative treatment

Over-the-counter preparations containing aloe (*Aloe barbadensis*) are an effective treatment for sunburn, easing pain and inflammation while also relieving dryness of the skin. A variety of topical herbal remedies applied as lotions, poultices, or compresses may also help relieve the effects of sunburn. Calendula (*Calendula officinalis*) is one of the most frequently recommended to reduce inflammation. Apple cider vinegar applied to the burn area also can help in relieving the pain of sunburns.

Prognosis

Moderately burned skin should heal within a week. While the skin will heal after sunburn, the risk of skin cancer increases with exposure and subsequent burns. Even one bad burn in childhood carries an increased risk of skin cancer.

Prevention

Everyone from age six months on should use a water-resistant sunscreen with a sun protective factor (SPF) of at least 15. Apply at least an ounce 15–30 minutes before going outside. It should be reapplied every two hours (more often after swimming). Babies should be kept completely out of the sun for the first six

months of life, because their skin is thinner than older children. Sunscreens have not been approved for infants.

In addition, people should:

- limit sun exposure to 15 minutes the first day, even if the weather is hazy, slowly increasing exposure daily
- reapply sunscreen every two hours (more often if sweating or swimming)
- reapply waterproof sunscreen after swimming more than 80 minutes, after toweling off, or after perspiring heavily
- avoid the sun between 10 a.m. and 3 p.m.
- use waterproof sunscreen on legs and feet, since the sun can burn even through water
- wear an opaque shirt in water, because reflected rays are intensified

If using a sunscreen under SPF 15, simply applying more of the same SPF won't prolong allowed time in the sun. Instead, patients should use a higher SPF in order to lengthen exposure safely. A billed cap protects 70% of the face; a wide-brimmed hat is better. People at very high risk for skin cancer can wear clothing that blocks almost all UV rays, but most people can simply wear white cotton summer-weight clothing with a tight weave.

Resources

BOOKS

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ORGANIZATIONS

- American Academy of Dermatology (AAD), P. O. Box 4014, Schaumburg, IL, 60168-4014, (866) 503-7546, <http://www.aad.org>.
National Institute of Arthritis and Musculoskeletal and Skin Diseases, National Institutes of Health, 1 AMS Circle,

Bethesda, MD, 20892, (877) 226-4267, <http://www.niams.nih.gov>.
 American Burn Association, 625 N. Michigan Ave., Suite 2550, Chicago, IL, 60611, (800) 548-2876, <http://www.ameriburn.org>.

Carol A. Turkington
 Ken R. Wells

Sunscreens

Definition

Sunscreens are products applied to the skin to protect against the harmful effects of the sun's ultraviolet (UV) rays.

Purpose

Everyone needs a little sunshine. About 15 minutes of exposure a day helps the body make vitamin D, which is important for healthy bones and teeth. But longer exposure may cause many problems, from wrinkles to skin **cancer**. One particularly deadly form of skin cancer, **malignant melanoma**, has been on the rise in recent decades, as **tanning** has become more popular. Over the same period, scientists have warned that the thin layer of ozone that protects life on Earth from the sun's ultraviolet radiation is being depleted. This allows more UV radiation to get through, adding to the risk of overexposure.

Sunscreens help protect against the sun's damaging effects. The sun gives off two kinds of ultraviolet radiation, called UVA and UVB. Ultraviolet B (UVB) rays are thought to cause most basal cell and squamous cell skin cancers. Ultraviolet A (UVA) rays penetrate the skin more deeply than UVB rays which may lead to more skin damage.

Some medical experts are concerned that sunscreens give people a false sense of security, allowing them to stay in the sun longer than they should. Although sunscreens protect the skin from burning, they may not protect against other kinds of damage. A number of studies suggest that people who use sunscreens may actually increase their risk of melanoma because they spend too much time in the sun. This does not mean that people should stop using sunscreens. It means that they should not rely on sunscreens *alone* for protection. According to the American Academy of Dermatology, sunscreens should be one part of sun protection, along with wide-brimmed hats and tightly-woven clothing that covers the arms and legs.

Sunscreens are also recommended for patients with **rosacea** or other skin disorders that are aggravated by exposure to sunlight.

Description

Many brands of sunscreens are available, containing a variety of ingredients. The active ingredients work by absorbing, reflecting, or scattering some or all of the sun's rays. Most sunscreen products contain combinations of ingredients.

Sunscreens are usually grouped into two major categories, namely chemical absorbers and physical blockers. Chemical absorber compounds include avobenzone, padimate O, octyl methoxycinnamate, octisalate, and octocrylene. Physical blocker compounds include titanium dioxide and zinc oxide. The chief drawback of physical blockers is their tendency to leave a white film on the skin, causing many people to use less of the product than they should for full sun protection.

Sunscreen products are sold as lotions, creams, gels, oils, sprays, sticks, and lip balms, and can be bought without a physician's prescription.

The U.S. Food and Drug Administration (FDA) has required sunscreen products to carry a sun protection factor (SPF) rating on their labels since 1999. This number tells the consumer how well the sunscreen protects against burning. The higher the number, the longer a person can stay in the sun without burning. However, SPF ratings apply only to protection from UVB rays.

UVA rays damage the skin without burning. In the United States, the FDA has had no mandatory criteria related to labeling of the effectiveness of sunscreens in blocking UVA rays. However, new proposed labeling criteria which went into effect in October, 2010 requires companies to label their products' effectiveness in protecting against UVA rays by using a one to four star rating system. A product with a one star rating offers the lowest protection while a four star rating indicates the highest level of protection against UVA rays. Products that do not provide any protection against UVA rays would be labeled without a star and must state on the label "no UVA protection." The star rating is to be prominently displayed near the SPF rating on the product label.

Products that contain the ingredients zinc oxide, titanium dioxide, avobenzone, and mexoryl provide protection against UVA rays.

KEY TERMS

Hair follicle—A tiny pit in the skin from which hair grows.

Melanoma—A rapidly spreading and deadly form of cancer that usually occurs on the skin.

Ozone—A gas found in the atmosphere. A layer of ozone about 15 mi (24 km) above Earth's surface helps protect living things from the damaging effects of the sun's ultraviolet rays.

Pus—Thick, whitish or yellowish fluid that forms in infected tissue.

Rosacea—A chronic skin disease characterized by persistent redness of the skin and periodic outbreaks of pustules, usually affecting the middle third of the face.

Ultraviolet rays—Invisible light rays with a wavelength shorter than that of visible light but longer than that of x rays.

Recommended dosage

One should be sure to read the instructions that come with a product. Some products need to be applied as long as one or two hours before sun exposure. Others should be applied 15–30 minutes before exposure, and reapplied frequently during exposure.

People should apply sunscreen liberally to all exposed parts of the skin, including hands, feet, nose, ears, neck, scalp (if the hair is thin or very short), and eyelids. Users should take care not to get sunscreen in the eyes, as it can cause irritation. People should also use a lip balm containing sunscreen to protect the lips, and reapply sunscreen liberally every one to two hours—more frequently when perspiring heavily or after swimming.

Precautions

Sunscreen alone will not provide full protection from the sun. When possible, one should wear a hat, long pants, long-sleeved shirts or blouses, and sunglasses. Adults should consider applying protective wear for children which is designed to cover the child from the neck to the knees with sun-protective factors. Try to stay out of the sun between 10 a.m. and 2 p.m. (11 a.m. to 3 p.m. Daylight Saving Time), when the sun's rays are strongest. The sun can damage the skin even on cloudy days, so get in the habit of using a sunscreen every day. Be especially careful at high elevations or in areas with surfaces that reflect the sun's rays, such as sand, water, concrete, or

snow. Check online at www.epa.gov/sunwise/uvindex.html to determine the UV index in your area on any particular day.

Sunlamps, tanning beds, and tanning booths were once thought to be safer than the sun, because they give off mainly UVA rays. However, UVA rays are now known to cause serious skin damage and exposure to UVA rays does increase the risk of developing melanoma. Health experts strongly advise people not to use these tanning devices.

People with fair skin, blond, red or light-brown hair, and blue or light-colored eyes are at greatest risk for developing skin cancer. So are people with many large skin **moles**. These people should avoid exposure to the sun as much as possible. However, even dark-skinned people, including African Americans and Hispanic Americans, may suffer skin damage from the sun and should be careful about exposure.

Other groups of people who should minimize sun exposure are those who have had organ transplants or recent **plastic surgery**. Patients who have received organ transplants have a greatly increased risk of developing skin cancer, and the facial skin of people who have had face lifts or similar plastic surgery procedures sunburns more easily than intact skin.

Sunscreens should not be used on infants under six months of age because of the risk of side effects. Instead, children this young should be kept out of the sun. Children over six months should be protected with clothing and sunscreens of at least SPF 15, preferably lotions. Sunscreens containing alcohol should not be used on children because they may irritate the skin.

Older people who stay out of the sun and use sunscreens may not produce enough vitamin D in their bodies. They may need to increase the vitamin D in their **diets** by including foods such as fortified milk and salmon. A health care professional can help decide if this precaution is necessary.

Anyone who has had unusual reactions to any sunscreen ingredients in the past should check with a physician or pharmacist before using a sunscreen. The physician or pharmacist should also be told about any **allergies** to foods, dyes, preservatives, or other substances, especially the following:

- artificial sweeteners
- anesthetics such as benzocaine, procaine, or tetracaine
- diabetes medicine taken by mouth
- hair dyes
- sulfa medicines
- water pills
- cinnamon flavoring

People with skin conditions or diseases should check with their physicians before using a sunscreen. This is especially true of people with conditions that get worse with exposure to light.

Side effects

The most common side effects are drying or tightening of the skin. This problem does not need medical attention unless it does not improve.

Other side effects are rare, but possible. If any of the following symptoms occur, check with a physician as soon as possible:

- acne
- burning, itching, or stinging of the skin
- redness or swelling of the skin
- rash, with or without blisters that ooze and become crusted
- pain in hairy parts of body
- pus in hair follicles

Interactions

Anyone who is using a prescription or nonprescription (over-the-counter) drug that is applied to the skin should check with a physician before using a sunscreen.

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ORGANIZATIONS

American Academy of Dermatology (AAD), PO Box 4014, Schaumburg, IL, 60168–4014, (866) 503-7546, <http://www.aad.org>.

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Sunstroke see **Heat disorders**

Superficial phlebitis see **Thrombophlebitis**

Superior vena cava syndrome

Definition

The superior vena cava is the major vein in the chest that carries blood from the upper part of the body in to the heart. A restriction of the blood flow (occlusion) through this vein can cause superior vena cava syndrome (SVCS).

Description

Superior vena cava syndrome is a partial occlusion of the superior vena cava. This leads to a lower than normal blood flow through this major vein. SVCS is also called superior mediastinal syndrome and/or superior vena cava obstruction.

Causes and symptoms

More than 95% of all cases of SVCS are associated with cancers involving the upper chest. The cancers most commonly associated with SVCS are advanced lung cancers, which account for nearly 80% of all cases of SVCS, and lymphoma. Cancers that have spread (metastasized) to the chest, such as metastatic **breast cancer** to the chest and metastatic **testicular cancer** to the chest have also been shown to cause SVCS.

Other causes of SVCS include: the formation of a blood clot in the superior vena cava, enlargement of the thyroid gland, **tuberculosis**, and **sarcoidosis**.

The symptoms of SVCS include:

- change in voice
- confusion
- cough
- enlargement of the veins in the upper body, particularly those in the arms
- headache

KEY TERMS

Metastasis—The spread of a cancer from one part of the body (where the cancer originated) to another part of the body.

Sarcoidosis—A disease of unknown origin in which there is chronic (recurrent) swelling in the lymph nodes and other tissues.

Superior vena cava—The major vein that carries blood from the upper body to the heart.

Thymoma—A tumor that originates in the thymus, a small gland just in front of the heart that produces hormones necessary for the development of certain components of the immune system.

- light-headedness
- shortness of breath
- swelling of the arms
- swelling of the face
- trouble swallowing

Diagnosis

SVCS should be considered in any **cancer** patient with swelling of the face and arms. This diagnosis can be confirmed by x ray, computerized tomography (CT) scan, or medical resonance imaging (MRI) of the chest that reveals a partial occlusion of the superior vena cava.

Treatment

Treatment of SVCS depends on the underlying cancer that is causing it. This treatment may include radiation, **chemotherapy**, or a combination of both. In some cases, surgical procedures may be performed to open (dilate) the vessel. These procedures are generally performed by a trained radiologist or vascular surgeon.

