

The GALE
ENCYCLOPEDIA
of MEDICINE

SECOND EDITION

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VOLUME

2

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The GALE ENCYCLOPEDIA of MEDICINE SECOND EDITION

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PLEASE READ—IMPORTANT INFORMATION

The *Gale Encyclopedia of Medicine 2* is a medical reference product designed to inform and educate readers about a wide variety of disorders, conditions, treatments, and diagnostic tests. The Gale Group believes the product to be comprehensive, but not necessarily definitive. It is intended to supplement, not replace, consultation with a physician or other healthcare practitioner. While the Gale Group has made substantial efforts to provide information that is accurate, comprehensive, and up-to-date, the Gale Group 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 medical 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 health care provider.

INTRODUCTION

The *Gale Encyclopedia of Medicine 2 (GEM2)* is a one-stop source for medical information on nearly 1,700 common medical disorders, conditions, tests, and treatments, including high-profile diseases such as AIDS, Alzheimer's disease, cancer, and heart attack. 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 2* fills a gap between basic consumer health resources, such as single-volume family medical guides, and highly technical professional materials.

SCOPE

Almost 1,700 full-length articles are included in the *Gale Encyclopedia of Medicine 2*, 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, eg. acetaminophen (Tylenol). Throughout the *Gale Encyclopedia of Medicine 2*, many prominent individuals 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
Description	Purpose
Causes and symptoms	Precautions
Diagnosis	Description
Treatment	Preparation
Alternative 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 2* 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.

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 *GEM2* 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 Group editor.

ABOUT THE CONTRIBUTORS

The essays were compiled by experienced medical writers, including physicians, pharmacists, nurses, and other health care professionals. *GEM2* medical advisors reviewed the completed essays to insure that they are appropriate, up-to-date, and medically accurate.

HOW TO USE THIS BOOK

The *Gale Encyclopedia of Medicine 2* 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 are also cross-referenced.
- A list of **key terms** are provided where appropriate to define unfamiliar terms or concepts.
- 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.

- **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 2* is enhanced with over 675 color images, including photos, charts, tables, and customized line drawings.

ADVISORY BOARD

A number of experts in the library and medical communities provided invaluable assistance in the formulation of this encyclopedia. Our advisory board performed a myriad of duties, from defining the scope of coverage to reviewing individual entries for accuracy and accessibility. The editor would like to express her appreciation to them.

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C

CABG surgery see **Coronary artery bypass graft surgery**

CAD see **Coronary artery disease**

Caffeine

Definition

Caffeine is a drug that stimulates the central nervous system.

Purpose

Caffeine makes people more alert, less drowsy, and improves coordination. Combined with certain **pain** relievers or medicines for treating **migraine headache**, caffeine makes those drugs work more quickly and effectively. Caffeine alone can also help relieve headaches. **Antihistamines** are sometimes combined with caffeine to counteract the drowsiness that those drugs cause. Caffeine is also sometimes used to treat other conditions, including breathing problems in newborns and in young babies after surgery.

Description

Caffeine is found naturally in coffee, tea, and chocolate. Colas and some other soft drinks contain it. Caffeine also comes in tablet and capsule forms and can be bought without a prescription. Over-the-counter caffeine brands include No Doz, Overtime, Pep-Back, Quick-Pep, Caffedrine, and Vivarin. Some pain relievers, medicines for migraine headaches, and antihistamines also contain caffeine.

Recommended dosage

Adults and children age 12 years and over

100–200 mg no more than every 3–4 hours. In timed-release form, the dose is 200–250 mg once a day.

Timed-release forms should not be taken less than six hours before bedtime.

Children under 12 years

Not recommended.

Other considerations

Avoid taking too much caffeine when it is being taken as an over-the-counter drug. Consider how much caffeine is being taken in from coffee, tea, chocolate, soft drinks, and other foods that contain caffeine. Check with a pharmacist or physician to find out how much caffeine is safe to use.

Precautions

Caffeine cannot replace sleep and should not be used regularly to stay awake as the drug can lead to more serious **sleep disorders**, like **insomnia**.

People who use large amounts of caffeine over long periods build up a tolerance to it. When that happens, they have to use more and more caffeine to get the same effects. Heavy caffeine use can also lead to dependence. If the person then stops using caffeine abruptly, withdrawal symptoms may occur. These can include throbbing headaches, **fatigue**, drowsiness, yawning, irritability, restlessness, vomiting, or runny nose. These symptoms can go on for as long as a week if caffeine is avoided. Then the symptoms usually disappear.

If taken too close to bedtime, caffeine can interfere with sleep. Even if it does not prevent a person from falling asleep, it may disturb sleep during the night.

The notion that caffeine helps people sober up after drinking too much alcohol is a myth. In fact, using caffeine and alcohol together is not a good idea. The combination can lead to an upset stomach, nausea, and vomiting.

Older people may be more sensitive to caffeine and thus more likely to have certain side effects, such as irritability, nervousness, **anxiety**, and sleep problems.

KEY TERMS

Arrhythmia—Abnormal heart rhythm.

Central nervous system—The brain, spinal cord and nerves throughout the body.

Fetus—A developing baby inside the womb.

Palpitation—Rapid, forceful, throbbing, or fluttering heartbeat.

Withdrawal symptoms—A group of physical or mental symptoms that may occur when a person suddenly stops using a drug to which he or she has become dependent.

Special conditions

Caffeine may cause problems for people with certain medical conditions or who are taking certain medicines.

ALLERGIES. Anyone with **allergies** to foods, dyes, preservatives, or to the compounds aminophylline, dyphylline, oxtriphylline, theobromine, or theophylline should check with a physician before using caffeine. Anyone who has ever had an unusual reaction to caffeine should also check with a physician before using it again.

PREGNANCY. Caffeine can pass from a pregnant woman's body into the developing fetus. Although there is no evidence that caffeine causes **birth defects** in people, it does cause such effects in laboratory animals given very large doses (equal to human doses of 12–24 cups of coffee a day). In humans, evidence exists that doses of more than 300 mg of caffeine a day (about the amount of caffeine in 2–3 cups of coffee) may cause **miscarriage** or problems with the baby's heart rhythm. Women who take more than 300 mg of caffeine a day during **pregnancy** are also more likely to have babies with low birth weights. Any woman who is pregnant or planning to become pregnant should check with her physician before using caffeine.

BREASTFEEDING. Caffeine passes into breast milk and can affect the nursing baby. Nursing babies whose mothers use 600 mg or more of caffeine a day may be irritable and have trouble sleeping. Women who are breastfeeding should check with their physicians before using caffeine.

OTHER MEDICAL CONDITIONS. Caffeine may cause problems for people with these medical conditions:

- peptic ulcer
- heart **arrhythmias** or **palpitations**

- heart disease or recent **heart attack** (within a few weeks)
- high blood pressure
- liver disease
- insomnia (trouble sleeping)
- anxiety or panic attacks
- agoraphobia (fear of being in open places)
- premenstrual syndrome (PMS)

USE OF CERTAIN MEDICINES. Using caffeine with certain other drugs may interfere with the effects of the drugs or cause unwanted—and possibly serious—side effects.

Side effects

At recommended doses, caffeine can cause restlessness, irritability, nervousness, shakiness, **headache**, light-headedness, sleeplessness, nausea, vomiting, and upset stomach. At higher than recommended doses, caffeine can cause excitement, agitation, anxiety, confusion, a sensation of light flashing before the eyes, unusual sensitivity to touch, unusual sensitivity of other senses, ringing in the ears, frequent urination, muscle twitches or **tremors**, heart arrhythmias, rapid heartbeat, flushing, and convulsions.

Interactions

Certain drugs interfere with the breakdown of caffeine in the body. These include **oral contraceptives** that contain estrogen, the antiarrhythmia drug mexiletine (Mexitil), the ulcer drug cimetidine (Tagamet), and the drug disulfiram (Antabuse), used to treat **alcoholism**.

Caffeine interferes with drugs that regulate heart rhythm, such as quinidine and propranolol (Inderal). Caffeine may also interfere with the body's absorption of iron. Anyone who takes iron supplements should take them at least an hour before or two hours after using caffeine.

Serious side effects are possible when caffeine is combined with certain drugs. For example, taking caffeine with the decongestant phenylpropanolamine can raise blood pressure. And very serious heart problems may occur if caffeine and **monoamine oxidase inhibitors** (MAO) are taken together. These drugs are used to treat **Parkinson's disease**, depression, and other psychiatric conditions. Consult with a pharmacist or physician about which drugs can interact with caffeine.

Because caffeine stimulates the nervous system, anyone taking other central nervous system (CNS) stimulants should be careful about using caffeine.

Nancy Ross-Flanigan

- CAH see **Congenital adrenal hyperplasia**
Caisson disease see **Decompression sickness**
Calcanal spurs see **Heel spurs**
Calcitonin see **Bone disorder drugs**
Calcium carbonate see **Antacids**

Calcium channel blockers

Definition

Calcium channel blockers are medicines that slow the movement of calcium into the cells of the heart and blood vessels. This, in turn, relaxes blood vessels, increases the supply of oxygen-rich blood to the heart, and reduces the heart's workload.

Purpose

Calcium channel blockers are used to treat high blood pressure, to correct abnormal heart rhythms, and to relieve the type of chest **pain** called **angina pectoris**. Physicians also prescribe calcium channel blockers to treat panic attacks and **bipolar disorder** (manic depressive illness) and to prevent **migraine headache**.

Precautions

Seeing a physician regularly while taking calcium channel blockers is important. The physician will check to make certain the medicine is working as it should and will watch for unwanted side effects. People who have high blood pressure often feel perfectly fine. However, they should continue to see their prescribing physician even when they feel well so that he can keep a close watch on their condition. They should also continue to take their medicine even when they feel fine.

Calcium channel blockers will not cure high blood pressure, but will help to control the condition. To avoid the serious health problems associated with high blood pressure, patients may have to take this type of medication for the rest of their lives. Furthermore, the blockers alone may not be enough. People with high blood pressure may also need to avoid certain foods and keep their weight under control. The health care professional who is treating the condition can offer advice as to what measures may be necessary. Patients being treated for high blood pressure should not change their **diets** without consulting their physicians.

Anyone taking calcium channel blockers for high blood pressure should not take any other prescription or over-the-counter medication without first checking with the prescribing physician, as some of these drugs may increase blood pressure.

Some people feel drowsy or less alert than usual when taking calcium channel blockers. Anyone who takes these drugs should not drive, use machines, or do anything else that might be dangerous until they have found out how the drugs affect them.

People who normally have chest pain when they **exercise** or exert themselves may not have the pain when they are taking calcium channel blockers. This could lead them to be more active than they should be. Anyone taking calcium channel blockers should therefore consult with the prescribing physician concerning how much exercise and activity may be considered safe.

Some people get headaches that last for a short time after taking a dose of this medication. This problem usually goes away during the course of treatment. If it does not, or if the headaches are severe, the prescribing physician should be informed.

Patients taking certain calcium channel blockers may need to check their pulse regularly, as the drugs may slow the pulse too much. If the pulse is too slow, circulation problems may result. The prescribing physician can show patients the correct way to check their pulse.

This type of medication may cause the gums to swell, bleed, or become tender. If this problem occurs, a medical physician or dentist should be consulted. To help prevent the problem, care should be taken when brushing and flossing the teeth. Regular dental check-ups and cleanings are also recommended.

Older people may be unusually sensitive to the effects of calcium channel blockers. This may increase the chance of side effects.

Special conditions

People with certain medical conditions or who are taking certain other medicines may develop problems if they also take calcium channel blockers. Before taking these drugs, the prescribing physician should be informed about any of these conditions:

ALLERGIES. Anyone who has had a previous unusual reaction to any calcium channel blocker should let his or her physician know before taking the drugs again. The physician should also be notified about any **allergies** to foods, dyes, preservatives, or other substances.

PREGNANCY. The effects of taking calcium channel blockers during **pregnancy** have not been studied in

KEY TERMS

Angina pectoris—A feeling of tightness, heaviness, or pain in the chest, caused by a lack of oxygen in the muscular wall of the heart.

Bipolar disorder—A severe mental illness, also known as manic depression, in which a person has extreme mood swings, ranging from a highly excited state—sometimes with a false sense of well-being—to depression.

Migraine—A throbbing headache that usually affects only one side of the head. Nausea, vomiting, increased sensitivity to light, and other symptoms often accompany migraine.

humans. However, in studies of laboratory animals, large doses of these drugs have been reported to cause **birth defects, stillbirth**, poor bone growth, and other problems when taken during pregnancy. Women who are pregnant or who may become pregnant should check with their physicians before using these drugs.

BREASTFEEDING. Some calcium channel blockers pass into breast milk, but there have been no reports of problems in nursing babies whose mothers were taking this type of medication. However, women who need to take this medicine and want to breastfeed their babies should check with their physicians.

OTHER MEDICAL CONDITIONS. Calcium channel blockers may worsen heart or blood vessel disorders.

The effects of calcium channel blockers may be greater in people with kidney or liver disease, as their bodies are slower to clear the drug from their systems.

Certain calcium channel blockers may also cause problems in people with a history of heart rhythm problems or with depression, **Parkinson's disease**, or other types of parkinsonism.

USE OF CERTAIN MEDICINES. Taking calcium channel blockers with certain other drugs may affect the way the drugs work or may increase the chance of side effects.

As with most medications, certain side effects are possible and some interactions with other substances may occur.

Side effects

Side effects are not common with this medicine, but some may occur. Minor discomforts, such as **dizziness**, lightheadedness, flushing, **headache**, and nausea, usual-

ly go away as the body adjusts to the drug and do not require medical treatment unless they persist or they are bothersome.

If any of the following side effects occur, the prescribing physician should be notified as soon as possible:

- breathing problems, coughing or wheezing
- irregular, fast, or pounding heartbeat
- slow heartbeat (less than 50 beats per minute)
- skin rash
- swollen ankles, feet, or lower legs

Other side effects may occur. Anyone who has unusual symptoms after taking calcium blockers should contact the prescribing physician.

Interactions

Calcium channel blockers may interact with a number of other medications. When this happens, the effects of one or both of the drugs may change or the risk of side effects may increase. Anyone who takes calcium channel blockers should not take any other prescription or non-prescription (over-the-counter) medicines without first checking with the prescribing physician. Substances that may interact with calcium channel blockers include:

- Diuretics (water pills). This type of medicine may cause low levels of potassium in the body, which may increase the chance of unwanted effects from some calcium channel blockers.
- Beta-blockers, such as atenolol (Tenormin), propranolol (Inderal), and metoprolol (Lopressor), used to treat high blood pressure, angina, and other conditions. Also, eye drop forms of **beta blockers**, such as timolol (Timoptic), used to treat **glaucoma**. Taking any of these drugs with calcium channel blockers may increase the effects of both types of medicine and may cause problems if either drug is stopped suddenly.
- Digitalis heart medicines. Taking these medicines with calcium channel blockers may increase the action of the heart medication.
- Medicines used to correct irregular heart rhythms, such as quinidine (Quinidex), disopyramide (Norpace), and procainamide (Procan, Pronestyl). The effects of these drugs may increase if used with calcium channel blockers.
- Anti-seizure medications such as carbamazepine (Tegretol). Calcium channel drugs may increase the effects of these medicines.
- Cyclosporine (Sandimmune), a medicine that suppresses the immune system. Effects may increase if this drug is taken with calcium channel blockers.
- Grapefruit juice may increase the effects of some calcium channel blockers.

The above list does not include every drug that may interact with calcium channel blockers. The prescribing physician or pharmacist will advise as to whether combining calcium channel blockers with any other prescription or nonprescription (over-the-counter) medication is appropriate or not.

Description

Calcium channel blockers are available only with a physician's prescription and are sold in tablet, capsule, and injectable forms. Some commonly used calcium channel blockers include amlopidine (Norvasc), diltiazem (Cardizem), isradipine (DynaCirc), nifedipine (Adalat, Procardia), nicardipine (Cardene), and verapamil (Calan, Isoptin, Verelan).

The recommended dosage depends on the type, strength, and form of calcium channel blocker and the condition for which it is prescribed. Correct dosage is determined by the prescribing physician and further information can be obtained from the pharmacist.

Calcium channel blockers should be taken as directed. Larger or more frequent doses should not be taken, nor should doses be missed. This medicine may take several weeks to noticeably lower blood pressure. The patient taking calcium channel blockers should keep taking the medicine, to give it time to work. Once it begins to work and symptoms improve, it should continue to be taken as prescribed.

This medicine should not be discontinued without checking with the prescribing physician. Some conditions may worsen when patients stop taking calcium channel blockers abruptly. The prescribing physician will advise as to how to gradually taper down before stopping the medication completely.

Risks

A report from the European Cardiology Society in 2000 found that patients taking certain calcium channel blockers had a 27% greater risk of **heart attack**, and a 26% greater risk of **heart failure** than patients taking other high blood pressure medicines. However, there are many patients affected by conditions that still make calcium channel blockers the best choice for them. The patient should discuss this issue with the prescribing physician.

Normal results

The expected result of taking a calcium channel blocker is to either correct abnormal heart rhythms, return blood pressure to normal, or relieve chest pain.

Resources

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National Heart, Lung and Blood Institute. <<http://www.nhlbi.nih.gov>>.

Deanna M. Swartout-Corbeil, R.N.

Calcium imbalance see **Hypercalcemia; Hypocalcemia**

Calcium polycarbophil see **Laxatives**

California flower essences see **Flower remedies**

Calluses see **Corns and calluses**

Calorie-modified diet see **Diets**

Calymmatobacteriosis see **Granuloma inguinale**

Campylobacter jejuni infection see **Campylobacteriosis**

Campylobacteriosis

Definition

Campylobacteriosis refers to infection by the group of bacteria known as *Campylobacter*. The term comes from the Greek word meaning "curved rod" referring to the bacteria's curved shape. The most common disease caused by these organisms is **diarrhea**, which most often affects children and younger adults. *Campylobacter* infections account for a substantial percent of food-borne illness encountered each year.

Description

There are over 15 different subtypes, all of which are curved Gram-negative rods. *C. jejuni* is the subtype that

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. This can worsen the condition of a patient with dysentery or colitis.

Fluoroquinolones—A relatively new group of antibiotics that have had good success in treating infections with many Gram-negative bacteria. One drawback is that they should not be used in children under 17 years of age, because of possible effect on bone growth.

Food-borne illness—A disease that is transmitted by eating or handling contaminated food.

Gram-negative—Refers to the property of many bacteria that causes them to not take up color with Gram's stain, a method which is used to identify bacteria. Gram-positive bacteria which take up the

stain turn purple, while Gram-negative bacteria which do not take up the stain turn red.

Guillain-Barré syndrome—Progressive and usually reversible paralysis or weakness of multiple muscles usually starting in the lower extremities and often ascending to the muscles involved in respiration. The syndrome is due to inflammation and loss of the myelin covering of the nerve fibers, often associated with an acute infection.

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 even 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.

most often causes gastrointestinal disease. However, some species such as *C. fetus* produce disease outside the intestine, particularly in those with altered immune systems, such as people with AIDS, cancer, and liver disease.

Campylobacter are often found in the intestine of animals raised for food produce and pets. Infected animals often have no symptoms. Chickens are the most common source of human infection. It is estimated that 1% of the general population is infected each year.

Causes and symptoms

Improper or incomplete food preparation is the most common way the disease is spread, with poultry accounting for over half the cases. Untreated water and raw milk are also potential sources.

The incubation period after exposure is from one to 10 days. A day or two of mild **fever**, muscle aches, and **headache** occur before intestinal symptoms begin. Diarrhea with or without blood and severe abdominal cramps are the major intestinal symptoms. The severity of symptoms is variable, ranging from only mild fever to **dehydration** and rarely **death** (mainly in the very young or old). The disease usually lasts about one week, but per-

sists longer in about 20% of cases. At least 10% will have a relapse, and some patients will continue to pass the bacteria for several weeks.

Complications

Dehydration is the most common complication. Especially at the extremes of age, this should be watched for and treated with either Oral Rehydration Solution or intravenous fluid replacement.

Infection may also involve areas outside the intestine. This is unusual, except for infections with *C. fetus*. *C. fetus* infections tend to occur in those who have diseases of decreased immunity such as AIDS, cancer, etc. This subtype is particularly adapted to protect itself from the body's defenses.

Areas outside the intestine that may be involved are:

- Nervous system involvement either by direct infection of the meninges (outer covering of the spinal and brain) or more commonly by producing the **Guillain-Barré syndrome** (progressive and reversible **paralysis** or weakness of many muscles). In fact, *Campylobacter* may be responsible for 40% of the reported cases of this syndrome.

- Joint inflammation can occur weeks later (leading to an unusual form of arthritis).
- Infection of vessels and heart valves is a special characteristic of *C. fetus*. Immunocompromised patients may develop repeated episodes of passage of bacteria into the bloodstream from these sites of infection.
- The gallbladder, pancreas, and bone may be affected.

Diagnosis

Campylobacter is only one of 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.

Treatment

The first aim of treatment is to keep up **nutrition** and avoid dehydration. Medications used to treat diarrhea by decreasing intestinal motility, such as Loperamide or Diphenoxylate are also useful, but should only be used with the advice of a physician. **Antibiotics** are of value, if started within three days of onset of symptoms. They are indicated for those with severe or persistent symptoms. Either an erythromycin type drug or one of the **fluoroquinolones** (such as ciprofloxacin) for five to seven days are the accepted therapies.

Prognosis

Most patients with *Campylobacter* infection rapidly recover without treatment. For certain groups of patients, infection becomes chronic and requires repeated courses of antibiotics.

Prevention

Good hand washing technique as well as proper preparation and cooking of food is the best way to prevent infection.

Resources

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ORGANIZATIONS

Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. (800) 311-3435, (404) 639-3311. <<http://www.cdc.gov>>.

OTHER

Centers for Disease Control. <<http://www.cdc.gov/nccdp/ph/dt/ddthome.htm>>.

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Cancer

Definition

Cancer is not just one disease, but a large group of almost one hundred diseases. Its two main characteristics are uncontrolled growth of the cells in the human body and the ability of these cells to migrate from the original site and spread to distant sites. If the spread is not controlled, cancer can result in **death**.

Description

One out of every four deaths in the United States is from cancer. It is second only to heart disease as a cause of death in the states. About 1.2 million Americans are diagnosed with cancer annually; more than 500,000 die of cancer annually.

Cancer can attack anyone. Since the occurrence of cancer increases as individuals age, most of the cases are seen in adults, middle-aged or older. Sixty percent of all cancers are diagnosed in people who are older than 65 years of age. The most common cancers are skin cancer, lung cancer, **colon cancer**, **breast cancer** (in women), and **prostate cancer** (in men). In addition, cancer of the kidneys, ovaries, uterus, pancreas, bladder, rectum, and blood and lymph node cancer (leukemias and lymphomas) are also included among the 12 major cancers that affect most Americans.

Cancer, by definition, is a disease of the genes. A gene is a small part of DNA, which is the master molecule of the cell. Genes make "proteins," which are the ultimate workhorses of the cells. It is these proteins that allow our bodies to carry out all the many processes that permit us to breathe, think, move, etc.

Throughout people's lives, the cells in their bodies are growing, dividing, and replacing themselves. Many genes produce proteins that are involved in controlling the processes of cell growth and division. An alteration (mutation) to the DNA molecule can disrupt the genes and produce faulty proteins. This causes the cell to become abnormal and lose its restraints on growth. The abnormal cell begins to divide uncontrollably and eventually forms a new growth known as a "tumor" or neoplasm (medical term for cancer meaning "new growth").

In a healthy individual, the immune system can recognize the neoplastic cells and destroy them before they get a chance to divide. However, some mutant cells may escape immune detection and survive to become tumors or cancers.

Tumors are of two types, benign or malignant. A benign tumor is not considered cancer. It is slow growing, does not spread or invade surrounding tissue, and once it is removed, it doesn't usually recur. A malignant tumor, on the other hand, is cancer. It invades surrounding tissue and spreads to other parts of the body. If the cancer cells have spread to the surrounding tissues, then, even after the malignant tumor is removed, it generally recurs.

A majority of cancers are caused by changes in the cell's DNA because of damage due to the environment. Environmental factors that are responsible for causing the initial mutation in the DNA are called carcinogens, and there are many types.

There are some cancers that have a genetic basis. In other words, an individual could inherit faulty DNA from his parents, which could predispose him to getting cancer. While there is scientific evidence that both factors (environmental and genetic) play a role, less than 10% of all cancers are purely hereditary. Cancers that are known to have a hereditary link are breast cancer, colon cancer, **ovarian cancer**, and uterine cancer. Besides genes, certain physiological traits could be inherited and could contribute to cancers. For example, inheriting fair skin makes a person more likely to develop skin cancer, but only if they also have prolonged exposure to intensive sunlight.

There are several different types of cancers:

- Carcinomas are cancers that arise in the epithelium (the layers of cells covering the body's surface and lining the internal organs and various glands). Ninety percent of human cancers fall into this category. Carcinomas can be

subdivided into two types: adenocarcinomas and squamous cell carcinomas. Adenocarcinomas are cancers that develop in an organ or a gland, while squamous cell carcinomas refer to cancers that originate in the skin.

- Melanomas also originate in the skin, usually in the pigment cells (melanocytes).
- Sarcomas are cancers of the supporting tissues of the body, such as bone, muscle and blood vessels.
- Cancers of the blood and lymph glands are called leukemias and lymphomas respectively.
- Gliomas are cancers of the nerve tissue.

Causes and symptoms

The major risk factors for cancer are: tobacco, alcohol, diet, sexual and reproductive behavior, infectious agents, family history, occupation, environment and pollution.

According to the estimates of the American Cancer Society (ACS), approximately 40% of the cancer deaths in 1998 will be due to tobacco and excessive alcohol use. An additional one-third of the deaths will be related to diet and **nutrition**. Many of the one million skin cancers that are expected to be diagnosed in 1998 will be due to over-exposure to ultraviolet light from the sun's rays.

Tobacco

Eighty to ninety percent of the lung cancer cases occur in smokers. **Smoking** has also been shown to be a contributory factor in cancers of upper respiratory tract, esophagus, larynx, bladder, pancreas, and probably liver, stomach, and kidney as well. Recently, scientists have also shown that second-hand smoke (or passive smoking) can increase one's risk of developing cancer.

Alcohol

Excessive consumption of alcohol is a risk factor in certain cancers, such as **liver cancer**. Alcohol, in combination with tobacco, significantly increases the chances that an individual will develop mouth, pharynx, larynx and esophageal cancers.

Diet

Thirty-five percent of all cancers are due to dietary causes. Excessive intake of fat leading to **obesity** has been associated with cancers of the breast, colon, rectum, pancreas, prostate, gall bladder, ovaries and uterus.

Sexual and reproductive behavior

The human papilloma virus, which is sexually transmitted, has been shown to cause cancer of the cervix.

Having too many sex partners and becoming sexually active early has been shown to increase one's chances of contracting this disease. In addition, it has also been shown that women who don't have children or have children late in life have an increased risk for both ovarian and breast cancer.

Infectious agents

In the last 20 years, scientists have obtained evidence to show that approximately 15% of the world's cancer deaths can be traced to viruses, bacteria, or parasites. The most common cancer-causing pathogens and the cancers associated with them are shown in table form.

Family history

Certain cancers like breast, colon, ovarian and uterine cancer recur generation after generation in some families. A few cancers, such as the **eye cancer** "retinoblastoma," a type of colon cancer, and a type of breast cancer known as "early-onset breast cancer," have been shown to be linked to certain genes that can be tracked within a family. It is therefore possible that inheriting particular genes makes a person susceptible to certain cancers.

Occupational hazards

There is evidence to prove that certain occupational hazards account for 4% of all cancer deaths. For example, asbestos workers have an increased incidence of lung cancer. Similarly, a higher likelihood of getting **bladder cancer** is associated with dye, rubber and gas workers; skin and lung cancer with smelters, gold miners and arsenic workers; leukemia with glue and varnish workers; liver cancer with PVC manufacturers; and lung, bone and bone marrow cancer with radiologists and uranium miners.

Environment

Radiation is believed to cause 1–2% of all cancer deaths. Ultra-violet radiation from the sun accounts for a majority of melanoma deaths. Other sources of radiation are x rays, radon gas, and ionizing radiation from nuclear material.

Pollution

Several studies have shown that there is a well-established link between asbestos and cancer. Chlorination of water may account for a small rise in cancer risk. However, the main danger from pollution occurs when dangerous chemicals from the industries escape into the surrounding environment. It has been estimated that 1% of cancer deaths are due to air, land and water pollution.

Frequency Of Cancer-Related Death

Cancer Site	Number of Deaths Per Year
Lung	160,100
Colon and rectum	56,500
Breast	43,900
Prostate	39,200
Pancreas	28,900
Lymphoma	26,300
Leukemia	21,600
Brain	17,400
Stomach	13,700
Liver	13,000
Esophagus	11,900
Bladder	12,500
Kidney	11,600
Multiple myeloma	11,300

Cancer is a progressive disease, and goes through several stages. Each stage may produce a number of symptoms. Some symptoms are produced early and may occur due to a tumor that is growing within an organ or a gland. As the tumor grows, it may press on the nearby nerves, organs and blood vessels. This causes **pain** and some pressure which may be the earliest warning signs of cancer.

Despite the fact that there are several hundred different types of cancers, producing very different symptoms, the ACS has established the following seven symptoms as possible warning signals of cancer:

- changes in the size, color, or shape of a wart or a mole
- a sore that does not heal
- persistent **cough**, hoarseness, or **sore throat**
- a lump or thickening in the breast or elsewhere
- unusual bleeding or discharge
- chronic **indigestion** or difficulty in swallowing
- any change in bowel or bladder habits

Many other diseases, besides cancer, could produce the same symptoms. However, it is important to have these symptoms checked, as soon as possible, especially if they linger. The earlier a cancer is diagnosed and treated, the better the chance of it being cured. Many cancers such as breast cancer may not have any early symptoms. Therefore, it is important to undergo routine screening tests such as breast self-exams and mammograms.

Diagnosis

Diagnosis begins with a thorough **physical examination** and a complete medical history. The doctor will observe, feel and palpate (apply pressure by touch) different parts of the body in order to identify any variations from the normal size, feel and texture of the organ or tissue.

As part of the physical exam, the doctor will inspect the oral cavity or the mouth. By focusing a light into the mouth, he will look for abnormalities in color, moisture, surface texture, or presence of any thickening or sore in the lips, tongue, gums, the hard palate on the roof of the mouth, and the throat. To detect **thyroid cancer**, the doctor will observe the front of the neck for swelling. He may gently manipulate the neck and palpate the front and side surfaces of the thyroid gland (located at the base of the neck) to detect any nodules or tenderness. As part of the physical examination, the doctor will also palpate the lymph nodes in the neck, under the arms and in the groin. Many illnesses and cancers cause a swelling of the lymph nodes.

The doctor may conduct a thorough examination of the skin to look for sores that have been present for more than three weeks and that bleed, ooze, or crust; irritated patches that may itch or hurt, and any change in the size of a wart or a mole.

Examination of the female pelvis is used to detect cancers of the ovaries, uterus, cervix, and vagina. In the visual examination, the doctor looks for abnormal discharges or the presence of sores. Then, using gloved hands the physician palpates the internal pelvic organs such as the uterus and ovaries to detect any abnormal masses. Breast examination includes visual observation where the doctor looks for any discharge, unevenness, discoloration, or scaling. The doctor palpates both breasts to feel for masses or lumps.

For males, inspection of the rectum and the prostate is also included in the physical examination. The doctor inserts a gloved finger into the rectum and rotates it slowly to feel for any growths, tumors, or other abnormalities. The doctor also conducts an examination of the testes, where the doctor observes the genital area and looks for swelling or other abnormalities. The testicles are palpated to identify any lumps, thickening or differences in the size, weight and firmness.

If the doctor detects an abnormality on physical examination, or the patient has some symptom that could be indicative of cancer, the doctor may order diagnostic tests.

Laboratory studies of sputum (sputum cytology), blood, urine, and stool can detect abnormalities that may indicate cancer. Sputum cytology is a test where the phlegm that is coughed up from the lungs is microscopically examined. It is often used to detect lung cancer. A blood test for cancer is easy to perform, usually inexpensive and risk-free. The blood sample is obtained by a lab technician or a doctor by inserting a needle into a vein and is relatively painless. Blood tests can be either specific or non-specific. Often times, in certain cancers, the cancer cells release particular proteins (called **tumor markers**) and blood tests can be used to detect the pres-

ence of these tumor markers. However, with a few exceptions, tumor markers are not used for routine screening of cancers, because several non-cancerous conditions also produce positive results. Blood tests are generally more useful in monitoring the effectiveness of the treatment, or in following the course of the disease and detecting recurrent disease.

Imaging tests such as **computed tomography scans** (CT scans), **magnetic resonance imaging** (MRI), ultrasound and fiberoptic scope examinations help the doctors determine the location of the tumor even if it is deep within the body. Conventional x rays are often used for initial evaluation, because they are relatively cheap, painless and easily accessible. In order to increase the information obtained from a conventional x ray, air or a dye (such as barium or iodine) may be used as a contrast medium to outline or highlight parts of the body.

The most definitive diagnostic test is the biopsy, wherein a piece of tissue is surgically removed for microscope examination. Besides confirming a cancer, the biopsy also provides information about the type of cancer, the stage it has reached, the aggressiveness of the cancer and the extent of its spread. Since a biopsy provides the most accurate analysis, it is considered the gold standard of diagnostic tests.

Screening examinations conducted regularly by healthcare professionals can result in the detection of cancers of the breast, colon, rectum, cervix, prostate, testis, tongue, mouth, and skin at early stages, when treatment is more likely to be successful. Some of the routine screening tests recommended by the ACS are **sigmoidoscopy** (for colorectal cancer), **mammography** (for breast cancer), pap smear (for **cervical cancer**), and the PSA test (for prostate cancer). Self-examinations for cancers of the breast, testes, mouth, and skin can also help in detecting the tumors before the symptoms become serious.

A recent revolution in molecular biology and cancer genetics has contributed a great deal to the development of several tests designed to assess one's risk of getting cancers. These new techniques include **genetic testing**, where molecular probes are used to identify mutations in certain genes that have been linked to particular cancers. At present, however, there are a lot of limitations to genetic testing and its utility appears ambiguous, emphasizing the need to develop better strategies for early detection.

Treatment

The aim of cancer treatment is to remove all or as much of the tumor as possible and to prevent the recurrence or spread of the primary tumor. While devising a treatment plan for cancer, the likelihood of curing the

Common Pathogens And The Cancers Associated With Them

Causative Agent	Type of Cancer
Viruses	
Papillomaviruses	Cancer of the cervix
Hepatitis B virus	Liver cancer
Hepatitis C virus	Liver cancer
Epstein-Barr virus	Burkitt's lymphoma
Cancers of the upper pharynx	Hodgkin's lymphoma, Non-Hodgkin's lymphoma, Gastric cancers
Human immunodeficiency virus (HIV)	Kaposi's sarcoma lymphoma
Bacteria	
Helicobacter pylori	Stomach cancer lymphomas

cancer has to be weighed against the side effects of the treatment. If the cancer is very aggressive and a cure is not possible, then the treatment should be aimed at relieving the symptoms and controlling the cancer for as long as possible.

Cancer treatment can take many different forms, and it is always tailored to the individual patient. The decision on which type of treatment is the most appropriate depends on the type and location of cancer, the extent to which it has already spread, the patient's age, sex, general health status and personal treatment preferences. The major types of treatment are: surgery, radiation, **chemotherapy**, immunotherapy, hormone therapy, and bone-marrow transplantation.

Surgery

Surgery is the removal of a visible tumor and is the most frequently used cancer treatment. It is most effective when a cancer is small and confined to one area of the body.

Surgery can be used for many purposes.

- Treatment. Treatment of cancer by surgery involves removal of the tumor to cure the disease. This is typically done when the cancer is localized to a discrete area. Along with the cancer, some part of the normal surrounding tissue is also removed to ensure that no cancer cells remain in the area. Since cancer usually spreads via the lymphatic system, adjoining lymph nodes may be examined and sometimes they are removed as well.
- Preventive surgery. Preventive or prophylactic surgery involves removal of an abnormal looking area that is likely to become malignant over time. For example, 40% of the people with a colon disease known as **ulcerative colitis**, ultimately die of colon cancer. Rather than live with the fear of developing colon cancer, these people may choose to have their colons removed and reduce the risk significantly.

- Diagnostic purposes. The most definitive tool for diagnosing cancer is a biopsy. Sometimes, a biopsy can be performed by inserting a needle through the skin. However, at other times, the only way to obtain some tissue sample for biopsy is by performing a surgical operation.
- Cytoreductive surgery is a procedure where the doctor removes as much of the cancer as possible, and then treats the remaining with **radiation therapy** or chemotherapy or both.
- Palliative surgery is aimed at curing the symptoms, not the cancer. Usually, in such cases, the tumor is so large or has spread so much that removing the entire tumor is not an option. For example, a tumor in the abdomen may be so large that it may press on and block a portion of the intestine, interfering with digestion and causing pain and vomiting. "Debulking surgery" may remove a part of the blockage and relieve the symptoms. In tumors that are dependent on hormones, removal of the organs that secrete the hormones is an option. For example, in prostate cancer, the release of testosterone by the testicles stimulates the growth of cancerous cells. Hence, a man may undergo an "orchectomy" (removal of testicles) to slow the progress of the disease. Similarly, in a type of aggressive breast cancer, removal of the ovaries (**oophorectomy**) will stop the synthesis of hormones from the ovaries and slow the progression of the cancer.

Radiation

Radiation kills tumor cells. Radiation is used alone in cases where a tumor is unsuitable for surgery. More often, it is used in conjunction with surgery and chemotherapy. Radiation can be either external or internal. In the external form, the radiation is aimed at the tumor from outside the body. In internal radiation (also known as brachytherapy), a radioactive substance in the form of pellets or liquid is placed at the cancerous site by means of a pill, injection or insertion in a sealed container.

Chemotherapy

Chemotherapy is the use of drugs to kill cancer cells. It destroys the hard-to-detect cancer cells that have spread and are circulating in the body. Chemotherapeutic drugs can be taken either orally (by mouth) or intravenously, and may be given alone or in conjunction with surgery, radiation or both.

When chemotherapy is used before surgery or radiation, it is known as primary chemotherapy or “neoadjuvant chemotherapy.” An advantage of neoadjuvant chemotherapy is that since the cancer cells have not been exposed to anti-cancer drugs, they are especially vulnerable. It can therefore be used effectively to reduce the size of the tumor for surgery or target it for radiation. However, the toxic effects of neoadjuvant chemotherapy are severe. In addition, it may make the body less tolerant to the side effects of other treatments that follow such as radiation therapy. The more common use of chemotherapy is adjuvant therapy, which is given to enhance the effectiveness of other treatments. For example, after surgery, adjuvant chemotherapy is given to destroy any cancerous cells that still remain in the body.

Immunotherapy

Immunotherapy uses the body’s own immune system to destroy cancer cells. This form of treatment is being intensively studied in clinical trials and is not yet widely available to most cancer patients. The various immunological agents being tested include substances produced by the body (such as the interferons, interleukins, and growth factors), monoclonal antibodies and vaccines. Unlike traditional vaccines, cancer vaccines do not prevent cancer. Instead, they are designed to treat people who already have the disease. Cancer vaccines work by boosting the body’s immune system and training the immune cells to specifically destroy cancer cells.

Hormone therapy

Hormone therapy is standard treatment for some types of cancers that are hormone-dependent and grow faster in the presence of particular hormones. These include cancer of the prostate, breast, and uterus. Hormone therapy involves blocking the production or action of these hormones. As a result the growth of the tumor slows down and survival may be extended for several months or years.

Bone marrow transplantation

The bone marrow is the tissue within the bone cavities that contains blood-forming cells. Healthy bone marrow tissue constantly replenishes the blood supply

and is essential to life. Sometimes, the amount of drugs or radiation needed to destroy cancer cells also destroys bone marrow. Replacing the bone marrow with healthy cells counteracts this adverse effect. A bone marrow transplant is the removal of marrow from one person and the transplant of the blood-forming cells either to the same person or to someone else. Bone-marrow transplantation, while not a therapy in itself, is often used to “rescue” a patient, by allowing those with cancer to undergo very aggressive therapy.

Many different specialists generally work together as a team to treat cancer patients. An oncologist is a physician who specializes in cancer care. The oncologist provides chemotherapy, hormone therapy, and any other non-surgical treatment that does not involve radiation. The oncologist often serves as the primary physician and coordinates the patient’s treatment plan.

The radiation oncologist specializes in using radiation to treat cancer, while the surgical oncologist performs the operations needed to diagnose or treat cancer. Gynecologist-oncologists and pediatric-oncologists, as their titles suggest, are physicians involved with treating women’s and children’s cancers respectively. Many other specialists may also be involved in the care of a cancer patient. For example, radiologists specialize in the use of x rays, ultrasounds, computed tomography scans (CT scans), MRI imaging and other techniques that are used to diagnose cancer. Hematologists specialize in disorders of the blood and are consulted in case of blood cancers and bone marrow cancers. The samples that are removed for biopsy are sent to a laboratory, where a pathologist examines them to determine the type of cancer and extent of the disease. Only some of the specialists who are involved with cancer care have been mentioned above. There are many other specialties, and virtually any type of medical or surgical specialist may become involved with care of the cancer patient should it become necessary.

Alternative treatment

There are a multitude of alternative treatments available to help the person with cancer. They can be used in conjunction with, or separate from, surgery, chemotherapy, and radiation therapy. Alternative treatment of cancer is a complicated arena and a trained health practitioner should be consulted.

Although the effectiveness of complementary therapies such as **acupuncture** in alleviating cancer pain has not been clinically proven, many cancer patients find it safe and beneficial. Bodywork therapies such as massage and **reflexology** ease muscle tension and may alleviate the side effects such as **nausea and vomiting**. **Homeopathy** and herbal remedies used in Chinese traditional

herbal medicine have also been shown to alleviate some of the side effects of radiation and chemotherapy and are being recommended by many doctors.

Certain foods including many vegetables, fruits and grains are believed to offer protection against various cancers. However, isolation of the individual constituent of vegetables and fruits that are anti-cancer agents has proven difficult. In laboratory studies, **vitamins** such as A, C and E, as well as compounds such as isothiocyanates and dithiolthiones found in broccoli, cauliflower, and cabbage, and beta-carotene found in carrots have been shown to protect against cancer. Studies have shown that eating a diet rich in fiber as found in fruits and vegetables reduces the risk of colon cancer. **Exercise** and a low fat diet help control weight and reduce the risk of endometrial, breast, and colon cancer.

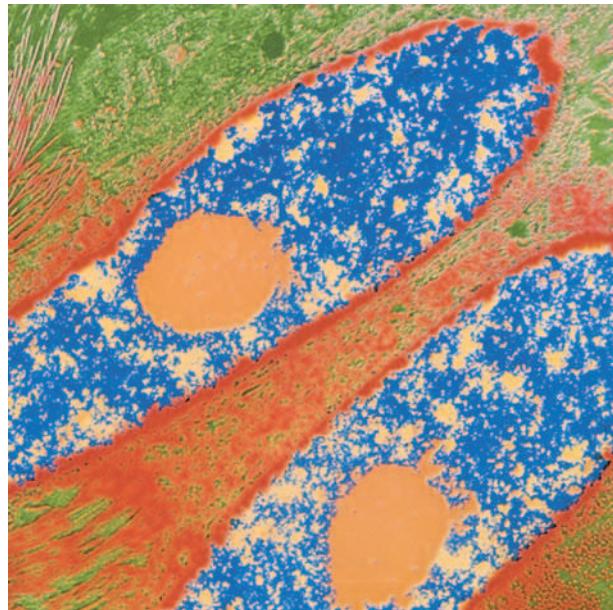
Certain drugs, which are currently being used for treatment, could also be suitable for prevention. For example, the drug tamoxifen (Nolvadex), which has been very effective against breast cancer, is currently being tested by the National Cancer Institute for its ability to prevent cancer. Similarly, retinoids derived from vitamin A are being tested for their ability to slow the progression or prevent head and neck cancers. Certain studies have suggested that cancer incidence is lower in areas where soil and foods are rich in the mineral selenium. More trials are needed to explain these intriguing connections.

Prognosis

“Lifetime risk” is the term that cancer researchers use to refer to the probability that an individual over the course of a lifetime will develop cancer or die from it. In the United States, men have a one in two lifetime risk of developing cancer, and for women the risk is one in three. Overall, African-Americans are more likely to develop cancer than whites. African-Americans are also 30% more likely to die of cancer than whites.

Most cancers are curable if detected and treated at their early stages. A cancer patient’s prognosis is affected by many factors, particularly the type of cancer the patient has, the stage of the cancer, the extent to which it has metastasized and the aggressiveness of the cancer. In addition, the patient’s age, general health status and the effectiveness of the treatment being pursued are also important factors.

To help predict the future course and outcome of the disease and the likelihood of recovery from the disease, doctors often use statistics. The five-year survival rates are the most common measures used. The number refers to the proportion of people with cancer who are expected to be alive, five years after initial diagnosis, compared



A transmission electron micrograph (TEM) of two spindle cell nuclei from a human sarcoma. Sarcomas are cancers of the connective tissue (bone, nerves, smooth muscle). (Photograph by Dr. Brian Eyden, Photo Researchers, Inc. Reproduced by permission.)

with a similar population that is free of cancer. It is important to note that while statistics can give some information about the average survival experience of cancer patients in a given population, it cannot be used to indicate individual prognosis, because no two patients are exactly alike.

Prevention

According to nutritionists and epidemiologists from leading universities in the United States, a person can reduce the chances of getting cancer by following some simple guidelines:

- eating plenty of vegetables and fruits
- exercising vigorously for at least 20 minutes every day
- avoiding excessive weight gain
- avoiding tobacco (even second hand smoke)
- decreasing or avoiding consumption of animal fats and red meats
- avoiding excessive amounts of alcohol
- avoiding the midday sun (between 11 A.M. and 3 P.M.) when the sun’s rays are the strongest
- avoiding risky sexual practices
- avoiding known carcinogens in the environment or work place

KEY TERMS

Benign—A growth that does not spread to other parts of the body. Recovery is favorable with treatment.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes.

Bone marrow—Spongy material that fills the inner cavities of the bones. The progenitors of all the blood cells are produced in this bone marrow.

Carcinogen—Any substance capable of causing cancer by mutating the cell's DNA.

Chemotherapy—Treatment with drugs that are anti cancer.

Epithelium—The layer of cells covering the body's surface and lining the internal organs and various glands.

Hormone therapy—Treatment of cancer by inhibiting the production of hormones such as testosterone and estrogen.

Immunotherapy—Treatment of cancer by stimulating the body's immune defense system.

Malignant—A general term for cells that can dislodge from the original tumor, invade and destroy other tissues and organs.

Metastasis—The spread of cancer from one part of the body to another.

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

Sore—An open wound or a bruise or lesion on the skin.

Tumor—An abnormal growth resulting from a cell that lost its normal growth control restraints and started multiplying uncontrollably.

X rays—High-energy radiation used in high doses, either to diagnose or treat disease.

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Cancer chemotherapy drugs see **Anticancer drugs**

Cancer therapy, definitive

Definition

Definitive **cancer** therapy is a treatment plan designed to potentially cure cancer using one or a combination of interventions including surgery, radiation, chemical agents, or biological therapies.

Purpose

The primary purpose of definitive care is to establish a cure and to destruct and remove all cancer cells from the infected person.

Surgery is not only a diagnostic tool, but also used for **tumor removal**. The surgeon usually identifies potential candidates for tumor removal and repairs intraoperatively (during the operation procedure). Surgery can be curative for some stomach, genital/urinary, thyroid, breast, skin, and central nervous system cancers. The best chance for a surgical cure is usually with the first opera-

Rosalyn Carson-DeWitt

tion. It is essential that the cancer surgeon (oncologic surgeon) be experienced in the specific procedure.

Radiation therapy is commonly administered to approximately 50% of cancer patients during the course of illness. It can be used as the sole method of cure for tumors in the mouth and neighboring structures in the oral cavity, vagina, prostate, cervix, esophagus, **Hodgkin's disease**, and certain types of cancer in the spinal cord and brain. Research and clinical trials have demonstrated that combination treatment is more effective than radiotherapy alone.

Chemotherapy is curative for only a small percentage of cancers. It is most effective for **choriocarcinoma**, cancer of the testis, some types of lymphomas, and cancer of skeletal muscles.

Biological therapies are a new and promising direction for cancer cures. Usually when cancer cells grow they manage to derive a blood supply that allows passage of nutrients promoting continuation of abnormal cancer growth. Research that focuses on destroying these blood vessels is called angiogenesis. Cutting off the blood supply has been shown to destroy tumors, since this stops the flow of essential nutrients required for cancer growth. Use of certain growth factors can also stimulate self-destructive pathways in cancer cells (apoptosis). **Gene therapy** is directed towards inhibiting specific cellular signals that promote cancer cell multiplication.

Precautions

Surgical resection requires an experienced surgeon, preoperative assessment, imaging studies, and delicate operative technique. Care should be taken during the procedure to avoid unnecessary tumor manipulation, which can cause cancer cells to infiltrate adjacent structures. If manipulation is excessive, cells can enter nearby areas for future re-growth. Accurate isolation of the tumor can also help to avoid contamination of the surgical area. Early ligation of the blood supply to the tumor is an essential component of a surgical cure.

Radiotherapy requires extensive treatment planning and imaging. Care must be taken to localize the cancer field while attempting to spare destruction of normal tissue. This requires image monitoring and exact positioning during radiation treatment sessions.

Chemotherapy usually causes destruction of normal cells, and cancer cells can become immune to chemical destruction. Side effects and patient tolerance issues are typically anticipated and dosages may have to be specifically altered. Very few chemotherapeutic agents offer curative responses.

Biological therapies may cause patient toxicity resulting in extensive side effects. This can occur since

the optimal dose may be exceedingly elevated above patient tolerance.

Description

Surgery

Surgical removal of the tumor must be performed with care and accuracy. The surgeon must avoid over manipulation of the surgical field. Too much movement within the area can cause cancer cell displacement into surrounding tissue. If this occurs and no further treatment is indicated, the tumor may grow again. The surgeon should also perform an assessment concerning tissue removal around the cancer site. Tissue around the site may not by inspection seem cancerous, but adjacent structures may have cancer cells and surrounding tissue removal is usually part of the operative procedure. Pieces of tumor and the surrounding area are analyzed microscopically during the operation for cell type. An adequate resection (removal of tissue) will reveal normal cells in the specimens analyzed from areas bordering the cancerous growth. Surgery can also help to decrease the tumor bulk and, along with other treatment measures, may provide a cure for certain cancers.

Not only can surgery be curative for some cancers, but it is an essential diagnostic tool that must be assessed intraoperatively since microscopic analysis will guide the surgeon concerning tumor and surrounding tissue removal. These diagnostic procedures include an aspiration biopsy, which inserts a needle to extract (aspirate) fluid contained inside a cancerous growth; a needle biopsy uses a specialized needle to obtain a core tissue specimen; an incision biopsy removes a section from a large tumor; and an excision biopsy removes the entire tumor. The surgeon can also take samples of neighboring lymph nodes. Cancer in surrounding lymph nodes is an important avenue for distant spread of cancer to other areas. If microscopic analysis determines the presence of cancer cells in lymph nodes then the surgeon may decide to perform a more aggressive surgical approach.

Radiation therapy

Similar to surgical intervention, radiotherapy is a localized treatment. It involves the administration of ionizing radiation to a solid tumor location. This generates reactive oxygen molecules, causing the destruction of DNA in local cells. There are three commonly used radiotherapy beams: gamma rays from a linear accelerator machine produce a focused beam; orthovoltage rays are of less energy, thus penetrate less and typically deliver higher doses to superficial tissues (efficient for treating skin cancers); and megavoltage rays are high energy producing beams and can penetrate deeply situated inter-

KEY TERMS

Bone marrow suppression—A decrease in cells responsible for providing immunity, carrying oxygen and those responsible for normal blood clotting.

DNA—The molecule responsible for cell multiplication.

Titrate—To analyze the best end point (for dose) for a medication.

nal organs, while sparing extensive skin damage. Two common routes can deliver radiation. Brachytherapy delivers radiation to a local area by placing radioactive materials within close proximity to the cancerous site. Teletherapy delivers radiation to a specific area using an external beam machine.

Chemotherapy

Curative chemotherapy usually requires multiple administrations of the chemical agent. Chemotherapy or systemic therapy is administered in the blood and circulates through the entire body. The choice of chemotherapeutic agents depends on the specific type of cancer. Chemotherapy is more commonly used for metastatic (malignant cancer which has spread to other areas beyond the primary site of cancer growth) disease, since very few cancers are cured by systemic therapy.

Biologic therapy

Biologic therapies primarily function to alter the patient's response to cancer. These treatments are mostly investigations and there are numerous research protocols studying the effects of biologic treatments. These protocols usually have strict admission criteria that may exclude potential candidates who can benefit from treatment. These treatments tend to stimulate specific immune cells or immune chemicals to destroy cancer cells.

Preparation

For all treatment modalities imaging studies, biopsy, and constant blood analysis is essential before, during, and after treatments. Surgical candidates should undergo extensive pre-operative evaluation with imaging studies, blood chemistry analysis, stabilized health status, and readiness of staff for any potential complications and cell biopsy analysis. Patients with other pre-existing chronic disease may require intensive post-operative monitoring.

For radiotherapy, the patient undergoes extensive imaging studies. Additional planning strategies include beam

localization to spare normal tissues, calibration of fractionated doses, and specific positioning during treatment sessions.

Patients who receive curative chemotherapy should be informed of possible side effects associated with the chemotherapeutic agent. Patients should also be informed of temporary lifestyle changes and medications that may offer some symptomatic relief.

Patients undergoing biologic therapies are usually advised of potential side effects, treatment cycles and specific tests for monitoring progress according to the specific research protocol.

Aftercare

Patients will typically be evaluated by imaging studies, blood analysis, **physical examination**, and health improvement. These follow-up visits usually occur at specific time intervals during the course of treatment. Surgical patients may require closer observation during the initial post-operative period to avoid potential complications. Reconstructive surgery can be considered to improve appearance and restore function. Certain surgical procedures (such as flaps and microsurgery of blood vessels) can restore new tissues to a previous surgery site.

Risks

Surgical risks

Surgical therapy can be both disfiguring and disabling. Many normal tissues can be adversely affected by radiotherapy. Side effects that commonly occur shortly after a treatment cycle include nausea, vomiting, **fatigue**, loss of appetite, and bone marrow suppression (a decrease in the cells that provide defense against infections and those which carry oxygen to cells).

Radiation risks

Radiotherapy can also cause difficulty swallowing, oral gum disease, and **dry mouth**. Additionally, radiation therapy can cause damage to local structures within the irradiated field.

Chemotherapy risks

Chemotherapy commonly causes bone marrow suppression. Additionally, a cell called platelets—important for normal blood clotting—may be significantly lowered, causing patients to bleed. This may be problematic enough to limit the treatment course. Bone marrow suppression can increase susceptibility to infection and also cause **infertility**. Patients commonly have bouts of **nausea and vomiting** shortly after a treatment session. Rapidly multiplying normal cells are also affected such as skin cells (causing blistering and ulceration) and hair cells (causing loss of hair, a condition called **alopecia**).

Biologic therapies risks

Biologic therapies can cause patients to develop suppression of cells that help the body fight against infection. Administration of certain chemicals that have anti-cancer effects can cause heart damage. Injection of killer immune cells (lymphokine-activated killer cells) may cause bone marrow suppression, and the host may reject the newly introduced cells.

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can also alleviate pain, bleeding, and obstruction of neighboring areas. Chemotherapy may be helpful to reduce tumor size and provide some reduction to metastatic disease. Long-term chemotherapy patients develop drug resistance, a situation that renders chemotherapeutic treatments ineffective. If this occurs patients are usually given a second line medication or, if admission criteria are met, they may participate in an experimental research protocol. Palliative treatments and terminal cancer in combination can cause many symptoms that can become problematic. These symptoms commonly include pain, nausea, vomiting, difficulty in breathing, **constipation**, dehydration, agitation, and **delirium**. The palliative treatment-planning goal focuses to reduce these symptoms.

Precautions

Surgery for the purpose of **tumor removal**, biopsy, or size reduction is associated with postoperative pain and local nerve damage, which may be both severe and difficult to alleviate. Chemotherapy and radiation therapy can also produce nerve damage and severe pain. Additionally, patients with malignant cancer are susceptible to infections like herpes, **pneumonia**, urinary tract infections, and wound **abscess**, all of which can cause severe pain. Pain associated with cancer and/or treatments can significantly impair the patient's capabilities for performing daily tasks and hence impair quality of life. These complications may negatively impact the patient's psychological well being.

Cancer therapy, palliative

Definition

Palliative **cancer** therapy is treatment specifically directed to help improve the symptoms associated with terminal cancer.

Purpose

Palliative care is directed to improving symptoms associated with incurable cancer. Care can include surgery, **radiation therapy**, **chemotherapy**, symptomatic treatments resulting from cancer, and side effects of treatment. The primary objective of palliative care is to improve the quality of remaining duration of life. Treatment usually involves a combination of modalities (multimodality approach) and numerous specialists are typically involved in the treatment planning process. Therapeutic planning usually involves meticulous coordination with the treatment team.

Surgery can be utilized for palliation after careful evaluation and planning. The use of surgery in these cases may reduce the tumor bulk and help improve the quality of life by relieving **pain**, alleviating obstruction, or controlling bleeding. Radiotherapy for terminal cancer patients

Description

Pain is one of the common symptoms associated with cancer. Approximately 75% of terminal cancer patients have pain. Pain is a subjective symptom and thus it cannot be measured using technological approaches. Pain can be assessed using numeric scales (from one to 10, one is rated as no pain while 10 is severe) or rating specific facial expressions associated with various levels of pain. The majority of cancer patients experience pain as a result of tumor mass that compresses neighboring nerves, bone, or soft tissues, or from direct nerve injury (neuropathic pain). Pain can occur from affected nerves in the ribs, muscles, and internal structures such as the abdomen (cramping type pain associated with obstruction). Many patients also experience various types of pain as a direct result of follow-up tests, treatments (surgery, radiation, and chemotherapy) and diagnostic procedures (i.e., biopsy).

Preparation

Patients are typically informed that their diagnosis is terminal and treatments are directed to improve quality of life for the remaining time and to minimize emotional suffering associated with pain.

KEY TERMS

Opioids—Narcotic pain killing medication.

World Health Organization (WHO)—An international organization concerned with world health and welfare.

A careful history is necessary to assess duration, severity, and location of pain. A **physical examination** may verify the presence of pain. Imaging analysis may further confirm the presence of potential causes of pain. The World Health Organization (WHO) recommends an analgesic ladder. This treatment approach provides medication selections based on previous analgesic use and severity of pain. The ladder starts with the use of non-opioid (non-morphine) drugs such as **aspirin**, acetaminophen, or non-steroidal anti-inflammatory medications for control of mild pain. Chronic pain must be treated with constant and consistently administered medication(s). The “take as needed” approach is not advised. Supplemental doses may be recommended in addition to the standard dose for circumstances that may worsen pain. Opioids (i.e., morphine and codeine) are the medications of choice for moderate to severe pain. Doses are adjusted to produce maximum pain relief while minimizing side effects. These medications are conveniently administered orally. Administering steroids can help reduce **nausea and vomiting**. Delirium and **anxiety** may be improved by psychoactive medications.

Aftercare

Care for palliation is continuous and consistent for the remainder of life. Patients who have less than six months of life remaining may choose a hospice to stop treatment and control pain.

Risks

Patients taking opioids for pain relief can develop tolerance and dependence. Tolerance develops when a patient requires increasing amounts of medication to produce pain reduction. Dependence shows characteristic withdrawal symptoms if medications are abruptly stopped. These symptoms can be avoided by tapering down doses in the event that these medications should be stopped.

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Cancer therapy, supportive

Definition

Supportive **cancer** therapy is the use of medicines to counteract unwanted effects of cancer treatment.

Purpose

Along with their beneficial effects, many cancer treatments produce uncomfortable and sometimes harmful side effects. For example, cancer drugs may cause nausea or vomiting. They may also destroy red or white blood cells, resulting in a low **blood count**. Fortunately, many of these side effects can be relieved with other medicines.

Description

Different kinds of drugs are used for different purposes in supportive cancer therapy. To relieve **nausea and vomiting**, a physician may prescribe dolasetron (Anzemet), granisetron (Kytril) or ondansetron (Zofran). Drugs called colony stimulating factors are used to help the bone marrow make new white blood cells to replace those destroyed by cancer treatment. Examples of colony stimulating factors are filgrastim (Neupogen) and sargramostim (Leukine). Another type of drug, epoetin (Epogen, Procrit), stimulates the bone marrow to make new red blood cells. It is a synthetically made version of human erythropoietin that is made naturally in the body and has the same effect on bone marrow.

Some physicians who treat cancer recommend that their patients use **marijuana** to relieve nausea and vomiting. This practice is controversial for several reasons. Using marijuana, even for medicinal purposes, is illegal in most states. Also, most of the evidence that marijuana effectively relieves nausea and vomiting comes from reports of people who have used it, not from carefully designed scientific studies. An oral medication that contains one of the active ingredients of marijuana is available with a physician's prescription and sometimes is used to treat nausea and vomiting in patients undergoing cancer treatment. However, the drug, dronabinol (Marinol), takes longer to work than smoked marijuana and may be difficult for patients with nausea and vomiting to swallow and keep down.

In 1997, the National Institutes of Health issued a report calling for more research into medical uses of marijuana. The panel of experts who wrote the report also recommended that researchers investigate other ways of getting the active ingredients of marijuana into the body, such as nasal sprays, skin patches and inhalers.

Patients who want to use marijuana to relieve side effects of cancer treatment should talk to their physicians and should carefully consider the benefits and risks, both medical and legal.

Recommended dosage

The recommended dosage depends on the type of supportive cancer therapy. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage.

Precautions

Dolasetron, granisetron and ondansetron

If severe nausea and vomiting occur after taking this medicine, check with a physician.

The use of ondansetron after abdominal surgery may cover up symptoms of stomach problems.

People with liver disease may be more likely to have side effects from ondansetron.

Colony stimulating factors

Certain cancer drugs reduce the body's ability to fight infections. Although colony stimulating factors help restore the body's natural defenses, the process takes time. Getting prompt treatment for infections is important, even while taking this medicine. Call the physician at the first sign of illness or infection, such as a **sore throat, fever** or chills.

Seeing a physician regularly while taking this medicine is important. This will give the physician a chance to make sure the medicine is working and to check for unwanted side effects.

People with certain medical conditions may have problems if they take colony stimulating factors. In people who have kidney disease, liver disease, or conditions caused by inflammation or immune system problems, colony stimulating factors may make these problems worse. People with heart disease may be more likely to have side effects such as water retention and heart rhythm problems when they take these drugs. And people with lung disease may be more likely to have **shortness of breath**. Anyone who has any of these medical conditions should check with his or her physician before using colony stimulating factors.

Epoetin

This medicine may cause seizures (convulsions), especially in people with a history of seizures. Anyone who takes these drugs should not drive, use machines or do anything else that might be dangerous if they have had a seizure.

Epoetin helps the body make new red blood cells, but it cannot do its job unless there is plenty of iron in the body. The physician may recommend taking iron supplements or certain **vitamins** that help get iron into the body. Follow the physician's orders to make sure the body has enough iron for this medicine to work. Do not take iron supplements unless they are prescribed by a physician.

In studies of laboratory animals, epoetin taken during **pregnancy** caused **birth defects**, including damage to the bones and spine. However, the drug has not been reported to cause problems in human babies whose mothers take it. Women who are pregnant or who may become pregnant should check with their physicians for the most up-to-date information on the safety of taking this medicine during pregnancy.

People with certain medical conditions may have problems if they take this medicine. For example, the chance of side effects may be greater in people with high blood pressure, heart or blood vessel disease or a history of blood clots. Epoetin may not work properly in people who have bone problems or sickle cell anemia.

Dronabinol

This medicine contains sesame oil and one of the active ingredients of marijuana. Anyone who has had allergic or unusual reactions to sesame oil or marijuana products should let his or her physician know before taking dronabinol.

KEY TERMS

Bipolar disorder—A severe mental illness in which a person has extreme mood swings, ranging from a highly excited state—sometimes with a false sense of well-being—to depression.

Bone marrow—Soft tissue that fills the hollow centers of bones. Blood cells and platelets (disk-shaped bodies in the blood that are important in clotting) are produced in the bone marrow.

Hallucination—A false or distorted perception of objects, sounds, or events that seems real. Hallucinations usually result from drugs or mental disorders.

Immune system—The body's natural defenses against disease and infection.

Inflammation—Pain, redness, swelling, and heat that usually develop in response to injury or illness.

Schizophrenia—A severe mental disorder in which people lose touch with reality and may have illogical thoughts, delusions, hallucinations, behavioral problems and other disturbances.

Sickle cell anemia—An inherited disorder in which red blood cells contain an abnormal form of hemoglobin, a protein that carries oxygen. The abnormal form of hemoglobin causes the red cells to become sickle- or crescent-shaped. The misshapen cells may clog blood vessels, preventing oxygen from reaching tissues and leading to pain, blood clots and other problems. Sickle cell anemia is most common in people of African descent and in people from Italy, Greece, India, and the Middle East.

Because dronabinol works on the central nervous system, it may add to the effects of alcohol and other drugs that slow down the central nervous system. Examples of these drugs are **antihistamines**, cold medicine, allergy medicine, sleep aids, medicine for seizures, tranquilizers, some **pain** relievers, and **muscle relaxants**. Dronabinol may also add to the effects of anesthetics, including those used for dental procedures. Anyone taking dronabinol should not drink alcohol and should check with his or her physician before taking any of the drugs listed above.

This drug makes some people feel drowsy, dizzy, lightheaded or “high,” with a sense of well-being. Because of these possible reactions, anyone who takes dronabinol should not drive, use machines or do anything else that might be dangerous until they have found out how the drug affects them. The **dizziness** and lightheadedness are especially likely when getting up after sitting

or lying down. Getting up gradually and holding onto something for support should lessen the problem.

In laboratory studies, giving high doses of dronabinol to pregnant animals increased the risk of the unborn baby's **death**. The medicine's effects on pregnant women have not been studied. Women who are pregnant or who may become pregnant should check with their physicians before taking this medicine.

Dronabinol passes into breast milk and may affect nursing babies whose mothers take the medicine. Women who are breastfeeding their babies should check with their physicians before using dronabinol.

Because of its possible mind-altering effects, dronabinol should be used with care in children and older people. Both children and older people should be watched carefully when they are taking this medicine.

Using dronabinol may worsen some medical conditions, including high blood pressure, heart disease, **bipolar disorder** and **schizophrenia**.

General precautions for all types of supportive cancer therapy

Anyone who previously has had unusual reactions to drugs used in supportive cancer therapy should let his or her physician know before taking the drugs again. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

Side effects

Dolasetron, granisetron and ondansetron

The most common minor side effects are **headache**, dizziness or lightheadedness, drowsiness, **dry mouth**, **diarrhea**, **constipation**, abdominal pain or stomach cramps and unusual tiredness or weakness. These problems usually do not require medical treatment.

Check with a physician as soon as possible if fever occurs after taking granisetron.

If any of these symptoms occur after taking ondansetron, check with a physician immediately:

- breathing problems or **wheezing**
- chest pain or tightness in chest
- skin rash, **hives** or **itching**

Colony stimulating factors

As this medicine starts to work, it may cause mild pain in the lower back or hips. This is nothing to worry about, and it will usually go away within a few days. If the pain is too uncomfortable, the physician may pre-

scribe a painkiller. Be sure to let the physician know if the painkiller does not help.

Other possible side effects include headache, joint or muscle pain, and skin rash or itching. These side effects usually go away as the body adjusts to the medicine and do not need medical treatment. If they continue or they interfere with normal activities, check with a physician.

Epoetin

This medicine may cause flu-like symptoms, such as muscle aches, bone pain, fever, chills, shivering, and sweating, within a few hours after it is taken. These symptoms usually go away within 12 hours. If they do not, or if they are troubling, check with a physician. Other possible side effects that do not need medical attention are diarrhea, nausea or vomiting, and tiredness or weakness.

Certain side effects should be brought to a physician's attention as soon as possible. These include headache, vision problems, increased blood pressure, fast heartbeat, weight gain, and swelling of the face, fingers, lower legs, ankles or feet.

Anyone who has chest pain or seizures after taking epoetin should check with a physician immediately.

Dronabinol

Side effects such as dizziness, drowsiness, confusion and clumsiness or unsteadiness usually do not need medical attention unless they are long-lasting or they interfere with normal activities.

Other side effects or signs of overdose should have immediate medical attention. These include:

- fast or pounding heartbeat
- constipation
- trouble urinating
- red eyes
- slurred speech
- mood changes, including depression, nervousness or **anxiety**
- confusion
- forgetfulness
- changes in sight, smell, taste, touch or hearing
- a sense that time is speeding up or slowing down
- **hallucinations**

General advice on side effects for all types of supportive cancer therapy

Other side effects are possible with any type of supportive cancer therapy. Anyone who has unusual symp-

toms during or after treatment with these drugs should get in touch with his or her physician.

Interactions

Anyone who has supportive cancer therapy should let the physician know all other medicines he or she is taking. Some combinations of drugs may interact, which may increase or decrease the effects of one or both drugs or may increase the risk of side effects. Ask the physician whether the possible interactions can interfere with drug therapy or cause harmful effects.

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Nancy Ross-Flanigan

Candida albicans infection see **Candidiasis**

Candidiasis

Definition

Candidiasis is an infection caused by a species of the yeast *Candida*, usually *Candida albicans*. This is a common cause of vaginal infections in women. Also, *Candida* may cause mouth infections in people with reduced immune function, or in patients taking certain **antibiotics**. *Candida* can be found in virtually all normal people but causes problems in only a fraction. In recent years, however, several serious categories of candidiasis have become more common, due to overuse of antibiotics, the rise of **AIDS**, the increase in organ transplantations, and the use of invasive devices (catheters, artificial joints and valves)—all of which increase a patient's susceptibility to infection.

Description

Vaginal candidiasis

Over one million women in the United States develop vaginal yeast infections each year. It is not life-threatening, but it can be uncomfortable and frustrating.

Oral candidiasis

This disorder, also known as thrush, causes white, curd-like patches in the mouth or throat.



This patient's tongue is infected with candidiasis. (Photograph by Edward H. Gill, Custom Medical Stock Photo. Reproduced by permission.)

Deep organ candidiasis

Also known as invasive candidiasis, deep organ candidiasis is a serious systemic infection that can affect the esophagus, heart, blood, liver, spleen, kidneys, eyes, and skin. Like vaginal and oral candidiasis, it is an opportunistic disease that strikes when a person's resistance is lowered, often due to another illness. There are many diagnostic categories of deep organ candidiasis, depending on the tissues involved.

Causes and symptoms

Vaginal candidiasis

Most women with vaginal candidiasis experience severe vaginal **itching**. They also have a discharge that often looks like cottage cheese and has a sweet or bread-like odor. The vulva and vagina can be red, swollen, and painful. Sexual intercourse can also be painful.

Oral candidiasis

Whitish patches can appear on the tongue, inside of the cheeks, or the palate. Oral candidiasis typically occurs in people with abnormal immune systems. These can include people undergoing **chemotherapy** for **cancer**, people taking immunosuppressive drugs to protect transplanted organs, or people with HIV infection.

Deep organ candidiasis

Anything that weakens the body's natural barrier against colonizing organisms—including stomach

surgery, **burns**, nasogastric tubes, and catheters—can predispose a person for deep organ candidiasis. Rising numbers of AIDS patients, organ transplant recipients, and other individuals whose immune systems are compromised help account for the dramatic increase in deep organ candidiasis in recent years. Patients with granulocytopenia (deficiency of white blood cells) are particularly at risk for deep organ candidiasis.

Diagnosis

Often clinical appearance gives a strong suggestion about the diagnosis. Generally, a clinician will take a sample of the vaginal discharge or swab an area of oral plaque, and then inspect this material under a microscope. Under the microscope, it is possible to see characteristic forms of yeasts at various stages in the lifecycle.

Fungal blood cultures should be taken for patients suspected of having deep organ candidiasis. Tissue biopsy may be needed for a definitive diagnosis.

Treatment

Vaginal candidiasis

In most cases, vaginal candidiasis can be treated successfully with a variety of over-the-counter antifungal creams or suppositories. These include Monistat, Gyne-Lotrimin, and Mycelex. However, infections often recur. If a woman has frequent recurrences, she should consult her doctor about prescription drugs such as Vagistat-1, Diflucan, and others.

Oral candidiasis

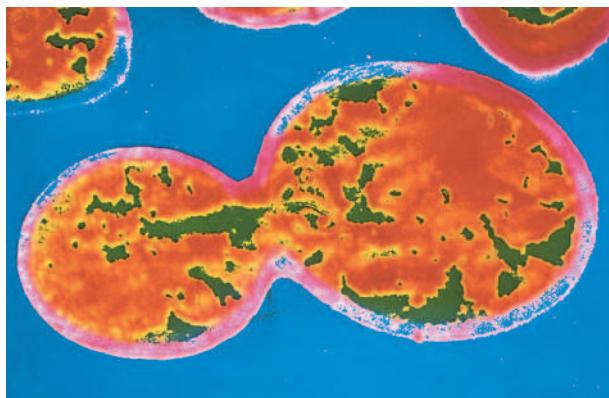
This is usually treated with prescription lozenges or mouthwashes. Some of the most-used prescriptions are nystatin mouthwashes (Nilstat or Nitrostat) and clotrimazole lozenges.

Deep organ candidiasis

The recent increase in deep organ candidiasis has led to the creation of treatment guidelines, including, but not limited to, the following: Catheters should be removed from patients in whom these devices are still present. Antifungal chemotherapy should be started to prevent the spread of the disease. Drugs should be prescribed based on a patient's specific history and defense status.

Alternative treatment

Home remedies for vaginal candidiasis include vinegar douches or insertion of a paste made from *Lactobacillus acidophilus* powder into the vagina. In theory,



A transmission electron microscopy (TEM) of *Candida albicans*. (Custom Medical Stock Photo. Reproduced by permission.)

these remedies will make the vagina more acidic and therefore less hospitable to the growth of *Candida*. Fresh garlic (*Allium sativum*) is believed to have antifungal action, so incorporating it into the diet or inserting a gauze-wrapped, peeled garlic clove into the vagina may be helpful. The insert should be changed twice daily. Some women report success with these remedies; they should try a conventional treatment if an alternative remedy isn't effective.

Prognosis

Vaginal candidiasis

Although most cases of vaginal candidiasis are cured reliably, these infections can recur. To limit recurrences, women may need to take a prescription anti-fungal drug such as terconazole (sold as Terazol) or take other anti-fungal drugs on a preventive basis.

Oral candidiasis

These infections can also recur, sometimes because the infecting *Candida* develops resistance to one drug. Therefore, a physician may need to prescribe a different drug.

Deep organ candidiasis

The prognosis depends on the category of disease as well as on the condition of the patient when the infection strikes. Patients who are already suffering from a serious underlying disease are more susceptible to deep organ candidiasis that spreads throughout the body.

Prevention

Because *Candida* is part of the normal group of microorganisms that co-exist with all people, it is impos-

KEY TERMS

Biopsy—The removal and examination of tissue from a live body.

Colonize—To become established in a host.

Granulocytopenia—A condition characterized by a deficiency of white blood cells.

Nasogastric—Tube inserted through the nasal passages into the stomach.

Opportunistic—Infection caused by microorganisms that are usually harmless, but which can cause disease when a host's resistance is lowered.

Systemic—Afflicting an entire body system or the body in general.

sible to avoid contact with it. Good vaginal hygiene and good **oral hygiene** might reduce problems, but they are not guarantees against candidiasis.

Because hospital-acquired (nosocomial) deep organ candidiasis is on the rise, people need to be made aware of it. Patients should be sure that catheters are properly maintained and used for the shortest possible time length. The frequency, length, and scope of courses of antibiotic treatment should also be cut back.

Resources

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Richard H. Lampert

Candidosis see **Candidiasis**

Canker sores

Definition

Canker sores are small sores or ulcers that appear inside the mouth. They are painful, self-healing, and can recur.

KEY TERMS

Inflammation—A local reaction to tissue injury or damage, usually characterized by pain, swelling, and redness.

Sore—A wound, lesion, or ulcer on the skin.

Ulcer—A site of damage to the skin or mucous membrane that is characterized by the formation of pus, death of tissue, and is frequently accompanied by an inflammatory reaction.

Description

Canker sores occur on the inside of the mouth, usually on the inside of the lips, cheeks, and/or soft palate. They can also occur on the tongue and in the throat. Often, several canker sores will appear at the same time and may be grouped in clusters. Canker sores appear as a whitish, round area with a red border. The sores are painful and sensitive to touch. The average canker sore is about one-quarter inch in size, although they can occasionally be larger. Canker sores are not infectious.

Approximately 20% of the U.S. population is affected with recurring canker sores, and more women than men get them. Women are more likely to have canker sores during their premenstrual period.

Canker sores are sometimes confused with cold sores. Cold sores are caused by herpes simplex virus. This disease, also known as oral herpes or **fever blisters**, can occur anywhere on the body. Most commonly, herpes infection occurs on the outside of the lips and the gums, and much less frequently on the inside the mouth. Cold sores are infectious.

Causes and symptoms

The exact cause of canker sores is uncertain, however, they seem to be related to a localized immune reaction. Other proposed causes for this disease are trauma to the affected areas from toothbrush scrapes, **stress**, hormones, and food **allergies**. Canker sores tend to appear in response to stress. The initial symptom is a tingling or mildly painful **itching** sensation in the area where the sore will appear. After one to several days, a small red swelling appears. The sore is round, and is a whitish color with a grayish colored center. Usually, there is a red ring of inflammation surrounding the sore. The main symptom is **pain**. Canker sores can be very painful, especially if they are touched repeatedly, e.g., by the tongue. They last for one to two weeks.

Diagnosis

Canker sores are diagnosed by observation of the blister. A distinction between canker sores and cold sores must be made because cold sores are infectious and the herpes infection can be transmitted to other people. The two sores can usually be distinguished visually and there are specific diagnostic tests for herpes infection.

Treatment

Since canker sores heal by themselves, treatment is not usually necessary. Pain relief remedies, such as topical anesthetics, may be used to reduce the pain of the sores. The use of corticosteroid ointments sometimes speeds healing. Avoidance of spicy or acidic foods can help reduce the pain associated with canker sores.

Alternative treatment

Alternative therapies for canker sores are aimed at healing existing sores and preventing their recurrence. Several herbal remedies, including calendula (*Calendula officinalis*), myrrh (*Commiphora molmol*), and goldenseal (*Hydrastis canadensis*), may be helpful in the treatment of existing sores. Compresses soaked in teas made from these herbs are applied directly to the sores. The tannic acid in a tea bag can also help dry up the sores when the wet tea bag is used as a compress. Taking dandelion (*Taraxacum officinale*) tea or capsules may help heal sores and also prevent future outbreaks. Since canker sores are often brought on by stress, such stress-relieving techniques as **meditation**, **guided imagery**, and certain **acupressure** exercises may help prevent canker sores or lessen their severity.

Prognosis

There is no cure for canker sores. They do not get larger or occur more frequently with age.

Resources

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John T. Lohr, PhD

Captopril see **Angiotensin-converting enzyme inhibitors**

Carbamazepine see **Anticonvulsant drugs**

Carbidopa see **Antiparkinson drugs**

Carbohydrate intolerance

Definition

Carbohydrate intolerance is the inability of the body to completely process the nutrient carbohydrate (a classification that includes sugars and starches) into a source of energy for the body, usually because of the deficiency of an enzyme needed for digestion. **Lactose intolerance**, the inability to digest the sugar found in milk, is widespread and affects up to 70% of the world's adult population.

Description

Carbohydrates are the primary source of energy and, along with fats and proteins, one of the three major nutrients in the human diet. Carbohydrates are classified according to their structure based on the number of basic sugar, or *saccharide* units they contain.

A monosaccharide is the simplest carbohydrate and called a simple sugar. Simple sugars include glucose (the form in which sugar circulates in the blood), fructose (found in fruit and honey), and galactose (produced by the digestion of milk). These simple sugars are important because they can be absorbed by the small intestine. Two simple sugars linked together make a disaccharide. The disaccharide sugars present in the diet are maltose (a product of the digestion of starch), sucrose (table sugar), and lactose (the sugar in milk). These disaccharides must be broken down by enzymes into two simple sugars so that they can be absorbed by the intestine. Polysaccharides are much more complex carbohydrates made up of many simple sugars, the most important of which are glycogen, which is stored in the liver, and starch.

Digestion of sugars

Digestion of food begins in the mouth, moves on to the stomach, and then into the small intestine. Along the way, specific enzymes are needed to process different types of sugars. An enzyme is a substance that acts as a catalyst to produce chemical changes without being changed itself. The enzymes lactase, maltase, and isomaltase (or sucrase) are needed to break down the disaccharides; when one or more is inadequate, the result is carbohydrate intolerance.

Types of intolerance

Carbohydrate intolerance can be primary or secondary. Primary deficiency is caused by an enzyme defect present at birth or developed over time. The most common is lactose intolerance. Secondary deficiencies are caused by a disease or disorder of the intestinal tract, and dis-

pear when the disease is treated. These include protein deficiency, **celiac disease**, and some intestinal infections.

Adult lactose intolerance is the most common of all enzyme deficiencies, and it is estimated that 30–50 million Americans have this condition. Some racial and ethnic populations are affected more than others. Lactose intolerance is found in as many as 75% of African Americans, Jewish Americans, Mexican Americans, and Native Americans, and in 90% of Asian Americans. Descendants of Northern Europeans and some Mediterranean peoples usually do not develop the condition. Deficiencies in enzymes other than lactase are extremely rare.

Causes and symptoms

Enzymes play an important role in breaking down carbohydrates into forms that can pass through the intestine and be used by the body. Usually they are named by adding *ase* to the name of the substance they act on, so lactase is the enzyme needed to process lactose. Cooked starch is broken down in the mouth to a disaccharide by amylase, an enzyme in the saliva. The disaccharides maltose, sucrose, and lactose cannot be absorbed until they have been separated into simple sugar molecules by their corresponding enzymes present in the cells lining the intestinal tract. If this process is not completed, digestion is interrupted.

Although not common, a deficiency in the enzymes needed to digest lactose, maltose, and sucrose is sometimes present at birth. Intestinal lactase enzymes usually decrease naturally with age, but this happens to varying degrees. Because of the uneven distribution of enzyme deficiency based on race and ethnic heritage, especially in lactose intolerance, genetics are believed to play a role in the cause of primary carbohydrate intolerance.

Digestive diseases such as celiac disease and tropical sprue (which affect absorption in the intestine), as well as intestinal infections and injuries, can reduce the amount of enzymes produced. In **cancer** patients, treatment with **radiation therapy** or **chemotherapy** may affect the cells in the intestine that normally secrete lactase, leading to intolerance.

The severity of the symptoms depends on the extent of the enzyme deficiency, and range from a feeling of mild bloating to severe **diarrhea**. In the case of a lactase deficiency, undigested milk sugar remains in the intestine, which is then fermented by the bacteria normally present in the intestine. These bacteria produce gas, cramping, bloating, a "gurgly" feeling in the abdomen, and flatulence. In a growing child, the main symptoms are diarrhea and a failure to gain weight. In an individual with lactase deficiency, gastrointestinal distress begins

KEY TERMS

Celiac disease—A disease, occurring in both children and adults, which is caused by a sensitivity to gluten, a protein found in grains. It results in chronic inflammation and shrinkage of the lining of the small intestine.

Digestion—The mechanical, chemical, and enzymatic process in which food is converted into the materials suitable for use by the body.

Enzyme—A substance produced by the body to assist in a chemical reaction. In carbohydrate intolerance, lack of an enzyme makes it impossible for one type of sugar to be broken down into a simpler form so that it can be absorbed by the intestines and used by the body.

Metabolism—All the physical and chemical changes that take place within an organism.

Nutrient—Food or another substance that supplies the body with the elements needed for metabolism.

Sugars—Those carbohydrates having the general composition of one part carbon, two parts hydrogen, and one part oxygen.

about 30 minutes to two hours after eating or drinking foods containing lactose. Food intolerances can be confused with food **allergies**, since the symptoms of nausea, cramps, bloating, and diarrhea are similar.

Sugars that aren't broken down into one of the simplest forms cause the body to push fluid into the intestines, which results in watery diarrhea (osmotic diarrhea). Diarrhea may sweep other nutrients out of the intestine before they can be absorbed, causing **malnutrition**.

Diagnosis

Carbohydrate intolerance can be diagnosed using oral tolerance tests. The carbohydrate being investigated is given by mouth in liquid form and several blood levels are measured and compared to normal values. This helps evaluate the individual's ability to digest the sugar.

To identify lactose intolerance in children and adults, the hydrogen breath test is used to measure the amount of hydrogen in the breath. The patient drinks a beverage containing lactose and the breath is analyzed at regular intervals. If undigested lactose in the large intestine (colon) is fermented by bacteria, various gases are produced. Hydrogen is absorbed from the intestines

and carried by the bloodstream into the lungs where it is exhaled. Normally there is very little hydrogen detectable in the breath, so its presence indicates faulty digestion of lactose.

When lactose intolerance is suspected in infants and young children, many pediatricians recommend simply changing from cow's milk to soy formula and watching for improvement. If needed, a stool sample can be tested for acidity. The inadequate digestion of lactose will result in an increase of acid in the waste matter excreted by the bowels and the presence of glucose.

Treatment

Carbohydrate intolerance caused by temporary intestinal diseases disappears when the condition is successfully treated. In primary conditions, no treatment exists to improve the body's ability to produce the enzymes, but symptoms can be controlled by diet.

Because the degree of lactose intolerance varies so much, treatment should be tailored for the individual. Young children showing signs of intolerance should avoid milk products; infants should switch to soy-based formula. Older children and adults can adjust their intake of lactose depending on how much and what they can tolerate. For some, a small glass of milk will not cause problems, while others may be able to handle ice cream or aged cheeses such as cheddar or Swiss, but not other dairy products. Generally, small amounts of lactose-containing foods taken throughout the day are better tolerated than a large amount consumed all at once.

For those individuals who are sensitive to even very small amounts of lactose, the lactase enzyme is available without a prescription. It comes in liquid form for use with milk. The addition of a few drops to a quart of milk will reduce the lactose content by 70% after 24 hours in the refrigerator. Heating the milk speeds up the process, and doubling the amount of lactase liquid will result in milk that is 90% lactose free. Chewable lactase enzyme tablets are also available. Three to six tablets taken before a meal or snack will aid in the digestion of solid foods. Lactose-reduced milk and other products are also available in stores. The milk contains the same nutrients as regular milk.

Because dairy products are an important source of calcium, people who reduce or severely limit their intake of dairy products may need to consider other ways to consume an adequate amount of calcium in their **diets**.

Prognosis

With good dietary management, individuals with carbohydrate intolerance can lead normal lives.

Prevention

Since the cause of the enzyme deficiency leading to carbohydrate intolerance is unknown, there is no way to prevent this condition.

Resources

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Karen Ericson, RN

Carbon monoxide poisoning

Definition

Carbon monoxide (CO) **poisoning** occurs when carbon monoxide gas is inhaled. CO is a colorless, odorless, highly poisonous gas that is produced by incomplete combustion. It is found in automobile exhaust fumes, faulty stoves and heating systems, fires, and cigarette smoke. Other sources include woodburning stoves, kerosene heaters, improperly ventilated water heaters and gas stoves, and blocked or poorly maintained chimney flues. CO interferes with the ability of the blood to carry oxygen. The result is **headache**, nausea, convulsions, and finally **death** by asphyxiation.

Description

Carbon monoxide, sometimes called coal gas, has been known as a toxic substance since the third century B.C. It was used for executions and suicides in early Rome. Today it is the leading cause of accidental poisoning in the United States. According to the *Journal of the American Medical Association*, 1,500 Americans die each year from accidental exposure to CO, and another 2,300 from intentional exposure (suicide). An additional 10,000 people seek medical attention after exposure to CO and recover.

Anyone who is exposed to CO will become sick, and the entire body is involved in CO poisoning. A developing fetus can also be poisoned if a pregnant woman breathes CO gas. Infants, people with heart or lung disease, or those with anemia may be more seriously affected. People such as underground parking garage attendants who are exposed to car exhausts in a confined area are more likely to be poisoned by CO. Firemen also run a higher risk of inhaling CO.

Causes and symptoms

Normally when a person breathes fresh air into the lungs, the oxygen in the air binds with a molecule called hemoglobin (Hb) that is found in red blood cells. This allows oxygen to be moved from the lungs to every part of the body. When the oxygen/hemoglobin complex reaches a muscle where it is needed, the oxygen is released. Because the oxygen binding process is reversible, hemoglobin can be used over and over again to pick up oxygen and move it throughout the body.

Inhaling carbon monoxide gas interferes with this oxygen transport system. In the lungs, CO competes with oxygen to bind with the hemoglobin molecule. Hemoglobin prefers CO to oxygen and accepts it more than 200 times more readily than it accepts oxygen. Not only does the hemoglobin prefer CO, it holds on to the CO much more tightly, forming a complex called carboxyhemoglobin (COHb). As a person breathes CO contaminated air, more and more oxygen transportation sites on the hemoglobin molecules become blocked by CO. Gradually, there are fewer and fewer sites available for oxygen. All cells need oxygen to live. When they don't get enough oxygen, cellular metabolism is disrupted and eventually cells begin to die.

The symptoms of CO poisoning and the speed with which they appear depend on the concentration of CO in the air and the rate and efficiency with which a person breathes. Heavy smokers can start off with up to 9% of their hemoglobin already bound to CO, which they regularly inhale in cigarette smoke. This makes them much more susceptible to environmental CO. The Occupational Safety and Health Administration (OSHA) has established a maximum permissible exposure level of 50 parts per million (ppm) over eight hours.

With exposure to 200 ppm for two to three hours, a person begins to experience headache, **fatigue**, nausea, and **dizziness**. These symptoms correspond to 15–25% COHb in the blood. When the concentration of COHb reaches 50% or more, death results in a very short time. Emergency room physicians have the most experience diagnosing and treating CO poisoning.

KEY TERMS

Carboxyhemoglobin (COHb)—Hemoglobin that is bound to carbon monoxide instead of oxygen.

Hemoglobin (Hb)—A molecule that normally binds to oxygen in order to carry it to our cells, where it is required for life.

Hypothermia—Development of a subnormal body temperature.

pH—A measurement of the acidity or alkalinity of a fluid. A neutral fluid, neither acid nor alkali, has a pH of 7.

The symptoms of CO poisoning in order of increasing severity include:

- headache
- shortness of breath
- dizziness
- fatigue
- mental confusion and difficulty thinking
- loss of fine hand-eye coordination
- nausea and vomiting
- rapid heart rate
- hallucinations
- inability to execute voluntary movements accurately
- collapse
- lowered body temperature (**hypothermia**)
- coma
- convulsions
- seriously low blood pressure
- cardiac and **respiratory failure**
- death

In some cases, the skin, mucous membranes, and nails of a person with CO poisoning are cherry red or bright pink. Because the color change doesn't always occur, it is an unreliable symptom to rely on for diagnosis.

Although most CO poisoning is acute, or sudden, it is possible to suffer from chronic CO poisoning. This condition exists when a person is exposed to low levels of the gas over a period of days to months. Symptoms are often vague and include (in order of frequency) fatigue, headache, dizziness, sleep disturbances, cardiac symptoms, apathy, nausea, and memory disturbances. Little is known about chronic CO poisoning, and it is often misdiagnosed.

Diagnosis

The main reason to suspect CO poisoning is evidence that fuel is being burned in a confined area, for example a car running inside a closed garage, a charcoal grill burning indoors, or an unvented kerosene heater in a workshop. Under these circumstances, one or more persons suffering from the symptoms listed above strongly suggests CO poisoning. In the absence of some concrete reason to suspect CO poisoning, the disorder is often misdiagnosed as **migraine headache**, **stroke**, psychiatric illness, **food poisoning**, alcohol poisoning, or heart disease.

Concrete confirmation of CO poisoning comes from a carboxyhemoglobin test. This blood test measures the amount of CO that is bound to hemoglobin in the body. Blood is drawn as soon after suspected exposure to CO as possible.

Other tests that are useful in determining the extent of CO poisoning include measurement of other arterial blood gases and pH; a complete **blood count**; measurement of other blood components such as sodium, potassium, bicarbonate, urea nitrogen, and lactic acid; an electrocardiogram (ECG); and a **chest x ray**.

Treatment

Immediate treatment for CO poisoning is to remove the victim from the source of carbon monoxide gas and get him or her into fresh air. If the victim is not breathing and has no pulse, **cardiopulmonary resuscitation (CPR)** should be started. Depending on the severity of the poisoning, 100% oxygen may be given with a tight fitting mask as soon as it is available.

Taken with other symptoms of CO poisoning, COHb levels of over 25% in healthy individuals, over 15% in patients with a history of heart or lung disease, and over 10% in pregnant women usually indicate the need for hospitalization. In the hospital, fluids and electrolytes are given to correct any imbalances that have arisen from the breakdown of cellular metabolism.

In severe cases of CO poisoning, patients are given hyperbaric oxygen therapy. This treatment involves placing the patient in a chamber breathing 100% oxygen at a pressure of more than one atmosphere (the normal pressure the atmosphere exerts at sea level). The increased pressure forces more oxygen into the blood. Hyperbaric facilities are specialized, and are usually available only at larger hospitals.

Prognosis

The speed and degree of recovery from CO poisoning depends on the length and duration of exposure to

the gas. The half-life of CO in normal room air is four to five hours. This means that, in four to five hours, half of the CO bound to hemoglobin will be replaced with oxygen. At normal atmospheric pressures, but breathing 100% oxygen, the half-life for the elimination of CO from the body is 50-70 minutes. In hyperbaric therapy at three atmospheres of pressure, the half-life is reduced to 20-25 minutes.

Although the symptoms of CO poisoning may subside in a few hours, some patients show memory problems, fatigue, confusion, and mood changes for two to four weeks after their exposure to the gas.

Prevention

Carbon monoxide poisoning is preventable. Particular care should be paid to situations where fuel is burned in a confined area. Portable and permanently installed carbon monoxide detectors that sound a warning similar to smoke detectors are available for under \$50. Specific actions that will prevent CO poisoning include:

- stop **smoking**. Smokers have less tolerance to environmental CO
- have heating systems and appliances installed by a qualified contractor to assure that they are properly vented and meet local building codes
- inspect and properly maintain heating systems, chimneys, and appliances
- do not use a gas oven or stove to heat the home
- do not burn charcoal indoors
- make sure there is good ventilation if using a kerosene heater indoors
- do not leave cars or trucks running inside the garage
- keep car windows rolled up when stuck in heavy traffic, especially if inside a tunnel

Resources

ORGANIZATIONS

American Lung Association. 1740 Broadway, New York, NY 10019. (800) 586-4872. <<http://www.lungusa.org>>.

OTHER

“Carbon Monoxide Headquarters.” *Wayne State University School of Medicine*. <<http://www.phypc.med.wayne.edu/>>.

Tish Davidson

Carbunculosis see **Boils**

Carcinoembryonic antigen test

Definition

The carcinoembryonic antigen (CEA) test is a laboratory blood study. CEA is a substance which is normally found only during fetal development, but may reappear in adults who develop certain types of **cancer**.

Purpose

The CEA test is ordered for patients with known cancers. The CEA test is most commonly ordered when a patient has a cancer of the gastrointestinal system. These include cancer of the colon, rectum, stomach (gastric cancer), esophagus, liver, or pancreas. It is also used with cancers of the breast, lung, or prostate.

The CEA level in the blood is one of the factors that doctors consider when determining the prognosis, or most likely outcome of a cancer. In general, a higher CEA level predicts a more severe disease, one that is less likely to be curable. But it does not give clear-cut information. The results of a CEA test are usually considered along with other laboratory and/or imaging studies to follow the course of the disease.

Once treatment for the cancer has begun, CEA tests have a valuable role in monitoring the patient’s progress. A decreasing CEA level means therapy is effective in fighting the cancer. A stable or increasing CEA level may mean the treatment is not working, and/or that the tumor is growing. It is important to understand that serial CEA measurements, which means several done over a period of time, are the most useful. A single test result is difficult to evaluate, but a number of tests, done weeks apart, shows trends in disease progression or regression.

Certain types of cancer treatments, such as hormone therapy for **breast cancer**, may actually cause the CEA level to go up. This elevation does not accurately reflect the state of the disease. It is sometimes referred to as a “flare response.” Recognition that a rise in CEA may be temporary and due to therapy is significant. If this possibility is not taken into account, the patient may be unnecessarily discouraged. Further, treatment that is actually effective may be stopped or changed prematurely.

CEA tests are also used to help detect recurrence of a cancer after surgery and/or other treatment has been completed. A rising CEA level may be the first sign of cancer return, and may show up months before other studies or patient symptoms would raise concern. Unfortunately, this does not always mean the recurrent cancer can be cured. For example, only a small percentage of patients with colorectal cancers and rising CEA levels will benefit from

another surgical exploration. Those with recurrence in the same area as the original cancer, or with a single metastatic tumor in the liver or lung, have a chance that surgery will eliminate the disease. Patients with more widespread return of the cancer are generally not treatable with surgery. The CEA test will not separate the two groups.

Patients who are most likely to benefit from non-standard treatments, such as bone marrow transplants, may be determined on the basis of CEA values, combined with other test results. CEA levels may be one of the criteria for determining whether the patient will benefit from more expensive studies, such as CT scan or MRI.

Precautions

The CEA test is not a screening test for cancer. It is not useful for detecting the presence of cancer. Many cancers do not produce an increased CEA level. Some noncancerous diseases, such as hepatitis, inflammatory bowel disease, **pancreatitis**, and obstructive pulmonary disease, may cause an elevated CEA level.

Description

Determination of the CEA level is a laboratory blood test. Obtaining a specimen of blood for the study takes only a few minutes. CEA testing should be covered by most insurance plans.

Preparation

No preparation is required.

Aftercare

None.

Risks

There are no complications or side effects of this test. However, the results of a CEA study should be interpreted with caution. A single test result may not yield clinically useful information. Several studies over a period of months may be needed.

Another concern is the potential for false positive as well as false negative results. A false positive result means the test shows an abnormal value when cancer is not present. A false negative means the test reveals a normal value when cancer actually is present.

Normal results

The absolute numbers which are considered normal vary from one laboratory to another. Any results reported

should come with information regarding the testing facility's normal range.

Abnormal results

A single abnormal CEA value may be significant, but must be regarded cautiously. In general, very high CEA levels indicate more serious cancer, with a poorer chance for cure. But some benign diseases and certain cancer treatments may produce an elevated CEA test. Cigarette **smoking** will also cause the CEA level to be abnormally high.

Resources

BOOKS

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Ellen S. Weber, MSN

Carcinoid tumors see **Neuroendocrine tumors**

Cardiac arrest see **Sudden cardiac death**

Cardiac arrhythmias see **Arrhythmias**

Cardiac blood pool scan

Definition

A cardiac blood pool scan is a non-invasive test that uses a mildly radioactive marker to observe the functioning of the left ventricle of the heart.

Purpose

The left ventricle is the main pump for distributing blood through the body. A cardiac blood pool scan is used to determine how efficiently the left ventricle is working. The scan can detect aneurysms of the left ventricle, motion abnormalities caused by damage to the heart wall, cardiac shunts between the left and right ventricle, and coronary occlusive artery disease.

Precautions

Pregnant women are the only patients who should not participate in a cardiac blood pool scan. However, the accuracy of the results may be affected if the patient moves during imaging, has had other recent nuclear scans, or has an irregular heartbeat.

Description

A cardiac blood pool scan is sometimes called equilibrium radionuclide angiography or gated (synchronized) cardiac blood pool imaging. A **multiple-gated acquisition (MUGA) scan** is a variation of this test.

To perform a cardiac blood pool scan, the patient lies under a special gamma scintillation camera that detects radiation. A protein tagged with a radioactive marker (usually technetium-99m) is injected into the patient's forearm.

The camera is synchronized with an electrocardiogram (ECG) to take a picture at specific times in the cycle of heart contraction and relaxation. When data from many sequential pictures is processed by a computer, a doctor can analyze whether the left ventricle is functioning normally.

The patient needs to remain silent and motionless during the test. Sometimes the patient is asked to **exercise**, then another set of pictures is taken for comparison. This test normally takes about 30 minutes.

Preparation

No changes in diet or medication are necessary. An ECG will probably be done before the test.

Aftercare

The patient may resume normal activities immediately.

Risks

Cardiac blood pool scans are a safe and effective way of measuring left ventricle function. The only risk is to the fetus of a pregnant woman.

Normal results

A computer is used to process the information from the test, then the results are analyzed by a doctor. A normally functioning left ventricle will contract symmetrically, show even distribution of the radioactively tagged protein, and eject about 55–65% of volume of blood it holds on each contraction.

Abnormal results

Patients with damage to the ventricle or heart wall will show an uneven distribution of the radiopharmaceutical. The volume of blood ejected in each contraction will be less than 55%.

KEY TERMS

Aneurysm—A sac or bulge that forms because of a weak spot in the wall of an artery or heart chamber.

Cardiac shunt—A defect in the wall of the heart that allows blood from different chambers to mix.

Coronary occlusive artery disease—Blockage of the arteries that supply blood to the heart; frequently a precursor to a heart attack.

Electrocardiogram (ECG)—A graph that shows the electrical charges that trigger the heart to contract. Heart abnormalities alter the graph, giving clues to the source of the abnormality.

Ventricle—One of the two bottom chambers of the heart (the heart has four chambers). The left ventricle acts as the body's main pump for blood.

Resources

BOOKS

"Cardiac Blood Pool Imaging." In *Illustrated Guide to Diagnostic Tests*, ed. J. A. Lewis. Springhouse, PA: Springhouse Corp., 1994.

Pagana, Kathleen Deska. *Mosby's Manual of Diagnostic and Laboratory Tests*. St. Louis: Mosby, Inc., 1998.

Tish Davidson

Cardiac catheterization

Definition

Cardiac catheterization (also called heart catheterization) is a diagnostic procedure which does a comprehensive examination of how the heart and its blood vessels function. One or more catheters are inserted through a peripheral blood vessel in the arm (antecubital artery or vein) or leg (femoral artery or vein) with x-ray guidance. This procedure gathers information such as adequacy of blood supply through the coronary arteries, blood pressures, blood flow throughout chambers of the heart, collection of blood samples, and x rays of the heart's ventricles or arteries.

A test that can be performed on either side of the heart, cardiac catheterization checks for different functions in both the left and right sides. When testing the heart's right side, tricuspid and pulmonary valve function

are evaluated, in addition to measuring pressures of and collecting blood samples from the right atrium, ventricle, and pulmonary artery. Left-sided heart catheterization is performed by way of a catheter through an artery which tests the blood flow of the coronary arteries, function of the mitral and aortic valves, and left ventricle.

Purpose

The primary reason for conducting a cardiac catheterization is to diagnose and manage persons known or suspected to have heart disease, a frequently fatal condition that leads to 1.5 million heart attacks annually in the United States.

Symptoms and diagnoses that may lead to performing this procedure include:

- chest **pain**, characterized by prolonged heavy pressure or a squeezing pain
- abnormal treadmill **stress test**
- myocardial infarction, also known as a **heart attack**
- congenital heart defects, or heart problems that originated from birth
- a diagnosis of valvular-heart disease
- a need to measure the heart muscle's ability to pump blood

Typically performed along with **angiography**, a technique of injecting a dye into the vascular system to outline the heart and blood vessels, a catheterization can aid in the visualization of any blockages, narrowing, or abnormalities in the coronary arteries. If these signs are visible, the cardiologist may assess the patient's need and readiness for coronary bypass surgery, or perhaps a less invasive approach, such as dilation of a narrowed blood vessel either surgically or with the use of a balloon (**angioplasty**).

When looking at the left side of the heart, fluoroscopic guidance also allows the following diagnoses to be assessed:

- enlargement of the left ventricle
- ventricular aneurysms (abnormal dilation of a blood vessel)
- narrowing of the aortic valve
- insufficiency of the aortic or mitral valve
- the detour of blood from one side of the heart to the other due to septal defects (also known as shunting)

Precautions

Cardiac catheterization is categorized as an "invasive" procedure which involves the heart, its valves, and

coronary arteries, in addition to a large artery in the arm or leg. Due to the nature of the test, it is important to evaluate for the following conditions before considering this procedure:

- A diagnosis of a bleeding disorder, poor kidney function, or debilitation. Any of these pre-existing conditions typically raises the risk of the catheterization procedure and may be reason to cancel the procedure.
- A diagnosis of heart valve disease. If this is detected, **antibiotics** may be given before the test to prevent inflammation of the membrane which lines the heart (endocarditis).

Description

To understand how a cardiac catheterization is able to diagnose and manage heart disease, the basic workings of the heart muscle must also be understood. Just as the body relies on a constant supply of blood to aid in its everyday functions, so does the heart. The heart is made up of an intricate web of blood vessels (coronary arteries) that ensure an adequate supply of blood rich in oxygen and nutrients. It is easy to see how an abnormality in any of these arteries can be detrimental to the heart's function. These abnormalities cause the heart's blood flow to decrease and result in the condition known as **coronary artery disease** or coronary insufficiency.

Catheterization is a valuable tool in detecting and treating abnormalities of the heart. Through the use of fluoroscopic (x ray) guidance, a catheter, which may resemble a balloon-tipped tube, is strung through the veins or arteries into the heart, so the cardiologist can monitor a body's various functions at each moment.

Generally a test that lasts two to three hours, a patient should expect the following prior to and during the catheterization procedure:

- A mild sedative may be given that will allow the patient to relax but remain conscious during the test.
- An intravenous needle will be inserted in the arm to administer medication. Electrodes will be attached to the chest to enable the painless procedure known as an electrocardiograph.
- Prior to inserting a catheter into an artery or vein in the arm or leg, the incision site will be made numb by injecting a local anesthetic. When the anesthetic is injected it may feel like a pin-prick followed by a quick stinging sensation. Pressure may also be experienced as the catheter travels through the blood vessel.
- After the catheter is guided into the coronary-artery system, a dye (also called a radiocontrast material) is injected to aid in the identification of any abnormalities

of the heart. During this time, the patient may experience a hot, flushed feeling or a quickly passing nausea. Coughing or breathing deeply aids in any discomfort.

- Medication may be given during the procedure if chest pain is experienced, and nitroglycerin may also be administered to allow expansion of the heart's blood vessels.
- When the test is complete, the physician will remove the catheter and close the skin with several sutures or tape.

Preparation

Prior to the cardiac catheterization procedure, it is important to relay information to the physician or nurse regarding **allergies** to shellfish (such as shrimp or scallops) which contain iodine, iodine itself, or the dyes that are commonly used in other diagnostic tests.

Because this procedure is categorized as a surgery, the patient will be instructed not to eat or drink anything for at least six hours prior to the test. Just before the test begins, the patient will urinate and change into a hospital gown, then lie flat on a padded table that may also be tilted in order for the heart to be examined from a variety of angles.

Aftercare

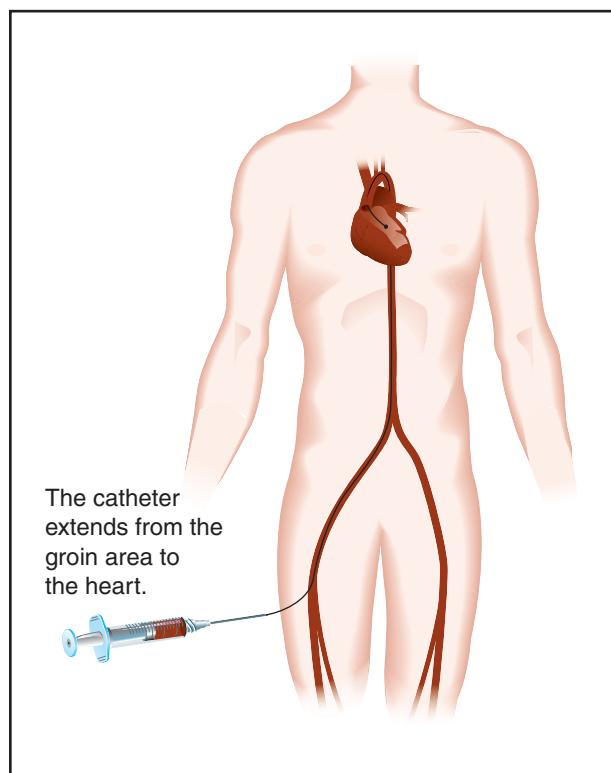
While cardiac catheterization may be performed on an outpatient basis, a patient may require close monitoring following the procedure while remaining in the hospital for at least 24 hours. The patient will be instructed to rest in bed for at least eight hours immediately after the test. If the catheter was inserted into a vein or artery in the leg or groin area, the leg will be kept extended for four to six hours. If a vein or artery in the arm was used to insert the catheter, the arm will need to remain extended for a minimum of three hours.

The patient should expect a hard ridge to form over the incision site that diminishes as the site heals. Bluish discoloration under the skin at the point of insertion should also be expected but fades in two weeks. It is also not uncommon for the incision site to bleed during the first 24 hours following surgery. If this should happen, the patient should apply pressure to the site with a clean tissue or cloth for 10–15 minutes.

Risks

Similar to all surgical procedures, the cardiac catheterization test does involve some risks. Complications that may occur during the procedure include

- cardiac **arrhythmias** (an irregular heart beat)
- pericardial tamponade (a condition that causes excess pressure in the pericardium which affects the heart due to accumulation of excess fluid)



The cardiac catheter runs from the groin to the heart. (Illustration by Argosy Inc.)

- the rare occurrence of myocardial infarction (heart attack) or **stroke** may also develop due to clotting or plaque rupture of one or more of the coronary or brain arteries.

Before left-side catheterization is performed, the anticoagulant medication heparin may be administered. This drug helps decrease the risk of the development of a blood clot in an artery (thrombosis) and blood clots traveling throughout the body (embolization).

The risks of the catheterization procedure increase in patients over the age of 60, those who have severe **heart failure**, or persons with serious **valvular heart disease**.

Normal results

Normal findings from a cardiac catheterization will indicate no abnormalities of heart chamber size or configuration, wall motion or thickness, the direction of blood flow, or motion of the valves. Smooth and regular outlines on the x ray indicate normal coronary arteries.

An essential part of the catheterization is measuring intracardiac pressures, or the pressure in the heart's chambers and vessels. Pressure readings that are higher

KEY TERMS

Aneurysm—An abnormal dilatation of a blood vessel, usually an artery. It can be caused by a congenital defect or weakness in the vessel's wall.

Angiography—In cardiac catheterization, a picture of the heart and coronary arteries is seen after injecting a radiopaque substance (often referred to as a dye) throughout the veins and arteries.

Angioplasty—An alternative to vascular surgery, a balloon catheter is used to mechanically dilate the affected area of the artery and enlarge the constricted or narrowed segment.

Aortic valve—The valve between the heart's left ventricle and ascending aorta that prevents regurgitation of blood back into the left ventricle.

Catheter—A tube made of elastic, elastic web, rubber, glass, metal, or plastic used to evacuate or inject fluids into the body. In cardiac catheterization, a long, fine catheter is used for passage through a blood vessel into the chambers of the heart.

Coronary bypass surgery—A surgical procedure which places a shunt to allow blood to travel from the aorta to a branch of the coronary artery at a point past an obstruction.

Left anterior descending coronary artery (LAD)—One of the heart's coronary artery branches from the left main coronary artery which supplies blood to the left ventricle.

Mitral valve—The bicuspid valve which is between the left atrium and left ventricle of the heart.

Pulmonary valve—The heart valve which is positioned between the right ventricle and the opening into the pulmonary artery.

Shunt—A passageway (or an artificially created passageway) that diverts blood flow from one main route to another.

Tricuspid valve—The right atrioventricular valve of the heart.

than normal are significant for a patient's overall diagnosis. The pressure readings that are lower, other than those which are produced as a result of **shock**, typically are not significant.

An ejection fraction, or a comparison of how much blood is ejected from the heart's left ventricle during its contraction phase with a measurement of blood remaining at the end of the left ventricle's relaxation phase, is also determined by performing a catheterization. The cardiologist will look for a normal ejection fraction reading of 60–70%.

Abnormal results

Cardiac catheterization provides valuable still and motion x-ray pictures of the coronary arteries that help in diagnosing coronary artery disease, poor heart function, disease of the heart valves, and septal defects (a defect in the septum, the wall that separates two heart chambers).

The most prominent sign of coronary artery disease is the narrowing or blockage in the coronary arteries, with narrowing that is greater than 70% considered significant. A clear indication for intervention (by angioplasty or surgery) is a finding of significant narrowing of the left main coronary artery and/or blockage or severe narrowing in the high, left anterior descending coronary artery.

A finding of impaired wall motion is an additional indicator of coronary artery disease, aneurysm, an enlarged heart, or a congenital heart problem. Using the findings from an ejection fraction test which measures wall motion, cardiologists look at an ejection fraction reading under 35% as increasing the risk of complications while also decreasing a successful long term or short term outcome with surgery.

Detecting the difference in pressure above and below the heart valve can verify heart valve disease. The greater narrowing correlates with the higher pressure difference.

To confirm septal defects, a catheterization measures oxygen content on both the left and right sides of the heart. The right heart pumps unoxygenated blood to the lungs, and the left heart pumps blood that contains oxygen from the lungs to the rest of the body. Right side elevated oxygen levels indicate left-to-right atrial or **ventricular shunt**. A left side that experiences decreased oxygen indicates a right-to-left shunt.

Resources

BOOKS

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- "Heart." In *Current Medical Diagnosis and Treatment, 1996*. 35th ed. Ed. Stephen McPhee, et al. Stamford: Appleton & Lange, 1995.
- The Patient's Guide to Medical Tests.* Ed. Barry L. Zaret, et al. Boston: Houghton Mifflin, 1997.

ORGANIZATIONS

- American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.
- National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

Beth A. Kapes

Cardiac compression see **Cardiac tamponade**

Cardiac conduction disorder see **Heart block**

Cardiac mapping see **Electrophysiology study of the heart**



This 40-year-old male is working out on a treadmill, monitored by his physician, following heart surgery. (Custom Medical Stock Photo. Reproduced by permission.)

Cardiac rehabilitation

Definition

Cardiac rehabilitation is a comprehensive exercise, education, and behavioral modification program designed to improve the physical and emotional condition of patients with heart disease.

Purpose

Heart attack survivors, bypass and angioplasty patients, and individuals with angina, congestive heart failure, and heart transplants are all candidates for a cardiac rehabilitation program. Cardiac rehabilitation is prescribed to control symptoms, improve exercise tolerance, and improve the overall quality of life in these patients.

Precautions

A cardiac rehabilitation program should be implemented and closely monitored by a trained team of healthcare professionals.

Description

Cardiac rehabilitation is overseen by a specialized team of doctors, nurses, and other healthcare professionals. Members of the cardiac rehabilitation team may include a dietitian or nutritionist, physical therapist, exercise physiologist, psychologist, vocational counselor, occupational therapist, and social worker. The program frequently begins in a hospital setting and continues on an outpatient basis after the patient is discharged over a period of six to 12 months.

Components of a cardiac rehabilitation program vary by individual clinical need, and each program will be carefully constructed for the patient by his or her rehabilitation team.

- Exercise. Exercise programs typically start out slowly, with simple range-of-motion arm and leg exercises. Walking and stair climbing soon follow. Blood pressure is carefully monitored before and after exercise ses-

KEY TERMS

Angina—Chest pain.