Alternative treatment

Since treatment of SVCS is aimed at treating the underlying disorder that is causing SVCS, alternative treatments must also focus on treating these underlying causes. Alternative treatments for cancer include **acupuncture**, **aromatherapy**, herbal remedies, **hydrotherapy**, hypnosis, and massage, among many others.

Prognosis

The prognosis depends on the underlying cause of SVCS. In cases of SVCS caused by lung cancers, the prognosis is generally rather poor since SVCS does not generally occur until the later stages of these diseases.

Prevention

SVCS may be prevented by early medical intervention to halt and/or reverse the cancer which, in a later stage, would have lead to SVCS.

Resources

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Beeson, Michael S. *eMedicine - Superior Vena Cava Syndrome*. May 12, 2001. (Accessed December 20, 2010) <http://www.emedicine.com/emerg/topic561.htm>.

ORGANIZATIONS

Lung Cancer Alliance, 888 16th St, NW, Suite 140, Washington, DC, 20006, (202) 463-2080, (800) 298-2436, info@lungcanceralliance.org, <http://www.lungcanceralliance.org>.

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Supportive cancer therapy see **Cancer therapy, supportive**

Surfactant

Definition

Surfactant is a complex naturally occurring substance made of six lipids (fats) and four proteins that is produced in the lungs. It can also be manufactured synthetically.

Purpose

Surfactant reduces the surface tension of fluid in the lungs and helps make the small air sacs in the lungs (alveoli) more stable. This keeps them from collapsing when an individual exhales. In preparation for breathing air, fetuses begin making surfactant while still in the womb. Babies that are born very prematurely often lack adequate surfactant and must receive surfactant replacement therapy immediately after birth in order to breathe.

Precautions

Babies are considered premature if they are born before 37 weeks gestation. Fetuses begin to produce surfactant between weeks 24 and 28. By about 35 weeks, most babies have enough naturally produced surfactant to keep the alveoli from collapsing. Babies born before 35 weeks, especially those born very prematurely (before 30 weeks), are likely to need surfactant replacement therapy. Over half the babies born before 28 weeks gestation need this treatment, while about one-third born between 32 and 36 weeks need supplemental surfactant. Some very premature infants may also need to be placed on a mechanical ventilator.

Description

The lungs consist of spongy tissue filled with air spaces called alveoli. In the alveoli, oxygen is taken up by the blood and carbon dioxide, a waste product of cellular metabolism, is released and exhaled. For efficient oxygen-carbon dioxide exchange to occur, the surface area of the alveoli must be as large as possible. Under normal conditions, when a person exhales, the alveoli would collapse into each other and form larger air sacs with less surface area. Surfactant prevents this collapse by reducing the surface tension of the fluids that line the lungs and helping to equalize the pressures between large and small air spaces.

Surface tension is a measure of the attraction molecules of a fluid have for each other. The attractive force pulls fluids into a shape with the smallest surface area. This is why a drop of water on a flat surface is rounded rather than flat. If the surface tension is lowered, the attraction among molecules of the fluid is decreased and the surface area of the fluid increases. For example, if a drop of detergent is added to a drop of water, the detergent reduces the surface tension and the drop of water flattens out.

In the lungs, surfactant reduces the surface tension and helps to maximize the surface area available for gas exchange. Without adequate surfactant, a baby works much harder to breathe, becomes exhausted, and does not get enough oxygen. Babies that do not have enough surfactant to breathe normally at birth are said to have infant **respiratory distress syndrome** (RDS) or hyaline membrane disease (HMD).

Babies with RDS are given replacement surfactant as soon as possible within the first six hours after birth. Manufactured surfactant is a white powder that is mixed with sterile water. It is given through a breathing tube (endotracheal tube) that is inserted in the baby's lungs. Multiple doses are usually required.

KEY TERMS

Alveolus (plural alveoli)—The terminal air sacs of the lungs where gas exchange occurs.

Hyaline membrane—A thin layer of cells that line the lung.

Surface tension—The attraction of molecules in a fluid for each other.

Surfactant replacement therapy continues until the baby's lungs have matured enough to make surfactant on their own. Some very premature babies are also put on mechanical respirators to help them breathe. Surfactant replacement therapy has reduced deaths due to respiratory distress by 50% since the early 1990s. This therapy is expensive, but it is normally covered by insurance.

Preparation

The administration of surfactant is often a neonatal emergency. The only way to prevent the need for surfactant replacement therapy is to prevent a premature birth. Mothers who are at known high risk to deliver prematurely are given drugs called **corticosteroids** toward the end of the **pregnancy** that stimulate the lungs of the fetus to mature and begin producing surfactant sooner. This helps reduce the need for surfactant replacement therapy. Although babies of all races may be born prematurely, **prematurity** is more common if the mother is diabetic, is carrying multiple fetuses, or has delivered a previous premature baby. The decision to use surfactant replacement therapy is based on the condition of the baby, its blood oxygen level, and degree of respiratory distress.

Aftercare

Babies receiving surfactant therapy are normally cared for by a neonatologist, a pediatrician that specializes in newborn care. Premature newborns often have other health problems in addition to RDS. Aftercare varies depending on their other health risks.

Risks

Delivery of surfactant requires inserting a breathing tube into the baby's lungs. Complications of this therapy include air leaking into the area between the chest wall and the lungs and air leaking into the sac around the heart. Some infants also develop chronic lung disease.

Normal results

Normally surfactant replacement therapy keeps the infant alive until the lungs start producing their own surfactant.

Abnormal results

Surfactant replacement therapy is very effective if begun within six hours after birth. When it fails, **death** may result.

Resources

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Surgical debridement see **Debridement**

Swallowing disorders

Definition

Swallowing disorders include a number of diseases and conditions that cause difficulty in passing food or liquid from the mouth to the stomach.

Demographics

Each year, about 10 million people in the United States require medical evaluation for swallowing problems. Some experts say that about 10% of Americans develop symptoms of swallowing disorders in adulthood. Elderly people are the most likely to have problems with swallowing.

Description

Although swallowing is normally automatic and unconscious on the part of an individual, it is a complex

process involving several phases and 29 muscles. Saliva helps soften food as it is chewed. The tongue helps move food to the back of the mouth, triggering a swallowing reflex that passes food through the pharynx. The epiglottis helps keep food from mistakenly going down the windpipe and directs it instead into the esophagus, which is the canal that carries food to the stomach. Swallowing disorders can occur at any phase in the swallowing process. The medical term for difficulty swallowing is dysphagia.

Causes and symptoms

Swallowing disorders often result from other conditions and diseases. For example, Parkinson's disease, **cerebral palsy**, **stroke**, **head injury**, and other central nervous system conditions can damage the muscles and nerves involved in swallowing. Some people are born with abnormalities in the swallowing structures, such as infants born with **cleft palate**.

Some cancers can lead to swallowing disorders. **Esophageal cancer** can cause narrowing and eventual blockage of the esophagus. Surgery and **radiation therapy** for **head and neck cancer** can restrict or weaken tongue motion, paralyze vocal cords, or cause muscle damage that affects swallowing. An inflamed esophagus, often resulting from **gastroesophageal reflux disease** (GERD), can cause painful or difficulty swallowing. Infections of the esophagus also can inflame it and cause it to narrow. Swallowing difficulty may result from **aging**, although researchers are not certain why.

The most common symptoms people report are **choking** and the feeling that food feels stuck in the throat. Other symptoms include needing to swallow many times to clear food from the mouth and throat, a "gurgly" wet sound to the voice after swallowing, having to clear the throat after eating, coughing, **pain** while swallowing, bringing food back up (regurgitation), food or acid backing up into the throat, unexpected weight loss, and not being able to swallow at all. Children also may gag during meals and may have excessive drooling or leaking of food or liquid from their mouths during meals. They may have difficulty breathing when eating or drinking, spit up frequently, and lag behind in weight gain. They also may have recurring **pneumonia** or respiratory infections.

Diagnosis

A physician should perform a full head and neck examination based on the patient's symptoms. Speech-language pathologists may aid in the diagnosis. Physicians also might order a swallowing test to study how the patient swallows. The patient will be asked to drink a

liquid with a contrast agent called barium that will show up on x rays of the throat and upper chest. The exam might be imaged with a technique called video fluoroscopy, which will take motion camera images in addition to still images. For this exam, the patient may be asked to swallow liquid, paste, and solids. A speech pathologist may work with the radiologist to perform this exam.

If the physician thinks the problem originates in the lower esophagus or has concerns about an abnormality in the esophagus, an **endoscopy** may be ordered. This test involves passing a thin, flexible instrument called an endoscope down the throat. The lighted endoscope helps the physician view the esophagus. Other tests may be used, including ultrasound.

Treatment

Treatment will depend on the cause of the swallowing problem. Special exercises may help strengthen the muscles used for chewing and swallowing. Problems originating in the mouth may be treated with artificial saliva, improved hydration, or better dental care. Esophageal problems will be treated depending on the cause. Patients with GERD will receive medications and instructions on how to better manage the disease. Esophageal **cancer** is a life-threatening disease that will involve coordinating care with an oncologist. Many patients will receive help with their disorders from speech pathologists. Special liquid **diets** may be ordered for patients who continue to have trouble chewing or swallowing. In severe cases, the patient may need a feeding tube that bypasses the part of the swallowing system that does not work.

Alternative treatment

Some herbs that may help improve swallowing include oil of peppermint and licorice. Valerian may be used as a tea. Homeopathic physicians may suggest some remedies aimed at improving bloating, **indigestion**, or **cough**. Alternative care should be sought from licensed practitioners and coordinated with physician care.

Prognosis

In many cases, these disorders can be corrected. If not treated, swallowing disorders can lead to serious complications, including **dehydration** and **malnutrition**. There also is a risk of food entering the airway (aspiration) as a person attempts to swallow, which can lead to aspiration pneumonia as food particles enter the lungs.

KEY TERMS

Cleft palate—An opening or hole in the roof of the mouth that occurs at birth when the roof fails to fully develop in the infant.

Epiglottis—A thin layer of cartilage behind the tongue that helps block food from entering the windpipe.

Pharynx—The muscular cavity that leads from the mouth and nasal passages to the larynx and esophagus.

Prevention

Many causes of swallowing disorders cannot be prevented. Slowly and fully chewing food helps. People with GERD should manage it to lower the risk of developing swallowing difficulties.

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American Academy of Otolaryngology—Head and Neck Surgery, 1650 Diagonal Rd., Alexandria, VA, 22314–2857, (703) 836-4444, <http://www.entnet.org>.

American Speech-Language Association (ASHA), 2200 Research Blvd., Rockville, MD, 20850-3289, (800) 638-8255, <http://www.asha.org>.

National Institute of Dental and Craniofacial Research (NIDCR), 45 Center Dr., Room 4AS19 MSC 6400, Bethesda, MD, 20892-6400, (301) 496-4261, <http://www.nidr.nih.gov>.
 National Institute of Neurological Disorders and Stroke (NINDS), PO Box 5801, Bethesda, MD, 20824, (800) 352-9424, <http://www.ninds.nih.gov/>.
 National Institutes of Health (NIH), 9000 Rockville Pike, Bethesda, MD, 20892, (301) 496-4000, <http://www.nih.gov>.
 National Library of Medicine, 8600 Rockville Pike, Bethesda, MD, 20894, <http://www.nlm.nih.gov>.
 National Stroke Association (NSA), 9707 E Easter Ln., Building B, Centennial, CO, 80112, (800) 787-6537, <http://www.stroke.org>.

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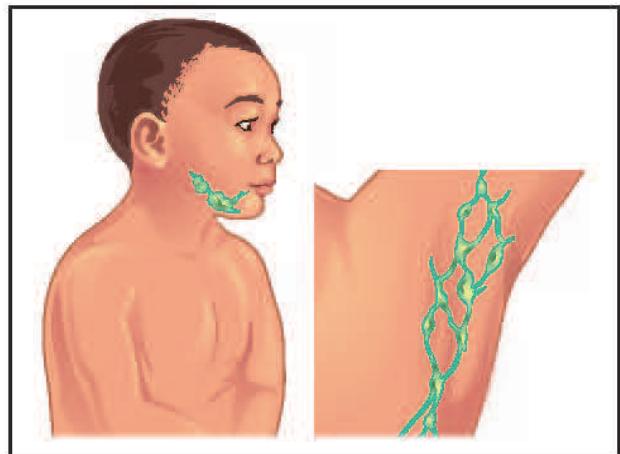
Swan-Ganz catheterization see **Pulmonary artery catheterization**

Sweating, excessive see **Hyperhidrosis**

Swimmer's ear see **Otitis externa**

Swimming pool conjunctivitis see **Inclusion conjunctivitis**

Swine flu see **H1N1 influenza**



Two common locations of swollen glands are in the neck and the armpit. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

cells that produce antibodies that recognize and bind to bacteria, viruses, and other foreign substances. Macrophages destroy these foreign invaders. There are about 600 lymph nodes located throughout the body, with the majority located in the head and neck region. Lymph glands occur in groups and filter the lymphatic fluid that drains from specific regions of the body, trapping and destroying bacteria, viruses, and other potentially harmful substances. They also help maintain the body's fluid balance.