Bypass surgery—A surgical procedure that grafts blood vessels onto arteries to reroute the blood flow around blockages in the arteries (arteriosclerosis).

sions, and patients are taught how to measure their heart rate and evaluate any possible cardiac symptoms during each session. Patients with advanced coronary disease may require continuous ECG monitoring throughout their exercise sessions. Once discharged from the hospital, the patient works with his cardiac team to create an individual exercise plan.

- **Diet.** Cardiac patients will work with a nutritionist or dietician to develop a low-fat, low-cholesterol diet plan. Patients with high blood pressure may be put on a salt-restricted diet and instructed to limit alcohol intake. Weight loss may also be a goal with obese cardiac patients.
- **Counseling.** A psychologist or social worker can help cardiac patients with issues that may be contributing to their heart condition, such as **stress** and **anxiety**. Relaxation techniques may be taught to patients to help them deal with these feelings. Cardiac patients frequently experience a period of depression, and group or individual counseling can be beneficial in overcoming these feelings. Vocational counselors can assist cardiac patients in returning to the workforce.
- **Education.** The patient and family should be fully educated on the physical limitations of the patient, his recommended diet and exercise plan, his emotional status, and the lifestyle changes required to improve the patient's overall health.
- **Smoking cessation.** Cardiac patients who smoke are twice as likely to have a heart attack in the following five years than non-smoking patients. These patients are strongly encouraged to enroll in a smoking cessation program, which typically includes patient education and behavioral counseling. Nicotine replacement therapy, which uses nicotine patches, nose spray, or gum to wean patients off of cigarettes, may also be part of the program. Antidepressants and anti-anxiety medication may be helpful in some cases.

Aftercare

Long-term maintenance is a critical feature of cardiac rehabilitation. Patients require support from their

healthcare team, family, and friends to continue the lifestyle changes they implemented during the rehabilitation period.

Risks

The risks of another heart attack during cardiac rehabilitation are slight, and greatly reduced by careful, continuous monitoring of the physical status of the patient.

Normal results

The outcome of the cardiac rehabilitation program depends on a number of variables, including patient follow-through, type and degree of heart disease, and the availability of an adequate support network for the patient. Patients who successfully complete the program will ideally reach an age-appropriate level of physical activity and be able to return to the workforce and/or other daily activities.

Resources

BOOKS

The American Heart Association. *American Heart Association Guide to Heart Attack Treatment, Recovery, and Prevention*. New York: Times Books, 1996.

DeBakey, Michael E., and Antonio Gotto Jr. *The New Living Heart*. Holbrook, MA: Adams Media Corporation, 1997.

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ORGANIZATIONS

American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

Paula Anne Ford-Martin

Cardiac tamponade

Definition

Cardiac tamponade occurs when the heart is squeezed by fluid that collects inside the sac that surrounds it.

Description

The heart is surrounded by a sac called the pericardium. When this sac becomes filled with fluid, the liquid presses on the heart, preventing the lower chambers of the heart from properly filling with blood.

Because the lower chambers (the ventricles) cannot fill with the correct amount of blood, less than normal amounts of blood reach the lungs and the rest of the body. This condition is very serious and can be fatal if not treated.

Causes and symptoms

Fluid can collect inside the pericardium and compress the heart when the kidneys do not properly remove waste from the blood, when the pericardium swells from unknown causes, from infection, or when the pericardium is damaged by **cancer**. Blunt or penetrating injury from trauma to the chest or heart can also result in cardiac tamponade when large amounts of blood fill the pericardium. Tamponade can also occur during heart surgery.

When the heart is compressed by the surrounding fluid, three conditions occur: a reduced amount of blood is pumped to the body by the heart, the lower chambers of the ventricles are filled with a less than normal amount of blood, and higher than normal blood pressures occur inside the heart, caused by the pressure of the fluid pushing in on the heart from the outside.

When tamponade occurs because of trauma, the sound of the heart beats can become faint, and the blood pressure in the arteries decreases, while the blood pressure in the veins increases.

In cases of tamponade caused by more slowly developing diseases, **shortness of breath**, a feeling of tightness in the chest, increased blood pressure in the large veins in the neck (the jugular veins), weight gain, and fluid retention by the body can occur.

Diagnosis

When cardiac tamponade is suspected, accurate diagnosis can be life-saving. The most accurate way to identify this condition is by using a test called an echocardiogram. This test uses sound waves to create an image of the heart and its surrounding sac, making it easy to visualize any fluid that has collected inside the sac.

Treatment

If the abnormal fluid buildup in the pericardial sac is caused by cancer or kidney disease, drugs used to treat these conditions can help lessen the amount of fluid collecting inside the sac. Drugs that help maintain normal blood pressure throughout the body can also help this condition; however, these drugs are only a temporary treatment. The fluid within the pericardium must be drained out to reduce the pressure on the heart and restore proper heart pumping.

The fluid inside the pericardium is drained by inserting a needle through the chest and into the sac itself. This

KEY TERMS

Pericardiocentesis—A procedure used to drain fluid out of the sac surrounding the heart. This is done by inserting a needle through the chest and into the sac.

allows the fluid to flow out of the sac, relieving the abnormal pressure on the heart. This procedure is called **pericardiocentesis**. In severe cases, a tube (catheter) can be inserted into the sac or a section of the sac can be surgically cut away to allow for more drainage.

Prognosis

This condition is life-threatening. However, drug treatments can be helpful, and surgical treatments can successfully drain the trapped fluid, though it may reaccumulate. Some risk of **death** exists with surgical drainage of the accumulated fluid.

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ORGANIZATIONS

- American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

Dominic De Bellis, PhD

Cardiac tumors see **Myxoma**

Cardiogenic shock see **Shock**

Cardiomyopathy see **Congestive cardiomyopathy; Restrictive cardiomyopathy**

Cardiopulmonary resuscitation (CPR)

Definition

Cardiopulmonary resuscitation (CPR) is a procedure to support and maintain breathing and circulation for a

person who has stopped breathing (respiratory arrest) and/or whose heart has stopped (cardiac arrest).

Purpose

CPR is performed to restore and maintain breathing and circulation and to provide oxygen and blood flow to the heart, brain, and other vital organs. CPR should be performed if a person is unconscious and not breathing. Respiratory and cardiac arrest can be caused by allergic reactions, an ineffective heartbeat, asphyxiation, breathing passages that are blocked, **choking**, drowning, drug reactions or overdoses, electric shock, exposure to cold, severe shock, or trauma. CPR can be performed by trained bystanders or healthcare professionals on infants, children, and adults. It should always be performed by the person on the scene who is most experienced in CPR.

Precautions

CPR should never be performed on a healthy person because it can cause serious injury to a beating heart by interfering with normal heartbeats.

Description

CPR is part of the emergency cardiac care system designed to save lives. Many deaths can be prevented by prompt recognition of the problem and notification of the emergency medical system (EMS), followed by early CPR, **defibrillation** (which delivers a brief electric shock to the heart in attempt to get the heart to beat normally), and advanced cardiac **life support** measures.

CPR must be performed within four to six minutes after cessation of breathing so as to prevent brain damage or **death**. It is a two-part procedure that involves rescue breathing and external chest compressions. To provide oxygen to a person's lungs, the rescuer administers mouth-to-mouth breaths, then helps circulate blood through the heart to vital organs by external chest compressions. Mouth-to-mouth breathing and external chest compression should be performed together, but if the rescuer is not strong enough to do both, the external chest compressions should be done. This is more effective than no resuscitation attempt, as is CPR that is performed "poorly."

When performed by a bystander, CPR is designed to support and maintain breathing and circulation until emergency medical personnel arrive and take over. When performed by healthcare personnel, it is used in conjunction with other basic and advanced life support measures.

According to the American Heart Association, early CPR and defibrillation combined with early advanced emergency care can increase survival rates for people

with a type of abnormal heart beat called **ventricular fibrillation** by as much as 40%. CPR by bystanders may prolong life during deadly ventricular fibrillation, giving emergency medical service personnel time to arrive.

However, many CPR attempts are not ultimately successful in restoring a person to a good quality of life. Often, there is brain damage even if the heart starts beating again. CPR is therefore not generally recommended for the chronically or terminally ill or frail elderly. For these people, it represents a traumatic and not a peaceful end of life.

Each year, CPR helps save thousands of lives in the United States. More than five million Americans annually receive training in CPR through American Heart Association and American Red Cross courses. In addition to courses taught by instructors, the American Heart Association also has an interactive video called Learning System, which is available at more than 500 healthcare institutions. Both organizations teach CPR the same way, but use different terms. These organizations recommend that family members or other people who live with people who are at risk for respiratory or cardiac arrest be trained in CPR. A hand-held device called a CPR Prompt is available to walk people trained in CPR through the procedure, using American Heart Association guidelines. CPR has been practiced for more than 40 years.

Performing CPR

The basic procedure for CPR is the same for all people, with a few modifications for infants and children to account for their smaller size.

PERFORMING CPR ON AN ADULT. The first step is to call the emergency medical system for help by telephoning 911; then to begin CPR, following these steps:

- The rescuer opens a person's airway by placing the head face up, with the forehead tilted back and the chin lifted. The rescuer checks again for breathing (three to five seconds), then begins rescue breathing (mouth-to-mouth artificial respiration), pinching the nostrils shut while holding the chin in the other hand. The rescuer's mouth is placed against the unconscious person's mouth with the lips making a tight seal, then gently exhales for about one to one and a half seconds. The rescuer breaks away for a moment and then repeats. The person's head is repositioned after each mouth-to-mouth breath.
- After two breaths, the rescuer checks the unconscious person's pulse by moving the hand that was under the person's chin to the artery in the neck (carotid artery). If the unconscious person has a heartbeat, the rescuer continues rescue breathing until help arrives or the per-

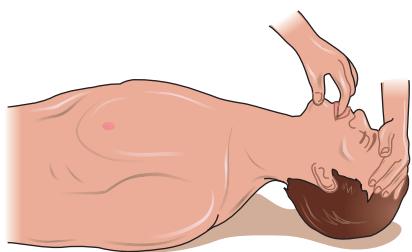


Figure A



Figure D

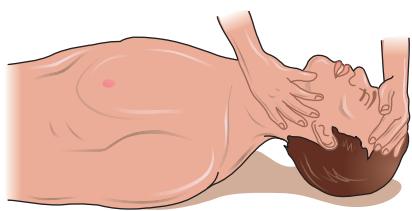


Figure B



Figure C



Figure E



Figure F

CPR in basic life support. Figure A: The victim should be flat on his back and his mouth should be checked for debris. Figure B: If the victim is unconscious, open airway, lift neck, and tilt head back. Figure C: If victim is not breathing, begin artificial breathing with four quick full breaths. Figure D: Check for carotid pulse. Figure E: If pulse is absent, begin artificial circulation by depressing sternum. Figure F: Mouth-to-mouth resuscitation of an infant. (Illustration by Electronic Illustrators Group.)

son begins breathing without assistance. If the unconscious person is breathing, the rescuer turns the person onto his or her side.

- If there is no heartbeat, the rescuer performs chest compressions. The rescuer kneels next to the unconscious person, placing the heel of one hand in the spot on the lower chest where the two halves of the rib cage come together. The rescuer puts one hand on top of the other on the person's chest and interlocks the fingers. The arms are straightened, the rescuer's shoulders are positioned directly above the hands on the unconscious person's chest. The hands are pressed down, using only the palms, so that the person's breastbone sinks in about 1.5–2 inches. The rescuer releases pressure without removing the hands, then repeats about 15 times per 10–15 second intervals.
- The rescuer tilts the unconscious person's head and returns to rescue breathing for one or two quick breaths. Then breathing and chest compressions are alternated for one minute before checking for a pulse. If the rescuer finds signs of a heartbeat and breathing, CPR is stopped. If the unconscious person is breathing but has no pulse, the chest compressions are continued. If the unconscious person has a pulse but is not breathing, rescue breathing is continued.
- For children over the age of eight, the rescuer performs CPR exactly as for an adult.

PERFORMING CPR ON AN INFANT OR CHILD UNDER THE AGE OF EIGHT. The procedures outlined above are followed with these differences:

- The rescuer administers CPR for one minute, then calls for help.
- The rescuer makes a seal around the child's mouth or infant's nose and mouth to give gentle breaths. The rescuer delivers 20 rescue breaths per minute, taking 1.5–2 seconds for each breath.
- Chest compressions are given with only one hand for a child and with two or three fingers for an infant. The breastbone is depressed only 1–1.5 in (2.5–3.8 cm) for a child and 0.5–1 in (1.3–2.5 cm) for an infant, and the rescuer gives at least 100 chest compressions per minute.

New developments in CPR

Some new ways of performing CPR have been tried. Active compression-decompression resuscitation, abdominal compression done in between chest compressions, and chest compression using a pneumatic vest have all been tested but none are currently recommended for routine use.

The active compression-decompression device was developed to improve blood flow from the heart, but clinical studies have found no significant difference in survival between standard and active compression-decompression CPR. Interposed abdominal counterpulsation, which requires two or more rescuers, one compressing the chest and the other compressing the abdomen, was developed to improve pressure and therefore blood flow. It has been shown in a small study to improve survival but more data is needed. A pneumatic vest, which circles the chest of an unconscious person and compresses it, increases pressure within the chest during external chest compression. The vest has been shown to improve survival in a preliminary study but more data is necessary for a full assessment.

Preparation

If a person suddenly becomes unconscious, a rescuer should call out for help from other bystanders, and then determine if the unconscious person is responsive by gently shaking the shoulder and shouting a question. Upon receiving no answer, the rescuer should call the emergency medical system. The rescuer should check to see whether the unconscious person is breathing by kneeling near the person's shoulders, looking at the person's chest, and placing a cheek next to the unconscious person's mouth. The rescuer should look for signs of breathing in the chest and abdomen, and listen and feel for signs of breathing through the person's lips. If no signs of breathing are present after three to five seconds, CPR should be started.

Aftercare

Emergency medical care is always necessary after successful CPR. Once a person's breathing and heartbeat have been restored, the rescuer should make the person comfortable and stay there until emergency medical personnel arrive. The rescuer can continue to reassure the person that help is coming and talk positively until professionals arrive and take over.

Risks

CPR can cause injury to a person's ribs, liver, lungs, and heart. However, these risks must be accepted if CPR is necessary to save the person's life.

Normal results

In many cases, successful CPR results in restoration of consciousness and life. Barring other injuries, a revived person usually returns to normal functions within a few hours of being revived.

Abnormal results

These include injuries incurred during CPR and lack of success with CPR. Possible sites for injuries include a person's ribs, liver, lungs, and heart. Partially successful CPR may result in brain damage. Unsuccessful CPR results in death.

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- American College of Osteopathic Emergency Physicians. 142 E. Ontario Street, Suite 550, Chicago, IL 60611. (312) 587-3709 or (800) 521-3709. Fax: (312) 587-9951. <<http://www.acoep.org>>.
- American Heart Association, National Center. 7272 Greenville Avenue, Dallas, TX 75231. (877) 242-4277. <http://www.americanheart.org/Heart_and_Stroke_A_Z_Guide/heim.html>.
- Heimlich Institute. PO Box 8858, Cincinnati, OH 45208. <heimlich@iglou.com>. <<http://www.heimlichinstitute.org/index.htm>>.
- National Safe Kids Campaign. 1301 Pennsylvania Avenue, Suite 1000, Washington, DC 20004-1707. <<http://pedscm.wustl.edu/All-Net/english/neurpage/protect/drown.htm>>.

KEY TERMS

Cardiac arrest—Temporary or permanent cessation of the heartbeat.

Cardiopulmonary—Relating to the heart and the lungs.

Defibrillation—A procedure to stop the type of irregular heart beat called ventricular fibrillation, usually by using electric shock.

Resuscitation—Bringing a person back to life after an apparent death or in cases of impending death.

Ventricular fibrillation—An irregular heartbeat where the heart beats very fast but ineffectively. Ventricular fibrillation is fatal if not quickly corrected.

OTHER

- American Heart Association. <<http://www.cpr-ecc.org/>> and <http://www.americanheart.org/Heart_and_Stroke_A_Z_Guide/cprns.html>.
- Columbia Presbyterian Medical Center. <http://cpmcnet.columbia.edu/texts/guide/hmg13_0001.html>.
- Learn CPR. <<http://www.learn-cpr.com>>.
- National Registry of Cardiopulmonary Resuscitation. <<http://www.nrcpr.org/>>.
- University of Washington School of Medicine. <<http://depts.washington.edu/learncpr/>>.

L. Fleming Fallon, Jr., MD, DrPH

Cardioversion

Definition

Cardioversion refers to the process of restoring the heart's normal rhythm by applying a controlled electric shock to the exterior of the chest.

Purpose

When the heart beats too fast, blood no longer circulates effectively in the body. Cardioversion is used to stop this abnormal beating so that the heart can begin normal rhythm and pump more efficiently.

Precautions

Not all unusual heart rhythms (called **arrhythmias**) are dangerous or fatal. Atrial fibrillation and atrial flutter often revert to normal rhythms without the need for car-

KEY TERMS

Atrial fibrillation—A condition in which the upper chamber of the heart quivers instead of pumping in an organized way.

Atrial flutter—A rapid pulsation of the upper chamber of the heart that interferes with normal function.

Ventricular fibrillation—A condition in which the lower chamber of the heart quivers instead of pumping in an organized way.

Ventricular tachycardia—A rapid heart beat, usually over 100 beats per minute.

dioversion. Healthcare providers may also try to correct the heart rhythm with medication or recommend a lifestyle change before trying cardioversion. However, **ventricular tachycardia** lasting more than 30 seconds and **ventricular fibrillation** require immediate cardioversion.

Description

Elective cardioversion is usually scheduled ahead of time. After arriving at the hospital, an intravenous (IV) catheter will be placed in the arm and oxygen will be given through a face mask. A short-acting general anesthetic will be administered through the vein. During the two or three minutes of anesthesia, the doctor will apply two paddles to the exterior of the chest and administer the electric shock. It may be necessary to give the shock two or three times to obtain normal rhythm.

Preparation

Medication to thin the blood is usually given for at least three weeks before elective cardioversion. Food intake should be stopped eight hours before the procedure.

Aftercare

Medical personnel will monitor the heart rhythm for a few hours, after which the patient is usually sent home. It is advisable to arrange for transportation home, because drowsiness may last several hours. The doctor may prescribe anti-arrhythmic medication to prevent the abnormal rhythm from returning.

Risks

Cardioverters have been in use for many years and the risks are few. Those unlikely risks that remain

include those instances when the device delivers greater or lesser power than expected or when power setting and control knobs are not set correctly. Unfortunately, in a number of cases, the heart prefers its abnormal rhythm and reverts to it despite cardioversion.

Normal results

Most cardioversions are successful and, at least for a time, restore the normal heart rhythm.

Resources

BOOKS

McGoon, Michael D., ed. *Mayo Clinic Heart Book: The Ultimate Guide to Heart Health*. New York: William Morrow and Co., Inc., 1993.

ORGANIZATIONS

American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

Dorothy Elinor Stonely

Carisoprodol see **Muscle relaxants**

Carotid artery surgery see **Endarterectomy**

Carotid Doppler ultrasound see **Doppler ultrasonography**

Carotid endarterectomy see **Endarterectomy**

Carotid sinus massage

Definition

Carotid sinus massage involves rubbing the large part of the arterial wall at the point where the common carotid artery, located in the neck, divides into its two main branches.

Purpose

Sinus, in this case, means an area in a blood vessel that is bigger than the rest of the vessel. This is a normal dilation of the vessel. Located in the neck just below the angle of the jaw, the carotid sinus sits above the point where the carotid artery divides into its two main branches. Rubbing the carotid sinus stimulates an area in the artery wall that contains nerve endings. These nerves respond to changes in blood pressure and are capable of slowing the heart rate. The response to this simple procedure often slows a rapid heart rate (for example, atrial

KEY TERMS

- Angina pectoris**—Chest pain usually caused by a lack of oxygen in the heart muscle.
- Arrhythmia**—Any deviation from a normal heart beat.
- Atrial fibrillation**—A condition in which the upper chamber of the heart quivers instead of pumping in an organized way.
- Atrial flutter**—Rapid, inefficient contraction of the upper chamber of the heart.
- Carotid artery**—One of the major arteries supplying blood to the head and neck.
- Tachycardia**—A rapid heart beat, usually over 100 beats per minute.

flutter or atrial tachycardia) and can provide important diagnostic information to the physician.

Description

The patient will be asked to lie down, with the neck fully extended and the head turned away from the side being massaged. While watching an electrocardiogram monitor, the doctor will gently touch the carotid sinus. If there is no change in the heart rate on the monitor, the pressure is applied more firmly with a gentle rotating motion. After massaging one side of the neck, the massage will be repeated on the other side. Both sides of the neck are never massaged at the same time.

Preparation

No special preparation is needed for carotid sinus massage.

Aftercare

No aftercare is required.

Risks

The physician must be sure there is no evidence of blockage in the carotid artery before performing the procedure. Massage in a blocked area might cause a clot to break loose and cause a **stroke**.

Normal results

Carotid sinus massage will slow the heart rate during episodes of atrial flutter, fibrillation, and some tachycar-

dias. It has been known to stop the arrhythmia completely. If the procedure is being done to help diagnose **angina pectoris**, massaging the carotid sinus may make the discomfort go away.

Resources

BOOKS

McGood, Michael D., ed. *Mayo Clinic Heart Book: The Ultimate Guide to Heart Health*. New York: William Morrow and Co., Inc., 1993.

Dorothy Elinor Stonely

Carpal tunnel syndrome

Definition

Carpal tunnel syndrome is a disorder caused by compression at the wrist of the median nerve supplying the hand, causing **numbness and tingling**.

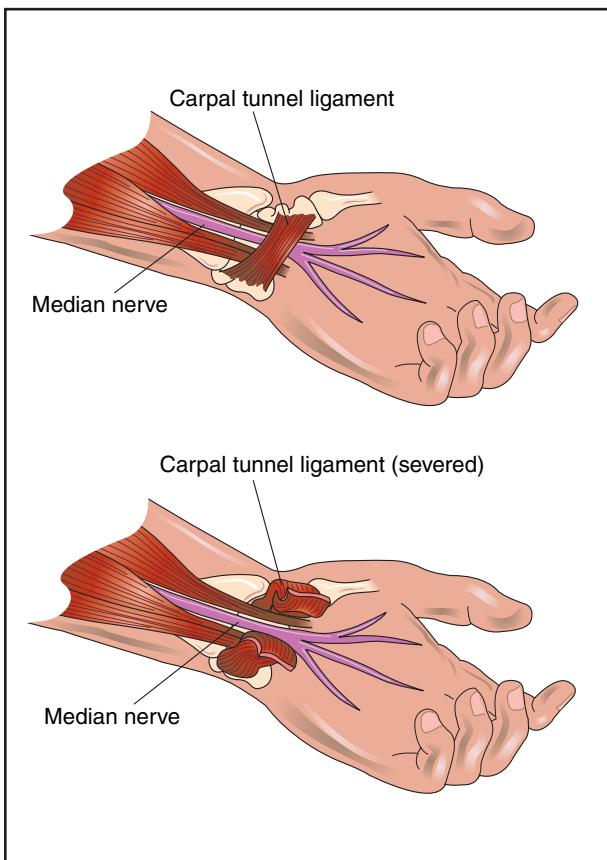
Description

The carpal tunnel is an area in the wrist where the bones and ligaments create a small passageway for the median nerve. The median nerve is responsible for both sensation and movement in the hand, in particular the thumb and first three fingers. When the median nerve is compressed, an individual's hand will feel as if it has "gone to sleep."

Women between the ages of 30 and 60 have the highest rates of carpal tunnel syndrome. Research has demonstrated that carpal tunnel syndrome is a very significant cause of missed work days due to **pain**. In 1995, about \$270 million was spent on sick days taken for pain from repetitive motion injuries.

Causes and symptoms

Compression of the median nerve in the wrist can occur during a number of different conditions, particularly those conditions which lead to changes in fluid accumulation throughout the body. Because the area of the wrist through which the median nerve passes is very narrow, any swelling in the area will lead to pressure on the median nerve. This pressure will ultimately interfere with the nerve's ability to function normally. **Pregnancy**, **obesity**, arthritis, certain thyroid conditions, diabetes, and certain pituitary abnormalities all predispose to carpal tunnel syndrome. Other conditions which increase the risk for carpal tunnel syndrome include some forms



The most severe cases of carpal tunnel syndrome may require surgery to decrease the compression of the median nerve and restore its normal function. This procedure involves severing the ligament that crosses the wrist, thus allowing the median nerve more room and decreasing compression. (Illustration by Electronic Illustrators Group.)

of arthritis and various injuries to the arm and wrist (including **fractures**, sprains, and dislocations). Furthermore, activities which cause an individual to repeatedly bend the wrist inward toward the forearm can predispose to carpal tunnel syndrome. Certain jobs which require repeated strong wrist motions carry a relatively high risk of carpal tunnel syndrome. Injuries of this type are referred to as "repetitive motion" injuries, and are more frequent among secretaries doing a lot of typing, people working at computer keyboards or cash registers, factory workers, and some musicians.

Symptoms of carpal tunnel syndrome include numbness, burning, tingling, and a prickly pin-like sensation over the palm surface of the hand, and into the thumb, forefinger, middle finger, and half of the ring finger. Some individuals notice a shooting pain which goes from the wrist up the arm, or down into the hand and fingers. With continued median nerve compression, an individual may begin to experience muscle weakness, making it dif-

ficult to open jars and hold objects with the affected hand. Eventually, the muscles of the hand served by the median nerve may begin to grow noticeably smaller (**atrophy**), especially the fleshy part of the thumb. Untreated, carpal tunnel syndrome may eventually result in permanent weakness, loss of sensation, or even **paralysis** of the thumb and fingers of the affected hand.

Diagnosis

The diagnosis of carpal tunnel syndrome is made in part by checking to see whether the patient's symptoms can be brought on by holding his or her hand in position with wrist bent for about a minute. Wrist x rays are often taken to rule out the possibility of a tumor causing pressure on the median nerve. A physician examining a patient suspected of having carpal tunnel syndrome will perform a variety of simple tests to measure muscle strength and sensation in the affected hand and arm. Further testing might include electromyographic or nerve conduction velocity testing to determine the exact severity of nerve damage. These tests involve stimulating the median nerve with electricity and measuring the resulting speed and strength of the muscle response, as well as recording speed of nerve transmission across the carpal tunnel.

Treatment

Carpal tunnel syndrome is initially treated with splints, which support the wrist and prevent it from flexing inward into the position which exacerbates median nerve compression. Some people get significant relief by wearing such splints to sleep at night, while others will need to wear the splints all day, especially if they are performing jobs which **stress** the wrist. Ibuprofen or other **nonsteroidal anti-inflammatory drugs** may be prescribed to decrease pain and swelling. When carpal tunnel syndrome is more advanced, injection of steroids into the wrist to decrease inflammation may be necessary.

The most severe cases of carpal tunnel syndrome may require surgery to decrease the compression of the median nerve and restore its normal function. Such a repair involves cutting that ligament which crosses the wrist, thus allowing the median nerve more room and decreasing compression. This surgery is done almost exclusively on an outpatient basis and is often performed without the patient having to be made unconscious. Careful injection of numbing medicines (local anesthesia) or nerve blocks (the injection of anesthetics directly into the nerve) create sufficient numbness to allow the surgery to be performed painlessly, without the risks associated with general anesthesia. Recovery from this type of surgery is usually quick and without complications.

KEY TERMS

Carpal tunnel—A passageway in the wrist, created by the bones and ligaments of the wrist, through which the median nerve passes.

Electromyography—A type of test in which a nerve's function is tested by stimulating a nerve with electricity, and then measuring the speed and strength of the corresponding muscle's response.

Median nerve—A nerve which runs through the wrist and into the hand. It provides sensation and some movement to the hand, the thumb, the index finger, the middle finger, and half of the ring finger.

Prognosis

Without treatment, continued pressure on the median nerve puts an individual at risk for permanent disability in the affected hand. Most people are able to control the symptoms of carpal tunnel syndrome with splinting and anti-inflammatory agents. For those who go on to require surgery, about 95% will have complete cessation of symptoms.

Prevention

Prevention is generally aimed at becoming aware of the repetitive motions which one must make which could put the wrist into a bent position. People who must work long hours at a computer keyboard, for example, may need to take advantage of recent advances in "ergonomics," which try to position the keyboard and computer components in a way that increases efficiency and decreases stress. Early use of a splint may also be helpful for people whose jobs increase the risk of carpal tunnel syndrome.

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Rosalyn Carson-DeWitt, MD

Casts see **Immobilization**

CAT scan see **Computed tomography scans**

Cat-bite infection see **Animal bite infections**

Cat-scratch disease

Definition

Cat-scratch disease is an uncommon infection that typically results from a cat's scratch or bite. Most sufferers experience only moderate discomfort and find that their symptoms clear up without any lasting harm after a few weeks or months. Professional medical treatment is rarely needed.

Description

Cat-scratch disease (also called cat-scratch **fever**) is caused by the *Bartonella henselae* bacterium, which is found in cats around the world and is transmitted from cat to cat by fleas. Researchers have discovered that large numbers of North American cats carry antibodies for the disease (meaning that the cats have been infected at some point in their lives). Some parts of North America have much higher rates of cat infection than others, however. *Bartonella henselae* is uncommon or absent in cold climates, which fleas have difficulty tolerating, but prevalent in warm, humid places such as Memphis, Tennessee, where antibodies were found in 71% of the cats tested. The bacterium, which remains in a cat's bloodstream for several months after infection, seems to be harmless to most cats, and normally an infected cat will not display any symptoms. Kittens (cats less than one year old) are more likely than adult cats to be carrying the infection.

Bartonella henselae can infect people who are scratched or (more rarely) bitten or licked by a cat. It cannot be passed from person to person. Although cats are popular pets found in about 30% of American households, human infection appears to be rare. One study estimated that for every 100,000 Americans there are only 2.5 cases of cat-scratch disease each year (2.5/100,000). It is also unusual for more than one family member to become ill; a Florida investigation discovered multiple cases in only 3.5% of the families studied. Children and teenagers appear to be the most likely victims of cat-scratch disease, although the possibility exists that

KEY TERMS

Acetaminophen—A drug for relieving pain and fever.

AIDS—Acquired immunodeficiency syndrome. A disease that attacks the immune system.

Antibiotics—A category of manufactured substances used to combat infection.

Antibodies—Special substances created by the body to combat infection.

Bacterium—A tiny organism. Some bacteria cause disease.

Hepatitis—A disease that inflames the liver.

Immune system—A body system that combats disease.

Immunocompromised—Having a damaged immune system.

Lymph nodes—Small, kidney-shaped organs that filter a fluid called lymph.

Pneumonia—A disease that inflames the lungs.

Pus—A thick yellowish or greenish fluid.

the disease may be more common among adults than previously thought.

Causes and symptoms

The first sign of cat-scratch disease may be a small blister at the site of a scratch or bite three to 10 days after injury. The blister (which sometimes contains pus) often looks like an insect bite and is usually found on the hands, arms, or head. Within two weeks of the blister's appearance, lymph nodes near the site of injury become swollen. Often the infected person develops a fever or experiences **fatigue** or headaches. The symptoms usually disappear within a month, although the lymph nodes may remain swollen for several months. Hepatitis, **pneumonia**, and other dangerous complications can arise, but the likelihood of cat-scratch disease posing a serious threat to health is very small. **AIDS** patients and other immunocompromised people face the greatest risk of dangerous complications.

Occasionally, the symptoms of cat-scratch disease take the form of what is called Parinaud's oculoglandular syndrome. In such cases, a small sore develops on the palpebral conjunctiva (the membrane lining the inner eyelid), and is often accompanied by **conjunctivitis** (inflammation of the membrane) and swollen lymph nodes in front of the ear. Researchers suspect that the first step in the develop-

ment of Parinaud's oculoglandular syndrome occurs when *Bartonella henselae* bacteria pass from a cat's saliva to its fur during grooming. Rubbing one's eyes after handling the cat then transmits the bacteria to the conjunctiva.

Diagnosis

A family doctor should be called whenever a cat scratch or bite fails to heal normally or is followed by a persistent fever or other unusual symptoms such as long-lasting bone or joint **pain**. The appearance of painful and swollen lymph nodes is another reason for consulting a doctor. When cat-scratch disease is suspected, the doctor will ask about a history of exposure to cats and look for evidence of a cat scratch or bite and swollen lymph nodes. A blood test for *Bartonella henselae* may be ordered to confirm the doctor's diagnosis.

Treatment

For otherwise healthy people, rest and over-the-counter medications for reducing fever and discomfort (such as **acetaminophen**) while waiting for the disease to run its course are usually all that is necessary. **Antibiotics** are prescribed in some cases, particularly when complications occur or the lymph nodes remain swollen and painful for more than two or three months, but there is no agreement among doctors about when and how they should be used. If a lymph node becomes very swollen and painful, the family doctor may decide to drain it.

Prognosis

Most people recover completely from a bout of cat-scratch disease. Further attacks are rare.

Prevention

Certain common-sense precautions can be taken to guard against the disease. Scratches and bites should be washed immediately with soap and water, and it is never a good idea to rub one's eyes after handling a cat without first washing one's hands. Children should be told not to play with stray cats or make cats angry. Immunocompromised people should avoid owning kittens, which are more likely than adult cats to be infectious. Because cat-scratch disease is usually not a life-threatening illness and people tend to form strong emotional bonds with their cats, doctors do not recommend getting rid of a cat suspected of carrying the disease.

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Howard Baker

Cat-scratch fever see **Cat-scratch disease**

Cataract surgery

Definition

Cataract surgery is a procedure performed to remove a cloudy lens from the eye; usually an intraocular lens is implanted at the same time.

Purpose

The purpose of cataract surgery is to restore clear vision. It is indicated when cloudy vision due to **cataracts** has progressed to such an extent that it interferes with normal daily activities.

Precautions

Cataract surgery is not performed on both eyes at once. To avoid risking blindness in both eyes in the event of infection or other catastrophe, the first eye is allowed to heal before the cataract is removed from the second eye.

The presence of cataracts can mask additional eye problems, such as retinal damage, that neither doctors nor patients are aware of prior to surgery. Since such conditions will continue to impair sight after cataract removal if they are not identified and treated, the eventual outcome of cataract surgery will depend on the outcome of other problems.

In 1997 and 1998, evidence that cataract surgery can contribute to the progression of age-related **macular degeneration** (ARMD) was published. ARMD is the degeneration of the central part of the retina. Accordingly, ARMD patients with cataracts must weigh the risks of the loss of central vision, within four or five years, against short-term improvement. When an ARMD patient chooses cataract surgery, the surgeon should shield the retina against bright light to protect it from possible light-induced damage during surgery and install an intraocular lens capable of absorbing ultraviolet and blue light, which seem to do the most damage.

KEY TERMS

Age-related macular degeneration (ARMD)—Degeneration of the macula (the central part of the retina where the rods and cones are most dense) that leads to loss of central vision in people over 60.

Cataract—Progressive opacity or clouding of an eye lens, which obstructs the passage of light to the retina.

Cornea—Clear outer covering of the front of the eye.

Intraocular lens—Lens made of silicone or plastic placed within the eye; can be corrective.

Retina—Innermost layer at the back of the eye, which contains light receptors, the rods and cones.

Description

There are two types of cataract surgery: intracapsular and extracapsular. Intracapsular surgery is the removal of both the lens and the thin capsule that surround them. This type of surgery was common before 1980, but has since been displaced by extracapsular surgery. Removal of the capsule requires a large incision and doesn't allow comfortable intraocular lens implantation. Thus, people who undergo intracapsular cataract surgery have long recovery periods and have to wear very thick glasses.

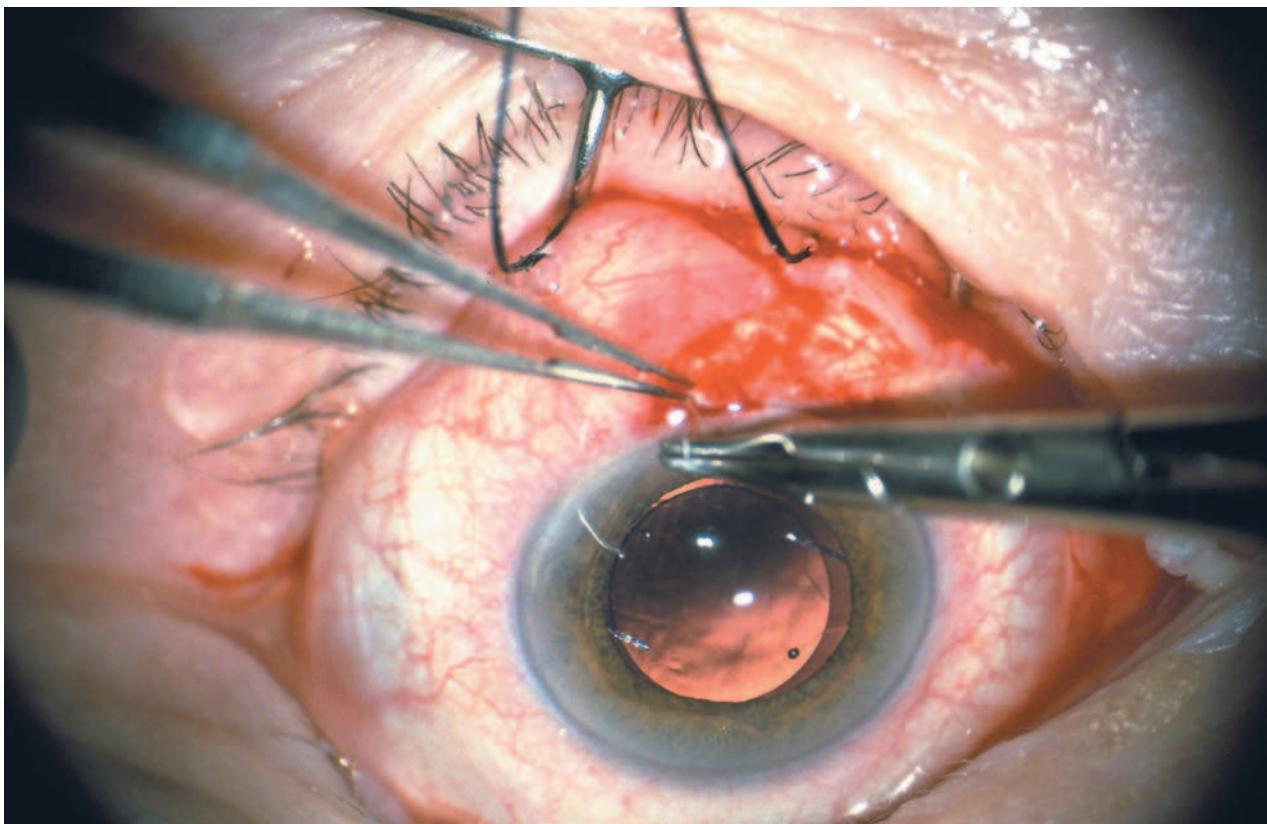
Extracapsular cataract surgery is the removal of the lens where the capsule is left in place. Each year in the United States, over a million cataracts are removed this way.

There are two methods for extracapsular cataract surgery. The usual technique is phacoemulsification. A tiny incision (about 0.12 in or 3 mm long) is made next to the cornea (the eye's outer covering), and an ultrasonic probe is used to break the cataract into minute pieces, which are then removed by suction. When the lens is too hard to be emulsified ultrasonically, the surgeon will use a different extracapsular technique requiring a larger incision. An incision about 0.37 in (9 mm) long is made, and the whole lens (without its capsule) is removed through the incision. Both kinds of extracapsular extraction leave the back of the capsule intact, so a silicone or plastic intraocular lens can be stably implanted in about the same location as the original lens.

The surgery takes about 30–60 minutes per eye.

Preparation

Patients must have a pre-operation **eye examination**, which will include ultrasound analysis to make sure the



Cataract surgery in progress. (Photograph by David Sutton/Zuma Images, The Stock Market. Reproduced by permission.)

retina (the innermost layer of the eye, containing the light receptors) is intact and also to measure eye curvature so that a lens with the proper correction can be implanted. The patient will also have a pre-operative **physical examination**. In addition, patients start a course of antibiotic eye drops or ointment the day before surgery.

Aftercare

Proper post-operative care is especially important after cataract surgery. Patients will need someone to drive them home after the surgery and should not bend over or do anything strenuous for about two weeks. They should refrain from rubbing the eye, should wear glasses to protect their eye, and should wear a shield while sleeping so the eye won't be rubbed or bumped accidentally. The patient will usually continue their antibiotic for two to three weeks and will also take anti-inflammatory medication for about the same length of time. If the patient experiences inflammation, redness, or **pain**, they should seek immediate medical treatment to avoid serious complications.

Risks

Cataract surgery itself is quite safe; over 90% of the time, there are no complications. Possible complications

include intraocular infection (endophthalmitis), central retinal inflammation (macular **edema**), post-operative **glaucoma**, **retinal detachment**, bleeding under the retina (choroidal hemorrhage), and tiny lens fragments in the back (vitreous) cavity of the eye, all of which can lead to loss of sight.

Normal results

Ordinarily, patients experience improved visual acuity and improved perception of the vividness of colors, leading to increased abilities in many activities, including reading, needlework, driving, golf, and tennis, for example. In addition, sometimes implanted corrective lenses eliminate the need for eyeglasses or contact lenses.

Resources

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ORGANIZATIONS

American Academy of Ophthalmology. 655 Beach Street, P.O. Box 7424, San Francisco, CA 94120-7424. <<http://www.eyenet.org>>.

American Society of Cataract and Refractive Surgery. 4000 Legato Road, Suite 850, Fairfax, VA 22033-4055. (703) 591-2220. <<http://www.ascrs.org>>.

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Lorraine Lica, PhD

Cataracts

Definition

A cataract is a cloudiness or opacity in the normally transparent crystalline lens of the eye. This cloudiness can cause a decrease in vision and may lead to eventual blindness.

Description

The human eye has several parts. The outer layer of the eyeball consists of a transparent dome-shaped cornea and an opaque, white sclera. The cornea and sclera help protect the eye. The next layer includes the iris, pupil, and ciliary body. The iris is the colored part of the eye and the pupil is the small dark round hole in the middle of the iris. The pupil and iris allow light into the eye. The ciliary body contains muscles that help in the eye’s focusing ability. The lens lies behind the pupil and iris. It is covered by a cellophane-like capsule. The lens is normally transparent, elliptical in shape, and somewhat elastic. This elasticity allows the lens to focus on both near and far objects. The lens is attached to the ciliary body by fibers (zonules of Zinn). Muscles in the ciliary body act on the zonules, which then change the shape of the lens. This process is called accommodation—the lens focuses images to help make vision clear. As people age, the lens hardens and changes shape less easily. As a result, the accommodation process becomes more difficult, making it harder to see things up close. This generally occurs around the age of 40 and continues until about age 65. The condition is called **presbyopia**. It is a normal condition of **aging**, generally resulting in the need for reading glasses.

The lens is made up of approximately 35% protein and 65% water. As people age, degenerative changes in the lens’ proteins occur. Changes in the proteins, water content, enzymes, and other chemicals are some of the reasons for the formation of a cataract.

The major areas of the lens are the nucleus, the cortex, and the capsule. The nucleus is in the center of the

lens, the cortex surrounds the nucleus, and the capsule is the outer layer. Opacities can occur in any area of the lens. Cataracts, then, can be classified according to location (nuclear, cortical, or posterior subcapular cataracts). The density and location of the cataract determines the amount of vision affected. If the cataract forms in the area of the lens directly behind the pupil, vision may be significantly impaired. A cataract that occurs on the outer edges or side of the lens will create less of a visual problem.

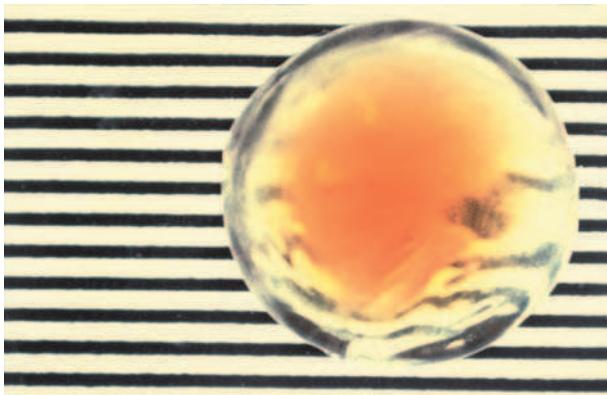
Cataracts in the elderly are so common that they are thought to be a normal part of the aging process. Between the ages of 52 and 64, there is a 50% chance of having a cataract, while at least 70% of those 70 and older are affected. Cataracts associated with aging (senile or age-related cataracts) most often occur in both eyes, with each cataract progressing at a different rate. Initially, cataracts may not affect vision. If the cataract remains small or at the periphery of the lens, the visual changes may be minor.

Cataracts that occur in people other than the elderly are much less common. Congenital cataracts occur very rarely in newborns. Genetic defects or an infection or disease in the mother during **pregnancy** are among the causes of congenital cataracts. Traumatic cataracts may develop after a foreign body or trauma injures the lens or eye. Systemic illnesses, such as diabetes, may result in cataracts. Cataracts can also occur secondary to other eye diseases—for example, an inflammation of the inner layer of the eye (**uveitis**) or **glaucoma**. Such cataracts are called complicated cataracts. Toxic cataracts result from chemical toxicity, such as steroid use. Cataracts can also result from exposure to the sun’s ultraviolet (UV) rays.

Causes and symptoms

Recent studies have been conducted to try to determine whether diet or the use of **vitamins** might have an effect on the formation of cataracts in older people. The results have been mixed, with some studies finding that there is a connection and other studies finding none. Much interest has been focused on the use of antioxidant supplements as a protection against cataracts. Antioxidant vitamins such as vitamins A, C, E and beta-carotene help the body clean-up oxygen-free radicals. Some vitamins are marketed specifically for the eyes. Patients should speak to their doctors about the use of such vitamins.

Smoking and alcohol intake have been implicated in cataract formation. Some studies have determined that a diet high in fat will increase the likelihood of cataract formation, while an increase in foods rich in antioxidants will reduce the incidence. More research is needed to determine if diet, smoking, alcohol consumption, or vitamins have any connection to the formation of cataracts.



A dense cataract on lens of eye. (Photograph by Margaret Cubberly, Phototake NYC. Reproduced by permission.)

There are several common symptoms of cataracts:

- gradual, painless onset of blurry, filmy, or fuzzy vision
- poor central vision
- frequent changes in eyeglass prescription
- changes in color vision
- increased glare from lights, especially oncoming headlights when driving at night
- “second sight” improvement in near vision (no longer needing reading glasses), but a decrease in distance vision
- poor vision in sunlight
- presence of a milky whiteness in the pupil as the cataract progresses.

Diagnosis

Both ophthalmologists and optometrists may detect and monitor cataract growth and prescribe prescription lenses for visual deficits. However, only an ophthalmologist can perform cataract extraction.

Cataracts are easily diagnosed from the reporting of symptoms, a visual acuity exam using an eye chart, and by examination of the eye itself. Shining a penlight into the pupil may reveal opacities or a color change of the lens even before visual symptoms have developed. An instrument called a slit lamp is basically a large microscope. This lets the doctor examine the front of the eye and the lens. The slit lamp helps the doctor determine the location of the cataract.

Some other diagnostic tests may be used to determine if cataracts are present or how well the patient may potentially see after surgery. These include a glare test, potential vision test, and contrast sensitivity test.

Treatment

For cataracts that cause no symptoms or only minor visual changes, no treatment may be necessary. Continued monitoring and assessment of the cataract is needed by an ophthalmologist or optometrist at scheduled office visits. Increased strength in prescription eyeglasses or contact lenses may be helpful. This may be all that is required if the cataract does not reduce the patient’s quality of life.

Cataract surgery—the only option for patients whose cataracts interfere with vision to the extent of affecting their daily lives—is the most frequently performed surgery in the United States. It generally improves vision in over 90% of patients. Some people have heard that a cataract should be “ripe” before being removed. A “ripe” or mature cataract is when the lens is completely opaque. Most cataracts are removed before they reach that stage. Sometimes cataracts need to be removed so that the doctor can examine the back of the eye more carefully. This is important in patients with diseases that may affect the eye. If cataracts are present in both eyes, only one eye at a time should be operated on. Healing occurs in the first eye before the second cataract is removed, sometimes as early as the following week. A final eyeglass prescription is usually given about four to six weeks after surgery. Patients will still need reading glasses. The overall health of the patient needs to be considered in making the decision to operate. However, age alone need not preclude effective surgical treatment of cataracts. People in their 90s can have successful return of vision after **cataract surgery**.

Surgery to remove cataracts is generally an outpatient procedure. A local anesthetic is used and the procedure lasts about an hour. Removal of the cloudy lens can be done by several different procedures. The three types of cataract surgery available are:

- Extracapsular cataract extraction. This type of cataract extraction is the most common. The lens and the front portion of the capsule are removed. The back part of the capsule remains, providing strength to the eye.
- Intracapsular cataract extraction. The lens and the entire capsule are removed. This method carries an increased risk for detachment of the retina and swelling after surgery. It is rarely used.
- Phacoemulsification. This type of extracapsular extraction needs a very small incision, resulting in faster healing. Ultrasonic vibration is applied to the lens to break it up into very small pieces which are then aspirated out of the eye with suction by the ophthalmologist.

A replacement lens is usually inserted at the time of the surgery. A plastic artificial lens called an intraocular lens (IOL) is placed in the remaining posterior lens capsule of the eye. When the intracapsular extraction

method is used, an IOL may be clipped onto the iris. Contact lenses and cataract glasses (aphakic lenses) are prescribed if an IOL was not inserted. A folding IOL is used when phacoemulsification is performed to accommodate the small incision.

Antibiotic drops to prevent infection and steroids to reduce inflammation are prescribed after surgery. An eye shield or glasses during the day will protect the eye from injury while it heals. During the night, an eye shield is worn. The patient returns to the doctor the day after surgery for assessment, with several follow-up visits over the next two months to monitor the healing process.

Prognosis

The success rate of cataract extraction is very high, with a good prognosis. A visual acuity of 20/40 or better may be achieved. If an extracapsular cataract extraction was performed, a secondary cataract may develop in the remaining back portion of the capsule. This can occur one to two years after surgery. YAG capsulotomy is most often used for this type of cataract. YAG stands for yttrium aluminum garnet, the name of the laser used for this procedure. This is a painless outpatient procedure and requires no incision. The laser beam makes a small opening in the remaining back part of the capsule, allowing light through.

In a very small percentage (3–5%) of surgical cataract extractions, complications occur. Infections, swelling of the cornea (**edema**), bleeding, **retinal detachment**, and the onset of glaucoma have been reported. Some problems may occur one to two days, or even several weeks, after surgery. Any haziness, redness, decrease in vision, nausea, or **pain** should be reported to the surgeon immediately.

Prevention

Preventive measures emphasize protecting the eyes from UV radiation by wearing glasses with a special coating to protect against UV rays. Dark lenses alone are not sufficient. The lenses must protect against UV light (specifically, UV-A and UV-B). Antioxidants may also provide some protection by reducing free radicals that can damage lens proteins. A healthy diet rich in sources of antioxidants, including citrus fruits, sweet potatoes, carrots, green leafy vegetables, and/or vitamin supplements may be helpful. When taking certain medications, such as steroids, more frequent eye exams may be necessary. Patients should speak to their doctors to see if medications may affect their eyes.

Resources

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KEY TERMS

Aphakia—Absence of the lens of the eye.

Ciliary body—A structure in the eye that contains muscles which will affect the focusing of the lens.

Glaucoma—Disease of the eye characterized by increased pressure of the fluid inside the eye. Untreated, glaucoma can lead to blindness.

Phacoemulsification—Surgical procedure to remove a cataract using sound waves to disintegrate the lens which is then removed by suction.

Retina—The innermost layer of the eyeball. Images focused onto the retina are then sent to the brain.

Ultraviolet radiation (UV)—Invisible light rays which may be responsible for sunburns, skin cancers, and cataract formation.

Uveitis—Inflammation of the uvea. The uvea is a continuous layer of tissue which consists of the iris, the ciliary body, and the choroid. The uvea lies between the retina and sclera.

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ORGANIZATIONS

American Academy of Ophthalmology. 655 Beach Street, P.O. Box 7424, San Francisco, CA 94120-7424. <<http://www.eyenet.org>>.

American Optometric Association. 243 North Lindbergh Blvd., St. Louis, MO 63141. (314) 991-4100. <<http://www.aoanet.org>>.

The Lighthouse. 111 East 59th St., New York, NY 10022. (800) 334-5497. <<http://www.lighthouse.org>>.

Prevent Blindness America. 500 East Remington Road, Schaumburg, IL 60173. (800) 331-2020. <<http://www.preventblindness.org>>.

Cynthia L. Frozena, RN

Catatonia

Definition

Catatonia is a condition marked by changes in muscle tone or activity associated with a large number of serious mental and physical illnesses. There are two distinct sets of symptoms that are characteristic of this condition. In catatonic stupor the individual experiences a

deficit of motor (movement) activity that can render him/her motionless. Catatonic excitement, or excessive movement, is associated with violent behavior directed toward oneself or others.

Features of catatonia may also be seen in Neuroleptic Malignant Syndrome (NMS) which is an uncommon (but potentially lethal) reaction to some medications used to treat major mental illnesses. NMS is considered a medical emergency since 25% of untreated cases result in **death**. Catatonia can also be present in individuals suffering from a number of other physical and emotional conditions such as drug intoxication, depression, and **schizophrenia**. It is most commonly associated with **mood disorders**.

Description

In catatonic stupor, motor activity may be reduced to zero. Individuals avoid bathing and grooming, make little or no eye contact with others, may be mute and rigid, and initiate no social behaviors. In catatonic excitement the individual is extremely hyperactive although the activity seems to have no purpose. Violence toward him/herself or others may also be seen.

NMS is observed as a dangerous side effect associated with certain neuroleptic (antipsychotic) drugs such as haloperidol (Haldol). It comes on suddenly and is characterized by stiffening of the muscles, **fever**, confusion and heavy sweating.

Catatonia can also be categorized as intrinsic or extrinsic. If the condition has an identifiable cause, it is designated as extrinsic. If no cause can be determined following **physical examination**, laboratory testing, and history taking, the illness is considered to be intrinsic.

Causes and symptoms

The causes of catatonia are largely unknown although research indicates that brain structure and function are altered in this condition. While this and other information point to a physical cause, none has yet been proven. A variety of medical conditions also may lead to catatonia including head trauma, cerebrovascular disease, **encephalitis**, and certain metabolic disorders. NMS is an adverse side effect of certain **antipsychotic drugs**.

A variety of symptoms are associated with catatonia. Among the more common are echopraxia (imitation of the gestures of others) and echolalia (parrot-like repetition of words spoken by others). Other signs and symptoms include violence directed toward him/herself, the assumption of inappropriate posture, selective **mutism**, negativism, facial grimaces, and animal-like noises.

Catatonic stupor is marked by immobility and a behavior known as *cerebral flexibilitas* (waxy flexibility) in which

KEY TERMS

Barbiturates—A group of medicines that slow breathing and lower the body temperature and blood pressure. They can be habit forming and are now used chiefly for anesthesia.

Benzodiazepines—This group of medicines is used to help reduce anxiety (especially before surgery) and to help people sleep.

Electroconvulsive therapy—This type of therapy is used to treat major depression and severe mental illness that does not respond to medications. A measured dose of electricity is introduced into the brain in order to produce a convulsion. Electroconvulsive therapy is safe and effective.

Mutism—The inability or refusal to speak.

Negativism—Behavior characterized by resistance, opposition, and refusal to cooperate with requests, even the most reasonable ones.

Neuroleptic drugs—Antipsychotic drugs, including major tranquilizers, used in the treatment of psychoses like schizophrenia.

the individual can be made to assume bizarre (and sometimes painful) postures that they will maintain for extended periods of time. The individual may become dehydrated and malnourished because food and liquids are refused. In extreme situations such individuals must be fed through a tube. Catatonic excitement is characterized by hyperactivity and violence; the individual may harm him/herself or others. On rare occasions, **isolation** or restraint may be needed to ensure the individual's safety and the safety of others.

Diagnosis

Recognition of catatonia is made on the basis of specific movement symptoms. These include odd ways of walking such as walking on tiptoes or ritualistic pacing, and rarely, hopping and skipping. Repetitive odd movements of the fingers or hands, as well as imitating the speech or movements of others also may indicate that catatonia is present. There are no laboratory or other tests that can be used to positively diagnose this condition, but medical and neurological tests are necessary to rule out underlying lesions or disorders that may be causing the symptoms observed.

Treatment

Treatment of catatonia includes medications such as benzodiazepines (which are the preferred treatment) and rarely **barbiturates**. Antipsychotic drugs may be appropri-

ate in some cases, but often cause catatonia to worsen. **Electroconvulsive therapy** may prove beneficial for clients who do not respond to medication. If these approaches are unsuccessful, treatment will be redirected to attempts to control the signs and symptoms of the illness.

Prognosis

Catatonia usually responds quickly to medication interventions.

Prevention

There is currently no known way to prevent catatonia because the cause has not yet been identified. Research efforts continue to explore possible origins. Avoiding excessive use of neuroleptic drugs can help minimize the risk of developing catatonic-like symptoms.

Resources

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Donald G. Barstow, RN

Catecholamines tests

Definition

Catecholamines is a collective term for the hormones epinephrine, norepinephrine, and dopamine. Manufactured chiefly by the chromaffin cells of the adrenal glands, these hormones are involved in readying the body for the “fight-or-flight” response (also known as the alarm reaction). When these hormones are released, the heart beats stronger and faster, blood pressure rises, more blood flows to the brain and muscles, the liver releases stores of energy as a sugar the body can readily use (glucose), the rate of breathing increases and airways widen, and digestive activity slows. These reactions direct more oxygen and fuel to the organs most active in responding to stress—mainly the brain, heart, and skeletal muscles.

Purpose

Pheochromocytoma (a tumor of the chromaffin cells of the adrenal gland) and tumors of the nervous system (neuroblastomas, ganglioneuroblastomas, and ganglioneuromas) that affect hormone production can cause excessive levels of different catecholamines to be secreted.

This results in constant or intermittent high blood pressure (**hypertension**). Episodes of high blood pressure may be accompanied by symptoms such as **headache**, sweating, **palpitations**, and **anxiety**. The catecholamines test can be ordered, then, to determine if high blood pressure and other symptoms are related to improper hormone secretion and to identify the type of tumor causing elevated catecholamine levels.

Description

The catecholamines test can be performed on either blood or urine. If performed on blood, the test may require one or two samples, depending on the physician’s request. The first blood sample will be drawn after the patient has been lying down in a warm, comfortable environment for at least 30 minutes. If a second sample is needed, the patient will be asked to stand for 10 minutes before the blood is drawn. Instead of a venipuncture, which can be stressful for the patient, possibly increasing catecholamine levels in the blood, a plastic or rubber tube-like device called a catheter may be used to collect the blood samples. The catheter would be inserted in a vein 24 hours in advance, eliminating the need for needle punctures at the time of the test.

It may take up to a week for a lab to complete testing of the samples. Because blood levels of catecholamines commonly go up and down in response to such factors as temperature, **stress**, postural change, diet, **smoking**, **obesity**, and many drugs, abnormally high blood test results should be confirmed with a 24-hour urine test. In addition, catecholamine secretion from a tumor may not be steady, but may occur periodically during the day, and potentially could be missed when blood testing is used. The urine test provides the laboratory with a specimen that reflects catecholamine production over an entire 24-hour period. If urine is tested, the patient or a healthcare worker must collect all the urine passed over the 24-hour period.

Preparation

It is important that the patient refrain from using certain medications, especially cold or allergy remedies, for two weeks before the test. Certain foods—including bananas, avocados, cheese, coffee, tea, cocoa, beer, licorice, citrus fruit, vanilla, and Chianti—must be avoided for 48 hours prior to testing. However, people should be sure to get adequate amounts of vitamin C before the test, because this vitamin is necessary for catecholamine formation. The patient should be **fasting** (nothing to eat or drink) for 10 to 24 hours before the blood test and should not smoke for 24 hours beforehand. Some laboratories may call for additional restrictions. As much as possible, the patient should try to avoid excessive physi-

KEY TERMS

Dopamine—Dopamine is a precursor of epinephrine and norepinephrine.

Epinephrine—Epinephrine, also called adrenaline, is a naturally occurring hormone released by the adrenal glands in response to signals from the sympathetic nervous system. These signals are triggered by stress, exercise, or by emotions such as fear.

Ganglioneuroma—A ganglioneuroma is a tumor composed of mature nerve cells.

Neuroblastoma—Neuroblastoma is a tumor of the adrenal glands or sympathetic nervous system. Neuroblastomas can range from being relatively harmless to highly malignant.

Norepinephrine—Norepinephrine is a hormone secreted by certain nerve endings of the sympathetic nervous system, and by the medulla (center) of the adrenal glands. Its primary function is to help maintain a constant blood pressure by stimulating certain blood vessels to constrict when the blood pressure falls below normal.

Pheochromocytoma—A pheochromocytoma is a tumor that originates from the adrenal gland's chromaffin cells, causing overproduction of catecholamines, powerful hormones that induce high blood pressure and other symptoms.

cal **exercise** and emotional stress before the test, because either may alter test results by causing increased secretion of epinephrine and norepinephrine.

Patients collecting their own 24-hour urine samples will be given a container with special instructions. The urine samples must be refrigerated.

Risks

Risks for the blood test are minimal, but may include slight bleeding from the venipuncture site, **fainting** or feeling lightheaded after blood is drawn, or blood accumulating under the puncture site (hematoma). There are no risks for the urine test.

Normal results

Reference ranges are laboratory-specific, vary according to methodology of testing, and differ between blood and urine samples. If testing is done by the method called High Performance Liquid Chromatography (HPLC), typical values for blood and urine follow.

Reference ranges for blood catecholamines

Supine (lying down): Epinephrine less than 50 pg/mL, norepinephrine less than 410 pg/mL, and dopamine less than 90 pg/mL. Standing: Values for blood specimens taken when the subject is standing are higher than the ranges for supine posture for norepinephrine and epinephrine, but not for dopamine.

Reference ranges for urine catecholamines

Epinephrine 0–20 microgram per 24 hours; norepinephrine 15–80 microgram per 24 hours; dopamine 65–400 microgram per 24 hours.

Abnormal results

Depending on the results, high catecholamine levels can indicate different conditions and/or causes:

- High catecholamine levels can help to verify pheochromocytoma, **neuroblastoma**, or ganglioneuroma. An aid to diagnosis is the fact that an adrenal medullary tumor (pheochromocytoma) secretes epinephrine, whereas ganglioneuroma and neuroblastoma secrete norepinephrine.
- Elevations are possible with, but do not directly confirm, thyroid disorders, low blood sugar (**hypoglycemia**), or heart disease.
- Electroshock therapy, or **shock** resulting from hemorrhage or exposure to toxins, can raise catecholamine levels.
- In the patient with normal or low baseline catecholamine levels, failure to show an increase in the sample taken after standing suggests an autonomic nervous system dysfunction (the division of the nervous system responsible for the automatic or unconscious regulation of internal body functioning).

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Janis O. Flores

Catheter ablation

Definition

Catheter ablation of an irregular heartbeat involves having a tube (a catheter) inserted into the heart through



During catheter ablation, a long flexible tube called a catheter is inserted into a vein in the patient's groin and guided toward the heart. A special x-ray machine called a fluoroscope helps the electrophysiologist visualize correct placement. (Photograph by Collette Placek. Reproduced by permission.)

which electrical energy is sent to either reset the heartbeat or stop the heart from beating so a mechanical pacemaker can be put in place.

Purpose

Irregular heartbeats can occur in healthy people without causing any dangerous symptoms or requiring medical attention. Slight changes in the normal patterns of heartbeats often reset themselves without notice.

But when the heartbeat is greatly disrupted—either because of traumatic injury, disease, **hypertension**, surgery, or reduced blood flow to the heart caused by blockages in the blood vessels that nourish the heart—the condition must be recognized and treated immediately. Otherwise, it can be fatal.

Various drugs can be used to control and help reset these abnormal heart rhythms (**arrhythmias**). The tech-

nique of catheter ablation (meaning tube-guided removal) is used to interrupt the abnormal contractions in the heart, allowing normal heart beating to resume. **Atrial fibrillation and flutter** and **Wolff-Parkinson-White syndrome** are two of the most common disorders treated with catheter ablation.

Precautions

The improper correction of abnormal heartbeats can cause additional arrhythmias and can be fatal. Abnormalities in different areas of the heart cause different types of irregular heartbeats; the type of arrhythmia must be clearly defined before this procedure can be properly done.

Description

Catheter ablation involves delivering highly focused heat (or radio frequency energy) to specific areas of the

KEY TERMS

Fluoroscope—A specialized x-ray machine used to visualize the placement of the catheter when attempting to correct irregular heartbeats.

Pacemaker—An electrical device that has electrodes attached to the heart to electrically stimulate the heart to beat normally. Pacemakers can be internal (placed under the skin) or external, with the electrodes placed on the skin or threaded through a tube placed into the heart.

heart. Radio frequency energy is very rapidly alternating electrical current that is produced at the tip of the catheter that is placed inside the heart. At the same time as the catheter is inserted, a second electrode is placed on the patient's skin. When the catheter is energized, the body conducts the energy from the catheter's tip, through the heart and to the electrode on the skin's surface, completing the circuit.

Although very little electricity is given off by the catheter, the instrument does generate a large amount of heat. This heat is absorbed by the heart tissue, causing a small localized burn and destroying the tissue in contact with the catheter tip; in this way, small regions of heart tissue are burned in a controlled manner. This controlled destruction of small sections of heart muscle actually kills the nerve cells causing the irregular heartbeat, stopping the nerve signals that are passing through this section of the heart. This usually causes the irregular heartbeat to be reset into a normal heartbeat.

Preparation

People can undergo this procedure by having general anesthesia or by taking medicines to make them relaxed and sleepy (sedatives) along with painkillers. Once the type of irregular heartbeat is identified and these medicines are given, the catheter is inserted through a blood vessel and into the heart. Importantly, correct placement of the catheter is visualized by using a specialized type of x-ray machine called a fluoroscope.

Aftercare

Being sure the patient is comfortable during and after this procedure is very important. However, because each person may have a different arrhythmia and possibly other medical problems as well, each patient's needs must be evaluated individually.

Risks

Overall, fewer than 5% of people having this procedure experience complications. The most common complications are usually related to blood vessel injury when the catheter is inserted and to different heart-related problems due to the moving of the catheter within the heart. However, in general, this technique is safe and can control many different heart arrhythmias.

Normal results

Depending upon the type of irregular heartbeat being treated, either the normal heartbeat resumes after treatment or the ability of the heart to beat on its own is lost, requiring the insertion of a pacemaker to stimulate the heart to beat regularly.

Abnormal results

Additional irregular heartbeats can occur as a result of this procedure, as can damage to the blood vessels that feed the heart. Because this procedure requires the use of the x-ray machine called a fluoroscope, there is exposure to x-ray radiation, but it's doubtful that this is harmful in adult patients. The risk versus benefit is considered with pediatric patients.

Resources

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American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

Dominic De Bellis, PhD

Cat's cry syndrome see **Cri du chat syndrome**

CBC see **Blood count**

- CEA test see **Carcinoembryonic antigen test**
CEB see **Chronic fatigue syndrome**
Cefaclor see **Cephalosporins**
Cefadroxil see **Cephalosporins**
Cefixime see **Cephalosporins**
Cefprozil see **Cephalosporins**
Cefurox see **Cephalosporins**

patients with celiac disease are considered at higher risk for the disorder.

Because celiac disease has a hereditary influence, close relatives (especially first degree relatives, such as children, siblings, and parents) have a higher risk of being affected with the condition. The chance that a first degree relative of someone with celiac disease will have the disease is about 10%.

As more is learned about celiac disease, it becomes evident that it has many variations which may not produce typical symptoms. It may even be clinically "silent," where no obvious problems related to the disease are apparent.

Celiac disease

Definition

Celiac disease is a disease of the digestive system that damages the small intestine and interferes with the absorption of nutrients from food.

Description

Celiac disease occurs when the body reacts abnormally to gluten, a protein found in wheat, rye, barley, and possibly oats. When someone with celiac disease eats foods containing gluten, that person's immune system causes an inflammatory response in the small intestine, which damages the tissues and results in impaired ability to absorb nutrients from foods. The inflammation and malabsorption create wide-ranging problems in many systems of the body. Since the body's own immune system causes the damage, celiac disease is classified as an "autoimmune" disorder. Celiac disease may also be called sprue, nontropical sprue, gluten sensitive enteropathy, celiac sprue, and adult celiac disease.

Celiac disease may be discovered at any age, from infancy through adulthood. The disorder is more commonly found among white Europeans or in people of European descent. It is very unusual to find celiac disease in African or Asian people. The exact incidence of the disease is uncertain. Estimates vary from one in 5000, to as many as one in every 300 individuals with this background. The prevalence of celiac disease seems to be different from one European country to another, and between Europe and the United States. This may be due to differences in diet and/or unrecognized disease. A recent study of random blood samples tested for celiac disease in the US showed one in 250 testing positive. It is clearly underdiagnosed, probably due to the symptoms being attributed to another problem, or lack of knowledge about celiac disease by physicians and laboratories. Because of the known genetic component, relatives of

Causes and symptoms

Celiac disease can run in families and has a genetic basis, although the pattern of inheritance is complicated. The type of inheritance pattern that celiac disease follows is called multifactorial (caused by many factors, both genetic and environmental). Researchers think that several factors must exist in order for the disease to occur. The patient must have a genetic predisposition to develop the disorder. Then, something in their environment acts as a stimulus, or "trigger," to their immune system, causing the disease to become active for the first time. For conditions with multifactorial inheritance, people without the genetic predisposition are less likely to develop the condition with exposure to the same triggers. Or, they may require more exposure to the stimulus before developing the disease than someone with a genetic predisposition. Some of the things which may provoke a reaction include surgery, especially gastrointestinal surgery; a change to a low fat diet, which has an increased number of wheat-based foods; **pregnancy**; **childbirth**; severe emotional **stress**; or a viral infection. This combination of genetic susceptibility and an outside agent leads to celiac disease.

Each person with celiac disease is affected differently. When food containing gluten reaches the small intestine, the immune system begins to attack a substance called gliadin, which is found in the gluten. The resulting inflammation causes damage to the delicate finger-like structures in the intestine, called villi, where food absorption actually takes place. The patient may experience a number of symptoms related to the inflammation and the chemicals it releases, and/or the lack of ability to absorb nutrients from food, which can cause **malnutrition**.

The most commonly recognized symptoms of celiac disease relate to the improper absorption of food in the gastrointestinal system. Many patients with gastrointestinal symptoms will have **diarrhea** and fatty, greasy,

unusually foul-smelling stools. The patient may complain of excessive gas (flatulence), distended abdomen, weight loss, and generalized weakness. Not all people have digestive system complications; some people only have irritability or depression. Irritability is one of the most common symptoms in children with celiac disease.

Not all patients have these problems. Unrecognized and therefore untreated celiac disease may cause or contribute to a variety of other conditions. The decreased ability to digest, absorb, and utilize food properly (malabsorption) may cause anemia (low red **blood count**) from iron deficiency or easy bruising from a lack of vitamin K. Poor mineral absorption may result in **osteoporosis**, or “brittle bones,” which may lead to bone **fractures**. Vitamin D levels may be insufficient and bring about a “softening” of bones (osteomalacia), which produces **pain** and bony deformities, such as flattening or bending. Defects in the tooth enamel, characteristic of celiac disease, may be recognized by dentists. Celiac disease may be discovered during medical tests performed to investigate **failure to thrive** in infants, or lack of proper growth in children and adolescents. People with celiac disease may also experience **lactose intolerance** because they don’t produce enough of the enzyme lactase, which breaks down the sugar in milk into a form the body can absorb. Other symptoms can include muscle cramps, **fatigue**, delayed growth, tingling or numbness in the legs (from nerve damage), pale sores in the mouth (called aphthus ulcers), tooth discoloration, or missed menstrual periods (due to severe weight loss).

A distinctive, painful skin rash, called **dermatitis herpetiformis**, may be the first sign of celiac disease. Approximately 10% of patients with celiac disease have this rash, but it is estimated that 85% or more of patients with the rash have the disease.

Many disorders are associated with celiac disease, though the nature of the connection is unclear. One type of epilepsy is linked to celiac disease. Once their celiac disease is successfully treated, a significant number of these patients have fewer or no seizures. Patients with **alopecia areata**, a condition where hair loss occurs in sharply defined areas, have been shown to have a higher risk of celiac disease than the general population. There appears to be a higher percentage of celiac disease among people with **Down syndrome**, but the link between the conditions is unknown.

Several conditions attributed to a disorder of the immune system have been associated with celiac disease. People with insulin dependent diabetes (type I) have a much higher incidence of celiac disease. One source estimates that as many as one in 20 insulin-dependent diabetics may have celiac disease. Patients with other conditions where celiac disease may be more commonly found

include those with juvenile chronic arthritis, some thyroid diseases, and IgA deficiency.

There is an increased risk of intestinal lymphoma, a type of **cancer**, in individuals with celiac disease. Successful treatment of the celiac disease seems to decrease the chance of developing lymphoma.

Diagnosis

Because of the variety of ways celiac disease can manifest itself, it is often not discovered promptly. Its symptoms are similar to many other conditions including irritable bowel syndrome, **Crohn’s disease**, **ulcerative colitis**, diverticulosis, intestinal infections, **chronic fatigue syndrome**, and depression. The condition may persist without diagnosis for so long that the patient accepts a general feeling of illness as normal. This leads to further delay in identifying and treating the disorder. It is not unusual for the disease to be identified in the course of medical investigations for seemingly unrelated problems. For example, celiac disease has been discovered during testing to find the cause of **infertility**.

If celiac disease is suspected, a blood test can be ordered. This test looks for the antibodies to gluten (called antigliadin, anti-endomysium, and antireticulin) that the immune system produces in celiac disease. Antibodies are chemicals produced by the immune system in response to substances that the body perceives to be threatening. Some experts advocate not just evaluating patients with symptoms, but using these blood studies as a screening test for high-risk individuals, such as those with relatives (especially first degree relatives) known to have the disorder. An abnormal result points towards celiac disease, but further tests are needed to confirm the diagnosis. Because celiac disease affects the ability of the body to absorb nutrients from food, several tests may be ordered to look for nutritional deficiencies. For example, doctors may order a test of iron levels in the blood because low levels of iron (anemia) may accompany celiac disease. Doctors may also order a test for fat in the stool, since celiac disease prevents the body from absorbing fat from food.

If these tests above are suspicious for celiac disease, the next step is a biopsy (removal of a tiny piece of tissue surgically) of the small intestine. This is usually done by a gastroenterologist, a physician who specializes in diagnosing and treating bowel disorders. It is generally performed in the office, or in a hospital’s outpatient department. The patient remains awake, but is sedated. A narrow tube, called an endoscope, is passed through the mouth, down through the stomach, and into the small intestine. A small sample of tissue is taken and sent to the laboratory for analysis. If it shows a pattern of tissue damage characteristic of celiac disease, the diagnosis is established.

The patient is then placed on a gluten-free diet (GFD). The physician will periodically recheck the level of antibody in the patient's blood. After several months, the small intestine is biopsied again. If the diagnosis of celiac disease was correct (and the patient followed the rigorous diet), healing of the intestine will be apparent. Most experts agree that it is necessary to follow these steps in order to be sure of an accurate diagnosis.

Treatment

The only treatment for celiac disease is a gluten-free diet. This may be easy for the doctor to prescribe, but difficult for the patient to follow. For most people, adhering to this diet will stop symptoms and prevent damage to the intestines. Damaged villi can be functional again in three to six months. This diet must be followed for life. For people whose symptoms are cured by the gluten-free diet, this is further evidence that their diagnosis is correct.

Gluten is present in any product that contains wheat, rye, barley, or oats. It helps make bread rise, and gives many foods a smooth, pleasing texture. In addition to the many obvious places gluten can be found in a normal diet, such as breads, cereals, and pasta, there are many hidden sources of gluten. These include ingredients added to foods to improve texture or enhance flavor and products used in food packaging. Gluten may even be present on surfaces used for food preparation or cooking.

Fresh foods that have not been artificially processed, such as fruits, vegetables, and meats, are permitted as part of a GFD. Gluten-free foods can be found in health food stores and in some supermarkets. Mail-order food companies often have a selection of gluten-free products. Help in dietary planning is available from dieticians (healthcare professionals specializing in food and **nutrition**) or from support groups for individuals with celiac disease. There are many cookbooks on the market specifically for those on a GFD.

Treating celiac disease with a GFD is almost always completely effective. Gastrointestinal complaints and other symptoms are alleviated. Secondary complications, such as anemia and osteoporosis, resolve in almost all patients. People who have experienced lactose intolerance related to their celiac disease usually see those symptoms subside, as well. Although there is no risk and much potential benefit to this treatment, it is clear that avoiding all foods containing gluten can be difficult.

Experts emphasize the need for lifelong adherence to the GFD to avoid the long-term complications of this disorder. They point out that although the disease may have symptom-free periods if the diet is not followed, silent damage continues to occur. Celiac disease cannot be "outgrown" or cured, according to medical authorities.

KEY TERMS

Antibodies—Proteins that provoke the immune system to attack particular substances. In celiac disease, the immune system makes antibodies to a component of gluten.

Gluten—A protein found in wheat, rye, barley, and oats.

Villi—Tiny, finger-like projections that enable the small intestine to absorb nutrients from food.

Prognosis

Patients with celiac disease must adhere to a strict GFD throughout their lifetime. Once the diet has been followed for several years, individuals with celiac disease have similar mortality rates as the general population. However, about 10% of people with celiac disease develop a cancer involving the gastrointestinal tract (both carcinoma and lymphoma).

There are a small number of patients who develop a refractory type of celiac disease, where the GFD no longer seems effective. Once the diet has been thoroughly assessed to ensure no hidden sources of gluten are causing the problem, medications may be prescribed. Steroids or **immunosuppressant drugs** are often used to try to control the disease. It is unclear whether these efforts meet with much success.

Prevention

There is no way to prevent celiac disease. However, the key to decreasing its impact on overall health is early diagnosis and strict adherence to the prescribed gluten-free diet.

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ORGANIZATIONS

- American Celiac Society. 58 Musano Court, West Orange, NJ, 7052. (201) 325-8837.
- Celiac Disease Foundation. 13251 Ventura Blvd., Suite 1, Studio City, CA 91604-1838. (818) 990-2354.
- Celiac Sprue Association/United States of America (CSA/USA). PO Box 31700, Omaha, NE 68131-0700. (402) 558-0600.
- Gluten Intolerance Group. PO Box 23053, Seattle, WA, 98102-0353. (206) 325-6980.
- National Center for Nutrition and Dietetics. American Dietetic Association, 216 West Jackson Boulevard, Suite 800, Chicago, IL, 60606-6995. (800) 366-1655.

OTHER

- National Institute of Diabetes & Digestive & Kidney Diseases. <<http://www.niddk.nih.gov/health/digest/pubs/celiac/index.htm>>.

Amy Vance, MS, CGC

Cellulitis

Definition

Cellulitis is a spreading bacterial infection just below the skin surface. It is most commonly caused by *Streptococcus pyogenes* or *Staphylococcus aureus*.

Description

The word “cellulitis” actually means “inflammation of the cells.” Specifically, cellulitis refers to an infection of the tissue just below the skin surface. In humans, the skin and the tissues under the skin are the most common locations for microbial infection. Skin is the first defense against invading bacteria and other microbes. An infection can occur when this normally strong barrier is damaged due to surgery, injury, or a burn. Even something as small as a scratch or an insect bite allows bacteria to enter the skin, which may lead to an infection. Usually, the immune system kills any invading bacteria, but sometimes the bacteria are able to grow and cause an infection.

Once past the skin surface, the warmth, moisture, and nutrients allow bacteria to grow rapidly. Disease-causing bacteria release proteins called enzymes which cause tissue damage. The body’s reaction to damage is inflammation which is characterized by **pain**, redness, heat, and swelling. This red, painful region grows bigger as the infection and resulting tissue damage spread. An untreated infection may spread to the lymphatic system (**acute lymphangitis**), the lymph nodes (**lymphadenitis**), the bloodstream (**bacteremia**), or into deeper tissues. Cellulitis most often occurs on the face, neck, and legs.

Orbital cellulitis

A very serious infection, called orbital cellulitis, occurs when bacteria enter and infect the tissues surrounding the eye. In 50–70% of all cases of orbital cellulitis, the infection spreads to the eye(s) from the sinuses or the upper respiratory tract (nose and throat). Twenty-five percent of orbital infections occur after surgery on the face. Other sources of orbital infection include a direct infection from an eye injury, from a dental or throat infection, and through the bloodstream.

Infection of the tissues surrounding the eye causes redness, swollen eyelids, severe pain, and causes the eye to bulge out. This serious infection can lead to a temporary loss of vision, blindness, brain abscesses, inflammation of the brain and spinal tissues (**meningitis**), and other complications. Before the discovery of **antibiotics**, orbital cellulitis caused blindness in 20% of patients and **death** in 17% of patients. Antibiotic treatment has significantly reduced the incidence of blindness and death.

Causes and symptoms

Although other kinds of bacteria can cause cellulitis, it is most often caused by *Streptococcus pyogenes* (the bacteria which causes **strep throat**) and *Staphylococcus aureus*. *Streptococcus pyogenes* is the so-called “flesh-eating bacteria” and, in rare cases, can cause a dangerous, deep skin infection called necrotizing fasciitis. Orbital cellulitis may be caused by bacteria which cannot grow in the presence of oxygen (anaerobic bacteria). In children, *Haemophilus influenzae* type B frequently causes orbital cellulitis following a sinus infection.

Streptococcus pyogenes can be picked up from a person who has strep throat or an infected sore. Other cellulitis-causing bacteria can be acquired from direct contact with infected sores. Persons who are at a higher risk for cellulitis are those who have a severe underlying disease (such as **cancer**, diabetes, and kidney disease), are taking steroid medications, have a reduced immune system (because of **AIDS**, organ transplant, etc.), have been burned, have insect bites, have reduced blood circulation to limbs, or have had a leg vein removed for coronary bypass surgery. In addition, chicken pox, human or animal bite **wounds**, skin wounds, and recent surgery can put a person at a higher risk for cellulitis.

The characteristic symptoms of cellulitis are redness, warmth, pain, and swelling. The infected area appears as a red patch that gets larger rapidly within the first 24 hours. A thick red line which progresses towards the heart may appear indicating an infection of the lymph vessels (lymphangitis). Other symptoms which may occur include **fever**, chills, tiredness, muscle aches, and a

general ill feeling. Some people also experience nausea, vomiting, stiff joints, and hair loss at the infection site.

The characteristic symptoms of orbital cellulitis are eye pain, redness, swelling, warmth, and tenderness. The eye may bulge out and it may be difficult or impossible to move. Temporary loss of vision, pus drainage from the eye, chills, fever, headaches, vomiting, and a general ill feeling may occur.

Diagnosis

Cellulitis may be diagnosed and treated by a family doctor, an infectious disease specialist, a doctor who specializes in skin diseases (dermatologist), or in the case of orbital cellulitis, an eye doctor (ophthalmologist). The diagnosis of cellulitis is based mainly on the patient's symptoms. The patient's recent medical history is also used in the diagnosis.

Laboratory tests may be done to determine which kind of bacteria is causing the infection but these tests are not always successful. If the skin injury is visible, a sterile cotton swab is used to pick up a sample from the wound. If there is no obvious skin injury, a needle may be used to inject a small amount of sterile salt solution into the infected skin, and then the solution is withdrawn. The salt solution should pick up some of the bacteria causing the infection. A blood sample may be taken from the patient's arm to see if bacteria have entered the bloodstream. Also, a blood test may be done to count the number of white blood cells in the blood. High numbers of white blood cells suggest that the body is trying to fight a bacterial infection.

For orbital cellulitis, the doctor may often perform a special x-ray scan called **computed tomography scan** (CT). This scan enables the doctor to see the patient's head in cross-section to determine exactly where the infection is and see if any damage has occurred. A CT scan takes about 20 minutes.

Treatment

Antibiotic treatment is the only way to battle this potentially life-threatening infection. Mild to moderate cellulitis can be treated with the following antibiotics taken every four to eight hours by mouth:

- penicillins (Bicillin, Wycillin, Pen Vee, V-Cillin)
- erythromycin (E-Mycin, Ery-Tab)
- cephalexin (Biocef, Keflex)
- cloxacillin (Tegopen)

Other medications may be recommended, such as **acetaminophen** (Tylenol) or ibuprofen (Motrin, Advil) to relieve pain, and **aspirin** to decrease fever.



This person's lower leg is swollen and inflamed due to cellulitis. Cellulitis is a *Streptococcus* bacterial infection of the skin and the tissues beneath it. The face, neck, or legs are common sites of cellulitis. (Custom Medical Stock Photo. Reproduced by permission.)

A normally healthy person is usually not hospitalized for mild or moderate cellulitis. General treatment measures include elevation of the infected area, rest, and application of warm, moist compresses to the infected area. The doctor will want to see the patient again to make sure that the antibiotic treatment is effective in stopping the infection.

Persons at high risk for severe cellulitis will probably be hospitalized for treatment and monitoring. Antibiotics may be given intravenously to patients with severe cellulitis. Complications such as deep infection, or bone or joint infections, might require surgical drainage and a longer course of antibiotic treatment. Extensive tissue destruction may require plastic surgery to repair. In cases of orbital cellulitis caused by a sinus infection, surgery may be required to drain the sinuses.

Prognosis

Over 90% of all cellulitis cases are cured after seven to 10 days of antibiotic treatment. Persons with serious disease and/or those who are taking immunosuppressive drugs may experience a more severe form of cellulitis which can be life threatening. Serious complications include blood **poisoning** (bacteria growing in the blood stream), meningitis (brain and spinal cord infection), tissue death (necrosis), and/or lymphangitis (infection of the lymph vessels). Severe cellulitis caused by *Streptococcus pyogenes* can lead to destructive and life-threatening necrotizing fasciitis.

Prevention

Cellulitis may be prevented by wearing appropriate protective equipment during work and sports to avoid

KEY TERMS

Inflammation—A local, protective response to tissue injury. It is characterized by redness, warmth, swelling, and pain.

Necrotizing fasciitis—A destructive infection which follows severe cellulitis and involves the deep skin and underlying tissues.

Sinuses—Air cavities found in the bones of the head. The sinuses which are connected to the nose are prone to infection.

skin injury, cleaning cuts and skin injuries with antiseptic soap, keeping wounds clean and protected, watching wounds for signs of infection, taking the entire prescribed dose of antibiotic, and maintaining good general health. Persons with diabetes should try to maintain good blood sugar control.

Resources

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Belinda Rowland, PhD

Cell therapy

Definition

Cell therapy is the transplantation of human or animal cells to replace or repair damaged tissue and/or cells.

Purpose

Cell therapy has been used successfully to rebuild damaged cartilage in joints, repair spinal cord injuries, strengthen a weakened immune system, treat autoimmune diseases such as AIDS, and help patients with neu-

rological disorders such as Alzheimer's disease, Parkinson's disease, and epilepsy. Further uses have shown positive results in the treatment of a wide range of chronic conditions such as arteriosclerosis, congenital defects, and sexual dysfunction. The therapy has also been used to treat cancer patients at a number of clinics in Tijuana, Mexico, although this application has not been well supported with controlled clinical studies.

Description

Origins

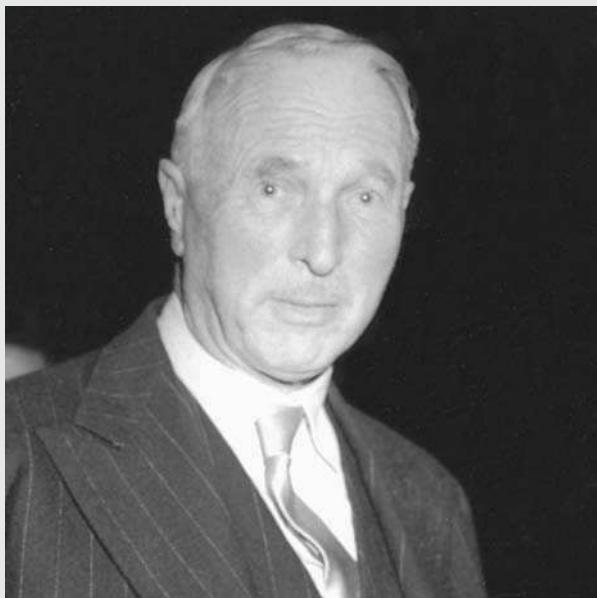
The theory behind cell therapy has been in existence for several hundred years. The first recorded discussion of the concept of cell therapy can be traced to Phillipus Aureolus Paracelsus (1493–1541), a German-Swiss physician and alchemist who wrote in his *Der grossen Wundartzney* ("Great Surgery Book") in 1536 that "the heart heals the heart, lung heals the lung, spleen heals the spleen; like cures like." Paracelsus and many of his contemporaries agreed that the best way to treat an illness was to use living tissue to restore the ailing. In 1667, at a laboratory in the palace of Louis XIV, Jean-Baptiste Denis (1640–1704) attempted to transfuse blood from a calf into a mentally ill patient—and since blood **transfusion** is, in effect, a form of cell therapy, this could be the first documented case of this procedure. However, the first recorded attempt at non-blood cellular therapy occurred in 1912 when German physicians attempted to treat children with **hypothyroidism**, or an underactive thyroid, with thyroid cells.

In 1931, Dr. Paul Niehans (1882–1971), a Swiss physician, became known as "the father of cell therapy" quite by chance. After a surgical accident by a colleague, Niehans attempted to transplant a patient's severely damaged parathyroid glands with those of a steer. When the patient began to rapidly deteriorate before the transplant could take place, Niehans decided to dice the steer's parathyroid gland into fine pieces, mix the pieces in a saline solution, and inject them into the dying patient. Immediately, the patient began to improve and, in fact, lived for another 30 years.

Cell therapy is, in effect, a type of organ transplant which has also been referred to as "live cell therapy," "xenotransplant therapy," "cellular suspensions," "glandular therapy," or "fresh cell therapy." The procedure involves the injection of either whole fetal xenogenic (animal) cells (e.g., from sheep, cows, pigs, and sharks) or cell extracts from human tissue. The latter is known as autologous cell therapy if the cells are extracted from and transplanted back into the same patient. Several different types of cells can be administered simultaneously.

Just as Paracelsus' theory of "like cures like," the types of cells that are administered correspond in some

PAUL NIEHANS (1882–1971)



(AP/Wide World Photos. Reproduced by permission.)

Paul Niehans was born and raised in Switzerland. His father, a doctor, was dismayed when he entered the seminary, but Niehans quickly grew dissatisfied with religious life and took up medicine after all. He first studied at Bern, then completed an internship in Zurich.

way with the organ or tissue in the patient that is failing. No one knows exactly how cell therapy works, but proponents claim that the injected cells travel to the similar organ from which they were taken to revitalize and stimulate that organ's function and regenerate its cellular structure. In other words, the cells are not species specific, but only organ specific. Supporters of cellular treatment believe that embryonic and fetal animal tissue contain active therapeutic agents distinct from **vitamins, minerals**, hormones, or enzymes.

Swedish researchers have successfully transplanted human fetal stem cells into human recipients, and the procedure is being investigated further as a possible treatment for repairing brain cells in Parkinson's patients. However, because the cells used in these applications must be harvested from aborted human fetuses, there is an ethical debate over their use.

Currently, applications of cell therapy in the United States is still in the research, experimental, and clinical trial stages. The U.S. Food and Drug Administration

Niehans enlisted in the Swiss Army in 1912. When war erupted in the Balkans, Niehans set up a hospital in Belgrade, Yugoslavia. The war provided him the opportunity to treat numerous patients, gaining a firsthand knowledge of the body and its workings.

Since 1913, Niehans had been intrigued with Alexis Carrel's experiments concerning the adaptive abilities of cells, though Niehans himself specialized in glandular transplants and by 1925 was one of the leading glandular surgeons in Europe.

Niehans referred to 1931 as the birth year of cellular therapy. That year, he treated a patient suffering from tetany whose parathyroid had been erroneously removed by another physician. Too weak for a glandular transplant, the patient was given injections of the parathyroid glands of an ox, and she soon recovered. Niehans made more injections, even experimenting on himself, and reported he could cure illnesses through injections of live cells extracted from healthy animal organs. He believed adding new tissue stimulated rejuvenation and recovery.

Niehans treated Pope Pius XII with his injections and was nominated to the Vatican Academy of Science following the pope's recovery.

Niehans remained a controversial figure throughout his life. As of 2000, the Clinique Paul Niehans in Switzerland, founded by his daughter, continues his work.

has approved the use of one cellular therapy technique for repairing damaged knee joints. The procedure involves removing healthy chondrocyte cells, the type of cell that forms cartilage, from the patient, culturing them in a laboratory for three to four weeks, and then transplanting them back into the damaged knee joint of the patient.

Preparations

There are several processes to prepare cells for use. One form involves extracting cells from the patient they are to be used on and then culturing them in a laboratory setting until they multiply to the level needed for transplant back into the patient. Another procedure uses freshly removed fetal animal tissue, which has been processed and suspended in a saline solution. The preparation of fresh cells then may be either injected immediately into the patient, or preserved by being freeze-dried or deep-frozen in liquid nitrogen before being injected. Cells may be tested for pathogens, such as bacteria, viruses, or parasites, before use.

KEY TERMS

Anaphylactic shock—A severe allergic reaction that causes blood pressure drop, racing heart, swelling of the airway, rash, and possibly convulsions.

Culturing—To grow cells in a special substance, or media, in the laboratory.

Encephalitis—Inflammation of the brain.

Precautions

Patients undergoing cell therapy treatments which use cells transplanted from animals or other humans run the risk of cell rejection, in which the body recognizes the cells as a foreign substance and uses the immune system's T-cells to attack and destroy them. Some forms of cell therapy use special coatings on the cells designed to trick the immune system into recognizing the new cells as native to the body.

There is also the chance of the cell solution transmitting bacterial or viral infection or other disease and parasites to the patient. Careful screening and testing of cells for pathogens can reduce this risk.

Many forms of cell therapy in the United States are still largely experimental procedures. Patients should approach these treatments with extreme caution, should inquire about their proven efficacy and legal use in the United States, and should only accept treatment from a licensed physician who should educate the patient completely on the risks and possible side effects involved with cell therapy. These same cautions apply for patients interested in participating in clinical trials of cell therapy treatments.

Side effects

Because cell therapy encompasses such a wide range of treatments and applications, and many of these treatments are still experimental, the full range of possible side effects of the treatments are not yet known. Anaphylactic **shock** (severe allergic reaction), immune system reactions, and **encephalitis** (inflammation of the brain) are just a few of the known reported side effects in some patients to date.

Side effects of the FDA-approved chondrocyte cell therapy used in knee joint repair may include tissue hypertrophy, a condition where too much cartilage grows in the joint where the cells were transplanted to and the knee joint begins to stiffen.

Research and general acceptance

There is a growing debate in the medical community over the efficacy and ethical implications of cell therapy. Much of the ethical debate revolves around the use of human fetal stem cells in treatment, and the fact that these cells must be harvested from aborted fetuses.

While some cell therapy procedures have had proven success in clinical studies, others are still largely unproven, including cell therapy for cancer treatment. Until more large, controlled clinical studies are performed on these procedures to either prove or disprove their efficacy, they will remain fringe treatments.

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ORGANIZATIONS

Center for Cell and Gene Therapy. Baylor College of Medicine. 1102 Bates St, Suite 1100, Houston, Texas 77030-2399. (713) 770-4663. <<http://wwwbcm.tmc.edu/genetherapy>>.

Paula Ford-Martin

Central Mississippi Valley disease see
Histoplasmosis

Central nervous system depressants

Definition

Central nervous system (CNS) depressants are drugs that can be used to slow down brain activity.

Purpose

CNS depressants may be prescribed by a physician to treat anxiety, muscle tension, pain, insomnia, acute stress reactions, panic attacks, and seizure disorders. In higher doses, some CNS depressants may be used as general anesthetics.

Description

Throughout history, humans have sought relief from anxiety and insomnia by using substances that depress brain activity and induce a drowsy or calming effect. CNS depressants include a wide range of drugs such as alcohol, narcotics, barbiturates (Amytal, Nembutal, Seconal), benzodiazepines (Ativan, Halcion, Librium, Valium, Xanax), chloral hydrate, and methaqualone (Quaaludes), as well as newer CNS depressants developed in the 1990s, such as Buspirone (Buspar) and Zolpidem (Ambien), which are thought to have the fewest sideeffects. Most CNS depressants activate a neurotransmitter called gamma-aminobutyric acid (GABA), which helps decrease brain activity. Street names for CNS depressants include Reds, Yellows, Blues, Ludes, Barbs, and Downers.

Precautions

Most CNS depressants have the potential to be physically and psychologically addictive. Alcohol is the most widely abused depressant. The body tends to develop tolerance for CNS depressants, and larger doses are needed to achieve the same effects. Withdrawal from some CNS depressants can be uncomfortable; for example, withdrawal from a depressant treating insomnia or anxiety can cause rebound insomnia or anxiety as the brain's activity bounces back after being suppressed. In some cases withdrawal can result in life-threatening seizures. Generally, depressant withdrawal should be undertaken under a physician's supervision. Many physicians will reduce the depressant dosage gradually, to give the body time to adjust. Certain CNS depressants such as barbiturates are easy to overdose on, since there is a relatively small difference between the optimal dose and an overdose. A small miscalculation can lead to coma, slowed breathing, and death. CNS depressants should be administered to elderly individuals with care, as these individuals have a reduced ability to metabolize CNS depressants.

Side Effects

Especially when taken in excess, CNS depressants can cause confusion and dizziness, and impair judgment, memory, intellectual performance, and motor coordination.

Interactions

CNS depressants should be used with other medications, such as antidepressant medications, only under a physician's supervision. Certain herbal remedies, such as Valerian and Kava, may dangerously exacerbate the effects of certain CNS depressants. Also, ingesting a combination of CNS depressants, such Valium and alcohol, for example, is not advised. When mixed together, CNS

KEY TERMS

GABA (gamma-aminobutyric acid)—A neurotransmitter that slows down the activity of nerve cells in the brain.

Neurotransmitter—A chemical compound in the brain that carries signals from one nerve cell to another.

depressants tend to amplify each other's effects, which can cause severely reduced heart rate and even death.

Resources

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American Society of Addiction Medicine. 4601 North Park Avenue, Arcade Suite 101, Chevy Chase, MD 20815. (301) 656-3920. <<http://www.asam.org>>.

National Clearinghouse for Alcohol and Drug Abuse Information (NCADI) Center for Substance Abuse Prevention. 5600 Fishers Lane, Rockville, MD 20857. (301) 443-0365. <<http://www.health.org>>.

National Institute on Drug Abuse. 6001 Executive Blvd, Bethesda, MD 20892. (301) 443-1124 <<http://www.nida.nih.gov>>.

Ann Quigley

Central nervous system infections

Definition

The central nervous system, or CNS, comprises the brain, the spinal cord, and associated membranes. Under some circumstances, bacteria may enter areas of the CNS. If this occurs, abscesses or empyemas may be established.

Description

In general, the CNS is well defended against infection. The spine and brain are sheathed in tough, protective membranes. The outermost membrane, the dura mater, and the next layer, the arachnoid, entirely encase the brain and spinal cord. However, these defenses are not absolute. In rare cases, bacteria gain access to areas within the CNS.

KEY TERMS

Abscess—A pus-filled area with definite borders.

Arachnoid—One of the membranes that sheathes the spinal cord and brain; the arachnoid is the second-layer membrane.

Cerebrospinal fluid—Fluid that is normally found in the spinal cord and brain. Abnormal levels of certain molecules in this fluid can indicate the presence of infection or damage to the central nervous system.

CT scan (computed tomography)—Cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures.

Dura mater—One of the membranes that sheathes the spinal cord and brain; the dura mater is the outermost layer.

Empyema—A pus-filled area with indefinite borders.

Lumbar puncture—A procedure in which a needle is inserted into the lower spine to collect a sample of cerebrospinal fluid.

MRI (magnetic resonance imaging)—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.

Bacterial infection of the CNS can result in abscesses and empyemas (accumulations of pus). Abscesses have fixed boundaries, but empyemas lack definable shape and size. CNS infections are classified according to the location where they occur. For example, a spinal epidural **abscess** is located above the dura mater, and a cranial subdural **empyema** occurs between the dura mater and the arachnoid.

As pus and other material from an infection accumulate, pressure is exerted on the brain or spinal cord. This pressure can damage the nervous system tissue, possibly permanently. Without treatment, a CNS infection is fatal.

Causes and symptoms

Typically, bacterial invasion results from the spread of a nearby infection; for example, a chronic sinus or middle ear infection can extend beyond its initial site. Bacteria may also be conveyed to the CNS from distant sites of infection by the bloodstream. In rare cases, head trauma or surgical procedures may introduce bacteria

directly into the CNS. However, the source of infection cannot always be identified.

Specific symptoms of a CNS infection hinge on its exact location, but may include severe **headache** or back **pain**, weakness, sensory loss, and a **fever**. An individual may report a stiff neck, nausea or vomiting, and tiredness or disorientation. There is a potential for seizures, **paralysis**, or **coma**.

Diagnosis

Physical symptoms, such as a fever and intense backache or a fever, severe headache, and stiff neck, raise the suspicion of a CNS infection. Blood tests may indicate the presence of an infection but do not pinpoint its location. CT scans or MRI scans of the brain and spine can provide definitive diagnosis, with an MRI scan being the most sensitive. A lumbar puncture and analysis of the cerebrospinal fluid can help diagnose an epidural abscess; however, the procedure can be dangerous in cases of subdural empyema.

Treatment

A two-pronged approach is taken to treat CNS infections. First, antibiotic therapy against an array of potential infectious bacteria is begun. The second stage involves surgery to drain the infected site. Although some CNS infections have been resolved with **antibiotics** alone, the more aggressive approach is often preferred. Surgery allows immediate relief of pressure on the brain or spinal cord, as well as an opportunity to collect infectious material for bacterial identification. Once the bacterial species is identified, drug therapy can be altered to a more specific antibiotic. However, surgery may not be an option in some cases, such as when there are numerous sites of infection or when infection is located in an inaccessible area of the brain.

Prognosis

The fatality rate associated with CNS infections ranges from 10% to as high as 40%. Some survivors experience permanent CNS damage, resulting in partial paralysis, speech problems, or seizures. Rapid diagnosis and treatment are essential for a good prognosis. With prompt medical attention, an individual may recover completely.

Prevention

Treatment for pre-existing infections, such as sinus or middle ear infections, may prevent some cases of CNS infection. However, since some CNS infections are of unknown origin, not all are preventable.

Resources

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Julia Barrett

small number of additional members of the CNS stimulant class do not fall into specific chemical groups.

Precautions

Amphetamines have a high potential for abuse. They should be used in weight reduction programs only when alternative therapies have been ineffective. Administration for prolonged periods may lead to drug dependence. These drugs are classified as schedule II under federal drug control regulations.

The amphetamines and their cogeners are contraindicated in advanced arteriosclerosis, symptomatic cardiovascular disease, and moderate to severe **hypertension** and **hyperthyroidism**. They should not be used to treat patients with hypersensitivity or idiosyncrasy to the sympathomimetic amines, or with **glaucoma**, a history of agitated states, a history of drug abuse, or during the 14 days following administration of monoamine oxidase (MAO) inhibitors.

Methylphenidate may lower the seizure threshold.

Benzphetamine is category X during **pregnancy**. Diethylpropion is category B. Other anorexiants have not been rated; however their use during pregnancy does not appear to be advisable. Safety for use of anorexiants has not been evaluated.

Amphetamines are all category C during pregnancy. Breastfeeding while receiving amphetamines is not recommended because the infant may experience withdrawal symptoms.

There have been reports that when used in children, methylphenidate and amphetamines may retard growth. Although these reports have been questioned, it may be suggested that the drugs not be administered outside of school hours (because most children have behavior problems in school), in order to permit full stature to be attained.

The most common adverse effects of CNS stimulants are associated with their primary action. Typical responses include overstimulation, **dizziness**, restlessness, and similar reactions. Rarely, hematologic reactions, including leukopenia, agranulocytosis, and bone marrow depression have been reported. Lowering of the seizure threshold has been noted with most drugs in this class.

Withdrawal syndrome

Abrupt discontinuation following prolonged high dosage results in extreme **fatigue**, mental depression and changes on the sleep EEG. This response is most evident with amphetamines, but may be observed with all CNS stimulants taken over a prolonged period of time.

Description

The majority of CNS stimulants are chemically similar to the neurohormone norepinephrine, and simulate the traditional "fight or flight" syndrome associated with sympathetic nervous system arousal. **Caffeine** is more closely related to the xanthines, such as theophylline. A

KEY TERMS

Agranulocytosis—An acute febrile condition marked by severe depression of the granulocyte-producing bone marrow, and by prostration, chills, swollen neck, and sore throat sometimes with local ulceration.

Anorexiant—A drug that suppresses appetite.

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.

Attention-deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

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.

Leucopenia—A condition in which the number of leukocytes circulating in the blood is abnormally low and which is most commonly due to a decreased production of new cells in conjunction with various infectious diseases or as a reaction to various drugs or other chemicals.

Pregnancy category—A system of classifying drugs according to their established risks for use during pregnancy. Category A: Controlled human studies have demonstrated no fetal risk. Category B: Animal studies indicate no fetal risk, but no human studies, or adverse effects in animals, but not in well-controlled human studies. Category C: No adequate human or animal studies, or adverse fetal effects in animal studies, but no available human data. Category D: Evidence of fetal risk, but benefits outweigh risks. Category X: Evidence of fetal risk. Risks outweigh any benefits.

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.

Resources

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ORGANIZATION

Children and Adults with Attention Deficit Disorders
(CH.A.D.D.). 499 N.W. 70th Avenue, Suite 109, Plantation, FL 33317. (305) 587-3700.

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Nancy Ross-Flanigan

Central retinal artery occlusion see
Retinopathies

Central retinal vein occlusion see
Retinopathies

Cephalosporins

Definition

Cephalosporins are medicines that kill bacteria or prevent their growth.

Purpose

Cephalosporins are used to treat infections in different parts of the body—the ears, nose, throat, lungs, sinuses, and skin, for example. Physicians may prescribe these drugs to treat **pneumonia**, **strep throat**, staph infections, **tonsillitis**, **bronchitis**, and **gonorrhea**. These drugs will *not* work for colds, flu, and other infections caused by viruses.

Description

Examples of cephalosporins are cefaclor (Ceclor), cefadroxil (Duricef), cefazolin (Ancef, Kefzol, Zolicef),

cefixime (Suprax), cefoxitin (Mefoxin), cefprozil (Cefzil), ceftazidime (Ceptaz, Fortaz, Tazicef, Tazideme), cefuroxime (Ceftin) and cephalexin (Keflex). These medicines are available only with a physician's prescription. They are sold in tablet, capsule, liquid, and injectable forms.

Recommended dosage

The recommended dosage depends on the type of cephalosporin. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage.

Always take cephalosporins exactly as directed by your physician. Never take larger, smaller, more frequent, or less frequent doses. Take the drug for exactly as long as directed—no more and no less. Do not save some doses of the drug to take for future infections. The medicine may not be right for other kinds of infections, even if the symptoms are the same. In addition, take all of the medicine to treat the infection for which it was prescribed. The infection may not clear up completely if too little medicine is taken. Taking this medicine for too long, on the other hand, may open the door to new infections that do not respond to the drug.

Some cephalosporins work best when taken on an empty stomach. Others should be taken after meals. Check with the physician who prescribed the medicine or the pharmacist who filled the prescription for instructions on how to take the medicine.

Precautions

Certain cephalosporins should not be combined with alcohol or with medicines that contain alcohol. Abdominal or stomach cramps, nausea, vomiting, facial flushing, and other symptoms may result within 15–30 minutes and may last for several hours. Do not drink alcoholic beverages or use other medicines that contain alcohol while being treated with cephalosporins and for several days after treatment ends.

Special conditions

People with certain medical conditions or who are taking certain other medicines can have problems if they take cephalosporins. Before taking these drugs, be sure to let the physician know about any of these conditions:

ALLERGIES. Severe allergic reactions to this medicine may occur. Anyone who is allergic to cephalosporins of any kind should not take other cephalosporins. Anyone who is allergic to penicillin should check with a physician before taking any cephalosporin. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

KEY TERMS

Bronchitis—Inflammation of the air passages of the lungs.

Colitis—Inflammation of the colon (large bowel).

Gonorrhea—A sexually transmitted disease (STD) that causes infection in the genital organs and may cause disease in other parts of the body.

Inflammation—Pain, redness, swelling, and heat that usually develop in response to injury or illness.

Phenylketonuria—(PKU) A genetic disorder in which the body lacks an important enzyme. If untreated, the disorder can lead to brain damage and mental retardation.

Pneumonia—A disease in which the lungs become inflamed. Pneumonia may be caused by bacteria, viruses, or other organisms, or by physical or chemical irritants.

Sexually transmitted disease—A disease that is passed from one person to another through sexual intercourse or other intimate sexual contact. Also called STD.

Staph infection—Infection with *Staphylococcus* bacteria. These bacteria can infect any part of the body.

Strep throat—A sore throat caused by infection with *Streptococcus* bacteria. Symptoms include sore throat, chills, fever, and swollen lymph nodes in the neck.

Tonsillitis—Inflammation of a tonsil, a small mass of tissue in the throat.

DIABETES. Some cephalosporins may cause false positive results on urine sugar tests for diabetes. People with diabetes should check with their physicians to see if they need to adjust their medication or their **diets**.

PHENYLKETONURIA. Oral suspensions of cefprozil contain phenylalanine. People with **phenylketonuria** (PKU) should consult a physician before taking this medicine.

PREGNANCY. Women who are pregnant or who may become pregnant should check with their physicians before using cephalosporins.

BREASTFEEDING. Cephalosporins may pass into breast milk and may affect nursing babies. Women who are breastfeeding and who need to take this medicine

should check with their physicians. They may need to stop breastfeeding until treatment is finished.

OTHER MEDICAL CONDITIONS. Before using cephalosporins, people with any of these medical problems should make sure their physicians are aware of their conditions:

- History of stomach or intestinal problems, especially colitis. Cephalosporins may cause colitis in some people.
- Kidney problems. The dose of cephalosporin may need to be lower.
- Bleeding problems. Cephalosporins may increase the chance of bleeding in people with a history of bleeding problems.
- Liver disease. The dose of cephalosporin may need to be lower.

USE OF CERTAIN MEDICINES. Taking cephalosporins with certain other drugs may affect the way the drugs work or may increase the chance of side effects.

Side effects

Get medical attention immediately if any of these symptoms develop while taking cephalosporins:

- shortness of breath
- Pounding heartbeat
- Skin rash or **hives**
- Severe cramps or **pain** in the stomach or abdomen
- **Fever**
- Severe watery or bloody **diarrhea** (may occur up to several weeks after stopping the drug)
- Unusual bleeding or bruising

Other rare side effects may occur. Anyone who has unusual symptoms during or after treatment with cephalosporins should get in touch with his or her physician.

Interactions

Some cephalosporins cause diarrhea. Certain diarrhea medicines, such as diphenoxylate-atropine (Lomotil), may make the problem worse. Check with a physician before taking any medicine for diarrhea caused by taking cephalosporins.

Birth control pills may not work properly when taken at the same time as cephalosporins. To prevent **pregnancy**, use other methods of birth control in addition to the pills while taking cephalosporins.

Taking cephalosporins with certain other drugs may increase the risk of excess bleeding. Among the drugs that may have this effect when taken with cephalosporins are:

- blood thinning drugs (anticoagulants) such as warfarin (Coumadin)
- blood viscosity reducing medicines such as pentoxifylline (Trental)
- the antiseizure medicines divalproex (Depakote) and valproic acid (Depakene)

Cephalosporins may also 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 cephalosporins should let the physician know all other medicines he or she is taking.

Nancy Ross-Flanigan

Cerebral abscess see **Brain abscess**

Cerebral amyloid angiopathy

Definition

Cerebral amyloid angiopathy (CAA) is also known as congophilic angiopathy or cerebrovascular **amyloidosis**. It is a disease of small blood vessels in the brain in which deposits of amyloid protein in the vessel walls may lead to **stroke**, brain hemorrhage, or **dementia**. Amyloid protein resembles a starch and is deposited in tissues during the course of certain chronic diseases.

Description

CAA may affect patients over age 45, but is most common in patients over age 65, and becomes more common with increasing age. Men and women are equally affected. In some cases, CAA is sporadic but it may also be inherited as an autosomal dominant condition (a form of inheritance in which only one copy of a gene coding for a disease need be present for that disease to be expressed; if either parent has the disease, a child has a 50% chance of inheriting the disease). CAA is responsible for 5–20% of brain hemorrhages and up to 30% of lobar hemorrhages localized to one lobe of the brain. CAA may be found during an **autopsy** in over one-third of persons over age 60, even though they may not have had brain hemorrhage, stroke, or other manifestations of the disease during life. In **Alzheimer's disease**, CAA is more common than in the general population, and may occur in more than 80% of patients over age 60.

Causes and symptoms

The cause of amyloid deposits in blood vessels in the brain in sporadic CAA is not known. In hereditary CAA,

genetic defects, typically on chromosome 21, allow accumulation of amyloid, a protein made up of units called beta-pleated sheet fibrils. The fibrils tend to clump together, so that the amyloid cannot be dissolved and builds up in the brain blood vessel walls. One form of amyloid fibril subunit proteins is the amyloid beta protein.

Different theories have been suggested for the source of amyloid beta protein in the brain. The systemic theory suggests that amyloid beta protein in the blood stream is deposited in blood vessels in the brain, causing weakness in the blood vessel wall and breakdown in the blood-brain barrier. Normally, the blood-brain barrier keeps proteins and other large molecules from escaping from the blood vessel to the brain tissue. When there is breakdown of the blood-brain barrier, amyloid beta protein leaks through the blood vessel wall, and is deposited in the brain substance, where it forms an abnormal structure called a neuritic plaque.

A second, more likely theory is that amyloid fibrils that form amyloid beta protein are produced by perivascular microglia, or support cells in contact with the brain blood vessel wall. The third theory is that the brain tissue gives rise to amyloid beta protein. Both the nerve cells and the glia are known to produce amyloid precursor protein, which increases with **aging** and with **cell stress**.

Bleeding into the brain may occur as tiny blood vessels carrying amyloid deposits become heavier and more brittle, and are therefore more likely to burst with minor trauma or with fluctuating blood pressure. Aneurysms, or ballooning of the blood vessel wall, may develop, and may also rupture as the stretched wall becomes thinner and is under more pressure. Amyloid deposits may destroy smooth muscle cells or cause inflammation in the blood vessel wall. This may also cause the blood vessel to break more easily.

The most common form of CAA is the sporadic form associated with aging. This type of CAA usually causes lobar hemorrhage, which may recur in different lobes of the brain. The frontal lobe (behind the forehead) and parietal lobe (behind the frontal lobe) are most often affected; the temporal lobe (near the temple) and occipital lobe (at the back of the brain) are affected less often; and the cerebellum (under the occipital lobe) is rarely affected. Approximately 10–50% of hemorrhages in sporadic CAA involve more than one lobe.