Lymph glands in children are normally about 0.4 in (1 cm) in diameter. They cannot be felt with the fingers unless they are swollen. The lymph glands that most often swell are those in the neck, under the jaw and chin, behind the ears, on the back of the head, and in the armpits and groin region. There are chains of lymph nodes on both sides of the front and back of the neck, as well as on each side of the neck. Swollen glands usually indicate that the numbers of lymphocytes in the glands have increased to produce more antibodies for fighting an infection.

Swollen glands

Definition

Swollen glands are enlarged lymph nodes. Lymph nodes are an integral part of the immune system and, although swollen glands can have many causes, they most often indicate that the body is fighting an infection. Swollen glands are also known as **lymphadenitis** or lymphadenopathy.

Demographics

Swollen glands are very common. They are one of the most frequent reasons for medical visits, especially by children. Swollen glands, especially in the neck region, are more common in children than in adults because children tend to have more viral infections than adults.

Description

Lymph glands or nodes are small, round or bean-shaped clusters of immune system cells called lymphocytes and macrophages. Lymphocytes are white blood

Risk factors

Infection is the major risk factor for swollen glands. However, many other diseases and conditions can put a child at risk for swollen glands.

Causes and symptoms

There are numerous causes of swollen glands. An infection—especially a viral infection such as the common cold—is by far the most frequent cause. Infections that can cause swollen glands include:

- viral infections, such as flu, mononucleosis (mono), chicken pox, measles, rubella, mumps, and cytomegalovirus (CMV)
- bacterial infections, such as strep throat caused by the *Streptococcus pyogenes* bacterium, Lyme disease spread by ticks, tuberculosis (TB), and cat scratch fever
- viral and bacterial sexually transmitted infections (STIs), including syphilis
- parasitic infections such as toxoplasmosis
- mouth sores
- gingivitis, an infection of the gums
- an infected (abscessed or impacted) tooth
- skin infections
- ear infections
- tonsillitis
- infections from cuts, wounds, or animal or insect bites or stings
- a boil or abscess from an infected hair follicle or sweat gland

Other causes of swollen glands include:

- various cancers, especially leukemia, Hodgkin's disease, non-Hodgkin's lymphoma, and breast cancer
- cancers that have spread (metastasized) to lymph nodes
- immune system disorders, such as lupus, rheumatoid arthritis, and HIV/AIDS
- side effects of vaccines, such as immunizations against malaria or typhoid
- side effects of certain medications, such as phenytoin (Dilantin)

The location and type of infection or other condition determine which glands swell. Infections or tumors in the head, mouth, or neck are particularly likely to cause swollen glands:

- A sore throat often causes glands in the neck to swell.
- A tooth, gum, or other mouth infection may cause swollen glands in the jaw or neck.
- Swollen glands at the base of the neck and above the collarbone are indicative of a chest infection or, rarely, a tumor in the chest.
- An infection on the arm may cause glands in the armpit to swell.
- An infected animal bite can cause swollen glands above the wound.

Swollen glands that are soft, tender, and move easily are usually signs of infection or inflammation. Serious infections can cause swollen glands to become very hard and tender. Sudden, painful swelling is

usually caused by an injury or early-stage infection. Swollen glands that are caused by **cancer** or a tumor tend to develop gradually and painlessly and remain swollen. They usually do not have other signs of inflammation, such as redness or tenderness. Swelling of glands throughout the body can be caused by a systemic infection, such as mononucleosis or HIV/AIDS, or an immune disorder, such as lupus or **rheumatoid arthritis**.

Swollen glands may be two or three times their normal size and can be readily felt with the fingers. Sometimes the swelling can be seen through the skin. A child's lymph node is considered to be enlarged if it is more than 0.4 in (1 cm) in diameter. Other symptoms of swollen glands include:

- a lump
- tenderness or pain when a gland is pressed
- red, warm, swollen skin over a lymph node
- symptoms of an underlying infection, such as fever, runny nose, mouth sores, or a sore throat
- a swollen limb, which can indicate an enlarged lymph node that is causing a blockage in the lymph system, but is too deep inside the body to feel through the skin

A child with swollen glands should be seen by a pediatrician if:

- the swelling and tenderness last for more than five days
- glands throughout the body appear swollen
- glands enlarge rapidly
- skin over a swollen gland turns red or purple
- the child has a fever above 101°F (38.3°C)
- the child is tired or lethargic or has no appetite

Diagnosis

Examination

During an examination, the healthcare provider feels all of the palpable lymph nodes—those that can be felt through the skin. The lymph glands are examined for size, texture, tenderness, warmth, and firmness. The location of the swollen glands may help diagnose the underlying cause. The **physical examination** includes any other symptoms that are present. Sometimes it is possible to see signs of infection or injury near the swollen gland. The healthcare provider will take a medical history that includes any exposure to infectious agents, recent injuries, and medications.

KEY TERMS

Antibody—A specific protein produced by the immune system in response to a specific foreign protein called an antigen.

Lymph nodes—Lymph glands; small, round or pea-shaped tissue masses that are distributed along the lymphatic vessels and contain lymphocytes that filter the lymphatic fluid.

Lymphatic system—Consists of lymphatic fluid and vessels, lymph nodes, lymphocytes, and the thymus, spleen, tonsils, and bone marrow. The part of the circulatory system that is responsible for immune responses, that scavenges fluids and proteins and returns them to the blood, and that removes debris and foreign substances.

Lymphocyte—A type of white blood cell that functions in the immune response, including antibody-producing B cells and T cells.

Macrophage—A type of immune system cell that engulfs and destroys antigens and presents them to other immune system cells.

Tests

Infections may require growing (culturing) a sample of a bodily fluid or secretion to identify the causative agent and to select an appropriate antibiotic in the case of a bacterial infection. Sometimes an infected lymph node is drained and cultured. A **tuberculosis** skin test or specific tests to diagnose mononucleosis may be required.

Blood tests to help diagnose the underlying cause of swollen glands may include:

- a complete blood count (CBC)
- a blood differential, which measures the percentages of each type of white blood cell
- liver function tests
- kidney function tests

Procedures

Procedures for diagnosing the underlying cause of swollen glands may include a:

- chest x ray
- liver-spleen scan
- computerized tomography (CT) scan of the affected area

Sometimes a **lymph node biopsy** is necessary for diagnosis, especially for diagnosis of a tumor or a fungal infection. A fine-needle aspiration (FNA), in which a thin, hollow needle is inserted into the node to remove or aspirate cells, may be performed in the physician's office or by a surgeon or radiologist. Ultrasound may be used to guide the needle. A surgical biopsy removes some or all of a lymph node through an incision and is performed under local or **general anesthesia**. The cells or tissue are sent to a laboratory for examination under a microscope.

Treatment

Traditional

Swollen glands most often return to normal on their own once the underlying infection is resolved. Likewise, swollen glands caused by a **vaccination** usually resolve on their own. Swelling caused by a medication may require changing the dose or type of drug. If the gland itself is infected, surgical drainage may be required. Swollen glands caused by an underlying condition such as cancer or an immune system disorder require treatment of the underlying disease.

Drugs

Swollen glands can be treated with over-the-counter **pain** relievers, such as **acetaminophen** (Tylenol) or a nonsteroidal anti-inflammatory drug (NSAID), such as ibuprofen (Advil, Motrin) or naproxen (Aleve). Children should not be given **aspirin** without consulting a physician, due to the unlikely but serious risk of developing **Reye's syndrome**. Swollen glands caused by a bacterial infection are usually treated with **antibiotics**.

Alternative

As with traditional treatments, alternative treatments for swollen glands depend on the cause. For example, many alternative practitioners recommend vitamin C and zinc or herbal remedies, such as *Echinacea* spp., for treating common colds.

Home remedies

A tender or painful swollen gland can be treated with warm, wet compresses. A washcloth wrung out with warm water should be applied for 20–30 minutes, three or four times per day. The heat and moisture can relieve discomfort, increase circulation to the area, localize the infection, and encourage healing. If possible, the gland should be exposed to the air. Swollen glands should not be rubbed, squeezed, scratched, or otherwise irritated. Bed rest also can be helpful for treating swollen glands.

Prognosis

Swollen glands are rarely serious. Soreness in swollen glands usually disappears within a couple of days without treatment. Swollen glands often return to their normal size as soon as an infection, such as a cold, has resolved. However, it may be several weeks after an infection has cleared before the glands return to their normal size. When swollen glands result from a serious infection or other underlying condition, they may remain enlarged for a long period.

Prevention

Although there are no specific preventative measures for the less frequent causes of swollen glands, there are measures that can help prevent swollen glands caused by infection. Frequent and thorough hand washing, especially during cold and flu season, is one of the best ways to prevent infection. Proper cleaning of **wounds** and prompt treatment with antibiotics, if required, can prevent swollen glands. Measures for preventing skin infections include:

- keeping the skin clean with mild soap or cleanser and lukewarm water
- rinsing thoroughly and gently patting the skin dry after washing
- avoiding irritating skin products
- using water-based and oil-free or hypoallergenic skincare products that do not clog pores
- washing as soon as possible after sweating
- wearing soft, cotton clothing or moleskin under sports equipment to avoid irritation
- avoiding squeezing, scratching, draining, or puncturing swollen glands, so as not to inflame or irritate the glands or push infection deeper into the skin

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ORGANIZATIONS

American Academy of Family Physicians, 11400 Tomahawk Creek Parkway, Leawood, KS, 66211-2680, (913) 906-6000, (800) 274-6000, (913) 906-6075, <http://www.aafp.org>.

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Margaret Alic, PhD

Sydenham's chorea

Definition

Sydenham's chorea is an acute but self-limited movement disorder that occurs most commonly in children between the ages of 5 and 15, and occasionally in pregnant women. It is closely associated with **rheumatic fever** following a throat infection. The disorder is named for Thomas Sydenham (1624–1689), an English doctor who first described it in 1686. Other names for Sydenham's chorea include simple chorea, chorea minor, acute chorea, rheumatic chorea, juvenile chorea, and St. Vitus' dance. The English word "chorea" itself comes from the Greek word *choreia*, which means "dance." The disorder takes its name from the rapid involuntary jerking or twitching movements of the patient's face, limbs, and upper body.

Description

Sydenham's chorea is a disorder that occurs in children and is associated with **rheumatic fever**. Rheumatic fever is an acute **infectious disease** caused by certain types of streptococci bacteria. It usually starts with **strep throat** or **tonsillitis**. These types of streptococci are able to cause disease throughout the body. The most serious damage caused by rheumatic fever is to the valves in the heart. At one time, rheumatic fever

was the most common cause of damaged heart valves, and it still is in most developing countries around the world. Rheumatic fever and rheumatic heart disease are still present in the industrialized countries, but the incidence has dropped substantially.

Both acute rheumatic fever and Sydenham's chorea are relatively uncommon disorders in the United States. According to the Centers for Disease Control and Prevention (CDC), only 1–3% of people with streptococcal throat infections develop acute rheumatic fever (ARF); thus the incidence of ARF in the United States is thought to be about 0.5 per 100,000 patients between 5 and 17 years of age.

With regard to age, the incidence of Sydenham's chorea is higher in childhood and adolescence than in adult life. It occurs more frequently in females than in males; the gender ratio is thought to be about 2 F: 1 M. Since the peak incidence of rheumatic fever in North America occurs in late winter and spring, Sydenham's chorea is more likely to occur in the summer and early fall. There is no evidence that the disorder selectively affects specific racial or ethnic groups.

Rheumatic fever may appear in several different forms. Sydenham's chorea is one of five major criteria for the diagnosis of rheumatic fever. There are also four minor criteria and two types of laboratory tests associated with the disease. The "Jones criteria" define the diagnosis. They require laboratory evidence of a streptococcal infection plus two or more of the criteria. The laboratory evidence may be identification of streptococci from a **sore throat** or antibodies to streptococcus in the blood. The most common criteria are arthritis and heart disease, occurring in half to three-quarters of the patients. Sydenham's chorea, characteristic nodules under the skin, and a specific type of skin rash occur only 10% of the time.

About 20% of patients diagnosed with Sydenham's chorea experience a recurrence of the disorder, usually within two years of the first episode. Most women who develop Sydenham's during **pregnancy** have a history of acute rheumatic fever in childhood or of using birth control pills containing estrogen.

Causes and symptoms

Sydenham's is caused by certain types of streptococci called Group A beta-hemolytic streptococci or GAS bacteria. In general, streptococci are spherical-shaped anaerobic bacteria that occur in pairs or chains. GAS bacteria belong to a subcategory known as pyogenic streptococci, which means that the infections they cause produce pus. These particular germs seem to be able to create an immune response that attacks the

body's own tissues along with the germs. Those tissues are joints, heart valves, skin, and brain.

The initial throat infection that leads to Sydenham's chorea is typically followed by a symptom-free period of 1–5 weeks. The patient then develops an acute case of rheumatic fever (ARF), an inflammatory disease that affects multiple organ systems and tissues of the body. In most patients, ARF is characterized by fever, arthritis in one or more joints, and carditis, or inflammation of the heart. In about 20% of patients, however, Sydenham's chorea is the only indication of ARF. Sydenham's is considered a delayed complication of rheumatic fever; it may begin as late as 12 months after the initial sore throat, and it may start only after the patient's temperature and other physical signs have returned to normal. The average time interval between the pharyngitis and the first symptoms of Sydenham's, however, is eight or nine weeks.

It is difficult to describe a "typical" case of Sydenham's chorea because the symptoms vary in speed of onset as well as severity. Most patients have an acute onset of the disorder, but in others, the onset is insidious, which means that the symptoms develop slowly and gradually. In some cases, the child's physical symptoms are present for 4–5 weeks before they become severe enough for the parents to consult a doctor. In other cases, emotional or psychiatric symptoms precede the clumsiness and involuntary muscular movements that characterize the disorder. The psychiatric symptoms that may develop in patients with Sydenham's chorea are one reason why it is sometimes categorized as a PANDAS disorder. PANDAS stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections.

Behavioral or emotional disturbances that have been observed with Sydenham's include:

- frequent mood changes
- episodes of uncontrollable crying
- behavioral regression; that is, acting like much younger children
- mental confusion
- general irritability
- difficulty concentrating
- impulsive behavior

Some researchers think that children who have had Sydenham's are at increased risk of developing **obsessive-compulsive disorder** (OCD). OCD is characterized by obsessions, which are unwanted recurrent thoughts, images, or impulses, and by compulsions, which are repetitive rituals, mental acts, or behaviors. Obsessions in children often take the form of fears of intruders or

KEY TERMS

Arthralgia—Joint pain.

Chorea—A term that is used to refer to rapid, jerky, involuntary movements of the limbs or face that characterize several different disorders of the nervous system, including chorea of pregnancy and Huntington's chorea as well as Sydenham's chorea.

Electrocardiogram—Mapping the electrical activity of the heart.

Insidious—Developing in a stealthy or gradual manner. Sydenham's chorea may have an insidious onset.

PANDAS disorders—A group of childhood disorders associated with such streptococcal infections as scarlet fever and "strep throat." The acronym stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections. Sydenham's chorea is considered a PANDAS disorder.

Pharyngitis—Inflammation of the throat, accompanied by dryness and pain. Pharyngitis caused by a

streptococcal infection is the usual trigger of Sydenham's chorea.

Rheumatic fever—Chiefly childhood disease marked by fever, inflammation, joint pain, and Sydenham's chorea. It is often recurrent and can lead to heart valve damage.

St. Vitus' dance—Another name for Sydenham's chorea. St. Vitus was a fourth-century martyr who became the patron saint of dancers and actors during the Middle Ages. He was also invoked for protection against nervous disorders, epilepsy, and the disease that bears his name.

Streptococcus (plural, streptococci)—A genus of spherical-shaped anaerobic bacteria occurring in pairs or chains. Sydenham's chorea is considered a complication of a streptococcal throat infection.

Tonsillitis—Inflammation of the tonsils, which are in the back of the throat.

harm coming to a family member. Compulsions may include such acts as counting silently, washing the hands over and over, insisting on keeping items in a specific order, checking repeatedly to make sure a door is locked, and similar behaviors.

Diagnosis

Because rheumatic fever is such a damaging disease, a complete evaluation should be done whenever it is suspected. This includes cultures for streptococci, blood tests, and usually an electrocardiogram (heartbeat mapping to detect abnormalities).

The diagnosis of Sydenham's is also based on the doctor's observation of the patient's involuntary movements. Unlike tics, the movements associated with chorea are not repetitive; and unlike the behavior of hyperactive children, the movements are not intentional. The recent onset of the movements rules out a diagnosis of **cerebral palsy**. If the doctor suspects Sydenham's, he or she may ask the patient to stick out the tongue and keep it in that position, or to squeeze the doctor's hand. Many patients with Sydenham's cannot hold their mouth open and keep the tongue out for more than a second or two. Another characteristic of Sydenham's is an inability to grip with a steady pressure; when the patient squeezes the doctor's hand, the strength of the grip will increase and decrease in an erratic fashion. This characteristic is sometimes called the "milking sign."

Treatment

Suspected streptococcal infections must be treated. All the other manifestations of rheumatic fever, including Sydenham's chorea and excluding heart valve damage, remit with the acute disease and do not require treatment. Sydenham's chorea generally lasts for several months.

Most patients with Sydenham's chorea recover after a period of bed rest and temporary limitation of normal activities. In most cases the symptoms disappear gradually rather than stopping abruptly.

Most doctors recommend ongoing treatment with penicillin to prevent a recurrence of rheumatic fever or Sydenham's chorea, although there is some disagreement as to whether this treatment should continue for 5 years after an acute attack or for the rest of the patient's life. The penicillin may be given orally or by injection. Patients who cannot take penicillin may be given erythromycin or sulfadiazine.

Prognosis

Sydenham's chorea usually clears up without complications when the rheumatic fever is treated. The heart valve damage associated with rheumatic fever may lead to heart trouble and require a surgical valve repair or replacement.

In most cases of Sydenham's, the patient recovers completely, although a recurrence is possible. In a very few cases—about 1.5% of patients diagnosed with Sydenham's—there may be increasing muscle stiffness and loss of muscle tone resulting in disability. This condition is occasionally referred to as paralytic chorea

Prevention

All cases of strep throat in children should be treated with a full 10 days of **antibiotics** (penicillin or erythromycin). Treatment may best be delayed a day or two to allow the body to build up its own antibodies. In addition, for those who have had an episode of rheumatic fever or have damaged heart valves from any other cause, prophylactic antibiotics should be continued to prevent recurrence.

It is possible to eradicate dangerous GAS bacteria from a community by culturing everyone's throat and treating everyone who tests positive. This is worth doing wherever a case of rheumatic fever appears, but it is expensive and requires many resources.

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National Institute of Neurological Disorders and Stroke (NINDS), NIH Neurological Institute, P. O. Box 5801, Bethesda, MD, 20824, (301) 496-5751, (800) 352-9424, <http://www.ninds.nih.gov/>.

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Sympathectomy

Definition

Sympathectomy is a surgical procedure that destroys nerves in the sympathetic nervous system. The procedure is done to increase blood flow and decrease long-term **pain** in certain diseases that cause narrowed blood vessels. It can also be used to decrease excessive sweating. This surgical procedure cuts or destroys the sympathetic ganglia, collections of nerve cell bodies in clusters along the thoracic or lumbar spinal cord.

Purpose

The autonomic nervous system that controls unwilled (involuntary) body functions, such as breathing, sweating, and blood pressure, are divided into the sympathetic and the parasympathetic nervous systems. The sympathetic nervous system speeds the heart rate, narrows (constricts) blood vessels, and raises blood pressure. Blood pressure is controlled by means of nerve cells that run through sheaths around the arteries. The sympathetic nervous system can be described as the "fight or flight" system because it allows us to respond to danger by fighting off an attacker or by running away. When danger threatens, the sympathetic nervous system increases heart and respiratory rate, increases blood flow to muscles, and decreases blood flow to other areas, such as skin, digestive tract, and limb veins. The net effect is an increase in blood pressure.

Sympathectomy is performed to relieve intermittent constricting of blood vessels (**ischemia**) when the fingers, toes, ears, or nose are exposed to cold (Raynaud's phenomenon). In Raynaud's phenomenon, the affected extremities turn white, then blue, and red as the blood

supply is cut off. The color changes are accompanied by **numbness**, **tingling**, burning, and pain. Normal color and feeling are restored when heat is applied. The condition sometimes occurs without direct cause but it is more often caused by an underlying medical condition, such as **rheumatoid arthritis**. Sympathectomy is usually less effective when Raynaud's is caused by an underlying medical condition. Narrowed blood vessels in the legs that cause painful cramping (claudication) are also treated with sympathectomy.

Sympathectomy may be helpful in treating **reflex sympathetic dystrophy** (RSD), a condition that sometimes develops after injury. In RSD, the affected limb is painful (causalgia) and swollen. The color, temperature, and texture of the skin change. These symptoms are related to prolonged and excessive activity of the sympathetic nervous system.

Because sweating is controlled by the sympathetic nervous system, sympathectomy is also effective in treating excessive sweating (**hyperhidrosis**) of the palms, armpits, or face.

Precautions

To determine whether sympathectomy is needed, a reversible block of the affected nerve cell (**ganglion**) should be done. A reversible ganglion block interrupts nerve impulses by means of steroid and anesthetic injected into it. If the block has a positive effect on pain and blood flow in the affected area, the sympathectomy will probably be helpful. The surgical procedure should be performed only if conservative treatment has not worked. Conservative treatment includes avoiding exposure to **stress** and cold, **physical therapy**, and medications.

Sympathectomy is most likely to be effective in relieving the pain of reflex sympathetic dystrophy if it is done soon after the injury occurs. However, increased benefit from early surgery should be balanced against time needed to promote spontaneous recovery and response to conservative treatment.

Description

Sympathectomy was traditionally done as an inpatient surgical procedure under **general anesthesia**. An incision was made on the mid-back, exposing the ganglia to be cut. Recent techniques are less invasive and may be done under **local anesthesia** and as outpatient surgery. If only one arm or leg is affected, it may be treated with a percutaneous radiofrequency technique. In this technique, the surgeon locates the ganglia by a combination of x ray and electrical stimulation. The

ganglia are destroyed by applying radio waves through electrodes on the skin.

Sympathectomy for hyperhidrosis can be done by making a small incision under the armpit and introducing air into the chest cavity. The surgeon inserts a fiber optic tube (endoscope) that projects an image of the operation on a video screen. The ganglia can then be cut with fine scissors attached to the endoscope. Laser beams can also be used to destroy the ganglia.

Preparation

As with any surgery, patients should discuss expected results and possible risks with their surgeons. They should tell their surgeons all medications they are taking and all their medical problems, and they should be in good general health. To improve general health, the patient may be asked to lose weight, give up **smoking** or alcohol, and get the proper sleep, diet, and **exercise**. Immediately before the surgery, patients will not be permitted to eat or drink, and the surgical site will be cleaned and scrubbed.

Aftercare

The surgeon will inform the patient about specific aftercare needed for the technique used. **Doppler ultrasonography**, a test using sound waves to measure blood flow, can help to determine whether sympathectomy has had a positive result.

Risks

Side effects of sympathectomy may include decreased blood pressure while standing, which may cause **fainting** spells. After sympathectomy in men, semen is sometimes ejaculated into the bladder, which may impair fertility. After a sympathectomy done by inserting an endoscope in the chest cavity, patients may experience chest pain with deep breathing. This problem usually disappears within two weeks. They may also experience **pneumothorax** (air in the chest cavity).

In 30% of cases, surgery for hyperhidrosis may cause increased sweating on the chest. In 2% of cases, this surgery causes increased sweating in other areas, including increased facial sweating while eating. Other complications occur less frequently. These complications include Horner's syndrome, a condition of the nervous system that causes the pupil of the eye to close, the eyelid to droop, and sweating to decrease on one side of the face. Other rare complications are nasal blockage and pain of the nerves supplying the skin between the ribs.

KEY TERMS

Causalgia—A severe burning sensation sometimes accompanied by redness and inflammation of the skin. Causalgia is caused by injury to a nerve outside the spinal cord.

Claudication—Cramping or pain in a leg caused by poor blood circulation. This condition is frequently caused by hardening of the arteries (atherosclerosis). Intermittent claudication occurs only at certain times, usually after exercise, and is relieved by rest.