Symptoms of lobar hemorrhage in CAA include sudden onset of **headache**, neurologic symptoms such as weakness, sensory loss, visual changes, or speech problems, depending on which lobe is involved; and decreased level of consciousness (a patient who is difficult to arouse), nausea, and vomiting. Sporadic CAA may be associated with symptoms unrelated to lobar

KEY TERMS

Amyloid—Amyloid protein resembles a starch and is deposited in tissues during the course of certain chronic diseases.

Ataxia—Problems with coordination and walking.

Autosomal dominant—A form of inheritance in which only one copy of a gene coding for a disease need be present for that disease to be expressed. If either parent has the disease, a child has a 50% chance of inheriting the disease.

Chromosome—A cellular structure containing genetic information in the form of DNA.

Dementia—Loss of memory and other higher functions, such as thinking or speech, lasting six months or more.

Hemorrhage—Bleeding, or escape of blood through ruptured or unruptured blood vessel walls.

Lobar hemorrhage—Bleeding into one of the lobes of the brain.

Seizure—Epileptic convulsion, fit, or attack.

Sporadic—A form of disease found in persons without a family history of the disease.

Spasticity—Limb stiffness related to disease of the brain or spinal cord.

Stroke—Sudden neurological deficit related to impaired blood supply to the brain.

hemorrhage. Petechial hemorrhages (tiny hemorrhages involving many small vessels) may produce recurrent, brief neurologic symptoms secondary to seizures or decreased blood flow, or may produce rapidly progressive dementia (loss of memory and other brain functions) that worsens in distinct steps rather than gradually. Over 40% of patients with hemorrhage secondary to CAA also have dementia.

Genetic factors play a role in certain types of CAA and in diseases associated with CAA:

- Dutch type of hereditary cerebral hemorrhage with amyloidosis (build up of amyloid protein in blood vessels): autosomal dominant, with a genetic mutation involving the amyloid precursor protein. Onset is at age 40–60 with headaches, brain hemorrhage often in the parietal lobe, strokes, and dementia. More than half of patients die from their first hemorrhage. Patients with the Dutch type of CAA may produce an abnormal anti-

- coagulant, or blood thinner, which makes hemorrhage more likely.
- Flemish type of hereditary cerebral hemorrhage with amyloidosis: autosomal dominant, with a mutation involving the amyloid precursor protein. Symptoms include brain hemorrhage or dementia.
 - Familial Alzheimer's disease: autosomal dominant, comprising 5–10% of all Alzheimer's disease cases (a brain disease in which **death** of nerve cells leads to progressive dementia).
 - **Down Syndrome:** caused by trisomy 21 (three rather than two copies of chromosome 21), causing excess amyloid precursor protein gene. Children with Down syndrome are mentally handicapped and may have heart problems.
 - Icelandic type of hereditary cerebral hemorrhage with amyloidosis: autosomal dominant, with mutation in the gene coding for cystatin C. Symptoms often begin at age 30–40 with multiple brain hemorrhages, dementia, **paralysis** (weakness), and death in 10–20 years. Headache occurs in more than half of patients, and seizures occur in one-quarter. Unlike most other forms of CAA, most hemorrhages involve the basal ganglia deep within the brain (Basal ganglia are islands of tissues in the cerebellum part of the brain.).
 - Familial oculo-leptomeningeal amyloidosis: autosomal dominant with unknown gene defect(s), described in Japanese, Italian, and North American families. Symptoms can include dementia, ataxia (problems with coordination), spasticity (limb stiffness), strokes, seizures, **peripheral neuropathy** (disease affecting the nerves supplying the limbs), migraine, spinal cord problems, blindness, and deafness. Brain hemorrhage is rare as the amyloid protein is deposited in blood vessels in the eye and meninges (brain coverings), but not in the brain itself. In Italian families with the disease, patients may be affected as early as 20–30 years of age.
 - British type of familial amyloidosis: autosomal dominant with unknown gene defect(s), associated with progressive dementia, spasticity, and ataxia. Brain stem, spinal cord, and cerebellum all exhibit amyloid deposits, but hemorrhage typically does not occur.

Diagnosis

As in most neurologic diseases, diagnosis is made most often from the patient's history, with careful inquiry into family history and the patient's onset and pattern of symptoms, as well as neurologic examination. Brain **computed tomography scan** (CT) or **magnetic resonance imaging** (MRI) may identify lobar hemorrhage, stroke, or petechial hemorrhages, and are important in

excluding arteriovenous malformation, **brain tumor**, or other causes of hemorrhage. **Angiography** (x-ray study of the interior of blood vessels and the heart) is not helpful in diagnosis of CAA, but may be needed to exclude aneurysm. **Brain biopsy** (surgical removal of a small piece of brain tissue) may show characteristic amyloid deposits, but is rarely performed, as the risk may not be justifiable in the absence of effective treatment for CAA. If diagnosis is uncertain, biopsy may be needed to rule out conditions which are potentially treatable. Definite diagnosis requires microscopic examination of brain tissue, either at biopsy, at autopsy, or at surgery when brain hemorrhage is drained. Lumbar puncture to examine cerebrospinal fluid proteins may show characteristic abnormalities, but is not part of the routine exam. In familial forms, genetic analysis may be helpful.

CAA with hemorrhage must be distinguished from other types of brain hemorrhage. In CAA, hemorrhage typically occurs in the lobar region, often ruptures into the subarachnoid space between the brain and its coverings, and occurs at night. In hemorrhage related to high blood pressure, hemorrhage is usually deeper within the brain, ruptures into the ventricles or cavities deep inside the brain, and occurs during daytime activities. Other causes of brain hemorrhage are **arteriovenous malformations**, trauma, aneurysms, bleeding into a brain tumor, **vasculitis** (inflammation of blood vessels), or bleeding disorders.

Treatment

Although there is no effective treatment for the underlying disease process of CAA, measures can be taken to prevent brain hemorrhage in patients diagnosed with CAA. High blood pressure should be treated aggressively, and even normal blood pressure can be lowered as much as tolerated without side effects from medications. Blood thinners such as Coumadin, antiplatelet agents such as **aspirin**, or medications designed to dissolve blood clots may cause hemorrhage in patients with CAA, and should be avoided if possible. If these medications are required for other conditions, such as heart disease, the potential benefits must be carefully weighed against the increased risks.

Seizures, or recurrent neurologic symptoms thought to be seizures, should be treated with anti-epileptic drugs, although Depakote (sodium valproate) should be avoided because of its antiplatelet effect. Anti-epileptic drugs are sometimes given to patients with large lobar hemorrhage in an attempt to prevent seizures, although the benefit of this is unclear.

Once brain hemorrhage has occurred, the patient should be admitted to a hospital (ICU) for neurologic monitoring and control of increased pressure within the brain, blood pressure control, and supportive medical

care. Antiplatelet agents and blood thinners should be discontinued and their effects reversed, if possible. Surgery may be needed to remove brain hemorrhage, although bleeding during surgery may be difficult to control.

CAA may be rarely associated with cerebral vasculitis, or inflammation of the blood vessel walls. In these cases treatment with steroids or immune system suppressants may be helpful. Without tissue examination, vasculitis cannot be diagnosed reliably, and probably coexists with CAA too rarely to justify steroid treatment in most cases.

Prognosis

Since CAA is associated with progressive blood vessel degeneration, and since there is no effective treatment, most patients have a poor prognosis. Aggressive neurosurgical management allows increased survival following lobar hemorrhage, but as of 1998, 20–90% of patients die from the first hemorrhage or its complications, which include progression of hemorrhage, brain **edema** (swelling) with herniation (downward pressure on vital brain structures), seizures, and infections such as **pneumonia**. Many survivors have persistent neurologic deficits related to the brain lobe affected by hemorrhage, and are at risk for additional hemorrhages, seizures, and dementia. Prognosis is worse in patients who are older, or who have larger hemorrhages or recurrent hemorrhages within a short time.

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Laurie Barclay, MD

Cerebral aneurysm

Definition

A cerebral aneurysm occurs at a weak point in the wall of a blood vessel (artery) that supplies blood to the

brain. Because of the flaw, the artery wall bulges outward and fills with blood. This bulge is called an aneurysm. An aneurysm can rupture, spilling blood into the surrounding body tissue. A ruptured cerebral aneurysm can cause permanent brain damage, disability, or death.

Description

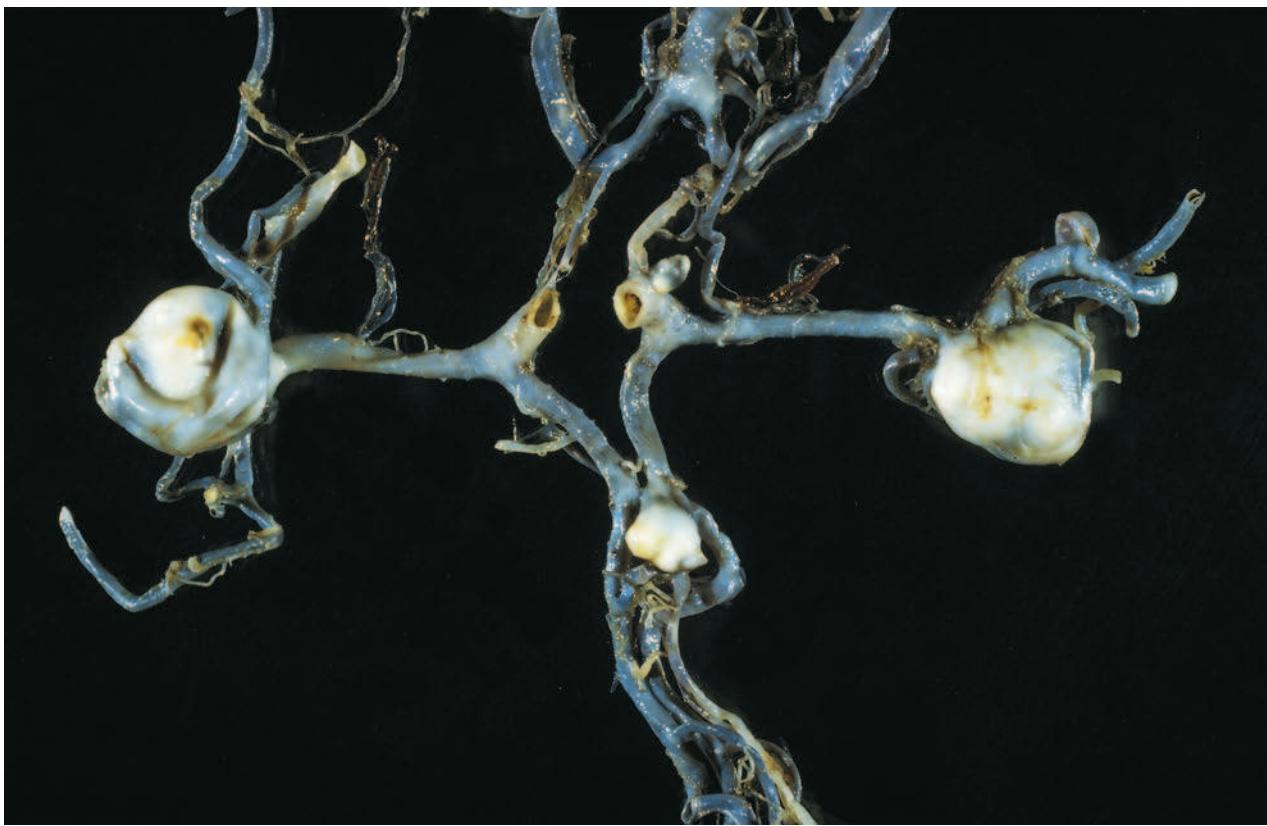
A cerebral aneurysm can occur anywhere in the brain. Aneurysms can have several shapes. The saccular aneurysm, once called a berry aneurysm, resembles a piece of fruit dangling from a branch. Saccular aneurysms are usually found at a branch in the blood vessel where they balloon out by a thin neck. Saccular cerebral aneurysms most often occur at the branch points of large arteries at the base of the brain. Aneurysms may also take the form of a bulge in one wall of the artery—a lateral aneurysm—or a widening of the entire artery—a fusiform aneurysm.

The greatest danger of aneurysms is rupture. Approximately 50–75% of stricken people survive an aneurysmal rupture. A ruptured aneurysm spills blood into the brain or into the fluid-filled area that surrounds the brain tissue. Bleeding into this area, called the subarachnoid space, is referred to as **subarachnoid hemorrhage** (SAH). About 25,000 people suffer a SAH each year. It is estimated that people with unruptured aneurysm have an annual 1–2% risk of hemorrhage. Under age 40, more men experience SAH. After age 40, more women than men are affected.

Most people who have suffered a SAH from a ruptured aneurysm did not know that the aneurysm even existed. Based on **autopsy** studies, medical researchers estimate that 1–5% of the population has some type of cerebral aneurysm. Aneurysms rarely occur in the very young or the very old; about 60% of aneurysms are diagnosed in people between ages 40 and 65.

Some aneurysms may have a genetic link and run in families. The genetic link has not been completely proven and a pattern of inheritance has not been determined. Some studies seem to show that first-degree relatives of people who suffered aneurysmal SAH are more likely to have aneurysms themselves. These studies reported that such immediate family members were four times more likely to have aneurysms than the general population. Other studies do not confirm these findings. Better evidence links aneurysms to certain rare diseases of the connective tissue. These diseases include **Marfan syndrome**, **pseudoxanthoma elasticum**, **Ehlers-Danlos syndrome**, and **fibromuscular dysplasia**. **Polycystic kidney disease** is also associated with cerebral aneurysms.

These diseases are also associated with an increased risk of aneurysmal rupture. Certain other conditions raise



Three aneurysms can be seen in this section of a cerebral artery removed from a human brain. (Photograph by Martin Rotker, Phototake NYC. Reproduced by permission.)

the risk of rupture, too. Most aneurysms that rupture are a half-inch or larger in diameter. Size is not the only factor, however, because smaller aneurysms also rupture. Cigarette **smoking**, excessive alcohol consumption, and recreational drug use (for example, use of **cocaine**) have been linked with an increased risk. The role, if any, of high blood pressure has not been determined. Some studies have implicated high blood pressure in aneurysm formation and rupture, but people with normal blood pressure also experience aneurysms and SAHs. High blood pressure may be a risk factor but not the most important one. **Pregnancy**, labor, and delivery also seem to increase the possibility that an aneurysm might rupture, but not all doctors agree. Physical exertion and use of oral contraceptives are not suspected causes for aneurysmal rupture.

Causes and symptoms

Cerebral aneurysms can be caused by brain trauma, infection, hardening of the arteries (**atherosclerosis**), or abnormal rapid cell growth (neoplastic disease), but most seem to arise from a congenital, or developmental, defect. These congenital aneurysms occur more frequent-

ly in women. Whatever the cause may be, the inner wall of the blood vessel is abnormally thin and the pressure of the blood flow causes an aneurysm to form.

Most aneurysms go unnoticed until they rupture. However, 10–15% of unruptured cerebral aneurysms are found because of their size or their location. Common warning signs include symptoms that affect only one eye, such as an enlarged pupil, a drooping eyelid, or **pain** above or behind the eye. Other symptoms are a localized **headache**, unsteady gait, a temporary problem with sight, double vision, or numbness in the face.

Some aneurysms bleed occasionally without rupturing. Symptoms of such an aneurysm develop gradually. The symptoms include headache, nausea, vomiting, neck pain, black-outs, ringing in the ears, **dizziness**, or seeing spots.

Eighty to ninety percent of aneurysms are not diagnosed until after they have ruptured. Rupture is not always a sudden event. Nearly 50% of patients who have aneurysmal SAHs also experience “the warning leak phenomenon.” Persons with warning leak symptoms have sudden, atypical headaches that occur days or weeks before the actual rupture. These headaches are

KEY TERMS

Congenital—Existing at birth.

Ehlers-Danlos syndrome—A rare inheritable disease of the connective tissue marked by very elastic skin, very loose joints, and very fragile body tissue.

Embolization—A technique to stop or prevent hemorrhage by introducing a foreign mass, such as an air-filled membrane (balloon), into a blood vessel to block the flow of blood.

Fibromuscular dysplasia—A disorder that causes unexplained narrowing of arteries and high blood pressure.

Magnetic resonance angiography—A noninvasive diagnostic technique that uses radio waves to map the internal anatomy of the blood vessels.

Marfan syndrome—An inheritable disorder that affects the skeleton, joints, and blood vessels. Major indicators are excessively long arms and legs, lax joints, and vascular defects.

Nimodipine (Nimotop)—A calcium-channel blocker, that is, a drug that relaxes arterial smooth muscle by slowing the movement of calcium across cell walls.

Polycystic kidney disease—An abnormal condition in which the kidneys are enlarged and contain many cysts.

Pseudoxanthoma elasticum—A hereditary disorder of the connective, or elastic, tissue marked by premature aging and breakdown of the skin and degeneration of the arteries that leads to hemorrhages.

Subarachnoid hemorrhage (SAH)—Loss of blood into the subarachnoid space, the fluid-filled area that surrounds the brain tissue.

Vasospasm—Narrowing of a blood vessel caused by a spasm of the smooth muscle of the vessel wall.

referred to as sentinel headaches. Nausea, vomiting, and dizziness may accompany sentinel headaches. Unfortunately, these symptoms can be confused with tension headaches or migraines, and treatment can be delayed until rupture occurs.

When an aneurysm ruptures, most victims experience a sudden, extremely severe headache. This headache is typically described as the worst headache of the victim's life. **Nausea and vomiting** commonly accompany the headache. The person may experience a short loss of consciousness or prolonged **coma**. Other common signs of a SAH include a stiff neck, **fever**, and a sensitivity to light. About 25% of victims experience neurological problems linked to specific areas of the brain, swelling of the brain due to fluid accumulation (**hydrocephalus**), or seizure.

Diagnosis

Based on the clinical symptoms, a doctor will run several tests to confirm an aneurysm or an SAH. A **computed tomography (CT)** scan of the head is the initial procedure. A **magnetic resonance imaging** test (MRI) may be done instead of a CT scan. MRI, however, is not as sensitive as CT for detecting subarachnoid blood. A CT scan can determine whether there has been a hemorrhage and can assist in pinpointing the location of the aneurysm. The scan is most useful when it is done within

72 hours of the rupture. Later scans may miss the signs of hemorrhage.

If the CT scan is negative for a hemorrhage or provides an unclear diagnosis, the doctor will order a **cerebrospinal fluid (CSF) analysis**, also called a lumbar puncture. In this procedure, a small amount of cerebrospinal fluid is removed from the lower back and examined for traces of blood and blood-breakdown products. If this test is positive, cerebral **angiography** is used to map the brain's blood vessels and the damaged area. The angiography is done to pinpoint the aneurysm's location. About 15% of people who experience SAH have more than one aneurysm. For this reason, angiography should include both the common carotid artery that feeds the front of the brain and the vertebral artery that feeds the base of the brain. Occasionally, the angiography fails to find the aneurysm and must be repeated. If seizures occur, **electroencephalography (EEG)** may be used to measure the electrical activity of the brain.

Treatment

Unruptured aneurysm

If an aneurysm has not ruptured and is not causing any symptoms, it may be left untreated. Because there is a 1–2% chance of rupture per year, the cumulative risk over a number of years may justify surgical treatment.

However, if the aneurysm is small or in a place that would be difficult to reach, or if the person who has the aneurysm is in poor health, the surgical treatment may be a greater risk than the aneurysm. Risk of rupture is higher for people who have more than one aneurysm. Unruptured aneurysm would probably be treated with a surgical procedure called the clip ligation, as described below.

Ruptured aneurysm

The primary treatment for a ruptured aneurysm involves stabilizing the victim's condition, treating the immediate symptoms, and promptly assessing further treatment options, especially surgical procedures. The patient may require mechanical ventilation, oxygen, and fluids. Medications may be given to prevent major secondary complications such as seizures, rebleeding, and vasospasm (narrowing of the affected blood vessel). Vasospasm decreases blood flow to the brain and causes the death of nerve cells. A drug such as nimodipine (Nimotop) may help prevent vasospasm by relaxing the smooth muscle tissue of the arteries. Even with treatment, however, vasospasm may cause stroke or death.

To prevent further hemorrhage from the aneurysm, it must be removed from circulation. In general, surgical procedures should be performed as soon as possible to prevent rebleeding. The chances that aneurysm will rebleed are greatest in the first 24 hours, and vasospasm usually does not occur until 72 hours or more after rupture. If the patient is in poor condition or if there is vasospasm or other complication, surgical procedures may be delayed. The preferred surgical method is a clip ligation in which a clip is placed around the base of the aneurysm to block it off from circulation. Surgical coating, wrapping, or trapping of the aneurysm may also be performed. These procedures do not completely remove the aneurysm from circulation, however, and there is some risk that it may rebleed in the future. Newer techniques that look promising include balloon embolization, a procedure that blocks the aneurysm with an inflatable membrane introduced by means of a catheter inserted through the artery.

Prognosis

An unruptured aneurysm may not cause any symptoms over an entire lifetime. Surgical clip ligation will ensure that it won't rupture, but it may be better to leave the aneurysm alone in some cases. Familial cerebral aneurysms may rupture earlier than those without a genetic link.

The outlook is not as good for a person who suffers a ruptured aneurysm. Fifteen to twenty-five percent of people who experience a ruptured aneurysm do not sur-

vive. An additional 25–50% die as a result of complications associated with the hemorrhage. Of the survivors, 15–50% suffer permanent brain damage and disability. These conditions are caused by the death of nerve cells. Nerve cells can be destroyed by the hemorrhage itself or by complications from the hemorrhage, such as vasospasm or hydrocephalus. Hydrocephalus, a dilatation (expansion) of the fluid-filled cavity surrounding the brain, occurs in about 15% of cases. Immediate medical treatment is vital to prevent further complications and brain damage in those who survive the initial rupture. Patients who survive SAH and aneurysm clipping are unlikely to die from events related to SAH.

Prevention

There are no known methods to prevent an aneurysm from forming. If an aneurysm is discovered before it ruptures, it may be surgically removed. CT or MRI angiography may be recommended for relatives of patients with familial cerebral aneurysms.

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Julia Barrett

Cerebral angiography see **Angiography**

Cerebral palsy

Definition

Cerebral palsy (CP) is the term used for a group of nonprogressive disorders of movement and posture caused by abnormal development of, or damage to, motor control centers of the brain. CP is caused by events before, during, or after birth. The abnormalities of muscle control that define CP are often accompanied by other neurological and physical abnormalities.

Description

Voluntary movement (walking, grasping, chewing, etc.) is primarily accomplished using muscles that are attached to bones, known as the skeletal muscles. Control of the skeletal muscles originates in the cerebral cortex, the largest portion of the brain. Palsy means **paralysis**, but may also be used to describe uncontrolled muscle movement. Therefore, cerebral palsy encompasses any disorder of abnormal movement and paralysis caused by abnormal function of the cerebral cortex. In truth, however, CP does not include conditions due to progressive disease or degeneration of the brain. For this reason, CP is also referred to as static (nonprogressive) encephalopathy (disease of the brain). Also excluded from CP are any disorders of muscle control that arise in the muscles themselves and/or in the peripheral nervous system (nerves outside the brain and spinal cord).

CP is not a specific diagnosis, but is more accurately considered a description—a description of a broad but defined group of neurological and physical problems.

The symptoms of CP and their severity are quite variable. Those with CP may have only minor difficulty with fine motor skills, such as grasping and manipulating items with their hands. A severe form of CP could involve significant muscle problems in all four limbs, **mental retardation**, seizures, and difficulties with vision, speech, and hearing.

Muscles that receive defective messages from the brain may be constantly contracted and tight (spastic), exhibit involuntary writhing movements (athetosis), or have difficulty with voluntary movement (dyskinesia). There can also be a lack of balance and coordination with unsteady movements (ataxia). A combination of any of these problems may also occur. Spastic CP and mixed CP constitute the majority of cases. Effects on the muscles can range from mild weakness or partial paralysis (pareisis), to complete loss of voluntary control of a muscle or group of muscles (plegia). CP is also designated by the number of limbs affected. For instance, affected muscles in one limb is monoplegia, both arms or both legs is

diplegia, both limbs on one side of the body is hemiplegia, and in all four limbs is quadriplegia. Muscles of the trunk, neck, and head may be affected as well.

CP can be caused by a number of different mechanisms at various times—from several weeks after conception, through birth, to early childhood. For many years, it was accepted that most cases of CP were due to brain injuries received during a traumatic birth, known as birth asphyxia. However, extensive research in the 1980s showed that only 5–10% of CP can be attributed to birth trauma. Other possible causes include abnormal development of the brain, prenatal factors that directly or indirectly damage neurons in the developing brain, premature birth, and brain injuries that occur in the first few years of life.

Advances in the medical care of premature infants in the last 20 years have dramatically increased the rate of survival of these fragile newborns. However, as gestational age at delivery and birth weight of a baby decrease, the risk for CP dramatically increases. A term **pregnancy** is delivered at 37–41 weeks gestation. The risk for CP in a preterm infant (32–37 weeks) is increased about five-fold over the risk for an infant born at term. Survivors of extremely preterm births (less than 28 weeks) face as much as a 50-fold increase in risk. About 50% of all cases of CP now being diagnosed are in children who were born prematurely.

Two factors are involved in the risk for CP associated with **prematurity**. First, premature babies are at higher risk for various CP-associated medical complications, such as intracerebral hemorrhage, infection, and difficulty in breathing, to name a few. Second, the onset of **premature labor** may be induced, in part, by complications that have already caused neurologic damage in the fetus. A combination of both factors almost certainly plays a role in some cases of CP. The tendency toward premature delivery runs in families, but the genetic mechanisms are far from clear.

An increase in multiple pregnancies in recent years, especially in the United States, is blamed on the increased use of fertility drugs. As the number of fetuses in a pregnancy increases, the risks for abnormal development and premature delivery also increase. Children from twin pregnancies have four times the risk of developing CP as children from singleton pregnancies, owing to the fact that more twin pregnancies are delivered prematurely. The risk for CP in a child of triplets is up to 18 times greater. Furthermore, recent evidence suggests that a baby from a pregnancy in which its twin died before birth is at increased risk for CP.

Approximately 500,000 children and adults in the United States have CP, and it is newly diagnosed in about 6,000 infants and young children each year. The inci-

dence of CP has not changed much in the last 20–30 years. Ironically, advances in medicine have decreased the incidence from some causes, Rh disease for example, but increased it from others, notably, prematurity and multiple pregnancies. No particular ethnic groups seem to be at higher risk for CP. However, people of disadvantaged background are at higher risk due to poorer access to proper prenatal care and advanced medical services.

Causes and symptoms

As noted, CP has many causes, making a discussion of the genetics of CP complicated. A number of hereditary/genetic syndromes have signs and symptoms similar to CP, but usually also have problems not typical of CP. Put another way, some hereditary conditions “mimic” CP. Isolated CP, meaning CP that is not a part of some other syndrome or disorder, is usually not inherited.

It might be possible to group the causes of CP into those that are genetic and those that are non-genetic, but most would fall somewhere in between. Grouping causes into those that occur during pregnancy (prenatal), those that happen around the time of birth (perinatal), and those that occur after birth (postnatal), is preferable. CP related to premature birth and multiple pregnancies (twins, triplets, etc., not “many pregnancies”) is somewhat different and considered separately.

Prenatal causes

Although much has been learned about human embryology in the last couple of decades, a great deal remains unknown. Studying prenatal human development is difficult because the embryo and fetus develop in a closed environment—the mother’s womb. However, the relatively recent development of a number of prenatal tests has opened a window on the process. Add to that more accurate and complete evaluations of newborns, especially those with problems, and a clearer picture of what can go wrong before birth is possible.

The complicated process of brain development before birth is susceptible to many chance errors that can result in abnormalities of varying degrees. Some of these errors will result in structural anomalies of the brain, while others may cause undetectable, but significant, abnormalities in how the cerebral cortex is “wired.” An abnormality in structure or wiring is sometimes hereditary, but is most often due to chance, or a cause unknown at this time. Whether and how much genetics played a role in a particular brain abnormality depends to some degree on the type of anomaly and the form of CP it causes.

Several maternal-fetal infections are known to increase the risk for CP, including **rubella** (German

measles, now rare in the United States), cytomegalovirus (CMV), and **toxoplasmosis**. Each of these infections is considered a risk to the fetus only if the mother contracts it for the first time during that pregnancy. Even in those cases, though, most babies will be born normal. Most women are immune to all three infections by the time they reach childbearing age, but a woman’s immune status can be determined using the so-called TORCH (for Toxoplasmosis, Rubella, Cytomegalovirus, and Herpes) test before or during pregnancy.

Just as a **stroke** can cause neurologic damage in an adult, so too can this type of event occur in the fetus. A burst blood vessel in the brain followed by uncontrolled bleeding (coagulopathy), known as intracerebral hemorrhage, could cause a fetal stroke, or a cerebral blood vessel could be obstructed by a clot (**embolism**). Infants who later develop CP, along with their mothers, are more likely than other mother-infant pairs to test positive for factors that put them at increased risk for bleeding episodes or blood clots. Some **coagulation disorders** are strictly hereditary, but most have a more complicated basis.

A teratogen is any substance to which a woman is exposed that has the potential to harm the embryo or fetus. Links between a drug or other chemical exposure during pregnancy and a risk for CP are difficult to prove. However, any substance that might affect fetal brain development, directly or indirectly, could increase the risk for CP. Furthermore, any substance that increases the risk for premature delivery and low birth weight, such as alcohol, tobacco, or **cocaine**, among others, might indirectly increase the risk for CP.

The fetus receives all nutrients and oxygen from blood that circulates through the placenta. Therefore, anything that interferes with normal placental function might adversely affect development of the fetus, including the brain, or might increase the risk for premature delivery. Structural abnormalities of the placenta, premature detachment of the placenta from the uterine wall (abruption), and placental infections (chorioamnionitis) are thought to pose some risk for CP.

Certain conditions in the mother during pregnancy might pose a risk to fetal development leading to CP. Women with autoimmune anti-thyroid or anti-phospholipid (APA) antibodies are at slightly increased risk for CP in their children. A potentially important clue uncovered recently points toward high levels of cytokines in the maternal and fetal circulation as a possible risk for CP. Cytokines are proteins associated with inflammation, such as from infection or **autoimmune disorders**, and they may be toxic to neurons in the fetal brain. More research is needed to determine the exact relationship, if any, between high levels of cytokines in pregnancy and

KEY TERMS

Asphyxia—Lack of oxygen. In the case of cerebral palsy, lack of oxygen to the brain.

Ataxia—A deficiency of muscular coordination, especially when voluntary movements are attempted, such as grasping or walking.

Athetosis—A condition marked by slow, writhing, involuntary muscle movements.

Cerebral palsy—Movement disability resulting from nonprogressive brain damage.

Coagulopathy—A disorder in which blood is either too slow or too quick to coagulate (clot).

Contracture—A tightening of muscles that prevents normal movement of the associated limb or other body part.

Cytokine—A protein associated with inflammation that, at high levels, may be toxic to nerve cells in the developing brain.

Diplegia—Paralysis affecting like parts on both sides of the body, such as both arms or both legs.

Dorsal rhizotomy—A surgical procedure that cuts nerve roots to reduce spasticity in affected muscles.

Dyskinesia—Impaired ability to make voluntary movements.

Hemiplegia—Paralysis of one side of the body.

Hypotonia—Reduced or diminished muscle tone.

Quadriplegia—Paralysis of all four limbs.

Serial casting—A series of casts designed to gradually move a limb into a more functional position.

Spastic—A condition in which the muscles are rigid, posture may be abnormal, and fine motor control is impaired.

Spasticity—Increased muscle tone, or stiffness, which leads to uncontrolled, awkward movements.

Static encephalopathy—A disease of the brain that does not get better or worse.

Tenotomy—A surgical procedure that cuts the tendon of a contracted muscle to allow lengthening.

CP. A woman has some risk of developing the same complications in more than one pregnancy, slightly increasing the risk for more than one child with CP.

Serious physical trauma to the mother during pregnancy could result in direct trauma to the fetus as well, or injuries to the mother could compromise the availability of nutrients and oxygen to the developing fetal brain.

Perinatal causes

Birth asphyxia significant enough to result in CP is now uncommon in developed countries. Tight nuchal cord (umbilical cord around the baby's neck) and prolapsed cord (cord delivered before the baby) are possible causes of birth asphyxia, as are bleeding and other complications associated with **placental abruption** and **placenta previa** (placenta lying over the cervix).

Infection in the mother is sometimes not passed to the fetus through the placenta, but is transmitted to the baby during delivery. Any such infection that results in serious illness in the newborn has the potential to produce some neurological damage.

Postnatal causes

The remaining 15% of CP is due to neurologic injury sustained after birth. CP that has a postnatal cause

is sometimes referred to as acquired CP, but this is only accurate for those cases caused by infection or trauma.

Incompatibility between the Rh blood types of mother and child (mother Rh negative, baby Rh positive) can result in severe anemia in the baby (**erythroblastosis fetalis**). This may lead to other complications, including severe **jaundice**, which can cause CP. Rh disease in the newborn is now rare in developed countries due to routine screening of maternal blood type and treatment of pregnancies at risk. The routine, effective treatment of jaundice due to other causes has also made it an infrequent cause of CP in developed countries. Rh blood type poses a risk for recurrence of Rh disease if treatment is not provided.

Serious infections that affect the brain directly, such as **meningitis** and **encephalitis**, may cause irreversible damage to the brain, leading to CP. A **seizure disorder** early in life may cause CP, or may be the product of a hidden problem that causes CP in addition to seizures. Unexplained (idiopathic) seizures are hereditary in only a small percentage of cases. Although rare in infants born healthy at or near term, intracerebral hemorrhage and brain embolism, like fetal stroke, are sometimes genetic.

Physical trauma to an infant or child resulting in brain injury, such as from abuse, accidents, or near

drowning/suffocation, might cause CP. Likewise, ingestion of a toxic substance such as lead, mercury, poisons, or certain chemicals could cause neurological damage. Accidental overdose of certain medications might also cause similar damage to the central nervous system.

By definition, the defect in cerebral function causing CP is nonprogressive. However, the symptoms of CP often change over time. Most of the symptoms of CP relate in some way to the aberrant control of muscles. To review, CP is categorized first by the type of movement/postural disturbance(s) present, then by a description of which limbs are affected, and finally by the severity of motor impairment. For example, spastic diplegia refers to continuously tight muscles that have no voluntary control in both legs, while athetoid quadraparesis describes uncontrolled writhing movements and muscle weakness in all four limbs. These three-part descriptions are helpful in providing a general picture, but cannot give a complete description of any one person with CP. In addition, the various “forms” of CP do not occur with equal frequency—spastic diplegia is seen in more individuals than is athetoid quadraparesis. CP can also be loosely categorized as mild, moderate, or severe, but these are very subjective terms with no firm boundaries between them.

A muscle that is tensed and contracted is hypertonic, while excessively loose muscles are hypotonic. Spastic, hypertonic muscles can cause serious orthopedic problems, including **scoliosis** (spine curvature), hip dislocation, or **contractures**. A contracture is shortening of a muscle, aided sometimes by a weak-opposing force from a neighboring muscle. Contractures may become permanent, or “fixed,” without some sort of intervention. Fixed contractures may cause postural abnormalities in the affected limbs. Clenched fists and contracted feet (*equinus* or *equinovarus*) are common in people with CP. Spasticity in the thighs causes them to turn in and cross at the knees, resulting in an unusual method of walking known as a “scissors gait.” Any of the joints in the limbs may be stiff (immobilized) due to spasticity of the attached muscles.

Athetosis and dyskinesia often occur with spasticity, but do not often occur alone. The same is true of ataxia. It is important to remember that “mild CP” or “severe CP” refers not only to the number of symptoms present, but also to the level of involvement of any particular class of symptoms.

Mechanisms that can cause CP are not always restricted to motor-control areas of the brain. Other neurologically based symptoms may include:

- mental retardation/learning disabilities
- behavioral disorders

- seizure disorders
- visual impairment
- hearing loss
- speech impairment (dysarthria)
- abnormal sensation and perception

These problems may have a greater impact on a child’s life than the physical impairments of CP, although not all children with CP are affected by other problems. Many infants and children with CP have growth impairment. About one-third of individuals with CP have moderate-to-severe mental retardation, one-third have mild mental retardation, and one-third have normal intelligence.

Diagnosis

The signs of CP are not usually noticeable at birth. Children normally progress through a predictable set of developmental milestones through the first 18 months of life. Children with CP, however, tend to develop these skills more slowly because of their motor impairments, and delays in reaching milestones are usually the first symptoms of CP. Babies with more severe cases of CP are usually diagnosed earlier than others.

Selected developmental milestones, and the ages for normally acquiring them, are given below. If a child does not acquire the skill by the age shown in parentheses, there is some cause for concern.

- sits well unsupported—six months (eight–10 months)
- babbles—six months (eight months)
- crawls—nine months (12 months)
- finger feeds, holds bottle—nine months (12 months)
- walks alone—12 months (15–18 months)
- uses one or two words other than *dada/mama*—12 months (15 months)
- walks up and down steps—24 months (24–36 months)
- turns pages in books; removes shoes and socks—24 months (30 months)

Children do not consistently favor one hand over the other before 12–18 months, and doing so may be a sign that the child has difficulty using the other hand. This same preference for one side of the body may show up as asymmetric crawling or, later on, favoring one leg while climbing stairs.

It must be remembered that children normally progress at somewhat different rates, and slow beginning accomplishment is often followed by normal development. Other causes for developmental delay—some benign, some serious—should be excluded before con-

sidering CP as the answer. CP is nonprogressive, so continued loss of previously acquired milestones indicates that CP is not the cause of the problem.

No one test is diagnostic for CP, but certain factors increase suspicion. The Apgar score measures a baby's condition immediately after birth. Babies that have low Apgar scores are at increased risk for CP. Presence of abnormal muscle tone or movements may indicate CP, as may the persistence of infantile reflexes. Imaging of the brain using ultrasound, x rays, MRI, and/or CT scans may reveal a structural anomaly. Some brain lesions associated with CP include scarring, cysts, expansion of the cerebral ventricles (**hydrocephalus**), periventricular leukomalacia (an abnormality of the area surrounding the ventricles), areas of dead tissue (necrosis), and evidence of an intracerebral hemorrhage or blood clot. Blood and urine biochemical tests, as well as genetic tests, may be used to rule out other possible causes, including muscle and peripheral nerve diseases, mitochondrial and metabolic diseases, and other inherited disorders. Evaluations by a pediatric developmental specialist and a geneticist may be of benefit.

Treatment

Cerebral palsy cannot be cured, but many of the disabilities it causes can be managed through planning and timely care. Treatment for a child with CP depends on the severity, nature, and location of the primary muscular symptoms, as well as any associated problems that might be present. Optimal care of a child with mild CP may involve regular interaction with only a physical therapist and occupational therapist, whereas care for a more severely affected child may include visits to multiple medical specialists throughout life. With proper treatment and an effective plan, most people with CP can lead productive, happy lives.

Therapy

Spasticity, muscle weakness, coordination, ataxia, and scoliosis are all significant impairments that affect the posture and mobility of a person with CP. Physical and occupational therapists work with the patient, and the family, to maximize the ability to move affected limbs, develop normal motor patterns, and maintain posture. "Assistive technology," things such as wheelchairs, walkers, shoe inserts, crutches, and braces, are often required. A speech therapist, and high-tech aids such as computer-controlled communication devices, can make a tremendous difference in the life of those who have speech impairments.

Medications

Before fixed contractures develop, muscle-relaxant drugs such as diazepam (Valium), dantrolene (Dantri-

um), and baclofen (Lioresal) may be prescribed. Botulinum toxin (Botox), a newer and highly effective treatment, is injected directly into the affected muscles. Alcohol or phenol injections into the nerve controlling the muscle are another option. Multiple medications are available to control seizures, and athetosis can be treated using medications such as trihexyphenidyl HCl (Artane) and benzotropine (Cogentin).

Surgery

Fixed contractures are usually treated with either serial casting or surgery. The most commonly used surgical procedures are tenotomy, tendon transfer, and dorsal rhizotomy. In tenotomy, tendons of the affected muscle are cut and the limb is cast in a more normal position while the tendon regrows. Alternatively, tendon transfer involves cutting and reattaching a tendon at a different point on the bone to enhance the length and function of the muscle. A neurosurgeon performing dorsal rhizotomy carefully cuts selected nerve roots in the spinal cord to prevent them from stimulating the spastic muscles. Neurosurgical techniques in the brain such as implanting tiny electrodes directly into the cerebellum, or cutting a portion of the hypothalamus, have very specific uses and have had mixed results.

Education

Parents of a child newly diagnosed with CP are not likely to have the necessary expertise to coordinate the full range of care their child will need. Although knowledgeable and caring medical professionals are indispensable for developing a care plan, a potentially more important source of information and advice is other parents who have dealt with the same set of difficulties. Support groups for parents of children with CP can be significant sources of both practical advice and emotional support. Many cities have support groups that can be located through the United Cerebral Palsy Association, and most large medical centers have special multidisciplinary clinics for children with developmental disorders.

Prognosis

Cerebral palsy can affect every stage of maturation, from childhood through adolescence to adulthood. At each stage, those with CP, along with their caregivers, must strive to achieve and maintain the fullest range of experiences and education consistent with their abilities. The advice and intervention of various professionals remains crucial for many people with CP. Although CP itself is not considered a terminal disorder, it can affect a person's lifespan by increasing the risk for certain med-

ical problems. People with mild cerebral palsy may have near-normal lifespans, but the lifespan of those with more severe forms may be shortened. However, over 90% of infants with CP survive into adulthood.

The cause of most cases of CP remains unknown, but it has become clear in recent years that birth difficulties are not to blame in most cases. Rather, developmental problems before birth, usually unknown and generally undiagnosable, are responsible for most cases. The rate of survival for preterm infants has leveled off in recent years, and methods to improve the long-term health of these at-risk babies are now being sought. Current research is also focusing on the possible benefits of recognizing and treating coagulopathies and inflammatory disorders in the prenatal and perinatal periods. The use of magnesium sulfate in pregnant women with preeclampsia or threatened preterm delivery may reduce the risk of CP in very preterm infants. Finally, the risk of CP can be decreased through good maternal **nutrition**, avoidance of drugs and alcohol during pregnancy, and prevention or prompt treatment of infections.

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- Epilepsy Foundation of America. 4351 Garden City Dr., Suite 406, Landover, MD 20785-2267. (301) 459-3700 or (800) 332-1000. <<http://www.epilepsysfoundation.org>>.
- March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (888) 663-4637. resource-center@modimes.org. <<http://www.modimes.org>>.
- National Easter Seal Society. 230 W. Monroe St., Suite 1800, Chicago, IL 60606-4802. (312) 726-6200 or (800) 221-6827. <<http://www.easter-seals.org>>.

National Institute of Neurological Disorders and Stroke. 31 Center Drive, MSC 2540, Bldg. 31, Room 8806, Bethesda, MD 20814. (301) 496-5751 or (800) 352-9424. <<http://www.ninds.nih.gov>>.

National Society of Genetic Counselors. 233 Canterbury Dr., Wallingford, PA 19086-6617. (610) 872-1192. <<http://www.nsge.org/GeneticCounselingYou.asp>>.

United Cerebral Palsy Association, Inc. (UCP). 1660 L St. NW, Suite 700, Washington, DC 20036-5602. (202) 776-0406 or (800) 872-5827. <<http://www.ucpa.org>>.

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Scott J. Polzin, MS

Cerebrospinal fluid (CSF) analysis

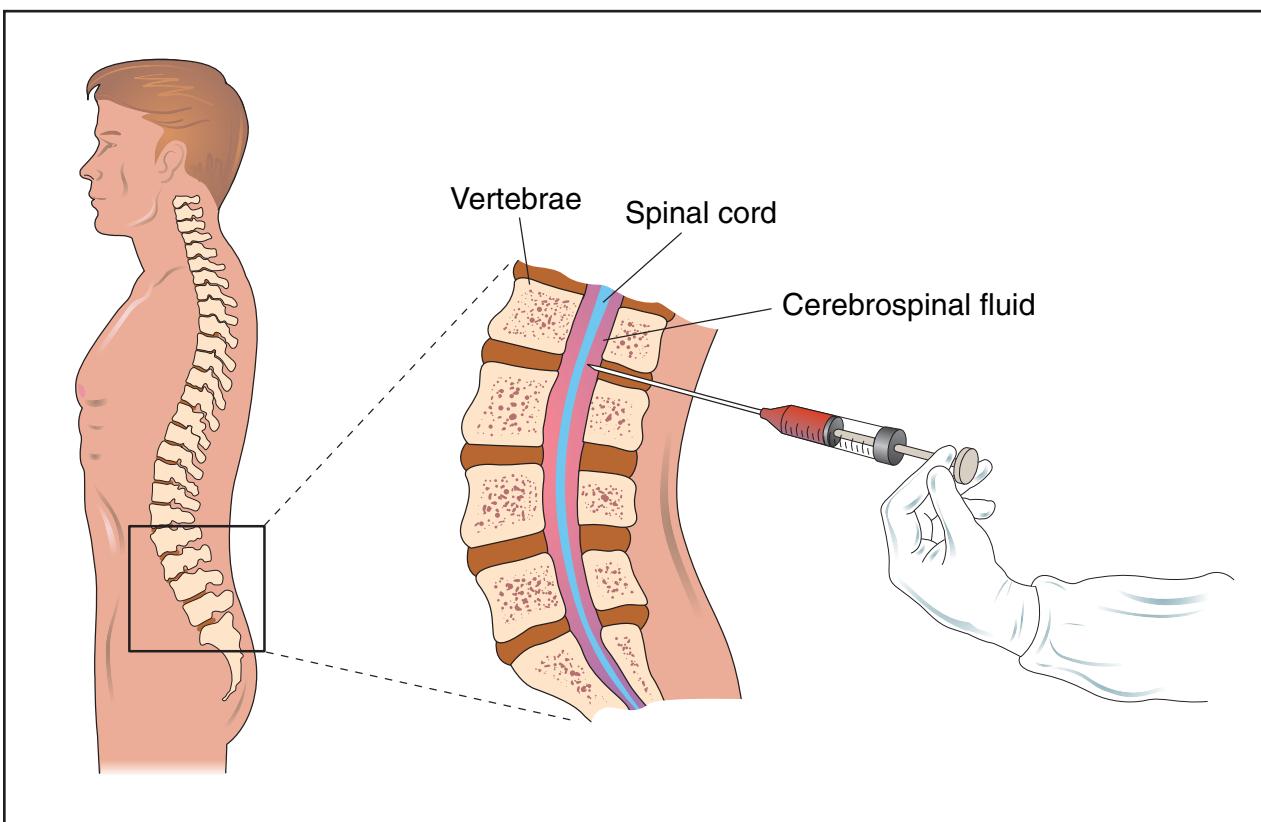
Definition

Cerebrospinal fluid (CSF) analysis is a laboratory test to examine a sample of the fluid surrounding the brain and spinal cord. This fluid is a clear, watery liquid that protects the central nervous system from injury and cushions it from the surrounding bone structure. It contains a variety of substances, particularly glucose (sugar), protein, and white blood cells from the immune system. The fluid is withdrawn through a needle in a procedure called a lumbar puncture.

Purpose

The purpose of a CSF analysis is to diagnose medical disorders that affect the central nervous system. Some of these conditions include:

- viral and bacterial infections, such as **meningitis** and **encephalitis**
- tumors or cancers of the nervous system
- syphilis, a sexually transmitted disease
- bleeding (hemorrhaging) around the brain and spinal cord
- multiple sclerosis, a disease that affects the myelin coating of the nerve fibers of the brain and spinal cord
- Guillain-Barré syndrome, an inflammation of the nerves.



During a lumbar puncture, or spinal tap, a procedure in which cerebrospinal fluid is aspirated, the physician inserts a hollow, thin needle in the space between two vertebrae of the lower back and slowly advances it toward the spine. The cerebrospinal fluid pressure is then measured and the fluid is withdrawn for laboratory analysis. (Illustration by Electronic Illustrators Group.)

Precautions

In some circumstances, a lumbar puncture to withdraw a small amount of CSF for analysis may lead to serious complications. Lumbar puncture should be performed only with extreme caution, and only if the benefits are thought to outweigh the risks, in certain conditions. For example, in people who have blood clotting (coagulation) or bleeding disorders, lumbar puncture can cause bleeding that can compress the spinal cord. If there is a large **brain tumor** or other mass, removal of CSF can cause the brain to droop down within the skull cavity (herniate), compressing the brain stem and other vital structures, and leading to irreversible brain damage or **death**. These problems are easily avoided by checking blood coagulation through a blood test and by doing a **computed tomography scan** (CT) or **magnetic resonance imaging** (MRI) scan before attempting the lumbar puncture. In addition, a lumbar puncture procedure should never be performed at the site of a localized skin infection on the lower back because the infection may be introduced into the CSF and may spread to the brain or spinal cord.

Description

The procedure to remove cerebrospinal fluid is called a lumbar puncture, or spinal tap, because the area of the spinal column used to obtain the sample is in the lumbar spine, or lower section of the back. In rare instances, such as a spinal fluid blockage in the middle of the back, a doctor may perform a spinal tap in the neck. The lower lumbar spine (usually between the vertebrae known as L4–5) is preferable because the spinal cord stops near L2, and a needle introduced below this level will miss the spinal cord and encounter only nerve roots, which are easily pushed aside.

A lumbar puncture takes about 30 minutes. Patients can undergo the test in a doctor's office, laboratory, or outpatient hospital setting. Sometimes it requires an inpatient hospital stay. If the patient has spinal arthritis, is extremely uncooperative, or obese, it may be necessary to introduce the spinal needle using x-ray guidance.

In order to get an accurate sample of cerebrospinal fluid, it is critical that a patient is in the proper position. The spine must be curved to allow as much space as possi-

KEY TERMS

Encephalitis—An inflammation or infection of the brain and spinal cord caused by a virus or as a complication of another infection.

Guillain-Barré syndrome—An inflammation involving nerves that affect the extremities. The inflammation may spread to the face, arms, and chest.

Immune system—Protects the body against infection.

Manometer—A device used to measure fluid pressure.

Meningitis—An infection or inflammation of the membranes or tissues that cover the brain and spinal cord, and caused by bacteria or a virus.

Multiple sclerosis—A disease that destroys the covering (myelin sheath) of nerve fibers of the brain and spinal cord.

Spinal canal—The cavity or hollow space within the spine that contains cerebrospinal fluid.

Vertebrae—The bones of the spinal column. There are 33 along the spine, with five (called L1-L5) making up the lower lumbar region.

ble between the lower vertebrae, or bones of the back, for the doctor to insert a lumbar puncture needle between the vertebrae and withdraw a small amount of fluid. The most common position is for the patient to lie on his or her side with the back at the edge of the exam table, head and chin bent down, knees drawn up to the chest, and arms clasped around the knees. (Small infants and people who are obese may need to curve their spines in a sitting position.) People should talk to their doctor if they have any questions about their position because it is important to be comfortable and to remain still during the entire procedure. In fact, the doctor will explain the procedure to the patient (or guardian) so that the patient can agree in writing to have it done (informed consent). If the patient is anxious or uncooperative, a short-acting sedative may be given.

During a lumbar puncture, the doctor drapes the back with a sterile covering that has an opening over the puncture site and cleans the skin surface with an antiseptic solution. Patients receive a local anesthetic to minimize any **pain** in the lower back.

The doctor inserts a hollow, thin needle in the space between two vertebrae of the lower back and slowly advances it toward the spine. A steady flow of clear cerebrospinal fluid, normally the color of water, will begin to

fill the needle as soon as it enters the spinal canal. The doctor measures the cerebrospinal fluid pressure with a special instrument called a manometer and withdraws several vials of fluid for laboratory analysis. The amount of fluid collected depends on the type and number of tests needed to diagnose a particular medical disorder.

In some cases, the doctor must remove and reposition the needle. This occurs when there is not an even flow of fluid, the needle hits bone or a blood vessel, or the patient reports sharp, unusual pain.

Preparation

Patients can go about their normal activities before a lumbar puncture. Experts recommend that patients relax before the procedure to release any muscle tension, since the lumbar puncture needle must pass through muscle tissue before it reaches the spinal canal. A patient's level of relaxation before and during the procedure plays a critical role in the test's success.

Aftercare

After the procedure, the doctor covers the site of the puncture with a sterile bandage. Patients must avoid sitting or standing and remain lying down for as long as six hours after the lumbar puncture. They should also drink plenty of fluid to help prevent lumbar puncture **headache**, which is discussed in the next section.

Risks

For most people, the most common side effect after the removal of CSF is a headache. This occurs in 10–30% of adult patients and in up to 40% of children. It is caused by a decreased CSF pressure related to a small leak of CSF through the puncture site. These headaches usually are a dull pain, although some people report a throbbing sensation. A stiff neck and nausea may accompany the headache. Lumbar puncture headaches typically begin within two days after the procedure and persist from a few days to several weeks or months.

Since an upright position worsens the pain, patients with a lumbar puncture headache can control the pain by lying in a flat position and taking a prescription or non-prescription pain relief medication, preferably one containing **caffeine**. In rare cases, the puncture site leak is “patched” using the patient’s own blood.

People should talk to their doctor about complications from a lumbar puncture. In most cases, this test to analyze CSF is a safe and effective procedure. Some patients experience pain, difficulty urinating, infection, or leakage of cerebrospinal fluid from the puncture site after the procedure.

Normal results

Normal CSF is clear and colorless. It may be cloudy in infections; straw- or yellow-colored if there is excess protein, as may occur with **cancer** or inflammation; blood-tinged if there was recent bleeding; or yellow to brown (xanthochromic) if caused by an older instance of bleeding.

A series of laboratory tests analyze the CSF for a variety of substances to rule out possible medical disorders of the central nervous system. The following are normal values for commonly tested substances:

- CSF pressure: 50–180 mmH₂O
- glucose: 40–85 mg/dL
- protein: 15–50 mg/dL
- leukocytes (white blood cells) total less than 5 per mL
- lymphocytes: 60–70%
- monocytes: 30–50%
- neutrophils: none

Normally, there are no red blood cells in the CSF unless the needle passes through a blood vessel on route to the CSF. If this is the case, there should be more red blood cells in the first tube collected than in the last.

Abnormal results

Abnormal test result values in the pressure or any of the substances found in the cerebrospinal fluid may suggest a number of medical problems including a tumor or spinal cord obstruction; hemorrhaging or bleeding in the central nervous system; infection from bacterial, viral, or fungal microorganisms; or an inflammation of the nerves. It is important for patients to review the results of a cerebrospinal fluid analysis with their doctor and to discuss any treatment plans.

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American Academy of Neurology. 1080 Montreal Ave., St. Paul, MN 55116. (612) 695-1940. <<http://www.aan.com>>.

Martha Floberg Robbins

Cerebrovascular accident see **Stroke**

Cerebrovascular amyloidosis see **Cerebral amyloid angiopathy**

Cerumen impaction

Definition

Cerumen impaction is a condition in which earwax has become tightly packed in the external ear canal to the point that the canal is blocked.

Description

Cerumen impaction develops when earwax accumulates in the inner part of the ear canal and blocks the eardrum. It affects between 2–6% of the general population in the United States. Impaction does not happen under normal circumstances because cerumen is produced by glands in the outer part of the ear canal; it is not produced in the inner part. The cerumen traps sand or dust particles before they reach the ear drum. It also protects the outer part of the ear canal because it repels water. The slow movement of the outer layer of skin of the ear canal carries cerumen toward the outer opening of the ear. As the older cerumen reaches the opening of the ear, it dries out and falls away.

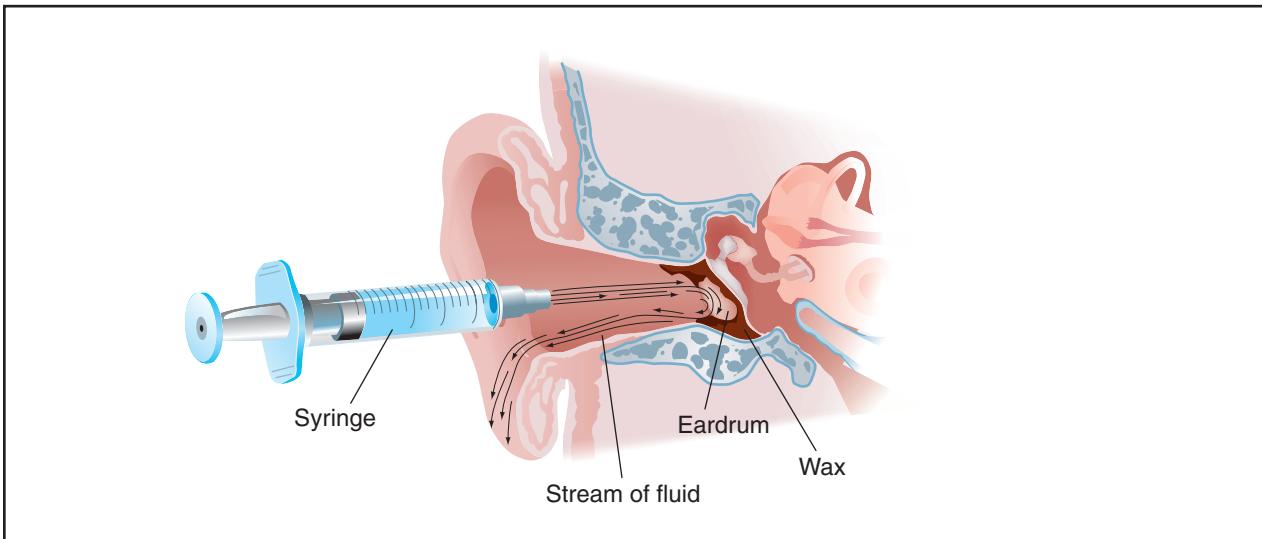
Causes and symptoms

Causes

Cerumen is most likely to become impacted when it is pushed against the eardrum by cotton-tipped applicators, hair pins, or other objects that people put in their ears; and when it is trapped against the eardrum by a hearing aid. Less common causes of cerumen impaction include overproduction of earwax by the glands in the ear canal, or an abnormally shaped ear canal.