Fiberoptics—In medicine, fiberoptics uses glass or plastic fibers to transmit light through a specially designed tube. The tube is inserted into organs or body cavities where it transmits a magnified image of the internal body structures.

Hyperhidrosis—Excessive sweating. Hyperhidrosis can be caused by heat, overactive thyroid glands, strong emotion, menopause, or infection.

Parasympathetic nervous system—The division of the autonomic (involuntary or unwilled) nervous system that slows heart rate, increases digestive and gland activity, and relaxes the sphincter muscles that close off body organs.

Percutaneous—Performed through the skin, from the Latin *per*, meaning through and *cutis*, meaning skin.

Pneumothorax—A collection of air or gas in the chest cavity that causes a lung to collapse. Pneumothorax may be caused by an open chest wound that admits air.

Normal results

Some studies report that sympathectomy relieves causalgia in as many as 75% of cases. The studies also show that it relieves hyperhidrosis in more than 90% of cases. The less invasive procedures cause very little scarring. Most patients stay in the hospital for less than one day and return to work within the week.

Resources

OTHER

The American Institute for Hyperhidrosis Page. <http://www.handsweat.com>.

Laurie Barclay, MD

Syncope see **Fainting**

Syndactyly see **Polydactyly and syndactyly**

Synergistic gangrene see **Flesh-eating disease**

Synovial fluid analysis see **Joint fluid analysis**

Synovial membrane biopsy see **Joint biopsy**

transmitted from a mother to her child either before or during birth. Untreated syphilis is a systemic, potentially fatal disease that can cause permanent damage to the heart and central nervous system.

Demographics

Syphilis has been a serious public health problem since at least the sixteenth century. Some estimates place the number of worldwide syphilis cases at about 50 million annually. However the incidence of syphilis varies greatly from one region to another and even within small geographical areas.

There were almost 41,000 new cases of syphilis reported in the United States in 2007. This included 11,466 cases of primary and secondary syphilis, 10,768 cases of early-latent syphilis, 18,256 cases of late-latent syphilis, and 430 cases of congenital syphilis in newborns. This is the highest number of new cases since 1997, but far less than the more than 135,000 new cases in 1990 and the more than 575,000 new cases reported in 1943. In 2007 the incidence of syphilis in the United States was 13.7 per 100,000 people, compared with 447 per 100,000 in 1943. These dramatic decreases are attributable to vastly improved treatment and prevention, as well as increased public awareness. The South accounts for almost half of all syphilis cases in the United States and only a small number of counties and urban areas account for the vast majority of cases.

In the United States syphilis primarily affects people aged 20–39. The highest numbers of cases are in women aged 20–24 and in men aged 35–39. Syphilis rates are highest among black Americans. In 2007 males

Syphilis

Definition

Syphilis is a sexually transmitted disease (STD) caused by the spirochete bacterium *Treponema pallidum*. The infection is acquired through direct—usually sexual—contact with a syphilis sore. It also can be



This patient has secondary syphilis, evidenced by the appearance of lesions on the skin. (Custom Medical Stock Photo, Inc. Reproduced by permission.)

accounted for six times the number of new syphilis cases as women. Men who have sex with men (MSM) account for more than 60% of cases. The increase in syphilis cases attributable to MSM increased from 4% in 2000 to 65% in 2007. Syphilis has been closely associated with HIV infection since the late 1980s. Syphilis makes it easier to transmit and acquire HIV. The incidence of syphilis is also high among crack **cocaine** users.

Description

Although the origin of syphilis remains controversial, evidence suggests that *Treponema* bacteria were brought to Europe from the New World with the return of Christopher Columbus's ships. Syphilis, which is also called lues from a Latin word meaning **plague**, was treated with mercury and other dangerous substances until World War I, when more effective treatments based on arsenic or bismuth were introduced. After World War II penicillin became available to cure syphilis and the incidence of the disease began to decline.

About 90% of syphilis cases are contracted through sexual contact, usually from people who are unaware that they have the disease. Syphilis is sometimes called the "great imposter," because its symptoms—when present at all—resemble those of various other diseases.

Syphilis is transmitted through direct contact with a syphilis sore, usually through vaginal, anal, or oral sex. Sores usually occur on the external genitals or in the

vagina, anus, or rectum, but also can occur on the lips and in the mouth. The chances of contracting syphilis during unprotected sex with a person who has an early stage of the disease are 30–50%. The bacteria also can be transmitted by touching infected sores or using contaminated needles to inject drugs. Babies of infected mothers can be born with congenital syphilis. Transmission through a blood **transfusion** is very rare because the bacterium cannot survive for more than 24 hours in stored blood and blood products are screened for the bacteria. Syphilis cannot be spread through toilet seats, doorknobs, swimming pools, hot tubs, bathtubs, shared clothing, or eating utensils. *T. pallidum* is easily killed by heat and drying.

Syphilis has both acute and chronic phases that produce a wide variety of symptoms affecting most of the body's organ systems. The range of symptoms makes it easy to ignore early signs or to confuse it with less serious diseases. Syphilis that is acquired through sexual contact has four stages—primary, secondary, latent, and tertiary. The bacteria can be spread by sexual contact during the first three stages. Although latent syphilis has few external symptoms, the disease continues to progress. Patients with tertiary syphilis cannot infect others.

Syphilis can be transmitted from an infected mother to her fetus through the placenta at any time during **pregnancy** or through contact with syphilitic ulcers during the birth process. The chances of infection depend on the stage of the mother's disease. Almost all infants born to mothers with untreated primary or secondary syphilis will be infected. If a woman has untreated syphilis acquired within four years of a pregnancy, there is an 80% risk that the newborn will have congenital syphilis. However the infection rate drops to 40% if the mother's disease is in the early-latent stage and to 6–14% if she has late-latent syphilis.

Untreated syphilis can have devastating consequences:

- Syphilis appears to increase a man's risk of developing prostate cancer in later life.
- Cardiovascular syphilis occurs in 10–15% of patients with tertiary syphilis.
- About 8% of those with untreated syphilis develop neurosyphilis 5–35 years after the onset of the primary infection. This central nervous system disease has both physical and psychiatric consequences. It affects men more frequently than women and Caucasians more frequently than blacks.
- General paresis, also called dementia paralytica, results from neurosyphilis and is most common in patients over age 40.

Risk factors

In the United States and Canada, populations at high risk for syphilis include:

- sexually abused children
- sexually active teenagers
- MSM
- women of childbearing age
- prisoners
- abusers of drugs or alcohol
- prostitutes of either sex and their customers
- those infected with another sexually transmitted infection (STI), including HIV/AIDS

Drug **abuse** can increase the risk of syphilis because of needle sharing and the exchange of sex for drugs. In addition, people abusing drugs or alcohol are more likely to engage in risky sexual practices.

Causes and symptoms

T. pallidum is a thin, spiral- or coil-shaped bacterium that enters the body through mucous membranes or breaks in the skin to cause primary syphilis. The first signs of infection often go unnoticed. After an incubation period of 10–90 days chancres may develop. These are small blister-like sores about 0.5 in (13 mm) in size. They resemble the ulcers of chlamydia infection, **genital herpes**, or skin tumors. Most chancres are on the genitals, but they also can develop on the breasts or lips or in the mouth. Rectal chancres are common in MSM. Chancres in a woman's vagina or on her cervix are easily overlooked. Chancres are not painful and disappear in three to six weeks without treatment. About 70% of patients with primary syphilis develop swollen lymph nodes near the chancre. The nodes may feel firm or rubbery but are not usually painful.

The disease continues to progress even in the complete absence of symptoms. Secondary syphilis begins between six weeks and six months after infection. Chancres may still be present but are usually healing. Secondary syphilis is a systemic infection marked by the eruption of skin **rashes** and ulcers in the mucous membranes. The skin rash may resemble other skin disorders such as drug reactions, **rubella**, **ringworm**, mononucleosis, or **pityriasis rosea**. Characteristics of a syphilis rash include:

- coppery color
- absence of pain or itching
- occurrence on the palms of the hands and soles of the feet

The skin eruptions may resolve in a few weeks or last as long as a year. Some patients develop

condylomata lata—weepy, pinkish or grey patches of flattened skin on moist areas of the body. The skin rashes, mouth and genital ulcers, and condylomata lata are all highly contagious.

About 50% of patients with secondary syphilis develop swollen lymph nodes in the armpits, groin, and neck; about 10% develop inflammations of the eyes, kidney, liver, spleen, bones, joints, or the meninges—membranes covering the brain and spinal cord. Patients also may have flu-like symptoms, including low **fever**, chills, loss of appetite, **headache**, runny nose, **sore throat**, and aching joints.

The latent phase of syphilis is divided into early latency, occurring less than two years after infection, and late latency. During early latency patients are at risk for spontaneous recurrences of the ulcers and skin rashes of secondary syphilis. In late latency these recurrences are much less common. Late latency can either resolve spontaneously or continue for the remainder of the patient's life.

About 35–40% of untreated syphilis cases progress to the tertiary stage. Tertiary syphilis can be either benign late syphilis or cardiovascular and/or neurosyphilis.

Benign late syphilis begins three to ten years after infection and is characterized by the development of gummas. These are rubbery tumor-like growths that usually involve the skin or long bones, but also can develop in the eyes, mucous membranes, throat, liver, or stomach lining. Gummas have become uncommon since the introduction of **antibiotics** for treating syphilis. Benign late syphilis is usually rapid in onset and responds well to treatment.

Cardiovascular syphilis develops between 10 and 25 years after infection and often occurs along with neurosyphilis. It usually begins as an inflammation of the arteries leading from the heart and causes heart attacks, scarring of the aortic valves, congestive **heart failure**, or an **aortic aneurysm**.

There are four types of neurosyphilis:

- Asymptomatic neurosyphilis causes no central nervous system symptoms but can be detected in the spinal fluid.
- Meningovascular neurosyphilis is characterized by changes in the blood vessels of the brain or inflammation of the meninges. It causes headaches, irritability, and visual problems. If the spinal cord is involved, the patient may experience weakness of the shoulder and upper arm muscles.
- Tabes dorsalis is a progressive degeneration of the spinal cord and nerve roots, causing a loss of perception of body position and orientation in space and resulting

KEY TERMS

Chancre—An open sore with a firm or hard base that is the initial skin ulcer of primary syphilis.

Condylomata lata—Highly infectious patches of weepy, pink or gray skin in moist areas of the body that occur during secondary syphilis.

Dark field—A microscopy technique in which light is directed at an oblique angle so that organisms appear bright against a dark background.

General paresis—An advanced form of neurosyphilis affecting personality and control of movement and possibly causing convulsions or partial paralysis.

Gumma—A rubbery swelling or tumor that heals slowly and leaves a scar and is a symptom of tertiary syphilis.

Jarisch-Herxheimer reaction—A temporary reaction to penicillin treatment for syphilis that includes fever, chills, and worsening of the skin rash or chancre.

Lues maligna—Areas of ulcerated and dying skin tissue that may occur with secondary syphilis, most frequently in HIV-positive patients.

Meninges—The membranes that cover the brain and spinal cord.

Miasm—In homeopathy, an inherited weakness or predisposition to disease. The syphilitic miasm is considered to be one of the most powerful.

Neurosyphilis—Syphilis of the central nervous system.

Nosode—A homeopathic remedy made from microbes, pus, or other disease material. Syphilinum is a nosode made from a diluted solution of killed *T. pallidum*.

Spirochete—A long, slender, coiled-shape bacterium, such as *T. pallidum* that causes syphilis.

Tabes dorsalis—A progressive deterioration of the spinal cord and spinal nerves that is associated with tertiary syphilis.

Treponema pallidum—The spirochete bacterium that causes syphilis.

in loss of muscle reflexes and difficulty walking. Patients may have shooting pains in the legs and periodic episodes of pain in the abdomen, throat, bladder, or rectum. Tabes dorsalis is sometimes called locomotor ataxia.

- General paresis affects the cortex of the brain, with slow, progressive memory loss, inability to concentrate, and loss of interest in self-care. Personality changes may include irresponsible behavior, depression, delusions of grandeur, or complete psychosis.

Syphilis sometimes mimics the symptoms of HIV/AIDS. Conversely HIV/AIDS appears to increase the severity of syphilis in patients suffering from both diseases and to speed the development or appearance of neurosyphilis. Patients with both syphilis and HIV/AIDS also are more likely to develop lues maligna, a skin condition that sometimes occurs in secondary syphilis and is characterized by areas of ulcerated and dying tissue.

Infants with early congenital syphilis have systemic symptoms that resemble those of secondary syphilis in adults. The central nervous system is affected in 40–60% of children with congenital syphilis. Symptoms include:

- skin rashes
- condylomata lata
- inflammation of the lungs

- persistent runny nose
- swollen lymph nodes
- jaundice
- enlargement of the spleen and liver
- anemia

Symptoms of late congenital syphilis develop after age two and include:

- facial deformities (saddle nose)
- Hutchinson's teeth (abnormal upper incisors)
- saber shins
- dislocated joints
- deafness
- mental retardation
- paralysis
- seizure disorders

Diagnosis

Examination

Diagnosis of syphilis is often delayed:

- The initial chancre may go unnoticed.
- There are wide variations in early symptoms.
- The incubation period varies greatly.

- Patients often do not connect their symptoms with recent sexual contact.

The skin rash of secondary syphilis is sometimes the first symptom to be diagnosed. Women may be diagnosed in the course of a routine gynecological exam. While taking a medical history, the physician will ask about recent sexual contacts to determine whether the patient falls into a high-risk group. Symptoms such as skin rashes or swollen lymph nodes will be noted with respect to the timing of a patient's sexual contacts.

Tests

The definitive diagnosis of syphilis depends on laboratory test results. Various tests also are used as screens for syphilis and for follow-up monitoring after treatment. Because of the long-term risks of untreated syphilis, groups of people are routinely screened for the disease:

- marriage-license applicants
- pregnant women
- children born to infected mothers
- patients with HIV/AIDS
- sexual contacts or partners of patients diagnosed with syphilis

Nontreponemal antigen tests are used as screens for syphilis. They measure the presence of reagin, an antibody formed in reaction to *T. pallidum*. In the venereal disease research laboratory (VDRL) test, a sample of the patient's blood serum is mixed with cardiolipin and cholesterol. The formation of clumps indicates a positive reaction. The serum sample can be diluted to determine the concentration of reagin in the patient's blood. The rapid plasma reagent (RPR) test is a kit in which the serum is mixed with cardiolipin on a plastic-coated card that can be examined with the naked eye. Nontreponemal antigen tests require interpretation and sometimes further testing. They can yield both false-negative and false-positive results. False negatives can occur when patients are tested too soon after exposure to syphilis, since it takes about 14–21 days for antibodies to become detectable after infection. False-positive results can be caused by other diseases, including mononucleosis, **malaria**, **leprosy**, **rheumatoid arthritis**, and lupus. Whereas the overall rate of false positives is 0.8%, the rate of false positives in HIV/AIDS patients is about 4%.

Treponemal antibody tests are used to rule out false-positive results on nontreponemal screening tests. They are more expensive and complicated than nontreponemal tests, but are very specific and sensitive. They measure the presence of antibodies that are specific for *T. pallidum*. These tests include:

- the microhemagglutination-*T. pallidum* (MHA-TP) test, in which sheep red blood cells are coated with *T. pallidum* antigen; the cells clump if the patient's blood contains specific antibodies against the antigen
- the fluorescent treponemal antibody absorption (FTA-ABS) test, in which antibodies in the blood are used to coat *T. pallidum* on a slide and a fluorescein dye causes the coated spirochetes to fluoresce under ultraviolet (UV) light
- the INNO-LIA test—the most accurate antibody test—which uses recombinant and peptide antigens derived from *T. pallidum*

T. pallidum also can be identified in samples of tissue or lymphatic fluid. Slides of fresh samples are examined under microscopic dark-field illumination or slides of dried smears are stained with fluorescein and viewed under UV light.

A high **white blood cell count** and elevated protein levels in the cerebrospinal fluid (CSF) may suggest neurosyphilis. VDRL or FTA-ABS tests of the CSF are used to diagnose:

- neurosyphilis
- congenital syphilis
- syphilis in HIV/AIDS patients
- patients who are not responding to treatment with penicillin

Patients who test positive for syphilis are tested for HIV infection at the time of diagnosis. All sexual partners of a diagnosed patient must be tested for syphilis.

Treatment

Drugs

Syphilis is treated with antibiotics, either injected intramuscularly (benzathine penicillin G or ceftriaxone) or administered orally (doxycycline, minocycline, tetracycline, or azithromycin). In the vast majority of cases a single dose of penicillin is sufficient to cure primary and secondary syphilis. Penicillin is less effective in treating later stages and additional doses may be necessary. Neurosyphilis is treated with a combination of aqueous crystalline penicillin G, benzathine penicillin G, or doxycycline. The levels of penicillin in the patient's body must be kept sufficiently high over a period of days or weeks because *T. pallidum* has a relatively long reproduction time. Follow-up blood tests should be performed every three months to confirm that the patient is completely cured.

Pregnant women with syphilis are treated with tetracycline as early in pregnancy as possible. Infants with proven or suspected congenital syphilis are treated with

either aqueous crystalline penicillin G or aqueous procaine penicillin G. Children who acquire syphilis after birth are treated with benzathine penicillin G.

Jarisch-Herxheimer reaction may occur during penicillin treatment for late-primary, secondary, or early-latent syphilis. The patient develops chills, fever, headache, and muscle pains within two to six hours after the penicillin injection and the chancre or rash temporarily worsens. The reaction lasts about one day and is thought to be an allergic reaction to the toxins that are released as massive numbers of spirochetes are destroyed.

Alternative

The historical link between homeopathy and syphilis is Hahnemann's theory of miasms, which labeled the syphilitic miasm as the second-oldest cause of constitutional weakness in humans. Homeopathic practitioners in the United States are banned from claiming that their treatments can cure syphilis. However because of the high incidence of syphilis in HIV/AIDS patients, some alternative practitioners claim that their homeopathic remedies for AIDS also are beneficial in treating syphilis. The most frequently suggested remedies are *Medorrhinum*, *Aurum*, *Mercurius vivus*, and *Syphilinum*. The use of *Mercurius vivus* reflects the historical use of mercury to treat syphilis. *Syphilinum* is in a class of homeopathic remedies called nosodes, which are made from disease material, such as bacteria, viruses, or pus. *Syphilinum* is made from a dilution of killed *T. pallidum*.

Certain outdated or discredited treatments for syphilis have resurfaced as alternative treatments for HIV/AIDS and cancer. Hyperthermia—inducing a fever to treat HIV/AIDS—originated as a treatment for syphilis, in which patients were infected with malaria in an attempt to kill *T. pallidum*. The Hoxsey treatment for cancer, which is no longer legally available in the United States, was developed in the 1920s by Harry Hoxsey and prescribed as a treatment for secondary and tertiary syphilis. The treatment consists of several chemical mixtures applied externally and a formula of nine herbs taken internally. The external formulation contains both arsenic and antimony, which were used to treat syphilis before the advent of antibiotics. The internal herbal formula includes *Phytolacca americana*, or pokeweed, which was used by Native Americans to treat syphilitic chancres, and *Stillingia sylvatica*, or queensroot, which was used in the past to treat syphilis. All of these components are potentially toxic and should not be used to treat syphilis.

Traditional Chinese medicine (TCM) and other alternative approaches emphasize the mental aspects of diseases such as syphilis. Alternative practitioners may

recommend mind-body medicine, **guided imagery**, and affirmations as adjuncts to antibiotic treatment for syphilis.

Home remedies

Although antibiotics are essential for the treatment of syphilis, recovery can be aided by good dietary habits, adequate sleep, **exercise**, and stress-reduction techniques. Skin rashes and ulcers should be kept clean and dry. Patients must abstain from sexual contact until their disease has been cured. Other people should not be exposed to fluid or discharges from chancres, skin ulcers, rashes, or condylomata lata.

Prognosis

Antibiotics—especially penicillin—cure early-stage syphilis quickly and effectively. Treatment failures do occur, especially in HIV/AIDS patients treated with penicillin. Patients also can be re-infected. Patients should be followed up with blood tests at one, three, six, and 12 months after treatment or until the results are negative. CSF should be tested after one year. Patients with primary and secondary syphilis who remain symptom-free and have negative blood tests for two years after treatment are usually considered cured. Patients with recurrences during the latency period should be tested for re-infection.

In patients with untreated syphilis:

- About 30% undergo spontaneous remission.
- About 30% have lifelong latency.
- About 40% develop potentially fatal tertiary forms of syphilis.

Proper treatment for maternal syphilis during the second and third trimesters of pregnancy reduces the risk of congenital syphilis in the infant from 90% to less than 2%. However nearly 50% of untreated fetuses die shortly before or after birth. Those who survive may appear normal at birth but show signs of infection between three and eight weeks of age.

Prevention

Prevention of syphilis depends on a combination of personal and public health measures. Patients with syphilis do not acquire lasting immunity against the disease, so they can be easily re-infected. The only reliable methods for preventing transmission of syphilis are sexual abstinence or a monogamous relationship between uninfected partners. **Condoms** reduce the risk of transmission but protect only the covered parts of the genitals. The general public needs to be informed about the transmission and early symptoms of syphilis.

and public health facilities must provide for adequate testing and treatment.

U.S. law requires the reporting of all syphilis cases to public health agencies. Sexual contacts of patients diagnosed with syphilis are traced and tested for the disease. This includes all contacts in the past three months for cases of primary syphilis and in the past year for cases of secondary syphilis. Neither patients nor their contacts should have sexual contact until they have been tested and treated. Patients should be informed about the disease and counseled regarding sexual behavior, safe sexual practices, and the importance of completing antibiotic treatment. In addition:

- Sexually active adolescents should be routinely screened for syphilis.
- Pregnant women should be tested for syphilis at the time of their first prenatal visit and again shortly before delivery.
- Many obstetricians and gynecologists recommend the routine screening of non-pregnant women.
- Because of the rising incidence of syphilis worldwide, many public health physicians recommend routine screening of immigrants, refugees, and international adoptees.

In 2006 the Centers for Disease Control and Prevention (CDC) announced an update to its “National Plan to Eliminate Syphilis in the United States:”

- investing and enhancing public health services and interventions for syphilis treatment
- prioritizing and targeting interventions for high-risk populations, such as African Americans and MSM
- improving the accountability of prevention efforts

Resources

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ORGANIZATIONS

American Social Health Association, P.O. Box 13827, Research Triangle Park, NC, 27709, (919) 361-8400, (800) 227-8922, (919) 361-8425, info@ashastd.org, <http://www.ashastd.org>.

National Institute of Allergy and Infectious Diseases (NIAID), Office of Communications and Public Liaison, 6610 Rockledge Drive, Bethesda, MD, 20892-66123, (866) 284-4107, <http://www3.niaid.nih.gov>.

U.S. Centers for Disease Control and Prevention (CDC), 1600 Clifton Road, Atlanta, GA, 30333, (800) CDC-INFO (232-4636), cdcinfo@cdc.gov, <http://www.cdc.gov>.

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Systemic antifungal drugs see **Antifungal drugs, systemic**

Systemic lupus erythematosus

Definition

Lupus is an autoimmune disorder that may damage body tissues and lead to widespread, chronic inflammation and pain during periods of worsened symptoms called flares. Lupus can cause problems with multiple body systems and organs, including the joints, skin,



A close-up view of a woman's face with a lesion caused by systemic lupus erythematosus (SLE). One characteristic of this autoimmune disease is a butterfly rash present across the cheeks and nose. (Custom Medical Stock Photo, Inc.)

Reproduced by permission.

kidneys, heart, lungs, and blood vessels. Although lupus cannot be cured, it is treatable with medications and other therapies.

Description

Lupus is an autoimmune disease. Normally, the white blood cells in the body's immune system protect a person from harmful substances called antigens. Antigens may include bacteria, viruses, foreign blood, **cancer** cells, and other toxins that could cause disease or infection. To defend the body against antigens, the body produces antibodies. In a person with a healthy immune system, the antibodies then destroy the antigens, keeping the person from getting sick.

In people with autoimmune diseases, however, the immune system cannot tell the difference between an antigen and healthy tissues. As a result, the body begins attacking its own healthy tissues. In people with lupus, the autoimmune response most often attacks the joints, skin, heart, lungs, kidneys, and blood and circulatory system.

There are several different types of lupus, including: systemic lupus erythematosus (SLE); **discoid lupus erythematosus** (DLE); and drug-induced lupus. The severity of lupus symptoms vary from person to person: in some people, the symptoms may be mild and involve

only the joints and skin; in others, the disease may be severe and cause joint, kidney, lung, heart, and bone complications.