Symptoms

The most important symptom of cerumen impaction is partial loss of hearing. Other symptoms are **itching**, **tinnitus** (noise or ringing in the ears), a sensation of fullness in the ear, and **pain**.



Ear wax is removed by flushing the ear canal with warm fluid. (Illustration by Argosy, Inc.)

Diagnosis

The diagnosis of impacted cerumen is usually made by examining the ear canal and eardrum with an otoscope, an instrument with a light attached that allows the doctor to look into the canal.

Treatment

Irrigation is the most common method of removing impacted cerumen. It involves washing out the ear canal with water from a commercial irrigator or a syringe with a catheter attached. Although some doctors use Water Piks to remove cerumen, most do not recommend them because the stream of water is too forceful and may damage the eardrum. The doctor may add a small amount of alcohol, hydrogen peroxide, or other antiseptic. The water must be close to body temperature; if it is too cold or too warm, the patient may feel dizzy or nauseated. After the ear has been irrigated, the doctor will apply antibiotic ear drops to protect the ear from infection.

Irrigation should not be used to remove cerumen if the patient's eardrum is ruptured or missing; if the patient has a history of chronic **otitis media** (inflammation of the middle ear) or a myringotomy (cutting the eardrum to allow fluid to escape from the middle ear); or if the patient has hearing in only one ear.

If irrigation cannot be used or fails to remove the cerumen, the patient is referred to an ear, nose, and throat (ENT) specialist. The specialist can remove the wax with a vacuum device or a curette, which is a small scoop-shaped surgical instrument.

Some doctors prescribe special ear drops, such as Cerumenex, to soften the wax. The most common side

effect of Cerumenex is an allergic skin reaction. Over-the-counter wax removal products include Debrox or Murine Ear Drops. A 3% solution of hydrogen peroxide may also be used. These products are less likely to irritate the skin of the ear.

Prognosis

In most cases, impacted cerumen is successfully removed by irrigation with no lasting side effects. Irrigation can, however, lead to infection of the outer or the middle ear if the patient has a damaged or absent ear drum. Patients who try to remove earwax themselves with hair pins or similar objects run the risk of perforating the ear drum or damaging the fragile skin covering the ear canal, causing bleeding and the risk of infection.

Prevention

The best method of cleaning the external ear is to wipe the outer opening with a damp washcloth folded over the index finger, without going into the ear canal itself. Two techniques have been recommended to prevent cerumen from reaccumulating in the ear. The patient may place two or three drops of mineral oil into each ear once a week, allow it to remain for two or three minutes, and rinse it out with warm water; or place two drops of Domeboro otic solution in each ear once a week after showering.

Patients who wear **hearing aids** should have their ears examined periodically for signs of cerumen accumulation.

Resources

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KEY TERMS

- Cerumen**—The medical term for earwax.
- Curette**—A small scoop-shaped surgical instrument that can be used to remove cerumen if irrigation does not work or cannot be used.
- Impaction**—A condition in which earwax has become tightly packed in the outer ear to the point that the external ear canal is blocked.
- Irrigation**—The technique of removing cerumen from the ear canal by flushing it with water.
- Myringotomy**—Surgical cutting of the ear drum to allow fluid to escape from the middle ear.
- Otitis media**—Inflammation of the middle ear. Patients who have had recurrent otitis media should not have cerumen removed by irrigation.
- Tinnitus**—A sensation of noise or ringing in the ears. Tinnitus may be a symptom of cerumen impaction.

Description

In the United States, cervical cancer is the fifth most common cancer among women aged 35–54, and the third most common cancer of the female reproductive tract. In some developing countries, it is the most common type of cancer. It generally begins as an abnormality in the cells on the outside of the cervix. The cervix is the lower part or neck of the uterus (womb). It connects the body of the uterus to the vagina (birth canal).

Approximately 90% of cervical cancers are squamous cell carcinomas. This type of cancer originates in the thin, flat, squamous cells on the surface of the ectocervix, the part of the cervix that is next to the vagina. (Squamous cells are the thin, flat cells of the surfaces of the skin and cervix and linings of various organs.) Another 10% of cervical cancers are of the adenocarcinoma type. This cancer originates in the mucus-producing cells of the inner or endocervix, near the body of the uterus. Occasionally, the cancer may have characteristics of both types and is called adenosquamous carcinoma or mixed carcinoma.

The initial changes that may occur in some cervical cells are not cancerous. However, these precancerous cells form a lesion called dysplasia or a squamous intraepithelial lesion (SIL), since it occurs within the epithelial or outer layer of cells. These abnormal cells can also be described as cervical intraepithelial neoplasia (CIN). Moderate to severe dysplasia may be called carcinoma in situ or non-invasive cervical cancer.

Dysplasia is a common condition and the abnormal cells often disappear without treatment. However, these precancerous cells can become cancerous. This may take years, although it can happen in less than a year. Eventually, the abnormal cells start to grow uncontrollably into the deeper layers of the cervix, becoming an invasive cervical cancer.

Although cervical cancer used to be one of the most common causes of cancer **death** among American women, in the past 40 years there has been a 75% decrease in mortality. This is primarily due to routine screening with Pap tests (Pap smear), to identify precancerous and early-invasive stages of cervical cancer. With treatment, these conditions have a cure rate of nearly 100%.

Worldwide, there are more than 400,000 new cases of cervical cancer diagnosed each year. The American Cancer Society (ACS) estimates that there will be 12,900 new cases of invasive cervical cancer diagnosed in the United States in 2001. More than one million women will be diagnosed with a precancerous lesion or non-invasive cancer of the cervix.

Older women are at the highest risk for cervical cancer. Although girls under the age of 15 rarely develop this

Cervical cancer

Definition

Cervical **cancer** is a disease in which the cells of the cervix become abnormal and start to grow uncontrollably, forming tumors.

cancer, the risk factor begins to increase in the late teens. Rates for carcinoma in situ peak between the ages of 20 and 30. In the United States, the incidence of invasive cervical cancer increases rapidly with age for African American women over the age of 25. The incidence rises more slowly for Caucasian women. However, women over age 65 account for more than 25% of all cases of invasive cervical cancer.

The incidence of cervical cancer is highest among poor women and among women in developing countries. In the United States, the death rates from cervical cancer are higher among Hispanic, Native American, and African American women than among Caucasian women. These groups of women are much less likely to receive regular Pap tests. Therefore, their cervical cancers usually are diagnosed at a much later stage, after the cancer has spread to other parts of the body.

Causes and symptoms

Human papilloma virus

Infection with the common human papilloma virus (HPV) is a cause of approximately 90% of all cervical cancers. There are more than 80 types of HPV. About 30 of these types can be transmitted sexually, including those that cause **genital warts** (papillomas). About half of the sexually transmitted HPVs are associated with cervical cancer. These “high-risk” HPVs produce a protein that can cause cervical epithelial cells to grow uncontrollably. The virus makes a second protein that interferes with tumor suppressors that are produced by the human immune system. The HPV-16 strain is thought to be a cause of about 50% of cervical cancers.

More than six million women in the United States have persistent HPV infections, for which there is no cure. Nevertheless, most women with HPV do not develop cervical cancer.

Symptoms of invasive cervical cancer

Most women do not have symptoms of cervical cancer until it has become invasive. At that point, the symptoms may include:

- unusual vaginal discharge
- light vaginal bleeding or spots of blood outside of normal menstruation
- **pain** or vaginal bleeding with sexual intercourse
- post-menopausal vaginal bleeding

Once the cancer has invaded the tissue surrounding the cervix, a woman may experience pain in the pelvic region and heavy bleeding from the vagina.

Diagnosis

The Pap test

Most often, cervical cancer is first detected with a **Pap test** that is performed as part of a regular pelvic examination. The vagina is spread with a metal or plastic instrument called a speculum. A swab is used to remove mucus and cells from the cervix. This sample is sent to a laboratory for microscopic examination.

The Pap test is a screening tool rather than a diagnostic tool. It is very efficient at detecting cervical abnormalities. The Bethesda System commonly is used to report Pap test results. A negative test means that no abnormalities are present in the cervical tissue. A positive Pap test describes abnormal cervical cells as low-grade or high-grade SIL, depending on the extent of dysplasia. About 5–10% of Pap tests show at least mild abnormalities. However, a number of factors other than cervical cancer can cause abnormalities, including inflammation from bacteria or yeast infections. A few months after the infection is treated, the Pap test is repeated.

Biopsy

Following an abnormal Pap test, a **colposcopy** is usually performed. The physician uses a magnifying scope to view the surface of the cervix. The cervix may be coated with an iodine solution that causes normal cells to turn brown and abnormal cells to turn white or yellow. This is called a Schiller test. If any abnormal areas are observed, a colposcopic biopsy may be performed. A biopsy is the removal of a small piece of tissue for microscopic examination by a pathologist.

Other types of cervical biopsies may be performed. An endocervical curettage is a biopsy in which a narrow instrument called a curette is used to scrape tissue from inside the opening of the cervix. A cone biopsy, or conization, is used to remove a cone-shaped piece of tissue from the cervix. In a cold knife cone biopsy, a surgical scalpel or laser is used to remove the tissue. A loop electrosurgical excision procedure (LEEP) is a cone biopsy using a wire that is heated by an electrical current. Cone biopsies can be used to determine whether abnormal cells have invaded below the surface of the cervix. They also can be used to treat many precancers and very early cancers. Biopsies may be performed with a local or general anesthetic. They may cause cramping and bleeding.

Diagnosing the stage

Following a diagnosis of cervical cancer, various procedures may be used to stage the disease (determine

how far the cancer has spread). For example, additional pelvic exams may be performed under anesthesia.

There are several procedures for determining if cervical cancer has invaded the urinary tract. With **cystoscopy**, a lighted tube with a lens is inserted through the urethra (the urine tube from the bladder to the exterior) and into the bladder to examine these organs for cancerous cells. Tissue samples may be removed for microscopic examination by a pathologist. **Intravenous urography** (intravenous pyelogram or IVP) is an x ray of the urinary system, following the injection of special dye. The kidneys remove the dye from the bloodstream and the dye passes into the ureters (the tubes from the kidneys to the bladder) and bladder. IVP can detect a blocked ureter, caused by the spread of cancer to the pelvic lymph nodes (small glands that are part of the immune system).

A procedure called proctoscopy or **sigmoidoscopy** is similar to cystoscopy. It is used to determine whether the cancer has spread to the rectum or lower large intestine.

Computed tomography (CT or CAT) scans, ultrasound, or other imaging techniques may be used to determine the spread of cancer to various parts of the body. With a CT scan, an x-ray beam rotates around the body, taking images from various angles. It is used to determine if the cancer has spread to the lymph nodes. **Magnetic resonance imaging** (MRI), which uses a magnetic field to image the body, sometimes is used for evaluating the spread of cervical cancer. Chest x rays may be used to detect cervical cancer that has spread to the lungs.

Treatment

Following a diagnosis of cervical cancer, the physician takes a medical history and performs a complete **physical examination**. This includes an evaluation of symptoms and risk factors for cervical cancer. The lymph nodes are examined for evidence that the cancer has spread from the cervix. The choice of treatment depends on the clinical stage of the disease.

The FIGO system of staging

The International Federation of Gynecologists and Obstetricians (FIGO) system usually is used to stage cervical cancer:

- Stage 0: Carcinoma in situ; non-invasive cancer that is confined to the layer of cells lining the cervix
- Stage I: Cancer that has spread into the connective tissue of the cervix but is confined to the uterus
- Stage IA: Very small cancerous area that is visible only with a microscope

- Stage IA1: Invasion area is less than 3 mm (0.13 in) deep and 7 mm (0.33 in) wide
- Stage IA2: Invasion area is 3–5 mm (0.13–0.2 in) deep and less than 7 mm (0.33 in) wide
- Stage IB: Cancer can be seen without a microscope or is deeper than 5 mm (0.2 in) or wider than 7 mm (0.33 in)
- Stage IB1: Cancer is no larger than 4 cm (1.6 in)
- Stage IB2: Stage IB cancer is larger than 4 cm (1.6 in)
- Stage II: Cancer has spread from the cervix but is confined to the pelvic region
- Stage IIA: Cancer has spread to the upper region of the vagina, but not to the lower one-third of the vagina
- Stage IIB: Cancer has spread to the parametrial tissue adjacent to the cervix
- Stage III: Cancer has spread to the lower one-third of the vagina or to the wall of the pelvis and may be blocking the ureters
- Stage IIIA: Cancer has spread to the lower vagina but not to the pelvic wall
- Stage IIIB: Cancer has spread to the pelvic wall and/or is blocking the flow of urine through the ureters to the bladder
- Stage IV: Cancer has spread to other parts of the body
- Stage IVA: Cancer has spread to the bladder or rectum
- Stage IVB: Cancer has spread to distant organs such as the lungs
- Recurrent: Following treatment, cancer has returned to the cervix or some other part of the body

In addition to the stage of the cancer, factors such as a woman's age, general health, and preferences may influence the choice of treatment. The exact location of the cancer within the cervix and the type of cervical cancer also are important considerations.

Treatment of precancer and carcinoma in situ

Most low-grade SILs that are detected with Pap tests revert to normal without treatment. Most high-grade SILs require treatment. Treatments to remove precancerous cells include:

- cold knife cone biopsy
- LEEP
- cryosurgery (freezing the cells with a metal probe)
- cauterization or diathermy (burning off the cells)
- laser surgery (burning off the cells with a laser beam)

These methods also may be used to treat cancer that is confined to the surface of the cervix (stage 0) and other

early-stage cervical cancers in women who may want to become pregnant. They may be used in conjunction with other treatments. These procedures may cause bleeding or cramping. All of these treatments require close follow-up to detect any recurrence of the cancer.

Surgery

A simple **hysterectomy** is used to treat some stages 0 and IA cervical cancers. Usually only the uterus is removed, although occasionally the fallopian tubes and ovaries are removed as well. The tissues adjoining the uterus, including the vagina, remain intact. The uterus may be removed either through the abdomen or the vagina.

In a radical hysterectomy, the uterus and adjoining tissues, including the ovaries, the upper region (1 in) of the vagina near the cervix, and the pelvic lymph nodes, are all removed. A radical hysterectomy usually involves abdominal surgery. However, it can be performed vaginally, in combination with a laparoscopic pelvic lymph node dissection. With **laparoscopy**, a tube is inserted through a very small surgical incision for the removal of the lymph nodes. These operations are used to treat stages IA2, IB, and IIA cervical cancers, particularly in young women. Following a hysterectomy, the tissue is examined to see if the cancer has spread and requires additional radiation treatment. Women who have had hysterectomies cannot become pregnant, but complications from a hysterectomy are rare.

If cervical cancer recurs following treatment, a pelvic exenteration (extensive surgery) may be performed. This includes a radical hysterectomy, with the additional removal of the bladder, rectum, part of the colon, and/or all of the vagina. Such operations require the creation of new openings for the urine and feces. A new vagina may be created surgically. Often the clitoris and other outer genitals are left intact.

Recovery from a pelvic exenteration may take six months to two years. This treatment is successful with 40–50% of recurrent cervical cancers that are confined to the pelvis. If the recurrent cancer has spread to other organs, radiation or **chemotherapy** may be used to alleviate some of the symptoms.

Radiation

Radiation therapy, which involves the use of high-dosage x rays or other high-energy waves to kill cancer cells, often is used for treating stages IB, IIA, and IIB cervical cancers, or in combination with surgery. With external-beam radiation therapy, the rays are focused on the pelvic area from a source outside the body. With implant or internal radiation therapy, a pellet of radioactive material is placed internally, near the tumor. Alternatively, thin needles may be used to insert the radioactive material directly into the tumor.

Radiation therapy to the pelvic region can have many side effects:

- skin reaction in the area of treatment
- fatigue
- upset stomach and loose bowels
- vaginal stenosis (narrowing of the vagina due to build-up of scar tissue) leading to painful sexual intercourse
- premature **menopause** in young women
- problems with urination

Chemotherapy

Chemotherapy, the use of one or more drugs to kill cancer cells, is used to treat disease that has spread beyond the cervix. Most often it is used following surgery or radiation treatment. Stages IIB, III, IV, and recurrent cervical cancers usually are treated with a combination of external and internal radiation and chemotherapy. The common drugs used for cervical cancer are cisplatin, ifosfamide, and fluorouracil. These may be injected or taken by mouth. The National Cancer Institute recommends that chemotherapy with cisplatin be considered for all women receiving radiation therapy for cervical cancer.

The side effects of chemotherapy depend on a number of factors, including the type of drug, the dosage, and the length of the treatment. Side effects may include:

- nausea and vomiting
- fatigue
- changes in appetite
- hair loss
- mouth or vaginal sores
- infections
- menstrual cycle changes
- premature menopause
- **infertility**
- bleeding or anemia (low red blood cell count)

With the exception of menopause and infertility, most of the side effects are temporary.

Alternative treatment

Biological therapy sometimes is used to treat cervical cancer, either alone or in combination with chemotherapy. Treatment with the immune-system pro-

KEY TERMS

Adenocarcinoma—Cervical cancer that originates in the mucus-producing cells of the inner or endocervix.

Biopsy—Removal of a small sample of tissue for examination under a microscope; used for the diagnosis and treatment of cervical cancer and precancerous conditions.

Carcinoma in situ—Cancer that is confined to the cells in which it originated and has not spread to other tissues.

Cervical intraepithelial neoplasia (CIN)—Abnormal cell growth on the surface of the cervix.

Cervix—Narrow, lower end of the uterus forming the opening to the vagina.

Colposcopy—Diagnostic procedure using a hollow, lighted tube (colposcope) to look inside the cervix and uterus.

Conization—Cone biopsy; removal of a cone-shaped section of tissue from the cervix for diagnosis or treatment.

Dysplasia—Abnormal cellular changes that may become cancerous.

Endocervical curettage—Biopsy performed with a curette to scrape the mucous membrane of the cervical canal.

Human papilloma virus (HPV)—Virus that causes abnormal cell growth (warts or papillomas); some types can cause cervical cancer.

Hysterectomy—Removal of the uterus.

Interferon—Potent immune-defense protein produced by viral-infected cells; used as an anti-cancer and anti-viral drug.

Laparoscopy—Laparoscopic pelvic lymph node dissection; insertion of a tube through a very small surgical incision to remove lymph nodes.

Loop electrosurgical excision procedure (LEEP)—Cone biopsy performed with a wire that is heated by electrical current.

Lymph nodes—Small round glands, located throughout the body, that filter the lymphatic fluid; part of the body's immune defense.

Pap test—Pap smear; removal of cervical cells to screen for cancer.

Pelvic exenteration—Extensive surgery to remove the uterus, ovaries, pelvic lymph nodes, part or all of the vagina, and the bladder, rectum, and/or part of the colon.

Squamous cells—Thin, flat cells on the surfaces of the skin and cervix and linings of various organs.

Squamous intraepithelial lesion (SIL)—Abnormal growth of squamous cells on the surface of the cervix.

Vaginal stenosis—Narrowing of the vagina due to a build-up of scar tissue.

tein interferon is used to boost the immune response. Biological therapy can cause temporary flu-like symptoms and other side effects.

Some research suggests that vitamin A (carotene) may help to prevent or stop cancerous changes in cells such as those on the surface of the cervix. Other studies suggest that **vitamins C** and E may reduce the risk of cervical cancer.

Prognosis

For cervical cancers that are diagnosed in the pre-invasive stage, the five-year-survival rate is almost 100%. When cervical cancer is detected in the early invasive stages, approximately 91% of women survive five years or more. Stage IVB cervical cancer is not considered to

be curable. The five-year-survival rate for all cervical cancers combined is about 70%. The death rate from cervical cancer continues to decline by about 2% each year. Women over age 65 account for 40–50% of all deaths from cervical cancer.

Prevention

Viral infections

Most cervical cancers are preventable. More than 90% of women with cervical cancer are infected with HPV. HPV infection is the single most important risk factor. This is particularly true for young women because the cells lining the cervix do not fully mature until age 18. These immature cells are more susceptible to cancer-causing agents and viruses.

Since HPV is a sexually-transmitted infection, sexual behaviors can put women at risk for HPV infection and cervical cancer. These behaviors include:

- sexual intercourse at age 16 or younger
- partners who began having intercourse at a young age
- multiple sexual partners
- sexual partners who have had multiple partners ("high-risk males")
- a partner who has had a previous sexual partner with cervical cancer

HPV infection may not produce any symptoms, so sexual partners may not know that they are infected. However, Pap tests can detect the infection. Condoms do not necessarily prevent HPV infection.

Infection with the human **immunodeficiency** virus (HIV) that causes acquired immunodeficiency syndrome (**AIDS**) is a risk factor for cervical cancer. Women who test positive for HIV may have impaired immune systems that cannot correct precancerous conditions. Furthermore, sexual behavior that puts women at risk for HIV infection, also puts them at risk for HPV infection. There is some evidence suggesting that another sexually transmitted virus, the **genital herpes** virus, also may be involved in cervical cancer.

Smoking

Smoking may double the risk of cervical cancer. Chemicals produced by tobacco smoke can damage the DNA of cervical cells. The risk increases with the number of years a woman smokes and the amount she smokes.

Diet and drugs

Diets that are low in fruits and vegetables increase the risk of cervical cancer. Women also have an increased risk of cervical cancer if their mothers took the drug diethylstilbestrol (DES) while they were pregnant. This drug was given to women between 1940 and 1971 to prevent miscarriages. Some statistical studies have suggested that the long-term use of **oral contraceptives** may slightly increase the risk of cervical cancer.

Pap tests

Most cases of cervical cancers are preventable, since they start with easily detectable precancerous changes. Therefore, the best prevention for cervical cancer is a regular Pap test. When precancerous changes are detected, appropriate treatment can prevent the development of invasive cancer. The ACS recommends that women have annual Pap tests beginning when they first start having sex or at age 18. Women who are past

menopause or some women with hysterectomies continue to require Pap tests.

The National Breast and Cervical Cancer Early Detection Program provides free or low-cost Pap tests and treatment for women without health insurance, for older women, and for members of racial and ethnic minorities. The program is administered through individual states, under the direction of the Centers for Disease Control and Prevention.

Special concerns

If a woman is diagnosed with very early-stage (IA) cervical cancer while pregnant, the physician usually will recommend a hysterectomy after the baby is born. For later-stage cancers, the **pregnancy** is terminated or the baby is removed by **cesarean section** as soon as it can survive outside the womb. This is followed by a hysterectomy and/or radiation treatment. For the most advanced stages of cervical cancer, treatment is initiated despite the pregnancy.

Many women with cervical cancer have hysterectomies, which are major surgeries. Although normal activities, including sexual intercourse, can be resumed in four-eight weeks, a woman may have emotional problems following a hysterectomy. A strong support system can help with these difficulties.

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Centers for Disease Control and Prevention. National Center for Chronic Disease Prevention and Health Promotion. Mail Stop K-64. 4770 Buford Highway NE, Atlanta, GA 30341-3717. (770) 488-4751. (888) 842-6355. <<http://www.cdc.gov/cancer>>.

EyesOnThePrize.Org. 446 S. Anaheim Hills Road, #108, Anaheim Hills, CA 92807. <<http://www.eyesontheprize.org>>.

Gynecologic Cancer Foundation. 401 North Michigan Avenue, Chicago, IL 60611. (800) 444-4441. (312) 644-6610. <<http://www.wcn.org/gcf/>>.

National Cancer Institute. Public Inquiries Office, Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD

20892-2580. (800) 4-CANCER. <<http://www.nci.nih.gov/>>. <<http://cancernet.nci.nih.gov>>.

National Cervical Cancer Coalition. 16501 Sherman Way, Suite #110, Van Nuys, CA 91406. (800) 685-5531. (818) 909-3849. <<http://www.nccc-online.org/>>.

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Cervical conization

Definition

Cervical conization is both a diagnostic and treatment tool used to detect and treat abnormalities of the cervix. It is also known as a cone biopsy or cold knife cone biopsy.

Purpose

Cervical conization is performed if the results of a cervical biopsy have found a precancerous condition in the cervix. The cervix is the small cylindrical organ at the lower part of the uterus, which separates the uterus from the vagina. Cervical conization also may be performed if there is an abnormal cervical smear test (**PAP test**). A biopsy is a diagnostic test in which tissue or cells are removed from the body and examined under a microscope, primarily to look for **cancer** or other abnormalities.

Precautions

As with any operation that is performed under general anesthesia, the patient must not eat or drink anything for six to eight hours before surgery.

Description

The patient lies on the table with her legs raised in stirrups, similar to the position when having a PAP test.

KEY TERMS

Biopsy—The removal of a small piece of living tissue for examination under a microscope.

PAP test—The short term for Papanicolaou test, this procedure tests a smear of cellular material scraped from the cervix and examined under a microscope to detect abnormal cells.

The patient is given general anesthesia, and the vagina is held open with an instrument called a speculum. Using a scalpel or laser the doctor removes a cone-shaped piece of the cervix containing the area with abnormal cells. The resulting crater is repaired by stitching flaps of tissue over the wound. Alternatively, the wound may be left open, and heat or freezing is used to stop bleeding.

Once the tissue has been removed, it is examined under a microscope for signs of cancer. If cancer is present, other tests will be needed. Surgery will be performed to remove the cervix and uterus (**hysterectomy**) and other treatments may be used as well. If the abnormal cells are precancerous, a laser can be used to destroy them.

Cold knife cone biopsy used to be the preferred treatment for removing abnormal cells in the cervix. Now, most cone biopsies are performed using **laser surgery**. Cold knife cone biopsy is generally used only for special situations. For example, if a biopsy did not remove all the abnormal cells, the cold knife cone procedure allows the physician to remove what's left.

Aftercare

An overnight stay in the hospital may be required. After the test, the patient may feel some cramps or discomfort for about a week. Women should not have sex, use tampons, or douche until after seeing their physician for a follow up appointment (a week or more after the procedure).

Risks

Because cone biopsies carry risks such as bleeding and problems with subsequent pregnancies, they have been replaced with newer technologies except in a few circumstances.

About one in 10 women experience bleeding from the vagina about two weeks after the biopsy. There is also a slight risk of infection or perforation of the uterus. In a few women, the cervical canal becomes narrowed or

completely blocked, which can later interfere with the movement of sperm. This can impair a woman's fertility.

If too much muscle tissue has been removed, the procedure can lead to an **incompetent cervix**, which can be a problem with subsequent pregnancies. An incompetent cervix cannot seal properly to maintain a **pregnancy**. If untreated, the condition increases the odds of **miscarriage or premature labor**.

Cervical conization also may temporarily alter cervical cells, which can make a Pap smear test hard to interpret accurately for three or four months.

Normal results

This procedure is only performed if an abnormality is known or suspected.

Abnormal results

The presence of precancerous or cancerous cells in the cervix.

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ORGANIZATIONS

Cancer Information Service. (800) 4-CANCER. <<http://www.rex.nci.nih.gov>>.

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Cervical disk disease

Definition

Cervical disk disease refers to a gradual deterioration of the spongy disks in the top part of the spine.

Description

The spine is made up of 33 bones called vertebrae separated by spongy rings of elastic material. These rings, known as disks, are often compared to **shock absorbers** because they help to cushion the vertebrae.

Just as importantly, they also make it possible to turn the head and neck. Over time, these disks slowly become flattened and less elastic due to everyday wear and tear. When this process occurs in the disks of the neck, it is referred to as cervical disk disease. Other general terms for this process include degenerative disk disease and intervertebral disk disease.

Cervical disk disease affects everyone to some degree, often without causing any bothersome symptoms. However, this condition can also lead to specific problems related to nerve functioning. For example, the outer edge of a disk may tear, allowing the gelatinous material inside to bulge outward (**herniated disk**). This can put pressure on nerves that exit the spine. Two adjacent vertebrae may rub together (sometimes resulting in bone spurs) that can also pinch these nerves. In other cases, the inner part of the ring may push on the spinal cord itself, which passes through the disk. Any of these situations can cause **pain** and limit movement. While symptoms primarily affect the neck, they can also occur in other parts of the body.

Causes and symptoms

Cervical disk disease is a gradual process that occurs with **aging**, though poor posture, repeated lifting, and tobacco use can hasten its course. Symptoms include pain when moving the neck and limited neck movement. The condition can also affect the hand, shoulder, and arm resulting in pain, numbness/tingling, and weakness. If the spinal cord itself is affected, these symptoms may occur in the legs. Loss of bowel or bladder control may also occur.

Diagnosis

Cervical disk disease is typically diagnosed by an orthopedist or a neurologist. After taking a medical history and conducting a **physical examination**, the doctor will recommend an imaging procedure to gather more information about the nature of the problem. This may include a CT scan, an MRI, or **myelography**. In addition, an electromyogram (EMG) may be used to evaluate the functioning of nerves in the arms, hands, or legs. Cervical disk disease is typically covered by medical insurance.

Treatment

Treatment usually involves physical therapy, several weeks of drug therapy with **nonsteroidal anti-inflammatory drugs** (NSAIDs), and limited use of a cervical collar (to reduce neck movement). Neck **traction** and **heat treatments** may also be recommended. In some

KEY TERMS

Bone spur—An overgrowth of bone.

Cervical—Relating to the top part of the spine that is composed of the seven vertebrae of the neck and the disks that separate them.

Computed tomography (CT) scan—An imaging procedure that produces a three-dimensional picture of organs or structures inside the body.

Myelography—An imaging procedure involving the injection of a radioactive dye into the fluid surrounding the spine. A myelography can be used to detect herniated disks, nerve root damage, and other problems affecting the cervical spine.

Neurologist—A doctor who specializes in disorders of the brain and central nervous system.

Orthopedist—A doctor who specializes in disorders of the musculoskeletal system.

Magnetic resonance imaging—A type of imaging that uses magnetic fields to generate a picture of internal structures.

cases, steroids or anesthetic drugs may be injected into the spinal canal to help alleviate symptoms. Aside from these measures, maintaining good posture and placing a pillow under the neck and head during sleep can be helpful. Treatment may last anywhere from several weeks to three months or more. Neck surgery is not usually advised unless other therapies have failed.

Alternative treatment

Acupuncture, therapeutic massage, and **yoga** are believed by some practitioners of alternative medicine to have generalized pain-relieving effects. However, any therapy that involves manipulating the neck is not recommended and should be approved by the primary doctor beforehand.

Prognosis

In most people symptoms go away within three months if not sooner. A smaller number may require surgery to correct the problem.

Prevention

While some degree of disk degeneration is inevitable, people can reduce their risk by practicing good posture (during sitting, standing, and lifting), performing neck-

stretching exercises, maintaining an ideal weight, and quitting **smoking**.

Resources

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ORGANIZATIONS

American Academy of Orthopaedic Surgeons. 6300 North River Road, Rosemont, IL 60018-4262. (800) 346-2267. <<http://www.aaos.org>>.

Greg Annussek

Cervical osteoarthritis see **Cervical spondylosis**

Cervical spondylosis

Definition

Cervical spondylosis refers to common age-related changes in the area of the spine at the back of the neck. With age, the vertebrae (the component bones of the spine) gradually form bone spurs, and their shock-absorbing disks slowly shrink. These changes can alter the alignment and stability of the spine. They may go unnoticed, or they may produce problems related to pressure on the spine and associated nerves and blood vessels. This pressure can cause weakness, numbness, and **pain** in various areas of the body. In severe cases, walking and other activities may be compromised.

Description

As it runs from the brain down the back, the spinal cord is protected by ringlike bones, called vertebrae, stacked one upon the other. The vertebrae are not in direct contact with one another, however. The intervening spaces are filled with structures called disks. The disks are made up of a tough, fibrous outer tissue with an inner core of elastic or gel-like tissue.

One of the most important functions of disks is protecting the vertebrae and the nerves and blood vessels between the vertebrae. The disks also lend flexibility to

the spinal cord, facilitating movements such as turning the head or bending the neck. As people age, disks gradually become tougher and more unyielding. Disks also shrink with age, which reduces the amount of padding between the vertebrae.

As the amount of padding shrinks, the spine loses stability. The vertebrae react by constructing osteophytes, commonly known as bone spurs. There are seven vertebrae in the neck; development of osteophytes on these bones is sometimes called cervical **osteoarthritis**. Osteophytes may help to stabilize the degenerating backbone and help protect the spinal cord.

By age 50, 25–50% of people develop cervical spondylosis; by 75 years of age, it is seen in at least 70% of people. Although shrunken vertebral disks, osteophyte growth, and other changes in their cervical spine may exist, many of these people never develop significant problems.

However, about 50% of people over age 50 experience neck pain and stiffness due to cervical spondylosis. Of these people, 25–40% have at least one episode of cervical radiculopathy, a condition that arises when osteophytes compress nerves between the vertebrae. Another potential problem occurs if osteophytes, degenerating disks, or shifting vertebrae narrow the spinal canal. This pressure compresses the spinal cord and its blood vessels, causing cervical spondylitic myelopathy, a disorder in which large segments of the spinal cord are damaged. This disorder affects fewer than 5% of people with cervical spondylosis. Symptoms of both cervical spondylitic myelopathy and cervical radiculopathy may be present in some people.

Causes and symptoms

As people age, shrinkage of the vertebral disks prompts the vertebrae to form osteophytes to stabilize the back bone. However, the position and alignment of the disks and vertebrae may shift despite the osteophytes. Symptoms may arise from problems with one or more disks or vertebrae.

Osteophyte formation and other changes do not necessarily lead to symptoms, but after age 50, half of the population experiences occasional neck pain and stiffness. As disks degenerate, the cervical spine becomes less stable, and the neck is more vulnerable to injuries, including muscle and ligament strains. Contact between the edges of the vertebrae can also cause pain. In some people, this pain may be referred—that is, perceived as occurring in the head, shoulders, or chest, rather than the neck. Other symptoms may include vertigo (a type of **dizziness**) or ringing in the ears.

The neck pain and stiffness can be intermittent, as can symptoms of radiculopathy. Radiculopathy refers to compression on the base, or root, of nerves that lead

away from the spinal cord. Normally, these nerves fit comfortably through spaces between the vertebrae. These spaces are called intervertebral foramina. As the osteophytes form, they can impinge on this area and gradually make the fit between the vertebrae too snug.

The poor fit increases the chances that a minor incident, such as overdoing normal activities, may place excess pressure on the nerve root, sometimes referred to as a pinched nerve. Pressure may also accumulate as a direct consequence of osteophyte formation. The pressure on the nerve root causes severe shooting pain in the neck, arms, shoulder, and/or upper back, depending on which nerve roots of the cervical spine are affected. The pain is often aggravated by movement, but in most cases, symptoms resolve within four-six weeks.

Cervical spondylosis can cause cervical spondylitic myelopathy through stenosis- or osteophyte-related pressure on the spinal cord. **Spinal stenosis** is a narrowing of the spinal canal—the area through the center of the vertebral column occupied by the spinal cord. Stenosis occurs because of misaligned vertebrae and out-of-place or degenerating disks. The problems created by spondylosis can be exacerbated if a person has a naturally narrow spinal canal. Pressure against the spinal cord can also be created by osteophytes forming on the inner surface of vertebrae and pushing against the spinal cord. Stenosis or osteophytes can compress the spinal cord and its blood vessels, impeding or choking off needed nutrients to the spinal cord cells; in effect, the cells starve to **death**.

With the death of these cells, the functions that they once performed are impaired. These functions may include conveying sensory information to the brain or transmitting the brain's commands to voluntary muscles. Pain is usually absent, but a person may experience leg numbness and an inability to make the legs move properly. Other symptoms can include clumsiness and weakness in the hands, stiffness and weakness in the legs, and spontaneous twitches in the legs. A person's ability to walk is affected, and a wide-legged, shuffling gait is sometimes adopted to compensate for the lack of sensation in the legs and the accompanying, realistic fear of falling. In very few cases, bladder control becomes a problem.

Diagnosis

Cervical spondylosis is often suspected based on the symptoms and their history. Careful neurological examination can help determine which nerve roots are involved, based on the location of the pain and numbness, and the pattern of weakness and changes in reflex responses. To confirm the suspected diagnosis, and to rule out other possibilities, imaging tests are ordered. The first test is an x ray. X rays reveal the presence of osteophytes, stenosis,

KEY TERMS

Alexander technique—A technique developed by Frederick Alexander that focuses on the variations in body posture, muscles, and breathing. Defects in these functions can lead to stress, nervous tension or possible loss of function.

Bone spur—Also called an osteophyte, it is an outgrowth or ridge that forms on a bone.

Cervical—Referring to structures within the neck.

Computed tomography myelography—This medical procedure combines aspects of computed tomography scanning and plain-film myelography. A CT scan is 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. Myelography involves injecting a water-soluble substance into the area around the spine to make it visible on x rays. In computed tomography myelography or CT myelography, the water-soluble substance is injected, but the imaging is done with a CT scan.

Disk—A ringlike structure that fits between the vertebrae in the spine to protect the bones, nerves, and blood vessels. The outer layer is a tough, fibrous tissue, and the inner core is composed of more elastic tissue.

Feldenkrais method—A therapy based on creating a good self image by correction and improvements of body movements.

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.

Myelopathy—A disorder in which the tissue of the spinal cord is diseased or damaged.

Orthosis—An external device, such as a splint or a brace, that prevents or assists movement.

Osteophyte—Also referred to as bone spur, it is an outgrowth or ridge that forms on a bone.

Radiculopathy—Sometimes referred to as a pinched nerve, it refers to compression of the nerve root—the part of a nerve between vertebrae. This compression causes pain to be perceived in areas to which the nerve leads.

Spine—A term for the backbone that includes the vertebrae, disks, and spinal cord as a whole.

Stenosis—A condition in which a canal or other passageway in the body is constricted.

Traction—A medical treatment that exerts a pulling or extending force. Used for cervical problems, it relieves pressure on structures between the vertebrae and muscular tension.

Vertebrae—The ringlike component bones of the spine.

constricted space between the vertebrae, and misalignment in the cervical spine—in short, an x ray confirms that a person has cervical spondylosis. To demonstrate that the condition is causing the symptoms, more details are needed. Other imaging tests, such as **magnetic resonance imaging (MRI)** and **computed tomography myelography**, help assess effects of cervical spondylosis on associated nerve tissue and blood vessels.

An MRI may be preferred, because it is a noninvasive procedure and does not require injecting a contrast medium as does computed tomography myelography. MRIs also have greater sensitivity for detecting disk problems and spinal cord involvement, and the test allows the physician to create images of a larger area from various angles. However, these images may not show enough detail about the vertebrae themselves. Computed tomography myelography yields a superior image of the bones involved in cervical spondylosis. Added ben-

efits include that it takes less time to perform and tends to be less expensive than an MRI. A good diagnosis may be reached with either a computed tomography myelography or an MRI, but sometimes complementary information from both tests is necessary. Nerve conduction velocity, electromyogram (EMG), and/or somatosensory evoked potential testing may help to confirm which nerve roots are involved.

Treatment

When possible, conservative treatment of symptoms is preferred. Conservative treatment begins with rest—either restricting normal activities to a less strenuous level or bed rest for three to five days. If rest is not adequate to relieve symptoms, a cervical orthosis may be prescribed, such as a soft cervical collar or stiffer neck brace to restrict neck movement and shift some of the head's weight from the neck to the shoulders. Cervical

traction may also be suggested, either at home with the advice of a physical therapist or in a health-care setting.

Pain is treated with **nonsteroidal anti-inflammatory drugs**, such as **aspirin** or ibuprofen. If these drugs are ineffective, a short-term prescription for **corticosteroids** or **muscle relaxants** may be given. For chronic pain, tricyclic antidepressants can be prescribed. Although these drugs were developed to treat depression, they are also effective in treating pain. Once any pain is resolved, exercises to strengthen neck muscle and preserve flexibility are prescribed.

If the pain is severe, a short treatment of epidural corticosteroids may be prescribed with discretion. A corticosteroid such as prednisone can be combined with an anaesthetic and injected with a long needle into the space between the damaged disk and the covering of the nerve and spinal cord. Injection into the cervical epidural space relieves severe pain that is not managed with conventional treatment. Frequent use of this treatment is not medically recommended and is used only if the more conservative therapy is not effective.

If pain is continuous and does not respond to conservative treatment, surgery may be suggested. Surgery is usually not recommended for neck pain, but it may be necessary to address radiculopathy and myelopathy. Surgery is particularly recommended for people who have already developed moderate to severe symptoms of myelopathy, although age or poor health may prohibit that recommendation. The specific details of the surgery depend on the structures involved, but the overall goal is to relieve pressure on the nerve root, spinal cord, or blood vessels and to stabilize the spine.

Alternative treatment

Alternative therapy is not meant to replace conventional medical treatment, but it can be a useful adjunct. Its main roles are to relieve tension, manage pain, and strengthen neck and back muscles. Massage is one way to relieve tension, and **yoga** provides the additional benefit of strengthening muscles. **Chiropractic** and **acupuncture** have been reported to relieve the pain associated with disk problems, although great care needs to be taken to avoid exacerbating them. Practitioners of the **Alexander technique** or the **Feldenkrais method** can provide instruction on correct posture and **exercise** that may help prevent further symptoms. Vitamin and mineral supplementation along with herbal therapies and **homeopathy** can help build and rebalance the weakened structure.

Prognosis

The gradual progression of cervical spondylosis cannot be stopped; however, it doesn't always cause

symptoms. For the individuals who do experience problems, conservative treatment is very effective in managing the symptoms. Nearly all people with neck pain, approximately 75% of persons with radiculopathy, and up to 50% of people with myelopathy find relief through therapy alone. For the remaining people with radiculopathy or myelopathy, surgery may be recommended. Surgery is deemed successful in 70–80% of cases.

Prevention

Since cervical spondylosis is part of the normal **aging** process, not much can be done to prevent it. It may be possible to ward off some or all of the symptoms by engaging in regular physical exercise and limiting occupational or recreational activities that place pressure on the head, neck, and shoulders. The best exercises for the health of the cervical spine are noncontact activities, such as swimming, walking, or yoga. Once symptoms have already developed, the emphasis is on symptom management rather than prevention.

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Julia Barrett

Cervicitis

Definition

Cervicitis is an inflammation of the cervix.

Description

Cervicitis is a inflammation of the cervix (the opening into the uterus). This inflammation can be chronic and may or may not have an identified cause.

KEY TERMS

Cryotherapy—Freezing the affected tissue.

Electrocoagulation—Using electrical current to cauterize the affected tissue.

LEEP—Loop Electrosurgical Excision Procedure.

Causes and symptoms

The most common cause of cervicitis is infection, either local or as a result of various **sexually transmitted diseases**, such as chlamydia or **gonorrhea**. Cervicitis can also be caused by birth control devices such as a cervical cap or diaphragm, or chemical exposure. Other risk factors include multiple sexual partners or cervical trauma following birth. In postmenopausal women, cervicitis is sometimes related to a lack of estrogen.

Although a woman may not notice any signs of infection, symptoms of cervicitis include the following:

- persistent unusual vaginal discharge
- abnormal bleeding, either between periods or following sexual intercourse
- painful sexual intercourse
- vaginal pain
- frequent need to urinate
- burning or **itching** in the vaginal area

Diagnosis

The standard method of diagnosing cervicitis is through a pelvic examination or a Pap smear. During the **pelvic exam**, the physician usually swabs the affected area, and then sends the tissue sample to a laboratory. The laboratory tries to identify the specific organism responsible for causing the cervicitis. A biopsy to take a sample of tissue from the affected area is sometimes required in order to rule out **cancer**. **Colposcopy**, a procedure used to look at the cervix under a microscope, may also be used to rule out cancer.

Treatment

The first course of treatment for cervicitis is usually **antibiotics**. If these medicines do not cure the cervicitis, other treatment options include:

- Loop Electrosurgical Excision Procedure (LEEP)
- cryotherapy

- electrocoagulation
- laser treatment

Prognosis

Cervicitis will usually be cured when the course of therapy is complete. Severe cases, however, may last for a few months, even after the therapy is complete. If the cervicitis was caused by a sexually transmitted disease, both partners should be treated with medication.

Prevention

Practicing safe sexual behavior, such as monogamy, is one way of lowering the prevalence of cervicitis. In addition, women who began sexual activity at a later age have been shown to have a lower incidence of cervicitis. Another recommendation is to use a latex **condom** consistently during intercourse. If the cervicitis is caused by any sexually transmitted disease, the patient is advised to notify all sexual partners.

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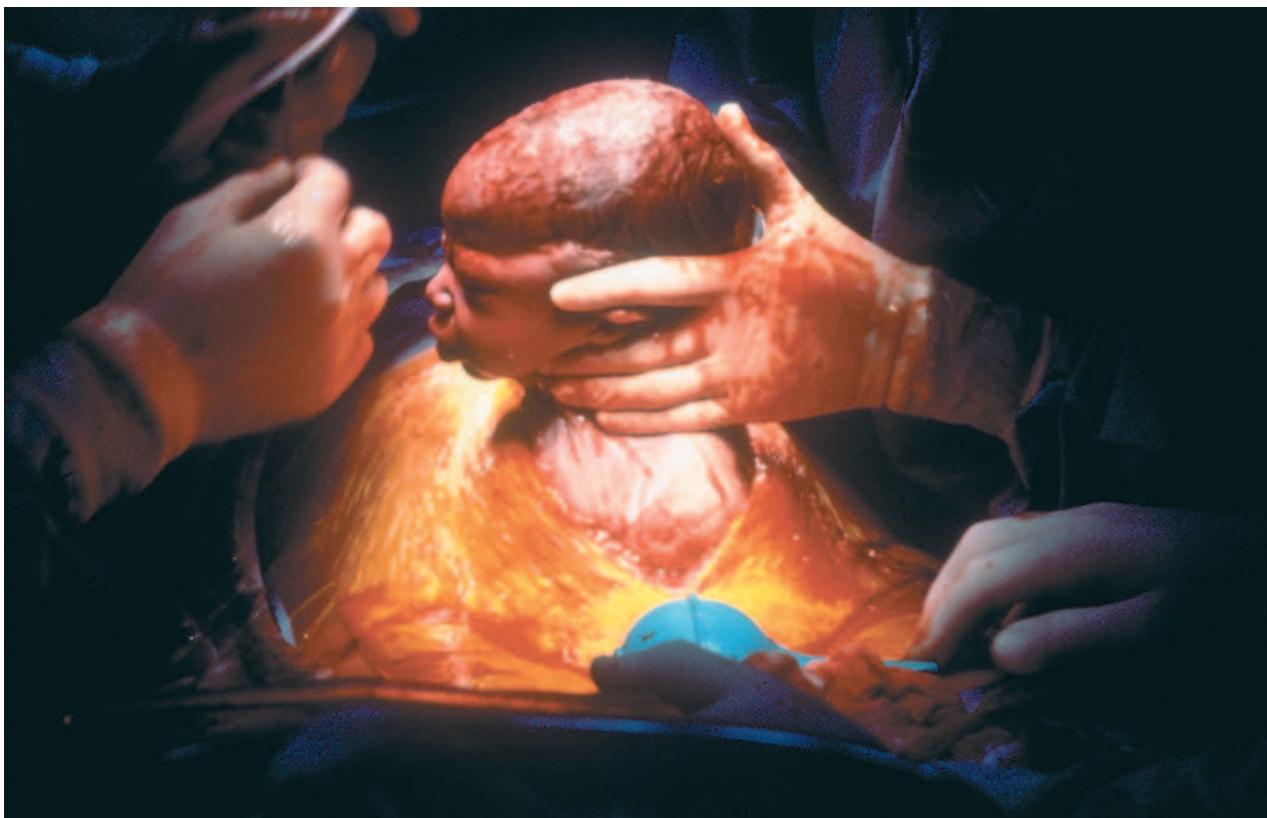
- American College of Obstetricians and Gynecologists. 409 12th Street, SW P.O. Box 96920, Washington, DC 20090-6920. (202) 863-2518. <<http://www.acog.org>>.

Kim Sharp, M.LN.

Cesarean section

Definition

A cesarean section is a surgical procedure in which incisions are made through a woman's abdomen and uterus to deliver her baby.



This baby is being delivered by cesarean section. (Photograph by John Smith, Custom Medical Stock Photo.)

Purpose

Cesarean sections, also called c-sections, are performed whenever abnormal conditions complicate labor and vaginal delivery, threatening the life or health of the mother or the baby. The procedure is performed in the United States on nearly one of every four babies delivered—more than 900,000 babies each year. The procedure is often used in cases where the mother has had a previous c-section. Dystocia, or difficult labor, is the other common cause of c-sections.

Difficult labor is commonly caused by one of the three following conditions: abnormalities in the mother's birth canal; abnormalities in the position of the fetus; or abnormalities in the labor, including weak or infrequent contractions.

Another major factor is fetal distress, a condition where the fetus is not getting enough oxygen. Fetal brain damage can result from oxygen deprivation. Fetal distress is often related to abnormalities in the position of the fetus or abnormalities in the birth canal, causing reduced blood flow through the placenta. Other conditions also can make c-section advisable, such as vaginal herpes, **hypertension**, and diabetes in the mother.

Precautions

There are several ways that obstetricians and other doctors diagnose conditions that may make a c-section necessary. Ultrasound testing reveals the positions of the baby and the placenta and may be used to estimate the baby's size and gestational age. Fetal heart monitors, in use since the 1970s, transmit any signals of fetal distress. Oxygen deprivation may be determined by checking the amniotic fluid for meconium (feces)—a lack of oxygen causes an unborn baby to defecate. Oxygen deprivation may also be determined by testing the pH of a blood sample taken from the baby's scalp; a pH of 7.25 or higher is normal, between 7.2 and 7.25 is suspicious, and below 7.2 is a sign of trouble.

When a c-section is being considered because labor is not progressing, the mother should first be encouraged to walk around to stimulate labor. Labor may also be stimulated with the drug oxytocin.

When a c-section is being considered because the baby is in a breech position, the doctor may first attempt to reposition the baby; this is called external cephalic version. The doctor may also try a vaginal breech delivery, depending on the size of the mother's pelvis, the size

KEY TERMS

Breech presentation—The condition in which the baby enters the birth canal with its buttocks or feet first.

Cephalopelvic disproportion (CPD)—The condition in which the baby's head is too large to fit through the mother's pelvis.

Classical incision—In a cesarean section, an incision made vertically along the uterus; this kind of incision makes a larger opening but also creates more bleeding, a greater chance of infection, and a weaker scar.

Dystocia—Failure to progress in labor, either because the cervix will not dilate (expand) further or (after full dilation) the head does not descend through the mother's pelvis.

Low transverse incision—Incision made horizontally across the lower end of the uterus; this kind of

incision is preferred for less bleeding and stronger healing.

Placenta previa—The placenta totally or partially covers the cervix, preventing vaginal delivery.

Placental abruption—Separation of the placenta from the uterine wall before the baby is born, cutting off blood flow to the baby.

Prolapsed cord—The umbilical cord is pushed into the vagina ahead of the baby and becomes compressed, cutting off blood flow to the baby.

Respiratory distress syndrome (RDS)—Difficulty breathing, found in infants with immature lungs.

Transverse presentation—The baby is laying sideways across the cervix instead of head first.

VBAC—Vaginal birth after cesarean.

of the baby, and the type of breech position the baby is in. However, a c-section is safer than a vaginal delivery when the baby is 8 lb (3.6 kg) or larger, in a breech position with the feet crossed, or in a breech position with the head hyperextended.

A woman should receive regular prenatal care and be able to alert her doctor to the first signs of trouble. Once labor begins, she should be encouraged to move around and to urinate. The doctor should be conservative in diagnosing dystocia (nonprogressive labor) and fetal distress, taking a position of "watchful waiting" before deciding to operate.

Description

The most common reason that a cesarean section is performed (in 35% of all cases, according to the United States Public Health Service) is that the woman has had a previous c-section. The "once a cesarean, always a cesarean" rule originated when the classical uterine incision was made vertically; the resulting scar was weak and had a risk of rupturing in subsequent deliveries. Today, the incision is almost always made horizontally across the lower end of the uterus (this is called a "low transverse incision"), resulting in reduced blood loss and a decreased chance of rupture. This kind of incision allows many women to have a vaginal birth after a cesarean (VBAC).

The second most common reason that a c-section is performed (in 30% of all cases) is difficult **childbirth** due to nonprogressive labor (dystocia). Uterine contrac-

tions may be weak or irregular, the cervix may not be dilating, or the mother's pelvic structure may not allow adequate passage for birth. When the baby's head is too large to fit through the pelvis, the condition is called cephalopelvic disproportion (CPD).

Another 12% of c-sections are performed to deliver a baby in a breech presentation: buttocks or feet first. Breech presentation is found in about 3% of all births.

In 9% of all cases, c-sections are performed in response to fetal distress. Fetal distress refers to any situation that threatens the baby, such as the umbilical cord getting wrapped around the baby's neck. This may appear on the fetal heart monitor as an abnormal heart rate or rhythm.

The remaining 14% of c-sections are indicated by other serious factors. One is prolapse of the umbilical cord: the cord is pushed into the vagina ahead of the baby and becomes compressed, cutting off blood flow to the baby. Another is **placental abruption**: the placenta separates from the uterine wall before the baby is born, cutting off blood flow to the baby. The risk of this is especially high in multiple births (twins, triplets, or more). A third factor is **placenta previa**: the placenta covers the cervix partially or completely, making vaginal delivery impossible. In some cases requiring c-section, the baby is in a transverse position, lying horizontally across the pelvis, perhaps with a shoulder in the birth canal.

The mother's health may make delivery by c-section the safer choice, especially in cases of maternal diabetes,

hypertension, **genital herpes**, Rh blood incompatibility, and preeclampsia (high blood pressure related to **pregnancy**).

Preparation

When a c-section becomes necessary, the mother is prepped for surgery. A catheter is inserted into her bladder and an intravenous (IV) line is inserted into her arm. Leads for monitoring the mother's heart rate, rhythm, and blood pressure are attached. In the operating room, the mother is given anesthesia—usually a regional anesthetic (epidural or spinal), making her numb from below her breasts to her toes. In some cases, a general anesthetic will be administered. Surgical drapes are placed over the body, except the head; these drapes block the direct view of the procedure.

The abdomen is washed with an anti-bacterial solution and a portion of the pubic hair may be shaved. The first incision opens the abdomen. Infrequently, it will be vertical from just below the navel to the top of the pubic bone, or more commonly, it will be a horizontal incision across and above the pubic bone (informally called a "bikini cut").

The second incision opens the uterus. In most cases a transverse incision is made. This is the favored type because it heals well and makes it possible for a woman to attempt a vaginal delivery in the future. The classical incision is vertical. Because it provides a larger opening than a low transverse incision, it is used in the most critical situations, such as placenta previa. However, the classical incision causes more bleeding, a greater risk of abdominal infection, and a weaker scar, so the low transverse incision is preferred.

Once the uterus is opened, the amniotic sac is ruptured and the baby is delivered. The time from the initial incision to birth is typically five minutes.

Once the umbilical cord is clamped and cut, the newborn is evaluated. The placenta is removed from the mother, and her uterus and abdomen are stitched closed (surgical staples may be used instead in closing the outermost layer of the abdominal incision). From birth through suturing may take 30–40 minutes. Thus the entire surgical procedure may be performed in less than one hour.

Aftercare

A woman who undergoes a c-section requires both the care given to any new mother and the care given to any patient recovering from major surgery. She should be offered **pain** medication that does not interfere with breastfeeding. She should be encouraged to get out of bed and walk around eight to 24 hours after surgery to stimu-

late circulation (thus avoiding the formation of blood clots) and bowel movement. She should limit climbing stairs to once a day, and avoid lifting anything heavier than the baby. She should nap as often as the baby sleeps, and arrange for help with the housework, meals, and care of other children. She may resume driving after two weeks, although some doctors recommend waiting for six weeks, the typical recovery period from major surgery.

Risks

Because a c-section is a surgical procedure, it carries more risk to both the mother and the baby. The maternal **death** rate is less than 0.02%, but that is four times the maternal death rate associated with vaginal delivery. However, many women have a c-section for serious medical problems. The mother is at risk for increased bleeding (because a c-section may result in twice the blood loss of a vaginal delivery) from the two incisions, the placental attachment site, and possible damage to a uterine artery. Complications occur in less than 10% of cases. The mother may develop infection of either incision, the urinary tract, or the tissue lining the uterus (endometritis). Less commonly, she may receive injury to the surrounding organs, like the bladder and bowel. When a general anesthesia is used, she may experience complications from the anesthesia. Very rarely, she may develop a wound hematoma at the site of either incision or other blood clots leading to pelvic **thrombophlebitis** (inflammation of the major vein running from the pelvis into the leg) or a pulmonary embolus (a blood clot lodging in the lung).

Normal results

The after-effects of a c-section vary, depending on the woman's age, physical fitness, and overall health. Following this procedure, a woman commonly experiences gas pains, incision pain, and uterine contractions—which are also common in vaginal delivery. Her hospital stay may be two to four days. Breastfeeding the baby is encouraged, taking care that it is in a position that keeps the baby from resting on the mother's incision. As the woman heals, she may gradually increase appropriate exercises to regain abdominal tone. Full recovery may be seen in four to six weeks.

The prognosis for a successful vaginal birth after a cesarean (VBAC) may be at least 75%, especially when the c-section involved a low transverse incision in the uterus and there were no complications during or after delivery.

Abnormal results

Of the hundreds of thousands of women in the United States who undergo a c-section each year, about 500

die from serious infections, hemorrhaging, or other complications. These deaths may be related to the health conditions that made the operation necessary, and not simply to the operation itself.

Undergoing a c-section may also inflict psychological distress on the mother, beyond hormonal mood swings and **postpartum depression** ("baby blues"). The woman may feel disappointment and a sense of failure for not experiencing a vaginal delivery. She may feel isolated if the father or birthing coach is not with her in the operating room, or if she is treated by an unfamiliar doctor rather than by her own doctor or midwife. She may feel helpless from a loss of control over labor and delivery with no opportunity to actively participate. To overcome these feelings, the woman must understand why the c-section was necessary. She must accept that she couldn't control the unforeseen events that made the c-section the optimum means of delivery, and recognize that preserving the health and safety of both her and her child was more important than her delivering vaginally. Women who undergo a c-section should be encouraged to share their feelings with others. Hospitals can often recommend support groups for such mothers. Women should also be encouraged to seek professional help if negative emotions persist.