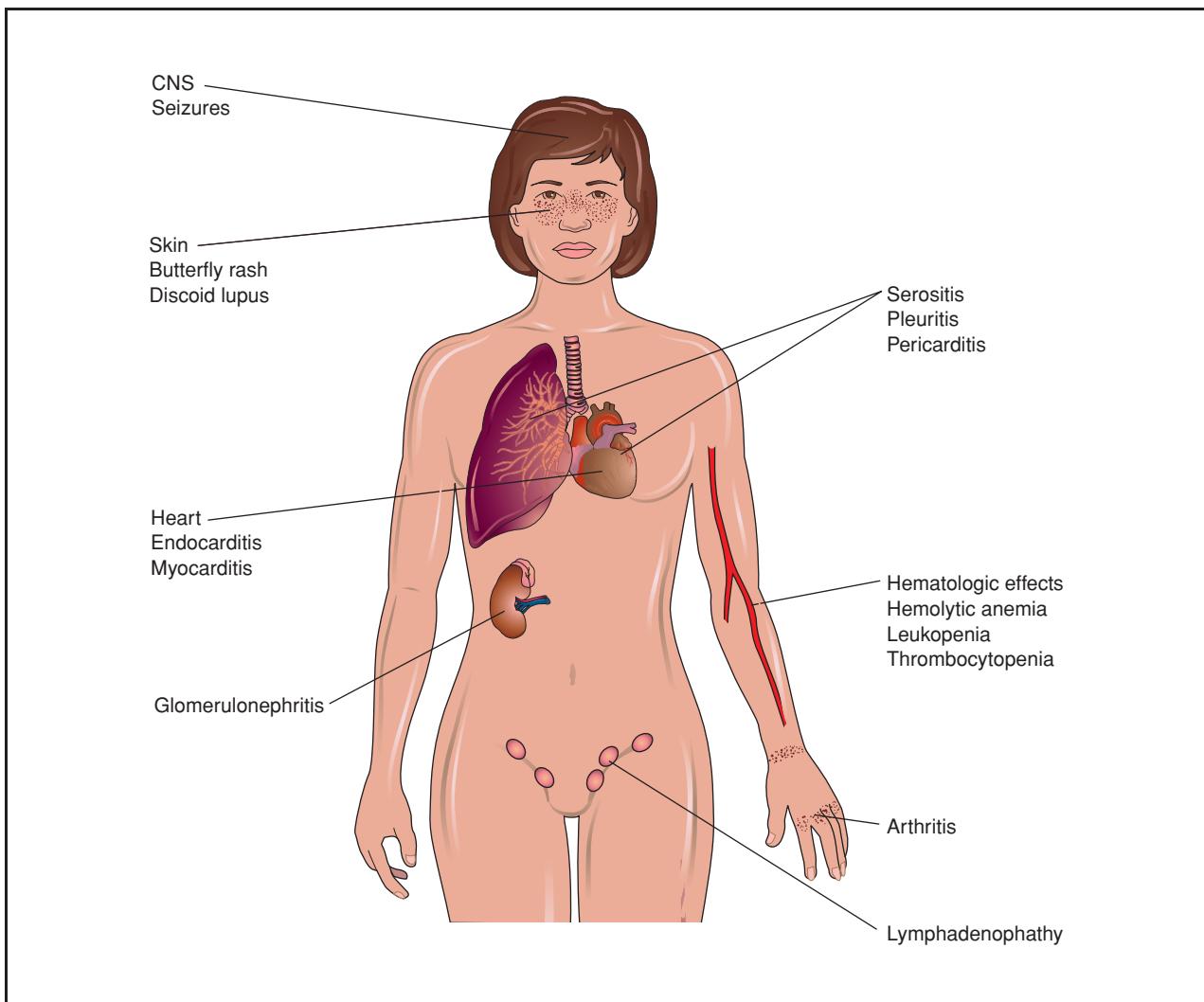
Systemic lupus usually involves multiple organs and body systems and is usually more severe than the DLE form of the disease. The majority, about 70% of all lupus cases involve this systemic form of the disease. People with SLE typically experience pain in the joints and muscles, **fatigue**, and skin **rashes**, which may come and go. These periods of more severe symptoms are called flares, whereas the milder periods of the disease are referred to as remission. People with SLE may experience kidney inflammation (**nephritis**), which can make it difficult for the body to remove toxins and other waste products. Lupus patients may be prone to develop **pneumonia** or inflammation of the chest cavity that makes it difficult to breathe. The disease may also contribute to central nervous system problems, including headaches, **dizziness**, seizures, behavior changes, and vision and memory difficulties. Having lupus also increases the risk of **atherosclerosis** (hardening of the arteries), **blood clots**, and deficiencies in red and white blood cells and platelets.

DLE, sometimes referred to as discoid or cutaneous lupus, primarily affects the skin and accounts for 10% of all lupus cases. People with this form of lupus typically develop a rash on the face, neck, and scalp but do not experience problems with the joints, kidneys, or heart. However, about 10% of people with DLE eventually develop SLE; doctors think that in these patients, the rash was an initial symptom of systemic inflammation.

Drug-induced lupus may also develop after a person takes certain prescription medications. People with this type of lupus tend to have symptoms similar to those of SLE, but the symptoms of lupus typically fade within days, weeks, or months of discontinuing the medications. Medications that may induce lupus include hydralazine (a drug used to treat high blood pressure) and procainamide (a drug used to treat irregular heart rhythms). About 4% of people who take these medications develop drug-induced lupus.

Demographics

Lupus affects people of all ages and races, but it is most common in women. Ninety percent of the 1.4 million diagnoses of lupus in the United States are made in women. Lupus is also more prevalent among younger women between 15 and 44 and women of particular ethnic groups. Hispanic/Latino, African American, and American Indian women are more likely to develop lupus than white women, and their symptoms tend to be more severe.



Systemic lupus erythematosus (SLE) is an autoimmune disease in which the individual's immune system attacks, injures, and destroys the body's own organs and tissues. Nearly every system of the body can be affected by SLE, as depicted in the illustration above. (Illustration by Electronic Illustrators Group. Reproduced by permission of Gale, a part of Cengage Learning.)

However, according to the Lupus Foundation of America, 15% of people with SLE develop it later in life—after age 55. Late onset lupus affects women eight times more than men, and is more prevalent in Caucasians, although it can occur in people of any race.

Drug-induced lupus is also more common in older adults because of the greater likelihood that they have other conditions (such as heart disease and high blood pressure) that require medication. When a person goes off the medication, the symptoms of drug-induced lupus eventually go away.

Causes and symptoms

The cause of SLE is unknown. Because the vast majority of patients are women, some research is being

done to determine what (if any) link the disease has to female hormones. SLE may have a genetic basis, although more than one gene is believed to be involved in the development of the disease. Because lupus often runs in families, researchers think there is a genetic component to the disease. However, other factors, including environment, **stress**, the use of certain medications, and exposure to sunlight, may also influence lupus development and exacerbate flares.

Symptoms of lupus depend on the type a person has and can vary widely. Some common symptoms of lupus include:

- a signature red rash or color change in the skin across the nose and cheeks (this is also called a malar rash; often it is in the shape of a butterfly)

- painful, swollen joints (arthritis) and glands
- fevers that can't be explained by illness
- pain in the chest when breathing
- extreme fatigue
- anemia (loss of red blood cells)
- hair loss
- sensitivity to the sun
- blood flow problems in the fingers when cold or stressed
- depression
- problems with memory or thinking clearly

For some people, mouth sores, seizures, **hallucinations**, and kidney problems signal lupus.

In people with late-onset lupus, symptoms tend to be milder and include arthritis, **pleurisy**, **pericarditis**, dry eyes and mouth, and muscle aches. In older adults, it may be harder to diagnose lupus because the symptoms mimic other diseases common in this age group, such as **rheumatoid arthritis**.

Lupus symptoms may come and go. These periods of worsened symptoms, called flares, may be triggered by spending time in the sun or during a time of emotional stress.

SLE has also been linked to a higher risk of developing **osteoporosis**, a disease that makes bones brittle and more likely to break. Osteoporosis may occur in lupus patients because the steroid medications often prescribed to reduce inflammation can lead to bone loss. Fatigue and pain in the joints and muscles also makes it more likely a person will remain inactive, which increases the likelihood of bone loss. Finally, lupus itself may contribute to weakened bones that are more likely to break.

According to the National Institute of Arthritis and Musculoskeletal and Skin Diseases, women with lupus may have more than five times the risk of a bone fracture from osteoporosis.

The severity of a patient's SLE varies over time. Patients may have periods with mild or no symptoms, followed by a flare. During a flare, symptoms increase in severity and new organ systems may become affected.

Many SLE patients have fevers, fatigue, muscle pain, weakness, decreased appetite, and weight loss. The spleen and lymph nodes are often swollen and enlarged. The development of other symptoms in SLE varies, depending on the organs affected.

- Joints. Joint pain and problems, including arthritis, are very common. About 90% of all SLE patients have these types of problems.

- Skin. A number of skin rashes may occur, including a red butterfly-shaped rash that spreads across the face. The "wings" of the butterfly appear across the cheekbones, and the "body" appears across the bridge of the nose. A discoid, or coin-shaped, rash causes red, scaly bumps on the cheeks, nose, scalp, ears, chest, back, and the tops of the arms and legs. The roof of the mouth may develop sore, irritated pits (ulcers). Hair loss is common. SLE patients tend to be very easily sunburned (photosensitive).
- Lungs. Inflammation of the tissues that cover the lungs and line the chest cavity causes pleuritis, with fluid accumulating in the lungs. The patient frequently experiences coughing and shortness of breath.
- Heart and circulatory system. Inflammation of the tissue surrounding the heart causes pericarditis; inflammation of the heart itself causes myocarditis. These heart problems may result in abnormal beats (arrhythmias), difficulty pumping the blood strongly enough (heart failure), or even sudden death. Blood clots often form in the blood vessels and may lead to complications.
- Nervous system. Headaches, seizures, changes in personality, and confused thinking (psychosis) may occur.
- Kidneys. The kidneys may suffer significant destruction, with serious life-threatening effects. They may become unable to adequately filter the blood, leading to kidney failure.
- Gastrointestinal system. Patients may experience nausea, vomiting, diarrhea, and abdominal pain. The lining of the abdomen may become inflamed (peritonitis).
- Eyes. The eyes may become red, sore, and dry. Inflammation of one of the nerves responsible for vision may cause vision problems, and blindness can result from inflammation of the blood vessels (vasculitis) that serve the retina.

Diagnosis

Obtaining a lupus diagnosis can be difficult for several reasons: the symptoms of lupus flare and disappear over long periods of time; the severity and type of lupus symptoms can vary widely from person to person; and finally, there is no one test that can diagnose the disease. Instead, doctors must rely on several diagnostic techniques to confirm a lupus diagnosis, including a detailed medical history, **physical examination**, blood and urine tests, and skin or kidney biopsies.

Many of the symptoms and laboratory test results of SLE patients are similar to those of patients with different diseases, including rheumatoid arthritis,

multiple sclerosis, and various nervous system and blood disorders.

When conducting a medical history, doctors may ask patients a variety of questions, such as:

- Have you had stiff, tender, and swollen joints? Is this worse in the morning?
- Do you ever feel extremely tired for days or weeks, even when you're getting plenty of sleep at night?
- Have you ever felt pain in your chest when taking deep breaths?
- Does your skin break out when you're in the sun, but not from sunburn?
- Have you had a rash across your nose and cheeks? Is it in the shape of a butterfly?

In addition to taking a thorough medical history, doctors will also conduct a physical examination. A physician may listen to the heart (in some lupus patients, doctors can hear a sound called a heart friction rub) and conduct a **neurological exam**.

Laboratory tests are also an integral part of the lupus diagnosis process. One test, called the antinuclear antibody (ANA) test, is often checked when a doctor suspects a person has lupus. In this test, a person's blood is checked for autoantibodies that are often present in the blood of people with lupus. Testing positive for ANA does not automatically mean a person has lupus, but it can help doctors make a diagnosis when considered with a person's physical symptoms. Other tests doctors may use to confirm a lupus diagnosis include the anti-double strand DNA (dsDNA), anti-Smith antibodies (Sm), **erythrocyte sedimentation rate** (ESR), and **C-reactive protein** binding. SLE patients tend to have low numbers of red blood cells (anemia) and low numbers of certain types of white blood cells. The ESR, a measure of inflammation in the body, tends to be quite elevated. Also, samples of tissue (biopsies) from affected skin and kidneys may show characteristics of the disease.

A test called the lupus erythematosus cell preparation (or LE prep) test is also performed. This test involves obtaining a sample of the patient's blood. Cells from the blood are damaged in the laboratory in order to harvest their nuclei. These damaged cells are then put together with the patient's blood serum, the liquid part of blood separated from the blood cells. Antinuclear antibodies within the patient's serum will clump together with the damaged nuclear material. A material called Wright's stain will cause these clumps to turn blue. These stained clumps are then reacted with some of the patient's white blood cells, which will essentially eat the clumps. LE cells are the white blood cells that contain the blue clumps.

This test will be positive in about 70–80% of all patients with SLE.

The American Rheumatism Association developed a list of symptoms used to diagnose SLE. Research supports the idea that people who have at least four of the eleven criteria (not necessarily simultaneously) are extremely likely to have SLE. The criteria are:

- butterfly rash
- discoid rash
- photosensitivity
- mouth ulcers
- arthritis
- inflammation of the lining of the lungs or the lining around the heart
- kidney damage, as noted by the presence of protein or other abnormal substances called casts in the urine
- seizures or psychosis
- the presence of certain types of anemia and low counts of particular white blood cells
- the presence of certain immune cells, anti-DNA antibodies, or a falsely positive test for syphilis
- the presence of antinuclear antibodies

If lupus is diagnosed, doctors may check a person's urine for signs of kidney problems, order chest x-rays for signs of inflammation in the lungs or heart, and have the patient's blood checked for problems with the white blood cells to see how far the disease has progressed.

Allopathic treatment

Several types of health care professionals may work together to treat an individual with lupus. Family doctors or internists, rheumatologists (specialists in rheumatic diseases), immunologists (specialists in immune system disorders), and other specialists may play a role in treating the lupus patient.

The treatment a person receives for lupus depends on the type of lupus and the extent and severity of the disease. Both over-the-counter and prescription medications may be recommended, such as:

- Nonsteroidal anti-inflammatory drugs (NSAIDS): These drugs, which include ibuprofen and naproxen, reduce inflammation and control pain, swelling, and fever. However, these medicines also may cause side effects such as nausea, heartburn, and diarrhea as well as liver, kidney, and neurological complications with prolonged use, so it is important that a person taking these drugs for lupus does so under the direction of a doctor.

- Antimalarial drugs: Antimalarial drugs such as hydroxychloroquine treat the fatigue, joint pain, rashes, and lung inflammation caused by lupus and may prevent flares from occurring. Side effects include nausea and, in rare cases, vision problems.
- Corticosteroids: A variety of corticosteroid medications, including prednisone, hydrocortisone, methylprednisolone, and dexamethasone, can suppress the inflammation often associated with lupus. Lupus patients take these drugs in pill form, apply creams to the skin, or receive corticosteroid injections. Despite their effectiveness, corticosteroid drugs do have short-term side effects, such as increased appetite and weight gain. Long-term side effects may include high blood pressure, weakened bones, artery damage, diabetes, and cataracts.
- Immunosuppressive agents: These drugs, including azathioprine, cyclophosphamide and mycophenolate mofetil, block the production of immune cells and are typically used in lupus patients who experience kidney or central nervous system problems. A person taking immunosuppressive drugs may experience nausea and vomiting, as well as bladder problems, hair loss, decreased fertility, and an increased risk of infection.

Doctors may also use arthritis drugs to help control symptoms of lupus and reduce the risk of flares.

Other treatments for SLE try to help specific symptoms. Clotting disorders will require blood thinners. Psychotic disorders will require specific medications. Kidney failure may require the blood to be cleaned outside the body through a machine (dialysis) or even a **kidney transplantation**.

Alternative treatment

Medications for lupus are costly, and many have the potential for serious adverse side effects. As a result, some patients turn to other therapies to relieve lupus symptoms.

Massage and **acupuncture** are just a few of the alternative and complementary therapies that may be used by lupus patients. Doctors may encourage lupus patients to get regular, gentle **exercise** during remission to increase joint flexibility and muscle strength. Stress management is key for people with SLE and such techniques as **meditation**, **hypnotherapy**, and **yoga** may be helpful in promoting relaxation.

In addition, some patients have tried dietary supplements in an attempt to alleviate lupus symptoms. Supplementation with **omega-3 fatty acids** found in fish oils could hold promise for lupus patients. In one

study of 60 people with SLE, daily doses of 3 grams of omega-3 fatty acids in the form of fish oil supplements over a 6-month period improved lupus symptoms. Not only did the supplements appear to relieve joint pain, but they also improved blood vessel function, researchers noted.

Other dietary suggestions include eating a whole foods diet with reduced amounts of red meat and dairy products in order to decrease pain and inflammation. **Food allergies** are believed either to contribute to SLE or to arise as a consequence of the digestive difficulties. Wheat, dairy products, and soy are the major offenders. An elimination/challenge diet can help identify the offending foods so that they can be avoided. Because alfalfa sprouts have been associated with the onset of flares in SLE, they should be avoided. Supplements that have been suggested to improve the health of SLE patients include **vitamins** B, C, and E, as well as selenium, zinc, magnesium, and a complete trace mineral supplement. Vitamin A is believed to help improve discoid skin rashes. Botanical medicine can help the entire body through immune modulation and **detoxification**, as well as assisting individual organs and systems. Homeopathy and flower essences can work deeply on the emotional level to help people with this difficult disease.

Nutrition/Dietetic concerns

There are no specific guidelines for people with lupus to follow, however, there are several nutritional considerations that may be impacted by a lupus diagnosis.

Lack of appetite and weight loss is common among people who have recently been diagnosed with lupus. Appetite and weight loss may be related to a person's symptoms of pain and fatigue, or they may be a side effect of common lupus medications. In addition, some people develop mouth sores when taking lupus medications, another factor that can make it difficult to eat. Older adults struggling with a lack of appetite should talk to their health care providers, who may recommend consulting with a registered dietitian who can suggest a diet that works best with the patient's needs and lifestyle.

Weight gain is also a common side effect of **corticosteroids**, drugs that are often used to treat the inflammation of lupus. If a person has gained weight after starting lupus medications, a registered dietitian (RD) can also help by devising a meal plan that incorporates nutritious foods that won't add excess weight. A RD can also help lupus patients work toward controlling high blood pressure and avoiding atherosclerosis. These cardiovascular

KEY TERMS

Antinuclear antibody (ANA) test—A test often used to look for autoantibodies that react against components of the nucleus of the body's cells. Many people with lupus test positive for ANA.

Arthritis—A condition characterized by inflamed, swollen, painful joints.

Autoimmune disorder—A disorder in which the body's antibodies mistake the body's own tissues for foreign invaders. The immune system then attacks and causes damage to these tissues.

Chromosomes—Spaghetti-like structures located within the nucleus (or central portion) of each cell. Chromosomes contain genes, structures that direct the growth and functioning of all the cells and systems in the body. Chromosomes are responsible for passing on hereditary traits from parents to child.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to prevent foreign organisms (bacteria, viruses, fungi, etc.) from invading the body.

Nephritis—Inflammation of the kidneys.

Osteoporosis—A condition that makes bones less dense and more likely to fracture.

Pericarditis—Inflammation of the sac around the heart.

Pleurisy—Chest pain that occurs when a person takes a deep breath.

Psychosis—Extremely disordered thinking with a poor sense of reality; may include hallucinations (seeing, hearing, or smelling things that are not really there).

complications may be more common in lupus patients, but a combination of a low-fat diet and exercise may reduce the risk of these common complications.

Also, lupus patients taking corticosteroids may need to take vitamin D and **calcium** supplements to counteract the bone-damaging effects of the disease and reduce the risk of osteoporosis.

In general, if medication use or lupus symptoms are making it difficult to eat, patients should consult with a doctor or nurse, who can provide additional information.

Prognosis

The prognosis for patients with SLE varies, depending on the organ systems most affected and the severity of inflammation. Some patients have long periods of time with mild or no symptoms. About 90–95% of patients are still living after 2 years with the disease. About 82–90% of patients are still living after 5 years with the disease. After 10 years, 71–80% of patients are still alive, and 63–75% are still alive after 20 years. The most likely causes of **death** during the first 10 years include infections and kidney failure. During years 11–20 of the disease, the most likely cause of death involves the development of abnormal blood clots.

Because SLE frequently affects women of childbearing age, **pregnancy** is an important issue. For pregnant SLE patients, about 30% of the pregnancies end in **miscarriage**. About 25% of all babies born to mothers with

SLE are premature. Most babies born to mothers with SLE are normal. However, a rare condition called neonatal lupus causes a baby of a mother with SLE to develop a skin rash, liver or blood problems, and a serious heart condition.

The Centers for Disease Control and Prevention estimate that more than 1,000 people die from lupus annually, and older adults, women, and blacks had the highest death rates among lupus patients. Following the doctor's instructions, taking medications exactly as they are prescribed, and getting help when symptoms flare can help lupus patients extend the quantity and quality of their lives.

Prevention

There are no known ways to avoid developing SLE. However, it is possible for a patient who has been diagnosed with SLE to prevent flares of the disease. Recommendations for improving general health to avoid flares include decreasing sun exposure, getting sufficient sleep, eating a healthy diet, decreasing stress, and exercising regularly. It is important for a patient to try to identify the early signs of a flare (like **fever**, increased fatigue, rash, **headache**). Some people believe that noticing and responding to these warning signs will allow a patient with SLE to prevent a flare, or at least to decrease its severity. In addition, getting regular health care and laboratory tests can help doctors note changes and make adjustments once a flare begins. Finally, because they are at risk for other complications from lupus, older adults should have their blood pressure

and cholesterol checked regularly. An annual **influenza** vaccine may also be recommended, and patients should reduce exposure to the sun and always wear sunscreen.

Caregiver concerns

Caregivers can help by learning the signs of their loved ones' flares and encouraging communication with the doctor when lupus symptoms occur.

Caregivers should keep in mind that uncontrolled bleeding, trouble breathing, **fainting**, confusion, chest pain, or seizures in a lupus patient are signs of a serious problem. These symptoms indicate their loved one with lupus needs immediate medical help.

Resources

BOOKS

Gorman, Sara. *Lupus: Despite Lupus: How to Live Well with a Chronic Illness*. New York: Four Legged Press, 2009.

Pigache, Philippe. *Positive Options for Living With Lupus: Self-Help and Treatment*. Alameda, CA: Hunter House Publishers, 2006.

Quintero Del Rio, Iris. *Lupus: A Patient's Guide to Diagnosis, Treatment and Lifestyle*. Munster, IN: Hilton Publishing, 2007.

Wallace, Daniel J. *The Lupus Book: A Guide for Patients and Their Families*. New York, NY: Oxford University Press, 2008.

PERIODICALS

Henderson, Shirley. "Living With Lupus: Although There Is No Cure, Many People are Making Lifestyle Adjustments to Fight the Disease and Improve Their Sense of Well-Being." *Ebony* (July 2007): 142(3).

Mahoney, Diana. "Recognize, Aggressively Treat Cutaneous Evidence of Lupus." *Family Practice News* (May 1, 2006): 28.

Seppa, N. "Self Help: Stem Cells Rescue Lupus Patients."

Science News (February 4, 2006): 67(2).

Stewart, Kimberly Lord. "The Wolf at the Door: Kimberly Lord Stewart Describes Her Battle With Lupus as She Explores the Fine Line Between Western Medicine and Alternative Therapies." *Better Nutrition* (July 2005): 30(4).

Walsh, Nancy. "Biologics Promising in Lupus, But More Research is Needed." *Family Practice News* (July 15, 2007): 35.

ORGANIZATIONS

Alliance for Lupus Research, 28 West 44th Street, Suite 501, New York, NY, 10036, (212) 218-2840, (800) 867-1743, info@lupusresearch.org, <http://www.lupusresearch.org/home.html>.

American College of Rheumatology, 1800 Century Place, Suite 250, Atlanta, GA, 30345, (404) 633-3777, <http://www.rheumatology.org>.

Lupus Canada, 590 Alden Road, Suite 211, Markham, Canada, ON, L3R 8N2, (905) 513-0004, (800) 661-1468 (Canada only), <http://www.lupuscanada.org>.

Lupus Foundation of America, 2000 L Street, N.W., Suite 710, Washington, DC, 20036, (202) 349-1155, (800) 558-0121, (202) 349-1156, <http://www.lupus.org/newsite/index.html>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases, 1 AMS Circle, Bethesda, MD, 20892-3675, (301) 495-4484, (877) 226-4267, (301) 718-6366, NIAMSinfo@mail.nih.gov, <http://www.niams.nih.gov>.

S.L.E. Lupus Foundation, 330 Seventh Avenue, Suite 1701, New York, NY, 10001, (212) 685-4118, (212) 545-1843, Lupus@LupusNY.org, <http://www.lupusny.org>.

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