Resources

ORGANIZATIONS

American Academy of Family Physicians. 8880 Ward Parkway, Kansas City, MO 64114. (816) 333-9700. <<http://www.aafp.org>>.

Childbirth.Org. <<http://www.childbirth.org>>.

International Cesarean Awareness Network. 1304 Kingsdale Ave., Redondo Beach, CA 90278. (310) 542-6400.

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (914) 428-7100. <<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. (800) 505-2742. <<http://www.nichd.nih.gov/sids/sids.htm>>.

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Bethany Thivierge

Cestodiasis see **Tapeworm diseases**

CFS see **Chronic fatigue syndrome**

CGD see **Chronic granulomatous disease**

Chagas' disease

Definition

Chagas' disease is named after Dr. Carlos Chagas who first found the organism in the early 1900s. It involves damage to the nerves that control the heart, digestive and other organs, and eventually leads to damage to these organs. Worldwide, Chagas' disease affects over 15 million persons, and kills 50,000 each year. Researchers believe that the parasite that causes the disease is only found in the Americas.

Description

When a person is infected with Chagas' disease, the parasite known as *Trypanosoma cruzi* first causes a mild, short-lived period of "acute" illness; then after a long period without symptoms, the effects of the infection begin to appear. The heart, esophagus, and colon are most frequently involved. These organs become unable to contract properly, and begin to stretch or dilate.

Causes and symptoms

T. cruzi is carried by insects or bugs known as reduviid or "kissing bugs." These insects are very common in Central and South America where they inhabit poorly constructed houses and huts. The insects deposit their waste material, exposing inhabitants to the parasites. The parasites then enter the body by way of a cut or via the eyes or mouth. *T. cruzi* can also be transmitted by blood **transfusion**. Eating uncooked, contaminated food or breastfeeding can also transmit the disease. The reduviids, in turn, become infected with the parasite by biting infected animals and humans.

There are three phases related to infection:

- Acute phase lasts about two months, with non-specific symptoms of low grade **fever**, **headache**, **fatigue**, and enlarged liver or spleen.
- Indeterminate phase lasts 10–20 years, during which time no symptoms occur, but the parasites are reproducing in various organs.
- Chronic phase is the stage when symptoms related to damage of major organs (heart, esophagus, colon) begin.

In the chronic phase, irregularities of heart rhythm, **heart failure**, and blood clots cause weakness, **fainting**, and even sudden **death**.

KEY TERMS

Achalasia—An esophageal disease of unknown cause, in which the lower sphincter or muscle is unable to relax normally, and leads to the accumulation of material within the esophagus.

Endoscopy—Exam using an endoscope (a thin flexible tube which uses a lens or miniature camera to view various areas of the gastrointestinal tract). When the procedure is performed to examine certain organs such as the bile ducts or pancreas, the organs are not viewed directly, but rather indirectly through the injection of x ray.

Parasite—An organism that lives on or in another and takes nourishment (food and fluids) from that organism.

Regurgitation—Flow of material back up the esophagus and into the throat or lungs.

Esophageal symptoms are related to difficulty with swallowing and chest **pain**. Because the esophagus does not empty properly, food regurgitates into the lungs causing **cough**, **bronchitis**, and repeated bouts of **pneumonia**. Inability to eat, weight loss, and **malnutrition** become a significant factor in affecting survival.

Involvement of the large intestine (colon) causes **constipation**, distention, and abdominal pain.

Diagnosis

The best way to diagnose acute infection is to identify the parasites in tissue or blood. Occasionally it is possible to culture the organism from infected tissue, but this process usually requires too much time to be of value. In the chronic phase, antibody levels can be measured. Efforts to develop new, more accurate tests are ongoing.

Treatment

In most cases treatment of symptoms is all that is possible. Present medications can reduce the duration and severity of an acute infection, but are only 50% effective, at best, in eliminating the organisms.

Cardiac effects are managed with **pacemakers** and medications. Esophageal complications require either endoscopic or surgical methods to improve esophageal emptying, similar to those used to treat the disorder known as **achalasia**. Constipation is treated by increasing fiber and bulk **laxatives**, or removal of diseased portions of the colon.

Prognosis

Those patients with gastrointestinal complications often respond to some form of treatment. Cardiac problems are more difficult to treat, particularly since transplant would rekindle infection.

Prevention

Visitors traveling to areas of known infection should avoid staying in mud, adobe, or similar huts. Mosquito nets and insect repellents are useful in helping to avoid contact with the bugs. Blood screening is not always effective in many regions where infection is common. It is necessary to carefully screen people who have emigrated from Central and South America before they make blood donations.

Resources

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David Kaminstein, MD

Chalazion see **Eyelid disorders**

Chancroid

Definition

Chancroid is a sexually transmitted disease caused by a bacterial infection that is characterized by painful sores on the genitals.

Description

Chancroid is an infection of the genitals that is caused by the bacterium *Haemophilus ducreyi*. Chancroid is a sexually transmitted disease, which means that it is spread from person to person almost always by sexual contact. However, there have been a few cases in which healthcare providers have become infected through contact with infected patients.

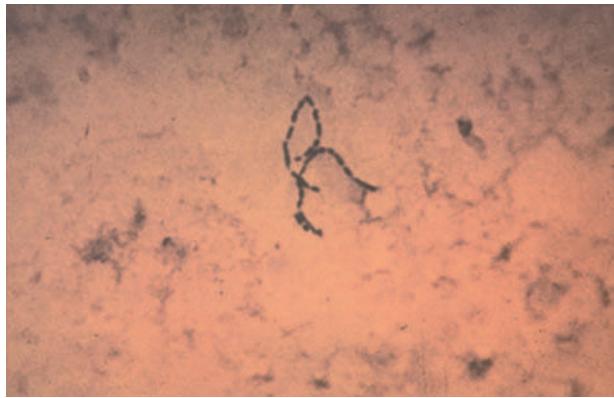
Common locations for chancroid sores (ulcers) in men are the shaft or head of the penis, foreskin, the groove behind the head of the penis, the opening of the penis, and the scrotum. In women, common locations are the labia majora (outer lips), labia minora (inner lips), perianal area (area around the anal opening), and inner thighs. It is rare for the ulcer(s) to be on the vaginal walls or cervix. In about 50% of the patients with chancroid, the infection spreads to either or both of the lymph nodes in the groin.

Chancroid is most commonly found in developing and third world countries. In the United States, the most common cause of genital ulcers is **genital herpes**, followed by **syphilis**, and then chancroid. As of 1997, there were fewer than 1,500 cases of chancroid in the United States per year and it occurred primarily in African Americans, Hispanic Americans, and Native Americans. There are occasional localized outbreaks of chancroid in the United States. In addition, the practice of exchanging sex for drugs has lead to a link between crack **cocaine** use and chancroid.

Even though the incidence of chancroid in the United States decreased in the 1990s, there is an alarming connection between chancroid and human **immunodeficiency** virus (HIV) infection. HIV causes **AIDS** (acquired immunodeficiency syndrome) and is easily spread from person to person through chancroid ulcers. Uncircumcised men with chancroid ulcers have a 48% risk of acquiring HIV from sexual contact. Women with chancroid ulcers are also at a greater risk of being infected with HIV during sexual contact. Genital ulcers seem to act as doorways for HIV to enter and exit.

Causes and symptoms

Haemophilus ducreyi is spread from person to person by vaginal, anal, and oral sexual contact. Uncircumcised men are about three times more likely than circumcised men to become infected following exposure to *Haemophilus ducreyi*. Having unprotected sex, exchanging sex for drugs, and having unprotected sex with a prostitute are other risk factors. Many cases of chancroid in the United States occur in persons who had traveled to countries where the disease is more common.



A close-up view of a chancroid specimen. (Custom Medical Stock Photo. Reproduced by permission.)

Chancroid occurs when *Haemophilus ducreyi* penetrates the skin through an injury, like a scratch or cut. Once past the skin surface, the warmth, moisture, and nutrients allow bacteria to grow rapidly. The first sign of chancroid is a small, red papule that occurs within three to seven days following exposure to the bacteria, but may take up to one month. Usually within one day, the papule becomes an ulcer. The chancroid ulcer is painful, bleeds easily, drains a grey or yellowish pus, and has sharply defined, ragged edges. They can vary in size from an eighth of an inch to two inches in diameter. Men usually have only one ulcer, but women often have four or more. Sometimes “kissing” ulcers occur when one ulcer spreads the bacterial infection to an opposite skin surface. For example, kissing ulcers can form on the lips of the labia majora. Alternatively, women may not have any external sores but may experience painful urination, intercourse, and/or bowel movements and may have a vaginal discharge or rectal bleeding.

Signs that the infection has spread to the lymph node appear about one week after the formation of the genital ulcer. Lymph nodes are small organs in the lymphatic system that filter waste materials from nearly every organ in the body. This lymph node infection is called “lymphadenitis” and the swollen, painful lymph node is called a “bubo.” The bubo, which appears as a red, spherical lump, may burst through the skin, releasing a thick pus and forming another ulcer.

Diagnosis

Chancroid may be diagnosed and treated by urologists (urinary tract doctors for men), gynecologists (for women), and infectious disease specialists. Part of the diagnosis of chancroid involves ruling out genital herpes and syphilis because genital ulcers are also symptoms of these diseases. The appearance of these three diseases

KEY TERMS

Bubo—A tender, swollen lymph node in the groin that may follow a chancroid ulcer.

Groin—The region of the body that lies between the abdomen and the thighs.

can be close enough to be confusing. However, the presence of a pus-filled lump in the groin of a patient with a genital ulcer is highly specific for chancroid.

For a clear-cut diagnosis of chancroid, *Haemophilus ducreyi* must be isolated from the ulcer. To do this, a sterile cotton swab is wiped over the ulcer to obtain a pus sample. In the laboratory, the sample is put into special media and placed in an incubator. *Haemophilus ducreyi* takes from two to five days to grow in the laboratory. In addition, the pus may be examined under the microscope to see which bacteria are in the ulcer. A sample of the pus may also be tested to see if the herpes virus is present. A blood sample will probably be taken from the patient's arm to test for the presence of antibodies to the bacteria that causes syphilis.

Treatment

The only treatment for chancroid is **antibiotics** given either once or for several days. Antibiotics taken by mouth for one to two weeks include erythromycin (E-Mycin, Ery-Tab), amoxicillin plus clavulanic acid (Augmentin), co-trimoxazole (Bactrim, Septra), or ciprofloxacin (Cipro). Antibiotics given in one dose include ceftriaxone (Rocephin), spectinomycin (Trobicin), co-trimoxazole, or ofloxacin (Floxin).

The ulcer(s) may be cleaned and soaked to reduce the swelling. Salt solution dressings may be applied to the ulcer(s) to reduce the spread of the bacteria and prevent additional ulcers. A serious infection of the foreskin may require **circumcision**. Pus would be removed from infected lymph nodes by using a needle and syringe. Very large buboes may require surgical drainage.

Prognosis

Without treatment, chancroid may either go away quickly or patients may experience the painful ulcers for many months. A complete cure is obtained with antibiotic treatment. Severe ulcers may cause permanent scars. Severe scarring of the foreskin may require circumcision. Urethral fistulas (abnormal passageways from the urine tube to the skin) may occur and requires corrective surgery.

Prevention

The best prevention for chancroid is to use a **condom** during sexual intercourse. Chancroid can also be prevented by abstinence (avoidance of any sexual contact) and by being in a monogamous relationship with a disease-free partner. To prevent the spread of chancroid, it is important that all sexual contacts of the patient are identified and treated.

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OTHER

Mayo Clinic Online. 5 Mar. 1998 <<http://www.mayohealth.org>>.

Belinda Rowland, PhD

Change of life see **Menopause**

Character disorders see **Personality disorders**

Charcoal, activated

Definition

Activated charcoal is a fine, black, odorless, and tasteless powder. It is made from wood or other materials that have been exposed to very high temperatures in an airless environment. It is then treated, or activated, to increase its ability to adsorb by reheating with oxidizing gas or other chemicals to break it into a very fine powder. Activated charcoal is pure carbon specially processed to make it highly adsorbent of particles and gases in the body's digestive system.

Activated charcoal has often been used since ancient times to cure a variety of ailments including **poisoning**.

Its healing effects have been well documented since as early as 1550 B.C. by the Egyptians. However, charcoal was almost forgotten until 15 years ago when it was rediscovered as a wonderful oral agent to treat most overdoses and toxins.

Description

Activated charcoal's most important use is for treatment of poisoning. It helps prevent the absorption of most poisons or drugs by the stomach and intestines. In addition to being used for most swallowed poisons in humans, charcoal has been effectively used in dogs, rabbits, rats, and other animals, as well. It can also adsorb gas in the bowels and has been used for the treatment of gas or **diarrhea**. Charcoal's other uses such as treatment of viruses, bacteria, bacterial toxic byproducts, snake venoms and other substances by adsorption have not been supported by clinical studies. By adding water to the powder to make a paste, activated charcoal can be used as an external application to alleviate **pain** and **itching** from **bites and stings**.

Poisons and drug overdoses

It is estimated that one million children accidentally overdose on drugs mistaken as candies or eat, drink, or inhale poisonous household products each year. Infants and toddlers are at the greatest risk for accidental poisoning. Activated charcoal is one of the agents most commonly used for these cases. It can absorb large amounts of poisons quickly. In addition, it is non-toxic, may be stored for a long time, and can be conveniently administered at home. Charcoal works by binding to irritating or toxic substances in the stomach and intestines. This prevents the toxic drug or chemical from spreading throughout the body. The activated charcoal with the toxic substance bound to it is then excreted in the stool without harm to the body. When poisoning is suspected the local poison control center should be contacted for instructions. They may recommend using activated charcoal, which should be available at home so that it can be given to the poisoned child or pet immediately. For severe poisoning, several doses of activated charcoal may be needed.

Intestinal disorders

In the past, activated charcoal was a popular remedy for gas. Even before the discovery of America by Europeans, Native Americans used powdered charcoal mixed with water to treat an upset stomach. Now charcoal is being rediscovered as an alternative treatment for this condition. Activated charcoal works like a sponge. Its huge surface area is ideal for soaking up different substances, including gas. In one study, people taking activated char-

KEY TERMS

Antidote—A remedy to counteract a poison or injury.

Adsorption—The binding of a chemical (e.g., drug or poison) to a solid material such as activated charcoal or clay.

coal after eating a meal with high gas-producing foods did not produce more gas than those who did not have these foods. Charcoal has also been used to treat other intestinal disorders such as diarrhea, **constipation**, and cramps. There are few studies to support these uses and there are also concerns that frequent use of charcoal may decrease absorption of essential nutrients, especially in children.

Other uses

Besides being a general antidote for poisons or remedy for gas, activated charcoal has been used to treat other conditions as well. Based on its ability to adsorb or bind to other substances, charcoal has been effectively used to clean skin **wounds** and to adsorb waste materials from the gastrointestinal tract. In addition, it has been used to adsorb snake venoms, viruses, bacteria, and harmful materials excreted by bacteria or fungi. However, because of lack of scientific studies, these uses are not recommended. Activated charcoal, when used together with other remedies such as aloe vera, acidophilus, and psyllium, helps to keep symptoms of **ulcerative colitis** under control. While charcoal shows some anti-aging activity in rats, it is doubtful if it can do the same for humans.

Recommended dosage

For poisoning

Activated charcoal is available without prescription. However, in case of accidental poisoning or **drug overdose** an emergency poison control center, hospital emergency room, or doctor's office should be called for advice. In case that both syrup of **ipecac** and charcoal are recommended for treatment of the poison, ipecac should be given first. Charcoal should not be given for at least 30 minutes after ipecac or until vomiting from ipecac stops. Activated charcoal is often mixed with a liquid before being swallowed or put into the tube leading to the stomach. Activated charcoal is available as 1.1 oz (33 ml) liquid bottles. It is also available in 0.5 oz (15 ml) container sizes and as slurry of charcoal pre-mixed in water or as a container in which water or soda pop is added. Keeping activated charcoal at home is a

good idea so that it can be taken immediately when needed for treatment of poisoning.

For acute poisoning, the dosage is as follows:

- Infants (under 1 year of age): 1 g/kg.
- Children (1–12 years of age): 15–30 g or 1–2 g/kg with at least 8 oz of water.
- Adults: 30–100 g or 1–2 g/kg with at least 8 oz of water.

For diarrhea or gas

A person can take charcoal tablets or capsules with water or sprinkle the content onto foods. The dosage for treatment of gas or diarrhea in adults is 520–975 mg after each meal and up to 5 g per day.

Precautions

Parents should keep activated charcoal on hand in case of emergencies.

Do not give charcoal together with syrup of ipecac. The charcoal will adsorb the ipecac. Charcoal should be taken 30 minutes after ipecac or after the vomiting from ipecac stops.

Some activated charcoal products contain sorbitol. Sorbitol is a sweetener as well as a laxative, therefore, it may cause severe diarrhea and vomiting. These products should not be used in infants.

Charcoal may interfere with the absorption of medications and nutrients such as **vitamins** or **minerals**. For uses other than for treatment of poisoning, charcoal should be taken two hours after other medications.

Charcoal should not be used to treat poisoning caused by corrosive products such as lye or other strong acids or petroleum products such as gasoline, kerosene, or cleaning fluids. Charcoal may make the condition worse and delay diagnosis and treatment. In addition, charcoal is also not effective if the poison is lithium, cyanide, iron, ethanol, or methanol.

Parents should not mix charcoal with chocolate syrup, sherbet, or ice cream, even though it may make charcoal taste better. These foods may prevent charcoal from working properly.

Activated charcoal may cause swelling or pain in the stomach. A doctor should be notified immediately. It has been known to cause problems in people with intestinal bleeding, blockage or those people who have had recent surgery. These patients should talk to their doctor before using this product.

Charcoal may be less effective in people with slow digestion.

Charcoal should not be given for more than three or four days for treatment of diarrhea. Continuing for longer periods may interfere with normal **nutrition**.

Charcoal should not be used in children under three years of age to treat diarrhea or gas.

Activated charcoal should be kept out of reach of children.

Side effects

Charcoal may cause constipation when taken for overdose or accidental poisoning. A laxative should be taken after the crisis is over.

Activated charcoal may cause the stool to turn black. This is to be expected.

Pain or swelling of the stomach may occur. A doctor should be consulted.

Interactions

Activated charcoal should not be mixed together with chocolate syrup, ice cream or sherbet. These foods prevent charcoal from working properly.

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Mai Tran

Charcot Marie Tooth disease

Definition

Charcot Marie Tooth disease (CMT) is the name of a group of inherited disorders of the nerves in the peripheral nervous system (nerves throughout the body

that communicate motor and sensory information to and from the spinal cord) causing weakness and loss of sensation in the limbs.

Description

CMT is named for the three neurologists who first described the condition in the late 1800s. It is also known as hereditary motor and sensory neuropathy, and is sometimes called peroneal muscular atrophy, referring to the muscles in the leg that are often affected. The age of onset of CMT can vary anywhere from young childhood to the 50s or 60s. Symptoms typically begin by the age of 20. For reasons yet unknown, the severity in symptoms can also vary greatly, even among members of the same family.

Although CMT has been described for many years, it is only since the early 1990s that the genetic cause of many of the types of CMT have become known. Therefore, knowledge about CMT has increased dramatically within a short time.

The peripheral nerves

CMT affects the peripheral nerves, those groups of nerve cells carrying information to and from the spinal cord. CMT decreases the ability of these nerves to carry motor commands to muscles, especially those furthest from the spinal cord located in the feet and hands. As a result, the muscles connected to these nerves eventually weaken. CMT also affects the sensory nerves that carry information from the limbs to the brain. Therefore people with CMT also have sensory loss. This causes symptoms such as not being able to tell if something is hot or cold or difficulties with balance.

There are two parts of the nerve that can be affected in CMT. A nerve can be likened to an electrical wire, in which the wire part is the axon of the nerve and the insulation surrounding it is the myelin sheath. The job of the myelin is to help messages travel very fast through the nerves. CMT is usually classified depending on which part of the nerve is affected. People who have problems with the myelin have CMT type 1 and people who have abnormalities of the axon have CMT type 2.

Specialized testing of the nerves, called nerve conduction testing (NCV), can be performed to determine if a person has CMT1 or CMT2. These tests measure the speed at which messages travel through the nerves. In CMT1, the messages move too slowly, but in CMT2 the messages travel at the normal speed.

Demographics

CMT has been diagnosed in people from all over the world. It occurs in approximately one in 2,500 people,

which is about the same incidence as **multiple sclerosis**. It is the most common type of inherited neurologic condition.

Signs and symptoms

CMT is caused by changes (mutations) in any one of a number of genes that carry the instructions to make the peripheral nerves. Genes contain the instructions for how the body grows and develops before and after a person is born. There are probably at least 15 different genes that can cause CMT. However, as of early 2001, many have not yet been identified.

CMT types 1 and 2 can be broken down into subtypes based upon the gene that is causing CMT. The subtypes are labeled by letters, so there is CMT1A, CMT1B, etc. Therefore, the gene with a mutation that causes CMT1A is different from that that causes CMT1B.

Types of CMT

CMT1A. The most common type of CMT is called CMT1A. It is caused by a mutation in a gene called peripheral myelin protein 22 (PMP22) located on chromosome 17. The job of this gene is to make a protein (PMP22) that makes up part of the myelin. In most people who have CMT, the mutation that causes the condition is a duplication (doubling) of the PMP22 gene. Instead of having two copies of the PMP22 gene (one on each chromosome) there are three copies. It is not known how this extra copy of the PMP22 gene causes the observed symptoms. A small percentage of people with CMT1A do not have a duplication of the PMP22 gene, but rather have a point mutation in the gene. A point mutation is like a typo in the gene that causes it to work incorrectly.

HEREDITARY NEUROPATHY WITH LIABILITY TO PRESSURE PALSIES (HNPP). HNPP is a condition that is also caused by a mutation in the PMP22 gene. The mutation is a deletion. Therefore, there is only one copy of the PMP22 gene instead of two. People who have HNPP may have some of the signs of CMT. However, they also have episodes where they develop weakness and problems with sensation after compression of certain pressure points such as the elbows or knee. Often these symptoms will resolve after a few days or weeks, but sometimes they are permanent.

CMT1B. Another type of CMT, called CMT1B, is caused by a mutation in a gene called myelin protein zero (MPZ) located on chromosome 1. The job of this gene is to make the layers of myelin stick together as they are wrapped around the axon. The mutations in this gene are point mutations because they involve a change (either deletion, substitution, or insertion) at one specific component of a gene.

CMTX. Another type of CMT, called CMTX, is usually considered a subtype of CMT1 because it affects the myelin, but it has a different type of inheritance than type 1 or type 2. In CMTX, the CMT-causing gene is located on the X chromosome and is called connexin 32 (Cx32). The job of this gene is to code for a class of protein called connexins that form tunnels between the layers of myelin.

CMT2. There are at least five different genes that can cause CMT type 2. Therefore, CMT2 has subtypes A, B, C, D and E. As of early 2001, scientists have narrowed in on the location of most of the CMT2 causing genes. However, the specific genes and the mutations have not yet been found for most types. Very recently, the gene for CMT2E has been found. The gene is called neurofilament-light (NF-L). Because it has just been discovered, not much is known about how mutations in this gene cause CMT.

CMT3. In the past a condition called Dejerine-Sottas disease was referred to as CMT3. This is a severe type of CMT in which symptoms begin in infancy or early childhood. It is now known that this is not a separate type of CMT and in fact people who have onset in infancy or early childhood often have mutations in the PMP22 or MPZ genes.

CMT4. CMT4 is a rare type of CMT in which the nerve conduction tests have slow response results. However, it is classified differently from CMT1 because it is passed through families by a different pattern of inheritance. There are five different subtypes and each has only been described in a few families. The symptoms in CMT4 are often severe and other symptoms such as deafness may be present. There are three different genes that have been associated with CMT4 as of early 2001. They are called MTMR2, EGR2, and NDRG1. More research is required to understand how mutations in these genes cause CMT.

Inheritance

CMT1A and 1B, HNPP, and all of the subtypes of CMT2 have autosomal dominant inheritance. Autosomal refers to the first 22 pairs of chromosomes that are the same in males and females. Therefore, males and females are affected equally in these types. In a dominant condition, only one gene of a pair needs to have a mutation in order for a person to have symptoms of the condition. Therefore, anyone who has these types has a 50%, or one in two, chance of passing CMT on to each of their children. This chance is the same for each **pregnancy** and does not change based on previous children.

CMTX has X-linked inheritance. Since males only have one X chromosome, they only have one copy of the Cx32 gene. Thus, when a male has a mutation in his Cx32

gene, he will have CMT. However, females have two X chromosomes and therefore have two copies of the Cx32 gene. If they have a mutation in one copy of their Cx32 genes, they will only have mild to moderate symptoms of CMT that may go unnoticed. This is because their normal copy of the Cx32 gene does make normal myelin.

Females pass on one or the other of their X chromosomes to their children—sons or daughters. If a woman with a Cx32 mutation passes her normal X chromosome, she will have an unaffected son or daughter who will not pass CMT on to his or her children. If the woman passes the chromosome with Cx32 mutation on she will have an affected son or daughter, although the daughter will be mildly affected or have no symptoms. Therefore, a woman with a Cx32 mutation has a 50%, or a one in two, chance of passing the mutation to her children: a son will be affected, and a daughter may only have mild symptoms.

When males pass on an X chromosome, they have a daughter. When they pass on a Y chromosome, they have a son. Since the Cx32 mutation is on the X chromosome, a man with CMTX will always pass the Cx32 mutation on to his daughters. However, when he has a son, he passes on the Y chromosome, and therefore the son will not be affected. Therefore, an affected male passes the Cx32 gene mutation on to all of his daughters, but to none of his sons.

CMT4 has autosomal recessive inheritance. Males and females are equally affected. In order for a person to have CMT4, they must have a mutation in both of their CMT-causing genes—one inherited from each parent. The parents of an affected person are called carriers. They have one normal copy of the gene and one copy with a mutation. Carriers do not have symptoms of CMT. Two carrier parents have a 25%, or one in four, chance of passing CMT on to *each* of their children.

The onset of symptoms is highly variable, even among members of the same family. Symptoms usually progress very slowly over a person's lifetime. The main problems caused by CMT are weakness and loss of sensation mainly in the feet and hands. The first symptoms are usually problems with the feet such as high arches and problems with walking and running. Tripping while walking and sprained ankles are common. Muscle loss in the feet and calves leads to "foot drop" where the foot does not lift high enough off the ground when walking. Complaints of cold legs are common, as are cramps in the legs, especially after **exercise**.

In many people, the fingers and hands eventually become affected. Muscle loss in the hands can make fine movements such as working buttons and zippers difficult. Some patients develop tremor in the upper limbs. Loss of sensation can cause problems such as numbness and the

inability to feel if something is hot or cold. Most people with CMT remain able to walk throughout their lives.

Diagnosis

Diagnosis of CMT begins with a careful neurological exam to determine the extent and distribution of weakness. A thorough family history should be taken at this time to determine if other people in the family are affected. Testing may also be performed to rule out other causes of neuropathy.

A nerve conduction velocity test should be performed to measure how fast impulses travel through the nerves. This test may show characteristic features of CMT, but it is not diagnostic of CMT. Nerve conduction testing may be combined with **electromyography** (EMG), an electrical test of the muscles.

A nerve biopsy (removal of a small piece of the nerve) may be performed to look for changes characteristic of CMT. However, this testing is not diagnostic of CMT and is usually not necessary for making a diagnosis.

Definitive diagnosis of CMT is made only by **genetic testing**, usually performed by drawing a small amount of blood. As of early 2001, testing is available to detect mutations in PMP22, MPZ, Cx32 and EGR2. However, research is progressing rapidly and new testing is often made available every few months. All affected members of a family have the same type of CMT. Therefore once a mutation is found in one affected member, it is possible to test other members who may have symptoms or are at risk of developing CMT.

Prenatal diagnosis

Testing during pregnancy to determine whether an unborn child is affected is possible if genetic testing in a family has identified a specific CMT-causing mutation. This can be done after 10–12 weeks of pregnancy using a procedure called **chorionic villus sampling** (CVS). CVS involves removing a tiny piece of the placenta and examining the cells. Testing can also be done by **amniocentesis** after 16 weeks gestation by removing a small amount of the amniotic fluid surrounding the baby and analyzing the cells in the fluid. Each of these procedures has a small risk of **miscarriage** associated with it, and those who are interested in learning more should check with their doctor or genetic counselor. Couples interested in these options should obtain **genetic counseling** to carefully explore all of the benefits and limitations of these procedures.

Treatment

There is no cure for CMT. However, physical and occupational therapy are an important part of CMT treatment. Physical therapy is used to preserve range of motion and minimize deformity caused by muscle shortening, or

contracture. Braces are sometimes used to improve control of the lower extremities that can help tremendously with balance. After wearing braces, people often find that they have more energy because they are using less energy to focus on their walking. Occupational therapy is used to provide devices and techniques that can assist tasks such as dressing, feeding, writing, and other routine activities of daily life. Voice-activated software can also help people who have problems with fine motor control.

It is very important that people with CMT avoid injury that causes them to be immobile for long periods of time. It is often difficult for people with CMT to return to their original strength after injury.

There is a long list of medications that should be avoided if possible by people diagnosed with CMT such as hydralazine (Apresoline), megadoses of vitamin A, B₆, and D, Taxol, and large intravenous doses of penicillin. Complete lists are available from the CMT support groups. People considering taking any of these medications should weigh the risks and benefits with their physician.

Prognosis

The symptoms of CMT usually progress slowly over many years, but do not usually shorten life expectancy. The majority of people with CMT do not need to use a wheelchair during their lifetime. Most people with CMT are able to lead full and productive lives despite their physical challenges.

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ORGANIZATIONS

- Charcot Marie Tooth Association (CMTA). 2700 Chestnut Parkway, Chester, PA 19013. (610) 499-9264 or (800) 606-CMTA. Fax: (610) 499-9267. <cmtassoc@aol.com>. <<http://www.charcot-marie-tooth.org>>.
CMT International. Attn: Linda Crabtree, 1 Springbank Dr. St. Catherine's, ONT L2S2K1. Canada (905) 687-3630. <<http://www.cmtint.org>>.
Muscular Dystrophy Association. 3300 East Sunrise Dr., Tucson, AZ 85718. (520) 529-2000 or (800) 572-1717. <<http://www.mdausa.org>>.

KEY TERMS

- Axon**—Skinny, wire-like extension of nerve cells.
- Myelin**—A fatty sheath surrounding nerves in the peripheral nervous system, which help them conduct impulses more quickly.
- Nerve conduction testing**—Procedure that measures the speed at which impulses move through the nerves.
- Neuropathy**—A condition caused by nerve damage. Major symptoms include weakness, numbness, paralysis, or pain in the affected area.
- Peripheral nerves**—Nerves throughout the body that carry information to and from the spinal cord.

Neuropathy Association. 60 E. 42nd St. Suite 942, New York, NY 10165. (212) 692-0662. <<http://www.neuropathy.org>>.

OTHER

- HNPP—Hereditary Neuropathy with liability to Pressure Palsies.** University of Washington, Seattle. <<http://www.hnpp.org>>.
- “GeneClinics.” <www.geneclinics.org>.
- OMIM—Online Mendelian Inheritance in Man.** <<http://www.ncbi.nlm.nih.gov/Omim>>.

Karen M. Krajewski, MS, CGC

Charcot's joints

Definition

Charcot's joints is a progressive degenerative disease of the joints caused by nerve damage resulting in the loss of ability to feel **pain** in the joint and instability of the joint.

Description

Charcot's joints, also called neuropathic joint disease, is the result of two conditions present in the joint. The first factor is the inability to feel pain in the joint due to nerve damage. The second factor is that injuries to the joint go unnoticed leading to instability and making the joint more susceptible to further injury. Repeated small injuries, strains and even **fractures** can go unnoticed until finally the joint is permanently destroyed. Loss of the protective sensation of pain is what leads to the disintegration of the joint and often leads to deformity in the joint.

Although this condition can affect any joint, the knee is the joint most commonly involved. In individuals with **diabetes mellitus**, the foot is most commonly affected. The disease can involve only one joint or it may affect two or three joints. More than three affected joints is very rare. In all cases, the specific joint(s) affected depends on the location of the nerve damage.

Causes and symptoms

Many diseases and injuries can interfere with the ability to feel pain. Conditions such as diabetes mellitus, spinal injuries and diseases, **alcoholism**, and even **syphilis** can all lead to a loss of the ability to feel pain in some areas. Lack of pain sensation may also be congenital.

The symptoms of Charcot's joints can go unnoticed for some time and may be confused with **osteoarthritis** in the beginning. Swelling and stiffness in a joint without the expected pain, or with less pain than would be expected, are the primary symptoms of this condition. As the condition progresses, however, the joint can become very painful due to fluid build-up and bony growths.

Diagnosis

Charcot's joints is suspected when a person with a disease that impairs pain sensation exhibits painless swelling and/or stiffness in a joint. Standard x rays will show damage to the joint, and may also show abnormal bone growth and calcium deposits. Floating bone fragments from previous injuries may also be visible.

Treatment

In the early stages of Charcot's joints, braces to stabilize the joints can help stop or minimize the damage. When the disease has progressed beyond braces, surgery can sometimes repair the joint. If the damage is extensive, an artificial joint may be necessary.

Prognosis

Treatment of the disease causing loss of pain perception may help to slow the damage to the joints.

Prevention

Preventing or effectively managing the underlying disease can slow or in some cases reverse joint damage, but the condition cannot be prevented.

Resources

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Dorothy Elinor Stonely

Charley horse see **Muscle spasms and cramps**

Chelation therapy

Definition

Chelation therapy is an intravenous treatment designed to bind heavy metals in the body in order to treat heavy metal toxicity. Proponents claim it also treats **coronary artery disease** and other illnesses that may be linked to damage from free radicals (reactive molecules).

Purpose

The benefits of EDTA chelation for the treatment of **lead poisoning** and excessively high calcium levels are undisputed. The claims of benefits for those suffering from **atherosclerosis**, coronary artery disease, and other degenerative diseases are more difficult to prove. Reported uses for chelation therapy include treatment of **angina**, **gangrene**, **arthritis**, **multiple sclerosis**, **Parkinson's disease**, **psoriasis**, and **Alzheimer's disease**. Improvement is also claimed for people experiencing diminished sight, hearing, smell, coordination, and sexual potency.

Description

Origins

The term chelation is from the Greek root word "chele," meaning "claw." Chelating agents, most commonly diamine tetraacetic acid (EDTA), were originally designed for industrial applications in the early 1900s. It was not until the World War II era that the potential for medical therapy was realized. The initial intent was to develop antidotes to poison gas and radioactive contaminants. The need for widespread therapy of this nature did not materialize, but more practical uses were found for chelation. During the following decade, EDTA chelation therapy became standard treatment for people suffering from lead **poisoning**. Patients who had received this treatment claimed to have other health improvements that could not be attributed to the lead removal only. Especially notable were comments from those who had previously suffered from intermittent claudication and angina. They reported suffering less **pain** and **fatigue**, with improved endurance, after chelation therapy. These reports stimulated further interest in the potential benefits of chelation therapy for people suffering from atherosclerosis and coronary artery disease.

If the preparatory examination suggests that there is a condition that could be improved by chelation therapy, and

there is no health reason why it shouldn't be used, then the treatment can begin. The patient is generally taken to a comfortable treatment area, sometimes in a group location, and an intravenous line is started. A solution of EDTA together with **vitamins** and **minerals** tailored for the individual patient is given. Most treatments take three to four hours, as the infusion must be given slowly in order to be safe. The number of recommended treatments is usually between 20 and 40. They are given one to three times a week. Maintenance treatments can then be given at the rate of once or twice a month. Maximum benefits are reportedly attained after approximately three months after a treatment series. The cost of therapy is considerable, but it is a fraction of the cost of an expensive medical procedure like cardiac bypass surgery. Intravenous vitamin C and mercury chelation therapies are also offered.

Preparations

A candidate for chelation therapy should initially have a thorough history and physical to define the type and extent of clinical problems. Laboratory tests will be done to determine whether there are any conditions present that would prevent the use of chelation. Patients who have pre-existing **hypocalcemia**, poor liver or kidney function, congestive **heart failure**, **hypoglycemia**, **tuberculosis**, clotting problems, or potentially allergic conditions are at higher risk for complications from chelation therapy. A Doppler ultrasound may be performed to determine the adequacy of blood flow in different regions of the body.

Precautions

It is important for people who receive chelation therapy to work with medical personnel who are experienced in the use of this treatment. Treatment should not be undertaken before a good physical, lifestyle evaluation, history, and any laboratory tests necessary are performed. The staff must be forthcoming about test results and should answer any questions the patient may have. Evaluation and treatment should be individualized and involve assessment of kidney function before each treatment with chelation, since the metals bound by the EDTA are excreted through the kidneys.

Although EDTA binds harmful, toxic metals like mercury, lead, and cadmium, it also binds some essential nutrients of the body, such as copper, iron, calcium, zinc, and magnesium. Large amounts of zinc are lost during chelation. Zinc deficiency can cause impaired immune function and other harmful effects. Supplements of zinc are generally given to patients undergoing chelation, but it is not known whether this is adequate to prevent deficiency. Also, chelation therapy does not replace proper **nutrition**, **exercise**, and appropriate medications or surgery for specific diseases or conditions.

KEY TERMS

Angina—Chest pain caused by reduced oxygen to the heart.

Atherosclerosis—Arterial disease characterized by fatty deposits on inner arterial walls.

Hypocalcemia—Low blood calcium.

Hypoglycemia—Low blood sugar.

Intermittent claudication—Leg pain and weakness caused by walking.

Thrombophlebitis—Inflammation of a vein together with clot formation.

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Judith Turner

Side effects

Side effects of chelation therapy are reportedly unusual, but are occasionally serious. Mild reactions may include, but are not limited to, local irritation at the infusion site, skin reactions, nausea, **headache**, **dizziness**, hypoglycemia, **fever**, leg cramps, or loose bowel movements. Some of the more serious complications reported have included hypocalcemia, kidney damage, decreased clotting ability, anemia, bone marrow damage, insulin shock, **thrombophlebitis** with **embolism**, and even rare deaths. However, some doctors feel that the latter groups of complications occurred before the safer method currently used for chelation therapy was developed.

Research and general acceptance

EDTA chelation is a highly controversial therapy. The treatment is approved by the United States Food and Drug Administration (FDA) for lead poisoning and seriously high calcium levels. However, for the treatment of atherosclerotic heart disease, EDTA chelation therapy is not endorsed by the American Heart Association (AHA), the FDA, the National Institutes of Health (NIH), or the American College of Cardiology. The AHA reports that there are no adequate, controlled, published scientific studies using currently approved scientific methods to support this therapy for the treatment of coronary artery disease. However, a pooled analysis from the results of over 70 studies showed positive results in all but one.

Resources

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Chemica see **Skin resurfacing**

Chemical debridement see **Debridement**

Chemabrasion see **Skin resurfacing**

Chemonucleolysis

Definition

Chemonucleolysis is a medical procedure that involves the dissolving of the gelatinous cushioning material in an intervertebral disk by the injection of an enzyme such as chymopapain.

Purpose

Between each vertebra lies a disk of cushioning material that keeps the spinal bones from rubbing together and absorbs some of the **shock** to the spine from body movements. In the center of the disk is soft, gelatinous material called the nucleus pulposus (NP). The NP is surrounded by a tough fibrous coating. Sometimes when the back is injured, this coating can weaken and bulge or tear to allow the NP to ooze out. When this happens, it is called a herniated nucleus pulposus (HNP), or—in common language—a **herniated disk**.

When the disk bulges or herniates, it can put pressure on nerves which originate in the spinal column, and go to other parts of the body. This causes lower back **pain**, and/or pain to the hips, legs, arms, shoulders, and neck, depending on the location of the herniated disk. Chemonucleolysis uses chymopapain, an enzyme derived from papyrus, to dissolve the disk material that has

KEY TERMS

Chymopapain—An enzyme from the milky white fluid of the papaya, used for medical purposes in chemonucleolysis.

Myelography—An x-ray test that evaluates the subarachnoid space of the spine.

Nucleus pulposus (NP)—an elastic, pulpy mass in the center of each vertebral disk.

been displaced because of injury. Herniated disks are the cause of only a small proportion of cases of lower back pain, and chemonucleolysis is appropriate for only some cases of HNP.

Chemonucleolysis is a conservative alternative to disk surgery. There are three types of disk injuries. A protruded disk is one that is intact but bulging. In an extruded disk, the fibrous wrapper has torn and the NP has oozed out, but is still connected to the disk. In a sequestered disk, a fragment of the NP has broken loose from the disk and is free in the spinal canal. Chemonucleolysis is effective on protruded and extruded disks, but not on sequestered disk injuries. In the United States, chymopapain chemonucleolysis is approved only for use in the lumbar (lower) spine. In other countries, it has also been used successfully to treat cervical (upper spine) hernias.

Other indications that a patient is a good candidate for chemonucleolysis instead of surgery include:

- the patient is 18–50 years of age
- leg pain is worse than lower back pain
- other conservative treatments have failed
- the spot where the herniated disk presses on the nerve has been pinpointed by **myelography**, computed tomography scan (CT scan), or **magnetic resonance imaging (MRI)**
- the patient wishes to avoid surgery

Precautions

There are some situations in which chemonucleolysis should not be performed. Chymopapain is derived from the papaya. About 0.3% of patients are allergic to chymopapain and go into life-threatening shock when exposed to the enzyme. Chemonucleolysis should not be performed on patients allergic to chymopapain or papaya. It also should not be done:

- when the patient is pregnant

- if the disk is sequestered
- if the patient has had several failed back operations
- if a spinal cord tumor is present
- if the patient has a neurological disease such as multiple sclerosis

Other conditions may affect the appropriateness of chemonucleolysis, including **hypertension**, **obesity**, diabetes, and a family history of stroke.

Description

A small gauge needle is placed in the center of the affected disk. Chymopapain is introduced into the disk. The patient needs to remain still.

Preparation

Patients will need tests such as a myelogram or CT scan to pinpoint the herniated disk. Some doctors medicate the patient 24 hours prior to the operation in order to decrease the chances of post-operative lower back stiffness.

Aftercare

Patients may feel lower back stiffness, which goes away in few weeks. Heavy lifting and sports activities should be avoided for at least three months.

Risks

The greatest risk is that the patient may be allergic to chymopapain. The **death** rate for chemonucleolysis is only 0.02%. Complications overall are five to 10 times less than with conventional surgery, and the failure rate is roughly comparable to the failure rate in conventional disk surgery.

Normal results

Many patients feel immediate relief from pain, but, in about 30% of patients, maximal relief takes six weeks. The long term (seven to 20 years) success rate averages about 75%, which is comparable to the success rate for conventional surgery.

Resources

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Tish Davidson

Chemotherapy

Definition

Chemotherapy is treatment of **cancer** with **anti-cancer drugs**.

Purpose

The main purpose of chemotherapy is to kill cancer cells. It is usually used to treat patients with cancer that has spread from the place in the body where it started (metastasized). Chemotherapy destroys cancer cells anywhere in the body. It even kills cells that have broken off from the main tumor and traveled through the blood or lymph systems to other parts of the body.

Chemotherapy can cure some types of cancer. In some cases, it is used to slow the growth of cancer cells or to keep the cancer from spreading to other parts of the body. When a cancer has been removed by surgery, chemotherapy may be used to keep the cancer from coming back (adjuvant therapy). Chemotherapy also can ease the symptoms of cancer, helping some patients to have a better quality of life.

Precautions

There are many different types of chemotherapy drugs. Oncologists, doctors who specialize in treating cancer, determine which drugs are best suited for each patient. This decision is based on the type of cancer, the patient's age and health, and other drugs the patient is taking. Some patients should not be treated with certain chemotherapy drugs. Age and other conditions may affect the drugs with which a person may be treated. Heart disease, kidney disease, and diabetes are conditions that may limit the choice of treatment drugs.

Description

More than 50 chemotherapy drugs are currently available to treat cancer and many more are being tested for their ability to destroy cancer cells. Most chemotherapy drugs interfere with the ability of cells to grow or multiply. Although these drugs affect all cells in the body, many useful treatments are most effective against rapidly growing cells. Cancer cells grow more quickly than most other body cells. Other cells that grow fast are cells of the bone marrow that produce blood cells, cells in the stomach and intestines, and cells of the hair follicles. Therefore, the most common side effects of chemotherapy are linked to their effects on other fast growing cells.

Types of chemotherapy drugs

Chemotherapy drugs are classified based on how they work. The main types of chemotherapy drugs are described below:

- Alkylating drugs kill cancer cells by directly attacking DNA, the genetic material of the genes. Cyclophosphamide is an alkylating drug.
- Antimetabolites interfere with the production of DNA and keep cells from growing and multiplying. An example of an antimetabolite is 5-fluorouracil (5-FU).
- Antitumor **antibiotics** are made from natural substances such as fungi in the soil. They interfere with important cell functions, including production of DNA and cell proteins. Doxorubicin and bleomycin belong to this group of chemotherapy drugs.
- Plant alkaloids prevent cells from dividing normally. Vinblastine and vincristine are plant alkaloids obtained from the periwinkle plant.
- Steroid hormones slow the growth of some cancers that depend on hormones. For example, tamoxifen is used to treat breast cancers that depend on the hormone estrogen for growth.

Combination chemotherapy

Chemotherapy is usually given in addition to other cancer treatments, such as surgery and **radiation therapy**. When given with other treatments, it is called adjuvant chemotherapy. An oncologist decides which chemotherapy drug or combination of drugs will work best for each patient. The use of two or more drugs together often works better than a single drug for treating cancer. This is called combination chemotherapy. Scientific studies of different drug combinations help doctors learn which combinations work best for each type of cancer.

How chemotherapy is given

Chemotherapy is administered in different ways, depending on the drugs to be given and the type of cancer. Doctors decide the dose of chemotherapy drugs considering many factors, among them being the patient's height and weight.

Chemotherapy may be given by one or more of the following methods:

- orally
- by injection
- through a catheter or port
- topically

Oral chemotherapy is given by mouth in the form a pill, capsule, or liquid. This is the easiest method and can usually be done at home.

Intravenous (IV) chemotherapy is injected into a vein. A small needle is inserted into a vein on the hand or lower arm. The needle is usually attached to a small tube called a catheter, which delivers the drug to the needle from an IV bag or bottle.

Intramuscular (IM) chemotherapy is injected into a muscle. Chemotherapy given by intramuscular injection is absorbed into the blood more slowly than IV chemotherapy. Because of this, the effects of IM chemotherapy may last longer than chemotherapy given intravenously. Chemotherapy may also be injected subcutaneously (SQ or SC), which means under the skin. Injection of chemotherapy directly into the cancer is called intralesional (IL) injection.

Chemotherapy may also be given by a catheter or port permanently inserted into a central vein or body cavity. A port is a small reservoir or container that is placed in a vein or under the skin in the area where the drug will be given. These methods eliminate the need for repeated injections and may allow patients to spend less time in the hospital while receiving chemotherapy. A common location for a permanent catheter is the external jugular vein in the neck. Intraperitoneal (IP) chemotherapy is administered into the abdominal cavity through a catheter or port. Chemotherapy given by catheter or port into the spinal fluid is called intrathecal (IT) administration. Catheters and ports may also be placed in the chest cavity, bladder, or pelvis, depending on the location of the cancer to be treated.

Topical chemotherapy is given as a cream or ointment applied directly to the cancer. This method is more common in treatment of certain types of skin cancer.

Treatment location and schedule

Patients may take chemotherapy at home, in the doctor's office, or as an inpatient or outpatient at the hospital. Most patients stay in the hospital when first beginning chemotherapy, so their doctor can check for any side effects and change the dose if needed.

How often and how long chemotherapy is given depends on the type of cancer, how patients respond to the drugs, patients' health and ability to tolerate the drugs, and the types of drugs given. Chemotherapy administration may take only a few minutes or may last as long as several hours. Chemotherapy may be given daily, weekly, or monthly. A rest period may follow a course of treatment before the next course begins. In combination chemotherapy, more than one drug may be



Patient undergoing high dose stem cell chemotherapy.
(Custom Medical Stock Photo. Reproduced by permission.)

given at a time, or they may be given alternately, one following the other.

Preparation

A number of medical tests are done before chemotherapy is started. The oncologist will determine how much the cancer has spread from the results of x rays and other imaging tests and from samples of the tumor taken during surgery.

Blood tests give the doctor important information about the function of the blood cells and levels of chemicals in the blood. A complete **blood count** (CBC) is commonly done before and regularly during treatment. The CBC shows the numbers of white blood cells, red blood cells, and platelets in the blood. Because chemotherapy affects the bone marrow, where blood cells are made, levels of these cells often drop during chemotherapy. The white blood cells and platelets are most likely to be affected by chemotherapy. A drop in the

white blood cell count means that the immune system cannot function properly. Low levels of platelets can cause a patient to bleed easily from a cut or other wound. A low red blood cell count can lead to anemia (deficiency of red blood cells) and **fatigue**.

When a chemotherapy treatment takes a long time, the patient may prepare for it by wearing comfortable clothes. Bringing a book to read or a tape to listen to may help pass the time and ease the **stress** of receiving chemotherapy. Some patients bring a friend or family member to provide company and support during treatment.

Sometimes, patients taking chemotherapy drugs known to cause nausea are given medications called anti-emetics before chemotherapy is administered. Anti-emetic drugs help to lessen feelings of nausea. Two anti-nausea medications that may be used are Kytril and Zofran.

Other ways to prepare for chemotherapy and help lessen nausea are:

- regularly eat nutritious foods and drink lots of fluids
- eat and drink normally until about two hours before chemotherapy
- eat high carbohydrate, low-fat foods and avoid spicy foods

Aftercare

Tips for helping to control side effects after chemotherapy include:

- follow any instructions given by the doctor or nurse
- take all prescribed medications
- eat small amounts of bland foods
- drink lots of fluids
- get plenty of rest

Some patients find it helps to breathe fresh air or get mild **exercise**, such as taking a walk.

Risks

Chemotherapy drugs are toxic to normal cells as well as cancer cells. A dose that will destroy cancer cells will probably cause damage to some normal cells. Doctors adjust doses to do the least amount of harm possible to normal cells. Some patients feel few or no side effects, and others may have more serious side effects. In some cases, a dose adjustment is all that is needed to reduce or stop a side effect.

Some chemotherapy drugs have more side effects than others. Some of the most common side effects are:

- **nausea and vomiting**

- loss of appetite
- hair loss
- anemia and fatigue
- infection
- easy bleeding or bruising
- sores in the mouth and throat
- neuropathy and other damage to the nervous system
- kidney damage

Nausea and vomiting are common, but can usually be controlled by taking **antinausea drugs**, drinking enough fluids, and avoiding spicy foods. Loss of appetite may be due to nausea or the stress of undergoing cancer treatment.

Some chemotherapy drugs cause hair loss, but it is almost always temporary.

Low blood cell counts caused by the effect of chemotherapy on the bone marrow can lead to anemia, infections, and easy bleeding and bruising. Patients with anemia have too few red blood cells to deliver oxygen and nutrients to the body's tissues. Anemic patients feel tired and weak. If red blood cell levels fall too low, a blood **transfusion** may be given.

Patients receiving chemotherapy are more likely to get infections. This happens because their infection-fighting white blood cells are reduced. It is important to take measures to avoid getting infections. When the white blood cell count drops too low, the doctor may prescribe medications called colony stimulating factors that help white blood cells grow. Neupogen and Leukine are two colony stimulants used as treatments to help fight infection.

Platelets are blood cells that make the blood clot. When patients do not have enough platelets, they may bleed or bruise easily, even from small injuries. Patients with low blood platelets should take precautions to avoid injuries. Medicines such as **aspirin** and other **pain** relievers can affect platelets and slow down the clotting process.

Chemotherapy can cause irritation and dryness in the mouth and throat. Painful sores may form that can bleed and become infected. Precautions to avoid this side effect include getting dental care before chemotherapy begins, brushing the teeth and gums regularly with a soft brush, and avoiding mouth washes that contain salt or alcohol.

Normal results

The main goal of chemotherapy is to cure cancer. Many cancers are cured by chemotherapy. It may be used in combination with surgery to keep a cancer from spread-

KEY TERMS

Adjuvant therapy—Treatment given after surgery or radiation therapy to prevent the cancer from coming back.

Alkaloid—A type of chemical commonly found in plants and often having medicinal properties.

Alykylating drug—A drug that kills cells by directly damaging DNA.

Antiemetic—A medicine that helps control nausea; also called an anti-nausea drug.

Antimetabolite—A drug that interferes with a cell's growth or ability to multiply.

Platelets—Blood cells that function in blood clotting.

ing to other parts of the body. Some widespread, fast-growing cancers are more difficult to treat. In these cases, chemotherapy may slow the growth of the cancer cells.

Doctors can tell if the chemotherapy is working by the results of medical tests. **Physical examination**, blood tests, and x rays are all used to check the effects of treatment on the cancer.

The possible outcomes of chemotherapy are:

- Complete remission or response. The cancer completely disappears. The course of chemotherapy is completed and the patient is tested regularly for a recurrence.
- Partial remission or response. The cancer shrinks in size but does not disappear. The same chemotherapy may be continued or a different combination of drugs may be tried.
- Stabilization. The cancer does not grow or shrink. Other therapy options may be explored. A tumor may stay stabilized for many years.
- Progression. The cancer continues to grow. Other therapy options may be explored.
- A secondary malignancy may develop from the one being treated, and that second cancer may need additional chemotherapy or other treatment.

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Toni Rizzo

Chest drainage therapy

Definition

Chest drainage therapy involves the removal of air, blood, pus, or other secretions from the chest cavity.

Purpose

Chest drainage therapy is done to relieve pressure on the lungs, and remove fluid that could promote infection. Installing a chest drainage tube can be either an emergency or a planned procedure.

Removing air or fluids from the chest involves the insertion of a tube through the skin and the muscles between the ribs, and into the chest cavity. This cavity is also called the pleural space. Insertion of this tube is called thoracostomy, and chest drainage therapy is sometimes called thoracostomy tube drainage.

Conditions that may need to be treated by chest drainage therapy include **emphysema** (air in the tissues of the lungs), **tuberculosis**, and spontaneous **pneumothorax** (air in the chest cavity) that causes more than a 25% collapse of the lung. Other conditions include **cancer** that causes excessive secretions, **empyema** (pus in the thoracic cavity), or **hemothorax** (blood in the thoracic cavity). Almost all chest drainage therapy is done to drain blood from the chest cavity after lung or heart surgery. In cases where the lung is collapsed, removing fluids by chest drainage therapy allows the lung to reinflate.

Oftentimes an x ray is performed prior to treatment to determine whether the problem is either fluid or air in

KEY TERMS

Empyema—Pus in the pleural cavity.

Hemothorax—Blood in the pleural cavity.

Pleural cavity—The area of the chest that includes the lining of the chest cavity, the space the lungs are located in, and the membrane covering of the lungs.

Spontaneous pneumothorax—Air in the chest cavity that occurs because of disease or other naturally occurring cause. Air and blood together in this space is called a pneumohemothorax.

the pleural space. Sometimes a procedure called **thoracentesis** is performed in an effort to avoid inserting a chest drainage tube. In this procedure a needle with a catheter is inserted into the pleural space and fluid is removed. When fluid continues to accumulate, chest drainage therapy is usually the next step. This is especially true when there is a lung infection underlying the fluid build-up.

Precautions

Chest drainage therapy is not done if a collapsed lung is not life-threatening. It also should be avoided for patients who have blood clotting problems.

Description

Most patients are awake when the chest drainage tube is inserted. They are given a sedative and a local anesthetic. Chest drainage tubes are usually inserted between the ribs. The exact location depends on the type of material to be drained and its location in the lungs.

An incision is made in the skin and through the muscles between the ribs. A chest tube is inserted and secured in place. The doctor connects one end of the tube to the chest drainage system.

The chest drainage system must remain sealed to prevent air from entering the chest cavity through the tube. One commonly used system is a water-seal drainage system, comprised of three compartments that collect and drain the fluid or air without allowing air to backflow into the tube. An alternative to this system is to connect the tube to a negative suction pump.

Once the tube and drainage system are in place, a chest x ray is done to confirm that the tube is in the right location, and that it is working. In some cases it may be

necessary to insert more than one tube to drain localized pockets of fluid that have accumulated.

Preparation

A chest x ray is usually done before the chest drainage tube is inserted. Sometimes fluid becomes trapped in isolated spaces in the lung, and it is necessary to do an ultrasound to determine where to locate the drainage tube. **Computed tomography scans (CT)** are useful in locating small pockets of fluids caused by cancer or tuberculosis.

Aftercare

Normally after the material has been removed from the chest cavity and the situation is resolved, the chest drainage tube is removed. In cases where the reason for the tube was air in the pleural cavity, the tube is clamped and left in place several hours before it is removed to make sure no more air is leaking into the space. If the patient is on mechanical ventilation, the tube is often left in place until a respirator is no longer necessary. Chest drainage therapy is usually done in conjunction with treating the underlying cause of the fluid build-up.

The fluid that has been drained is examined for bacterial growth, cancer cells, pus, and blood—to determine the underlying cause of the condition and appropriate treatment.

Risks

Problems can arise in the insertion of the tube if the membrane lining the chest cavity is thick or if it has many adhesions. The tube will not drain correctly if the chest cavity contains blood clots or thick secretions that are often associated with infections. Excessive bleeding may occur during the insertion and positioning of the tube. Infection may result from the procedure. **Pain** is also a common complication.

Normal results

The gas, pus, or blood is drained from the chest cavity, and the lungs reinflate or begin to function more efficiently. The site at which the tube was inserted heals normally.

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Tish Davidson

Chest pain see **Angina**

Chest physical therapy

Definition

Chest physical therapy is the term for a group of treatments designed to improve respiratory efficiency, promote expansion of the lungs, strengthen respiratory muscles, and eliminate secretions from the respiratory system.

Purpose

The purpose of chest physical therapy, also called chest physiotherapy, is to help patients breathe more freely and to get more oxygen into the body. Chest physical therapy includes postural drainage, chest percussion, chest vibration, turning, deep breathing exercises, and coughing. It is usually done in conjunction with other treatments to rid the airways of secretions. These other treatments include suctioning, nebulizer treatments, and the administration of expectorant drugs.

Chest physical therapy can be used with newborns, infants, children, and adults. People who benefit from chest physical therapy exhibit a wide range of problems that make it difficult to clear secretions from their lungs. Some people who may receive chest physical therapy include people with **cystic fibrosis** or neuromuscular diseases like **Guillain-Barré syndrome**, progressive muscle weakness (**myasthenia gravis**), or **tetanus**. People with lung diseases such as **bronchitis**, **pneumonia**, or chronic obstructive pulmonary disease (COPD) also benefit from chest physical therapy. People who are likely to aspirate their mucous secretions because of diseases such as **cerebral palsy** or **muscular dystrophy** also receive chest physical therapy, as do some people who are bedridden, confined to a wheelchair, or who cannot breathe deeply because of postoperative **pain**.

Precautions

Chest physical therapy should not be performed on people with

- bleeding from the lungs
- neck or head injuries
- fractured ribs

- collapsed lungs
- damaged chest walls
- tuberculosis
- acute asthma
- recent heart attack
- pulmonary embolism
- lung **abscess**
- active hemorrhage
- some spine injuries
- recent surgery, open **wounds**, or **burns**

Description

Chest physical therapy can be performed in a variety of settings including critical care units, hospitals, nursing homes, outpatient clinics, and at the patient's home. Depending on the circumstances, chest physical therapy may be performed by anyone from a respiratory care therapist to a trained member of the patient's family. Different patient conditions warrant different levels of training.

Chest physical therapy consists of a variety of procedures that are applied depending on the patient's health and condition. Hospitalized patients are reevaluated frequently to establish which procedures are most effective and best tolerated. Patients receiving long term chest physical therapy are reevaluated about every three months.

Turning

Turning from side to side permits lung expansion. Patients may turn themselves or be turned by a caregiver. The head of the bed is also elevated to promote drainage if the patient can tolerate this position. Critically ill patients and those dependent on mechanical respiration are turned once every one to two hours around the clock.

Coughing

Coughing helps break up secretions in the lungs so that the mucus can be suctioned out or expectorated. Patients sit upright and inhale deeply through the nose. They then exhale in short puffs or coughs. Coughing is repeated several times a day.

Deep breathing

Deep breathing helps expand the lungs and forces better distribution of the air into all sections of the lung. The patient either sits in a chair or sits upright in bed and inhales, pushing the abdomen out to force maximum amounts of air into the lung. The abdomen is then contracted, and the patient exhales. Deep breathing exercises are done several times each day for short periods.

KEY TERMS

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Percussion—This consists of rhythmically striking the chest wall with cupped hands. It is also called cupping, clapping, or tapotement. The purpose of percussion is to break up thick secretions in the lungs so that they can be more easily removed. Percussion is performed on each lung segment for one to two minutes at a time.

Postural drainage—This technique uses the force of gravity to assist in effectively draining secretions from the lungs and into the central airway where

they can either be coughed up or suctioned out. The patient is placed in a head or chest down position and is kept in this position for up to 15 minutes. Critical care patients and those depending on mechanical ventilation receive postural drainage therapy four to six times daily. Percussion and vibration may be performed in conjunction with postural drainage.

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Vibration—The purpose of vibration is to help break up lung secretions. Vibration can be either mechanical or manual. It is performed as the patient breathes deeply. When done manually, the person performing the vibration places his or her hands against the patient's chest and creates vibrations by quickly contracting and relaxing arm and shoulder muscles while the patient exhales. The procedure is repeated several times each day for about five exhalations.

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Preparation

The only preparation needed for chest physical therapy is an evaluation of the patient's condition and determination of which chest physical therapy techniques would be most beneficial.

Aftercare

Patients practice **oral hygiene** procedures to lessen the bad taste or odor of the secretions they spit out.

Risks

Risks and complications associated with chest physical therapy depend on the health of the patient. Although

chest physical therapy usually poses few problems, in some patients it may cause

- oxygen deficiency if the head is kept lowered for drainage
- increased intracranial pressure
- temporary low blood pressure
- bleeding in the lungs
- pain or injury to the ribs, muscles, or spine
- vomiting
- inhaling secretions into the lungs
- heart irregularities

Normal results

The patient is considered to be responding positively to chest physical therapy if some, but not necessarily all, of these changes occur:

- increased volume of sputum secretions
- changes in breath sounds
- improved vital signs
- improved chest x ray
- increased oxygen in the blood as measured by arterial blood gas values
- patient reports of eased breathing

Resources

PERIODICALS

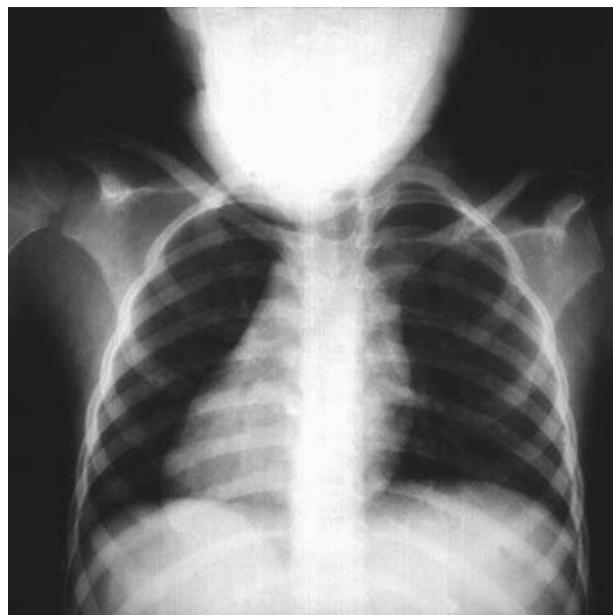
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ORGANIZATIONS

Cystic Fibrosis Foundation. 6931 Arlington Road, Bethesda, MD 20814. (800) 344-4823. <<http://www.cff.org>>.

Tish Davidson

Chest radiography see **Chest x ray**



A normal chest x ray of a child. (Photograph by Peter Berndt, M.D., P.A., Custom Medical Stock Photo. Reproduced by permission.)

that can penetrate the body and produce an image on an x-ray film. Another name for x ray is radiograph.

Purpose

Chest x rays are ordered for a wide variety of diagnostic purposes. In fact, this is probably the most frequently performed x ray. In some cases, chest x rays are ordered for a single check of an organ's condition, and at other times, serial x rays are ordered to compare to previous studies. Some common reasons for chest x rays include:

Pulmonary disorders

Chest films are frequently ordered to diagnose or rule out **pneumonia**. Other pulmonary disorders such as **emphysema** or **pneumothorax** (presence of air or gas in the chest cavity outside the lungs) may be detected or evaluated through the use of chest x ray.

Cancer

A chest x ray may be ordered by a physician to check for possible tumors of the lungs, thyroid, lymphoid tissue, or bones of the thorax. These may be primary tumors. X rays also check for secondary spread of **cancer** from one organ to another.

Cardiac disorders

While less sensitive than **echocardiography**, chest x ray can be used to check for disorders such as **congestive heart failure** or **pulmonary edema**.

KEY TERMS

Bronchi—Plural of bronchus. The air passages in the lungs through which inhaled air passes on its way to the lungs.

Diaphragm—The large muscle that is located between the abdomen and the chest area. The diaphragm aids in breathing.

Gastrointestinal—The digestive organs and structures, including the stomach and intestines.

Interstitial lung disease—About 180 diseases fall into this category of breathing disorders. Injury or foreign substances in the lungs, (such as asbestos fibers) as well as infections, cancers, or inherited disorders may cause the diseases. They can lead to breathing or heart failure.

Lymphoid—Tissues relating to the lymphatic system. A thin, yellowish fluid, called lymph fluid, travels throughout the body. The lymphatic system helps control fluids in the body.

Portable chest x ray—An x ray procedure taken by equipment that can be brought to the patient. The resulting radiographs may not be as high in quality as stationary x ray radiographs, but allow a technologist to come to the bedridden patient.

Pulmonary—Refers to the lungs and the breathing system and function.

Serial x rays—A number of x rays performed at set times in the disease progression or treatment intervals. The radiographs will be compared to one another to track changes.

Sternum—Also referred to as the breast bone, this is the long flat bone in the middle of the chest.

Thorax—The chest area, which runs between the abdomen and neck and is encased in the ribs.

X ray—A form of electromagnetic radiation with shorter wavelengths than normal light. X rays can penetrate most structures.

Other

Tuberculosis can be observed on chest x rays, as can cardiac disease and damage to the ribs or lungs. Chest x rays are used to see foreign bodies that may have been swallowed or inhaled, and to evaluate response to treatment for various diseases. Often the chest x ray is also used to verify correct placement of chest tubes or catheters.

Precautions

Pregnant women, particularly those in the first or second trimester, should not have chest x rays unless absolutely necessary. If the exam is ordered, women who are, or could possibly be, pregnant must wear a protective lead apron. Because the procedure involves radiation, care should always be taken to avoid overexposure, particularly for children. However, the amount of radiation from one chest x ray procedure is minimal.

Description

Routine chest x rays consist of two views, the frontal view (referred to as posterioranterior or PA), and the lateral (side) view. It is preferred that the patient stand for this exam, particularly when studying collection of fluid in the lungs.

During the actual time of exposure, the technologist will ask the patient to hold his or her breath. It is very important in taking a chest x ray to ensure there is no motion that could detract from the quality and sharpness of the film image. The procedure will only take a few minutes and the time patients must hold their breaths is a matter of a few seconds.

The chest x ray may be performed in a physician's office or referred to an outpatient radiology facility or hospital radiology department. In some cases, particularly for bedridden patients, a portable chest x ray may be taken. Portable films are sometimes of poorer quality than those taken with permanent equipment, but are the best choice for some patients or situations. Bedridden patients may be placed in an upright position as possible to get a clear picture, particularly of chest fluid.

Preparation

There is no advance preparation necessary for chest x rays. Once the patient arrives at the exam area, a hospital gown will replace all clothing on the upper body and all jewelry must be removed.

Aftercare

No aftercare is required by patients who have chest x rays.

Risks

The only risk associated with chest x ray is minimal exposure to radiation, particularly for pregnant women and children. Those patients should use protective lead aprons during the procedure. Technologists are cautioned to carefully check possible dislodging of any tubes or

monitors in the chest area from the patient's placement during the exam.

Normal results

A radiologist, or physician specially trained in the technique and interpretation of x rays, will evaluate the results. A normal chest x ray will show normal structures for the age and medical history of the patient. Findings, whether normal or abnormal, will be provided to the referring physician in the form of a written report.

Abnormal results

Abnormal findings on chest x rays are used in conjunction with a physician's physical exam findings, patient medical history and other diagnostic tests to reach a final diagnosis. For many diseases, chest x rays are more effective when compared to previous chest studies. The patient is asked to help the radiology facility in locating previous chest radiographs from other facilities.

Pulmonary disorders

Pneumonia shows up on radiographs as patches and irregular areas of density (from fluid in the lungs). If the bronchi, which are usually not visible, can be seen, a diagnosis of bronchial pneumonia may be made. Shifts or shadows in the hilum (lung roots) may indicate emphysema or a pulmonary **abscess**. Widening of the spaces between ribs suggests emphysema. Other pulmonary diseases may also be detected or suspected through chest x ray.

Cancer

In nearly all patients with lung cancer, some sort of abnormality can be seen on a chest radiograph. Hilar masses (enlargements at that part of the lungs where vessels and nerves enter) are one of the more common symptoms as are abnormal masses and fluid buildup on the outside surface of the lungs or surrounding areas. Interstitial lung disease, which is a large category of disorders, many of which are related to exposure of substances (such as asbestos fibers), may be detected on a chest x ray as fiberlike deposits, often in the lower portions of the lungs.

Other

Congestive heart failure and other cardiac diseases may be indicated on the view of a heart and lung in a chest radiograph. **Fractures** of the sternum and ribs are usually easily detected as breaks on the chest x ray. In some instances, the radiologist's view of the diaphragm may indicate an abdominal problem. Tuberculosis can

also be indicated by elevation of the diaphragm. Foreign bodies which may have been swallowed or inhaled can usually be located by the radiologist as they will look different from any other tissue or structure in the chest. Serial chest x rays may be ordered to track changes over a period of time.

Resources

ORGANIZATIONS

American Lung Association. 1740 Broadway, New York, NY 10019. (800) 586-4872. <<http://www.lungusa.org>>.

Emphysema Anonymous, Inc. P.O. Box 3224, Seminole, FL 34642. (813)391-9977.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

Teresa Norris, RN

Chickenpox

Definition

Chickenpox (also called varicella) is a common and extremely infectious childhood disease that also affects adults on occasion. It produces an itchy, blistery rash that typically lasts about a week and is sometimes accompanied by a **fever** or other symptoms. A single attack of chickenpox almost always confers lifelong immunity against the disease. Because the symptoms of chickenpox are easily recognized and in most cases merely unpleasant rather than dangerous, treatment can almost always be carried out at home. Severe complications can develop, however, and professional medical attention is essential in some circumstances.

Description

Before the varicella vaccine (Varivax) was released for use in 1995, virtually all of the four million children born each year in the United States contracted chickenpox, resulting in hospitalization in five of every 1,000 cases and 100 deaths. Chickenpox is caused by the varicella-zoster virus (a member of the herpes virus family), which is spread through the air or by direct contact with an infected person. Once someone has been infected with the virus, an incubation period of about 10–21 days passes before symptoms begin. The period during which infected people are able to spread the disease is believed to start one or two days before the rash breaks out and to continue until all the blisters have formed scabs, which usually happens four to seven days after the rash breaks

out but may be longer in adolescents and adults. For this reason, doctors recommend keeping children with chickenpox away from school for about a week. It is not necessary, however, to wait until all the scabs have fallen off.

Chickenpox has been a typical part of growing up for most children in the industrialized world (although this may change if the new varicella vaccine becomes more widely accepted). The disease can strike at any age, but by ages nine or 10 about 80–90% of American children have already been infected. U.S. children living in rural areas and many foreign-born children are less likely to be immune. Because almost every case of chickenpox, no matter how mild, leads to lifelong protection against further attacks, adults account for less than 5% of all cases in the United States. Study results reported by the Centers for Disease Control and Prevention (CDC) indicate that more than 90% of American adults are immune to the chickenpox virus. Adults, however, are much more likely than children to suffer dangerous complications. More than half of all chickenpox deaths occur among adults.

Causes and symptoms

A case of chickenpox usually starts without warning or with only a mild fever and a slight feeling of unwellness. Within a few hours or days small red spots begin to appear on the scalp, neck, or upper half of the trunk. After a further 12–24 hours the spots typically become itchy, fluid-filled bumps called vesicles, which continue to appear in crops for the next two to five days. In any area of skin, lesions of a variety of stages can be seen. These blisters can spread to cover much of the skin, and in some cases may also be found inside the mouth, nose, ears, vagina, or rectum. Some people develop only a few blisters, but in most cases the number reaches 250–500. The blisters soon begin to form scabs and fall off. Scarring usually does not occur unless the blisters have been scratched and become infected. Occasionally a minor and temporary darkening of the skin (called **hyperpigmentation**) is noticed around some of the blisters. The degree of itchiness can range from barely noticeable to extreme. Some chickenpox sufferers also have headaches, abdominal **pain**, or a fever. Full recovery usually takes five to 10 days after the first symptoms appear. Again, the most severe cases of the disease tend to be found among older children and adults.

Although for most people chickenpox is no more than a matter of a few days' discomfort, some groups are at risk for developing complications, the most common of which are bacterial infections of the blisters, **pneumonia**, **dehydration**, **encephalitis**, and hepatitis:

- Infants. Complications occur much more often among children less than one year old than among older children. The threat is greatest to newborns, who are more

at risk of **death** from chickenpox than any other group. Under certain circumstances, children born to mothers who contract chickenpox just prior to delivery face an increased possibility of dangerous consequences, including brain damage and death. If the infection occurs during early **pregnancy**, there is a small (less than 5%) risk of congenital abnormalities.

- Immunocompromised children. Children whose immune systems have been weakened by a genetic disorder, disease, or medical treatment usually experience the most severe symptoms of any group. They have the second-highest rate of death from chickenpox.
- Adults and children 15 and older. Among this group, the typical symptoms of chickenpox tend to strike with greater force, and the risk of complications is much higher than among young children.

Immediate medical help should always be sought when anyone in these high-risk groups contracts the disease.

Diagnosis

Where children are concerned, especially those with recent exposure to the disease, diagnosis can usually be made at home, by a school nurse, or by a doctor over the telephone if the child's parent or caregiver is unsure that the disease is chickenpox.

A doctor should be called immediately if:

- The child's fever goes above 102°F (38.9°C) or takes more than four days to disappear.
- The child's blisters appear infected. Signs of infection include leakage of pus from the blisters or excessive redness, warmth, tenderness, or swelling around the blisters.
- The child seems nervous, confused, unresponsive, or unusually sleepy; complains of a stiff neck or severe **headache**; shows signs of poor balance or has trouble walking; finds bright lights hard to look at; is having breathing problems or is coughing a lot; is complaining of chest pain; is vomiting repeatedly; or is having convulsions. These may be signs of **Reye's syndrome** or encephalitis, two rare but potentially very dangerous conditions.

Treatment

With children, treatment usually takes place in the home and focuses on reducing discomfort and fever. Because chickenpox is a viral disease, **antibiotics** are ineffective against it.

Applying wet compresses or bathing the child in cool or lukewarm water once a day can help the itch. Adding

four to eight ounces of baking soda or one or two cups of oatmeal to the bath is a good idea (oatmeal bath packets are sold by pharmacies). Only mild soap should be used in the bath. Patting, not rubbing, is recommended for drying the child off, to prevent irritating the blisters. Calamine lotion (and some other kinds of lotions) also help to reduce itchiness. Because scratching can cause blisters to become infected and lead to scarring, the child's nails should be cut short. Of course, older children need to be warned not to scratch. For babies, light mittens or socks on the hands can help guard against scratching.

If mouth blisters make eating or drinking an unpleasant experience, cold drinks and soft, bland foods can ease the child's discomfort. Painful genital blisters can be treated with anesthetic cream recommended by a doctor or pharmacist. Antibiotics are often prescribed if blisters become infected.

Fever and discomfort can be reduced by **acetaminophen** or another medication that does not contain **aspirin**. *Aspirin and any medications that contain aspirin or other salicylates must not be used with chickenpox, for they appear to increase the chances of developing Reye's syndrome.* The best idea is to consult a doctor or pharmacist if one is unsure about which medications are safe.

Immunocompromised chickenpox sufferers are sometimes given an antiviral drug called acyclovir (Zovirax). Studies have shown that Zovirax also lessens the symptoms of otherwise healthy children and adults who contract chickenpox, but the suggestion that it should be used to treat the disease among the general population, especially in children, is controversial.

Alternative treatment

Alternative practitioners seek to lessen the discomfort and fever caused by chickenpox. Like other practitioners, they suggest cool or lukewarm baths. Rolled oats (*Avena sativa*) in the bath water help relieve **itching**. (Place oats in a sock, run the bath, turn the sock to release the milky anti-itch properties.) Other recommended remedies for itching include applying aloe vera, witch hazel, or herbal preparations of rosemary (*Rosmarinus officinalis*) and calendula (*Calendula officinalis*) to the blisters. Homeopathic remedies are selected on a case by case basis. Some common remedy choices are tartar emetic (*Antimonium tartaricum*), windflower (*Pulsatilla*), poison ivy (*Rhus toxicodendron*), and sulphur.

Prognosis

Most cases of chickenpox run their course within a week without causing lasting harm. However, there is



A five-year-old girl with chickenpox. The first symptom of the disease is the rash that is evident on the girl's back and neck. The rash and the mild fever that accompanies it should disappear in a week or two. (Photograph by Jim Selby, Photo Researchers, Inc. Reproduced by permission.)

one long-term consequence of chickenpox that strikes about 20% of the population, particularly people 50 and older. Like all herpes viruses, the varicella-zoster virus never leaves the body after an episode of chickenpox, but lies dormant in the nerve cells, where it may be reactivated years later by disease or age-related weakening of the immune system. The result is **shingles** (also called herpes zoster), a very painful nerve inflammation, accompanied by a rash, that usually affects the trunk or the face for 10 days or more. Especially in the elderly, pain, called postherpetic **neuralgia**, may persist at the site of the shingles for months or years. As of 1998, two newer drugs for treatment of shingles are available. Both valacyclovir (Valtrex) and famciclovir (Famvir) stop the replication of herpes zoster when administered within 72 hours of appearance of the rash. The effectiveness of these two drugs in immunocompromised patients has not

KEY TERMS

Acetaminophen—A drug for relieving pain and fever. Tylenol is the most common example.

Acyclovir—An antiviral drug used for combating chickenpox and other herpes viruses. Sold under the name Zovirax.

Dehydration—Excessive water loss by the body.

Encephalitis—A disease that inflames the brain.

Hepatitis—A disease that inflames the liver.

Immune system—A biochemical complex that protects the body against pathogenic organisms and other foreign bodies.

Immunocompromised—Having a damaged immune system.

Pneumonia—A disease that inflames the lungs.

Pus—A thick yellowish or greenish fluid containing inflammatory cells. Usually caused by bacterial infection.

Reye's syndrome—A rare but often fatal disease that involves the brain, liver, and kidneys.

Salicylates—Substances containing salicylic acid, which are used for relieving pain and fever. Aspirin is the most common example.

Shingles—A disease (also called herpes zoster) that causes a rash and a very painful nerve inflammation. An attack of chickenpox will eventually give rise to shingles in about 20% of the population.

Trunk—That part of the body that does not include the head, arms, and legs.

Varicella-zoster immune globulin (VZIG)—A substance that can reduce the severity of chickenpox symptoms.

Varicella-zoster virus—The virus that causes chickenpox and shingles.

Varivax—A vaccine for the prevention of chickenpox.

Virus—A tiny particle that can cause infections by duplicating itself inside a cell using the cell's own software. Antibiotics are ineffective against viruses, though antiviral drugs exist for some viruses, including chickenpox.

been established, and Famvir is not recommended for patients under 18 years, as of 1998.

Prevention

A substance known as varicella-zoster immune globulin (VZIG), which reduces the severity of chickenpox symptoms, is available to treat immunocompromised children and others at high risk of developing complications. It is administered by injection within 96 hours of known or suspected exposure to the disease and is not useful after that. VZIG is produced as a gamma globulin from blood of recently infected individuals.

A vaccine for chickenpox became available in the United States in 1995 under the name Varivax. Varivax is a live, attenuated (weakened) virus vaccine. It has been proven to be 85% effective for preventing all cases of chickenpox and close to 100% effective in preventing severe cases. Side effects are normally limited to occasional soreness or redness at the injection site. CDC guidelines state that the vaccine should be given to all children (with the exception of certain high-risk groups) at 12–18 months of age, preferably when they

receive their measles-mumps-rubella vaccine. For older children, up to age 12, the CDC recommends **vaccination** when a reliable determination that the child in question has already had chickenpox cannot be made. Vaccination is also recommended for any older child or adult considered susceptible to the disease, particularly those, such as health care workers and women of child-bearing age, who face a greater likelihood of severe illness or transmitting infection. A single dose of the vaccine is sufficient for children up to age 12; older children and adults receive a second dose four to eight weeks later. In 1997 the cost of two adult doses of the vaccine in the United States was about \$80. Although this cost was not always covered by health insurance plans, children up to age 18 without access to the appropriate coverage could be vaccinated free of charge through the federal Vaccines for Children program. Varivax is not given to patients who already have overt signs of the disease. The vaccine is also not recommended for those women who are pregnant, or they should delay pregnancy for three months following a complete vaccination. The vaccine is useful when given early after exposure to chickenpox and, if given in the midst of the incubation period, it can be preventative. The Infectious Diseases Society of America stated in

2000 that immunization is recommended for all adults who have never had chickenpox.

While there was initial concern regarding the vaccine's safety and effectiveness when first released, the vaccination is gaining acceptance as numerous states require it for admittance into day care or public school. In 2000, 59% of toddlers in the United States were immunized; up from 43.2% in 1998. A study published in 2001 indicates that the varicella vaccine is highly effective when used in clinical practice. Although evidence has not ruled out a booster shot later in life, all research addressing the vaccine's effectiveness throughout its six-year use indicates that chickenpox may be the first human herpesvirus to be wiped out. Although initial concerns questioned if the vaccination might make shingles more likely, studies are beginning to show the effectiveness of the vaccine in reducing cases of that disease.

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ORGANIZATION

Centers for Disease Control and Prevention. National Immunization Hotline. 1600 Clifton Rd. NE, Atlanta, GA 30333. (800) 232-2522 (English). (800) 232-0233 (Spanish). <<http://www.cdc.gov>>.

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Beth Kapes

Child abuse

Definition

Child abuse is the blanket term for four types of child mistreatment: physical abuse, sexual abuse, emotional abuse, and neglect. In many cases children are the victims of more than one type of abuse. The abusers can be parents or other family members, caretakers such as teachers and babysitters, acquaintances (including other children), and (in rare instances) strangers.

Description

Prevalence of abuse

Child abuse was once viewed as a minor social problem affecting only a handful of United States children. However, in recent years it has received close attention from the media, law enforcement, and the helping professions, and with increased public and professional awareness has come a sharp rise in the number of reported cases. But because abuse is often hidden from view and its victims too young or fearful to speak out, experts suggest that its true prevalence is possibly much greater than the official data indicate. In 1996, more than three million victims of alleged abuse were reported to child protective services (CPS) agencies in the United States, and the reports were substantiated in more than one million cases. Put another way, 1.5% of the country's children were confirmed victims of abuse in 1996. Parents were the abusers in 77% of the confirmed cases, other relatives in 11%. Sexual abuse was more likely to be committed by males, whereas females were responsible for the majority of neglect cases. More than 1,000 United States children died from abuse in 1996.

Although experts are quick to point out that abuse occurs among all social, ethnic, and income groups, reported cases usually involve poor families with little education. Young mothers, single-parent families, and parental alcohol or drug abuse are also common in reported cases. Charles F. Johnson remarks, "More than 90% of abusing parents have neither psychotic nor criminal personalities. Rather they tend to be lonely, unhappy, angry, young, and single parents who do not plan their pregnancies, have little or no knowledge of child development, and have unrealistic expectations for child behavior." About 10%, or perhaps as many as 40%, of abusive parents were themselves physically abused as children, but most abused children do not grow up to be abusive parents.

Types of abuse

PHYSICAL ABUSE. Physical abuse is the nonaccidental infliction of physical injury to a child. The abuser is

usually a family member or other caretaker, and is more likely to be male. In 1996, 24% of the confirmed cases of United States child abuse involved physical abuse.

A rare form of physical abuse is **Munchausen syndrome** by proxy, in which a caretaker (most often the mother) seeks attention by making the child sick or appear to be sick.

SEXUAL ABUSE. Charles F. Johnson defines child sexual abuse as "any activity with a child, before the age of legal consent, that is for the sexual gratification of an adult or a significantly older child." It includes, among other things, sexual touching and penetration, persuading a child to expose his or her sexual organs, and allowing a child to view pornography. In most cases the child is related to or knows the abuser, and about one in five abusers are themselves underage. Sexual abuse was present in 12% of the confirmed 1996 abuse cases. An estimated 20–25% of females and 10–15% of males report that they were sexually abused by age 18.

EMOTIONAL ABUSE. Emotional abuse, according to Richard D. Krugman, "has been defined as the rejection, ignoring, criticizing, **isolation**, or terrorizing of children, all of which have the effect of eroding their self-esteem." Emotional abuse usually expresses itself in verbal attacks involving rejection, scapegoating, belittlement, and so forth. Because it often accompanies other types of abuse and is difficult to prove, it is rarely reported, and accounted for only 6% of the confirmed 1996 cases.

NEGLECT. Neglect—failure to satisfy a child's basic needs—can assume many forms. Physical neglect is the failure (beyond the constraints imposed by poverty) to provide adequate food, clothing, shelter, or supervision. Emotional neglect is the failure to satisfy a child's normal emotional needs, or behavior that damages a child's normal emotional and psychological development (such as permitting drug abuse in the home). Failing to see that a child receives proper schooling or medical care is also considered neglect. In 1996 neglect was the finding in 52% of the confirmed abuse cases.

Causes and symptoms

Physical abuse

The usual physical abuse scenario involves a parent who loses control and lashes out at a child. The trigger may be normal child behavior such as crying or dirtying a diaper. Unlike nonabusive parents, who may become angry at or upset with their children from time to time but are genuinely loving, abusive parents tend to harbor deep-rooted negative feelings toward their children.

Unexplained or suspicious **bruises** or other marks on the skin are typical signs of physical abuse, as are

burns. Skull and other bone **fractures** are often seen in young abused children, and in fact, head injuries are the leading cause of **death** from abuse. Children less than one year old are particularly vulnerable to injury from shaking. This is called **shaken baby syndrome** or shaken impact syndrome. Not surprisingly, physical abuse also causes a wide variety of behavioral changes in children.

Sexual abuse

John M. Leventhal observes, "The two prerequisites for this form of maltreatment include sexual arousal to children and the willingness to act on this arousal. Factors that may contribute to this willingness include alcohol or drug abuse, poor impulse control, and a belief that the sexual behaviors are acceptable and not harmful to the child." The chances of abuse are higher if the child is developmentally handicapped or vulnerable in some other way.

Genital or anal injuries or abnormalities (including the presence of **sexually transmitted diseases**) can be signs of sexual abuse, but often there is no physical evidence for a doctor to find. In fact, physical examinations of children in cases of suspected sexual abuse supply grounds for further suspicion only 15–20% of the time. **Anxiety**, poor academic performance, and suicidal conduct are some of the behavioral signs of sexual abuse, but are also found in children suffering other kinds of **stress**. Excessive masturbation and other unusually sexualized kinds of behavior are more closely associated with sexual abuse itself.

Emotional abuse

Emotional abuse can happen in many settings: at home, at school, on sports teams, and so on. Some of the possible symptoms include loss of self-esteem, sleep disturbances, headaches or stomachaches, school avoidance, and running away from home.

Neglect

Many cases of neglect occur because the parent experiences strong negative feelings toward the child. At other times, the parent may truly care about the child, but lack the ability or strength to adequately provide for the child's needs because he or she is handicapped by depression, drug abuse, **mental retardation**, or some other problem.

Neglected children often do not receive adequate nourishment or emotional and mental stimulation. As a result, their physical, social, emotional, and mental development is hindered. They may, for instance, be

Child Abuse: Signs And Symptoms

Although these signs do not necessarily indicate that a child has been abused, they may help adults recognize that something is wrong. The possibility of abuse should be investigated if a child shows a number of these symptoms, or any of them to a marked degree:

Sexual Abuse

- Being overly affectionate or knowledgeable in a sexual way inappropriate to the child's age
- Medical problems such as chronic itching, pain in the genitals, venereal diseases
- Other extreme reactions, such as depression, self-mutilation, suicide attempts, running away, overdoses, anorexia
- Personality changes such as becoming insecure or clinging
- Regressing to younger behavior patterns such as thumb sucking or bringing out discarded cuddly toys
- Sudden loss of appetite or compulsive eating
- Being isolated or withdrawn
- Inability to concentrate
- Lack of trust or fear someone they know well, such as not wanting to be alone with a babysitter
- Starting to wet again, day or night/nightmares
- Become worried about clothing being removed
- Suddenly drawing sexually explicit pictures
- Trying to be "ultra-good" or perfect; overreacting to criticism

Physical Abuse

- Unexplained recurrent injuries or burns
- Improbable excuses or refusal to explain injuries
- Wearing clothes to cover injuries, even in hot weather
- Refusal to undress for gym
- Bald patches
- Chronic running away
- Fear of medical help or examination
- Self-destructive tendencies
- Aggression towards others
- Fear of physical contact—shrinking back if touched
- Admitting that they are punished, but the punishment is excessive (such as a child being beaten every night to "make him/her study")
- Fear of suspected abuser being contacted

Emotional Abuse

- Physical, mental, and emotional development lags
- Sudden speech disorders
- Continual self-depreciation ("I'm stupid, ugly, worthless, etc.")
- Overreaction to mistakes
- Extreme fear of any new situation
- Inappropriate response to pain ("I deserve this")
- Neurotic behavior (rocking, hair twisting, self-mutilation)
- Extremes of passivity or aggression

Neglect

- Constant hunger
- Poor personal hygiene
- No social relationships
- Constant tiredness
- Poor state of clothing
- Compulsive scavenging
- Emaciation
- Untreated medical problems
- Destructive tendencies

A child may be subjected to a combination of different kinds of abuse. It is also possible that a child may show no outward signs and hide what is happening from everyone.

underweight, develop language skills less quickly than other children, and seem emotionally needy.

Diagnosis

Doctors and many other professionals who work with children are required by law to report suspected abuse to their state's Child Protective Services (CPS) agency. Abuse investigations are often a group effort

involving medical personnel, social workers, police officers, and others. Some hospitals and communities maintain child protection teams that respond to cases of possible abuse. Careful questioning of the parents is crucial, as is interviewing the child (if he or she is capable of being interviewed). The investigators must ensure, however, that their questioning does not further traumatize the child. A **physical examination** for signs of abuse or neglect is, of course, always

necessary, and may include x rays, blood tests, and other procedures.

Treatment

Notification of the appropriate authorities, treatment of the child's injuries, and protecting the child from further harm are the immediate priorities in abuse cases. If the child does not require hospital treatment, protection often involves placing him or her with relatives or in foster care. Once the immediate concerns are dealt with, it becomes essential to determine how the child's long-term medical, psychological, educational, and other needs can best be met, a process that involves evaluating not only the child's needs but also the family's (such as for drug abuse counseling or parental skills training). If the child has brothers or sisters, the authorities must determine whether they have been abused as well. On investigation, signs of physical abuse are discovered in about 20% of the brothers and sisters of abused children.

Prognosis

Child abuse can have lifelong consequences. Research shows that abused children and adolescents are more likely, for instance, to do poorly in school, suffer emotional problems, develop an antisocial personality, become promiscuous, abuse drugs and alcohol, and attempt suicide. As adults they often have trouble establishing intimate relationships. Whether professional treatment is able to moderate the long-term psychological effects of abuse is a question that remains unanswered.

Prevention

Government efforts to prevent abuse include home-visitor programs aimed at high-risk families and school-based efforts to teach children how to respond to attempted sexual abuse. Emotional abuse prevention has been promoted through the media.

When children reach age three, parents should begin teaching them about "bad touches" and about confiding in a suitable adult if they are touched or treated in a way that makes them uneasy. Parents also need to exercise caution in hiring babysitters and other caretakers. Any-one who suspects abuse should immediately report those suspicions to the police or his or her local CPS agency, which will usually be listed in the blue pages of the telephone book under Rehabilitative Services or Child and Family Services, or in the yellow pages. Round-the-clock crisis counseling for children and adults is offered by the Childhelp USA/IOF Foresters National Child Abuse Hotline. The National Committee to Prevent Child Abuse is an excellent source of information on the many

support groups and other organizations that help abused and at-risk children and their families. One of these organizations, National Parents Anonymous, sponsors 2,100 local self-help groups throughout the United States, Canada, and Europe. Telephone numbers for its local groups are listed in the white pages of the telephone book under Parents Anonymous or can be obtained by calling the national headquarters.

Resources

BOOKS

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Krugman, Richard D. "Child Abuse & Neglect." In *Pediatric Diagnosis & Treatment*, ed. William W. Hay Jr., et al. Stamford: Appleton & Lange, 1997.

Leventhal, John M. "Child Maltreatment: Neglect to Abuse." In *Rudolph's Pediatric*, ed. Abraham M. Rudolph, et al. Stamford: Appleton & Lange, 1996.

ORGANIZATIONS

Childhelp USA/IOF Foresters National Child Abuse Hotline. (800) 422-4453.

National Clearinghouse on Child Abuse and Neglect Information. P.O. Box 1182, Washington, DC 20013-1182. (800) 394-3366. <<http://www.calib.com/nccanch>>.

National Committee to Prevent Child Abuse. 200 S. Michigan Ave., 17th Floor, Chicago, IL 60604. (312) 663-3520. <<http://www.childabuse.org>>.

National Parents Anonymous. 675 W. Foothill Blvd., Suite 220, Claremont, CA 91711. (909) 621-6184.

Howard Baker

Child development see **Children's health**

Child safety see **Children's health**

Childbirth

Definition

Childbirth includes both labor (the process of birth) and delivery (the birth itself); it refers to the entire process as an infant makes its way from the womb down the birth canal to the outside world.

Description

Childbirth usually begins spontaneously, following about 280 days after conception, but it may be started by artificial means if the **pregnancy** continues past 42

weeks gestation. The average length of labor is about 14 hours for a first pregnancy and about eight hours in subsequent pregnancies. However, many women experience a much longer or shorter labor.

Labor can be described in terms of a series of phases.

First stage of labor

During the first phase of labor, the cervix dilates (opens) from 0–10 cm. This phase has an early, or latent, phase and an active phase. During the latent phase, progress is usually very slow. It may take quite a while and many contractions before the cervix dilates the first few centimeters. Contractions increase in strength as labor progresses. Most women are relatively comfortable during the latent phase and walking around is encouraged, since it naturally stimulates the process.

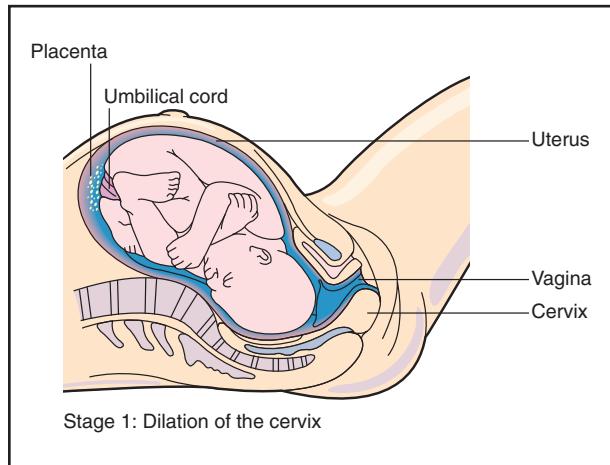
As labor begins, the muscular wall of the uterus begins to contract as the cervix relaxes and expands. As a portion of the amniotic sac surrounding the baby is pushed into the opening, it bursts under the pressure, releasing amniotic fluid. This is called “breaking the bag of waters.”

During a contraction, the infant experiences intense pressure that pushes it against the cervix, eventually forcing the cervix to stretch open. At the same time, the contractions cause the cervix to thin. During this first stage, a woman’s contractions occur more and more often and last longer and longer. The doctor or nurse will do a periodic **pelvic exam** to determine how the mother is progressing. If the contractions aren’t forceful enough to open the cervix, a drug may be given to make the uterus contract.

As **pain** and discomfort increase, women may be tempted to request pain medication. If possible, though, administration of pain medication or anesthetics should be delayed until the active phase of labor begins—at which point the medication will not act to slow down or stop the labor.

The active stage of labor is faster and more efficient than the latent phase. In this phase, contractions are longer and more regular, usually occurring about every two minutes. These stronger contractions are also more painful. Women who use the breathing exercises learned in childbirth classes find that these can help cope with the pain experienced during this phase. Many women also receive some pain medication at this point—either a short-term medication, such as Nubain or Numorphan, or an epidural anesthesia.

As the cervix dilates to 8–9 cm, the phase called the transition begins. This refers to the transition from the first phase (during which the cervix dilates from 0–10 cm) and the second phase (during which the baby is pushed out through the birth canal). As the baby’s head



Stage 1: Dilation of the cervix. (Illustration by Hans & Cassady.)

begins to descend, women begin to feel the urge to “push” or bear down. Active pushing by the mother should not begin until the second phase, since pushing too early can cause the cervix to swell or to tear and bleed. The attending healthcare practitioner should counsel the mother on when to begin to push.

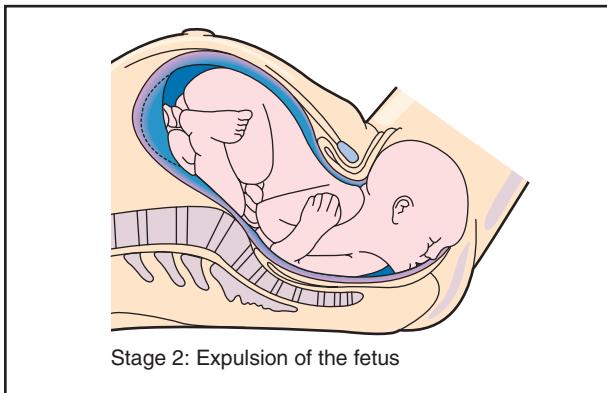
Second stage of labor

As the mother enters the second stage of labor, her baby’s head appears at the top of the cervix. Uterine contractions get stronger. The infant passes down the vagina, helped along by contractions of the abdominal muscles and the mother’s pushing. Active pushing by the mother is very important during this phase of labor. If an epidural anesthetic is being used, many practitioners recommend decreasing the amount administered during this phase of labor so that the mother has better control over her abdominal muscles.

When the top of the baby’s head appears at the opening of the vagina, the birth is nearing completion. First the head passes under the pubic bone. It fills the lower vagina and stretches the perineum (the tissues between the vagina and the rectum). This position is called “crowning,” since only the crown of the head is visible. When the entire head is out, the shoulders follow. The attending practitioner suctions the baby’s mouth and nose to ease the baby’s first breath. The rest of the baby usually slips out easily, and the umbilical cord is cut.

Episiotomy

As the baby’s head appears, the perineum may stretch so tight that the baby’s progress is slowed down. If there is risk of tearing the mother’s skin, the doctor may choose to make a small incision into the perineum to



Stage 2: Expulsion of the fetus. (Illustration by Hans & Cassidy.)

enlarge the vaginal opening. This is called an **episiotomy**. If the woman has not had an epidural or pudendal block, she will get a local anesthetic to numb the area. Once the episiotomy is made, the baby is born with a few pushes.

Third stage

In the final stage of labor, the placenta is pushed out of the vagina by the continuing uterine contractions. The placenta is pancake shaped and about 10 inches in diameter. It has been attached to the wall of the uterus and has served to convey nourishment from the mother to the fetus throughout the pregnancy. Continuing uterine contractions cause it to separate from the uterus at this point. It is important that all of the placenta be removed from the uterus. If it is not, the uterine bleeding that is normal after delivery may be much heavier.

Breech presentation

Approximately 4% of babies are in what is called the “breech” position when labor begins. In breech presentation, the baby’s head is not the part pressing against the cervix. Instead the baby’s bottom or legs are positioned to enter the birth canal instead of the head. An obstetrician may attempt to turn the baby to a head down position using a technique called version. This is only successful approximately half the time.

The risks of vaginal delivery with breech presentation are much higher than with a head-first presentation and the mother and attending practitioner will need to weigh the risks and make a decision on whether to deliver via a **cesarean section** or attempt a vaginal birth. The extent of the risk depends to a great extent on the type of breech presentation—of which there are three. Frank breech (the baby’s legs are folded up against its body) is the most common and the safest for vaginal delivery. The other types are

complete breech (in which the baby’s legs are crossed under and in front of the body) and footling breech (in which one leg or both legs are positioned to enter the birth canal) are not considered safe to attempt vaginal delivery.

Even in complete breech, other factors should be met before considering a vaginal birth. An ultrasound examination should be done to be sure the baby does not have an unusually large head and that the head is tilted forward (flexed) rather than back (hyperextended). Fetal monitoring and close observation of the progress of labor are also important. A slowing of labor or any indication of difficulty in the body passing through the pelvis should be an indication that it is safer to consider a cesarean section.

Forceps delivery

If the labor is not progressing as it should or if the baby appears to be in distress, the doctor may opt for a forceps delivery. A forceps is a spoon-shaped device that resembles a set of salad tongs. It is placed around the baby’s head so the doctor can pull the baby gently out of the vagina.

Forceps can be used after the cervix is fully dilated, and they might be required if:

- the umbilical cord has dropped down in front of the baby into the birth canal
- the baby is too large to pass through the birth canal unaided
- the baby shows signs of stress
- the mother is too exhausted to push

Before placing the forceps around the baby’s head, pain medication or anesthesia may be given to the mother. The doctor may use a catheter to empty the mother’s bladder, and may clean the perineal area with soapy water. Often an episiotomy is done before a forceps birth, although tears can still occur.

The obstetrician slides half of the forceps at a time into the vagina and around the side of the baby’s head to gently grasp the head. When both “tongs” are in place, the doctor pulls on the forceps to help the baby through the birth canal as the uterus contracts. Sometimes the baby can be delivered this way after the very next contraction.

The frequency of forceps delivery varies from one hospital to the next, depending on the experience of staff and the types of anesthesia offered at the hospital. Some obstetricians accept the need for a forceps delivery as a way to avoid cesarean birth. However, other obstetrical services don’t use forceps at all.

Complications from forceps deliveries can occur. Sometimes they may cause nerve damage or temporary

bruises to the baby's face. When used by an experienced physician, forceps can save the life of a baby in distress.

Vacuum-assisted birth

This method of helping a baby out of the birth canal was developed as a gentler alternative to forceps. Vacuum-assisted birth can only be used after the cervix is fully dilated (expanded), and the head of the fetus has begun to descend through the pelvis. In this procedure, the doctor uses a device called a vacuum extractor, placing a large rubber or plastic cup against the baby's head. A pump creates suction that gently pulls on the cup to ease the baby down the birth canal. The force of the suction may cause a bruise on the baby's head, but it fades away in a day or so.

The vacuum extractor is not as likely as forceps to injure the mother, and it leaves more room for the baby to pass through the pelvis. However, there may be problems in maintaining the suction during the vacuum-assisted birth, so forceps may be a better choice if it is important to remove the baby quickly.

Cesarean sections

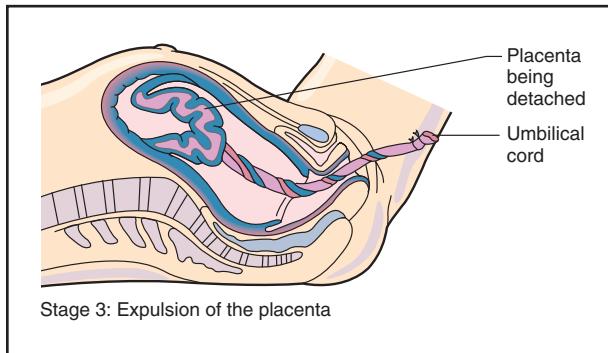
A cesarean section, also called a c-section, is a surgical procedure in which incisions are made through a woman's abdomen and uterus to deliver her baby.

Cesarean sections are performed whenever abnormal conditions complicate labor and vaginal delivery, threatening the life or health of the mother or the baby. The procedure is performed in the United States on nearly one of every four babies delivered—more than 900,000 babies each year. The procedure is used in cases where the mother has had a previous c-section and the area of the incision has been weakened. Dystocia, or difficult labor, is the another common reason for performing a c-section.

Difficult labor is commonly caused by one of the three following conditions: abnormalities in the mother's birth canal; abnormalities in the position of the fetus; abnormalities in the labor, including weak or infrequent contractions.

Another major factor is fetal distress, a condition where the fetus is not getting enough oxygen. Fetal brain damage can result from oxygen deprivation. Fetal distress is often related to abnormalities in the position of the fetus, or abnormalities in the birth canal, causing reduced blood flow through the placenta.

Other conditions also can make c-section advisable, such as vaginal herpes, **hypertension** (high blood pressure) and diabetes in the mother.



Stage 3: Expulsion of the placenta. (Illustration by Hans & Cassady.)

Causes and symptoms

One of the first signs of approaching childbirth may be a "bloody show," the appearance of a small amount of blood-tinged mucus released from the cervix as it begins to dilate. This is called the "mucus plug."

The most common sign of the onset of labor is contractions. Sometimes women have trouble telling the difference between true and false labor pains.

True labor pains:

- develop a regular pattern, with contractions coming closer together
- last from 15–30 seconds at the onset and get progressively stronger and longer (up to 60 seconds)
- may get stronger with physical activity
- occur high up on the abdomen, radiating throughout the abdomen and lower back

Another sign that labor is beginning is the breaking of the "bag of waters," the amniotic sac which had cushioned the baby during the pregnancy. When it breaks, it releases water in a trickle or a gush. Only about 10% of women actually experience this water flow in the beginning of labor, however. Most of the time, the rupture occurs sometime later in labor. If the amniotic sac doesn't rupture on its own, the doctor will break it during labor.

Some women have **diarrhea** or nausea as labor begins. Others notice a sudden surge of energy and the urge to clean or arrange things right before labor begins; this is known as "nesting."

Diagnosis

The onset of labor can be determined by measuring how much the cervix has dilated. The degree of dilation is estimated by feeling the opening cervix during a pelvic exam. Dilation is measured in centimeters, from zero to

KEY TERMS

Amniotic sac—The membranous sac that surrounds the embryo and fills with watery fluid as pregnancy advances.

Breech birth—Birth of a baby bottom-first, instead of the usual head first delivery. This can add to labor and delivery problems because the baby's bottom doesn't mold a passage through the birth canal as well as does the head.

Cervix—A small cylindrical organ about an inch or so long and less than an inch around that makes up the lower part and neck of the uterus. The cervix separates the body and cavity of the uterus from the vagina.

Embryo—The unborn child during the first eight weeks of its development following conception.

Gestation—The period from conception to birth, during which the developing fetus is carried in the uterus.

Perineum—The area between the thighs that lies behind the genital organs and in front of the anus.

Placenta—The organ that develops in the uterus during pregnancy and that links the blood supplies of mother and baby.

10. Contractions that cause the cervix to dilate are the sign of true labor.

Fetal monitoring

Fetal monitoring is a process in which the baby's heart rate is monitored for indicators of stress during labor and birth. There are several types of fetal monitoring.

A special stethoscope called a fetoscope may be used. This is a simple and non-invasive method.

The Doppler method uses ultrasound; it involves a handheld listening device that transmits the sounds of the heart rate through a speaker or into an attached ear piece. It can usually pick up the heart sounds 12 weeks after conception. This method offers intermittent monitoring. It allows the mother freedom to move about and is also useful during contractions.

Electronic fetal monitoring uses ultrasound and provides a view of the heartbeat in relationship to the mother's contractions. It can be used either continuously or intermittently. It is often used in high risk pregnancies, and is not often recommended for low risk ones

because it renders the mother immobile and requires interpretation.

Internal monitoring does not use ultrasound, is more accurate than electronic monitoring and provides continuous monitoring for the high risk mother. This requires the mother's water to be broken and that she be two to three centimeters dilated. It is used in high-risk situations only.

Telemetry monitoring is the newest type of monitoring. It uses radio waves transmitted from an instrument on the mother's thigh. The mother is able to remain mobile. It provides continuous monitoring and is used in high-risk situations.

Treatment

Most women choose some type of pain relief during childbirth, ranging from relaxation and imagery to drugs. The specific choice may depend on what's available, the woman's preferences, her doctor's recommendations, and how the labor is proceeding. All drugs have some risks and some advantages.

Regional anesthetics

Regional anesthetics include epidurals and spinals. In this technique, medication is injected into the space around the spinal nerves. Depending on the type of medications used, this type of anesthesia can block nerve signals, causing temporary pain relief, or a loss of sensation from the waist down. An epidural or spinal block can provide complete pain relief during cesarean birth.

An epidural is placed with the woman lying on her side or sitting up in bed with the back rounded to allow more space between the vertebrae. Her back is scrubbed with antiseptic, and a local anesthetic is injected in the skin to numb the site. The needle is inserted between two vertebrae and through the tough tissue in front of the spinal column. A catheter is put in place that allows continuous doses of anesthetic to be given.

This type of anesthesia provides complete pain relief, and can help conserve a woman's energy, since she can relax or even sleep during labor. This type of anesthesia does require an IV and fetal monitor. It may be harder for a woman to bear down when it comes time to push, although the amount of anesthesia can be adjusted as this stage nears.

Spinal anesthesia operates on the same principle as epidural anesthesia, and is used primarily in cases of c-section delivery. It is administered in the same way as an epidural, but the catheter is not left in place. The amount of anesthetic injected is large, since it must be injected at one time. Because of the anesthetic's effect on motor nerves, most women using it cannot push during delivery.

This is a disadvantage in labor, but not an issue during a c-section. Spinals provide quick and strong anesthesia and allow for major abdominal surgery with almost no pain.

Narcotics

Short-acting narcotics can ease pain and don't interfere with a woman's ability to push. However, they can cause **sedation**, **dizziness**, nausea, and vomiting. Narcotics cross the placenta and may slow down a baby's breathing; they can't be given too close to the time of delivery.

Natural childbirth and preparation for childbirth

There are several methods to prepare for childbirth. The one selected often depends on what is available through the healthcare provider. Overall, family involvement is receiving increased attention by the healthcare systems, and many hospitals now offer birthing rooms and maternity centers to help the entire family. There are several choices available for childbirth preparation.

Lamaze, or Lamaze-Pavlov, is the most common in the United States today. It was the first popular natural childbirth method, becoming popular in the 1960s. Breathing exercises and concentration on a focal point are practiced to allow mothers to control pain while maintaining consciousness. This allows the flow of oxygen to the baby and to the muscles in the uterus to be maintained. A partner coaches the mother throughout the birthing process.

The Read method, named for Dick Read, is a technique of breathing that was originated in the 1930s to help mothers deal with apprehension and tension associated with childbirth. This natural childbirth method uses different breathing for the different stages of childbirth.

The LeBoyer method stresses a relaxed delivery in a quiet, dim room. It attempts to avoid overstimulation of the baby and to foster mother-child bonding by placing the baby on the mother's abdomen and having the mother massage him or her immediately after the birth. Then the father washes the baby in a warm bath.

The Bradley method is called father-coached childbirth, because it focuses on the father serving as coach throughout the process. It encourages normal activities during the first stages of labor.

Resources

BOOKS

- Carlson, Karen J., Stephanie A. Eisenstat, and Terra Ziporyn. *The Harvard Guide to Women's Health*. Cambridge, MA: Harvard University Press, 1996.
- Cunningham, F. Gary, et.al. *Williams Obstetrics*. 20th ed. Stamford: Appleton & Lange, 1997.

Johnson, Robert V. *Mayo Clinic Complete Book of Pregnancy & Baby's First Year*. New York: William Morrow and Co., Inc., 1994.

Ryan, Kenneth J., Ross S. Berkowitz, and Robert L. Barbieri. *Kistner's Gynecology*. 6th ed. St. Louis: Mosby, 1995.

Tuteur, Amy B. *How Your Baby is Born*. Emeryville, CA: Ziff-Davis Press, 1994.

ORGANIZATIONS

American Academy of Husband-Coached Childbirth. P.O. Box 5224, Sherman Oaks, CA 91413. (800) 423-2397; in California (800) 422-4784.

American Society for Prophylaxis in Obstetrics/LAMAZE (ASP.O. /LAMAZE). 1840 Wilson Blvd., Ste. 204, Arlington, VA 22201. (800) 368-4404.

Childbirth Education Foundation. P.O. Box 5, Richboro, PA 18954. (215) 357-2792.

International Association of Parents and Professionals for Safe Alternatives in Childbirth. Rte. 1, Box 646, Marble Hill, MO 63764. (314) 238-2010.

International Childbirth Education Association. P.O. Box 20048, Minneapolis, MN 55420. (612) 854-8660.

Postpartum Support International. 927 North Kellogg Ave., Santa Barbara, CA 93111. (805) 967-7636.

Carol A. Turkington

Childhood disintegrative disorder see
Pervasive developmental disorders

Children's health

Definition

Children's health encompasses the physical, mental, emotional, and social well-being of children from infancy through adolescence.

Description

All children should have regular well-child check ups according to the schedule recommended by their physician or pediatrician. The American Academy of Pediatrics (AAP) advises that children be seen for well-baby check ups at two weeks, two months, four months, six months, nine months, twelve months, fifteen months, and eighteen months. Well-child visits are recommended at ages two, three, four, five, six, eight, ten, and annually thereafter through age 21.

In addition, an immunization schedule should be followed to protect against disease and infection. As of 2001, the AAP and the U.S. Centers for Disease Control (CDC) recommended that the following childhood immunizations be administered by age two:

KEY TERMS

Bipolar disorder—Manic depressive disorder. A mood disorder characterized by manic highs and depressive lows.

Child development—The process of physical, intellectual, emotional, and social growth that occurs from infancy through adolescence. Erik Erikson, Margaret Mahler, Sigmund Freud, and Jean Piaget are among the most well-known child development theorists.

CPR—Cardiopulmonary resuscitation. A first aid technique designed to stimulate breathing and blood flow through a combination of chest compressions and rescue breathing.

Immunization—Creating immunity to a disease through a vaccine injection that stimulates the production of antibodies.

Learning disabilities—An impairment of the cognitive processes of understanding and using spoken and written language that results in difficulties with

one or more academic skill sets (e.g., reading, writing, mathematics).

Motor skills—Controlled movement of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

Obsessive-compulsive disorder—Also known as OCD; a disorder characterized by obsessive thoughts (e.g., fear of contamination) and compulsive behaviors (e.g., repetitive hand washing) that cause distress and/or functional impairment.

Psychological tests—Written, verbal, or visual tasks that assess psychological functioning, intelligence, and/or personality traits.

Type 1 diabetes—A chronic immune system disorder in which the pancreas does not produce sufficient amounts of insulin, a hormone that enables cells to use glucose for energy. Also called juvenile diabetes, it must be treated with insulin injections.

- Hepatitis B. Three doses.
- Diphtheria, **Tetanus**, and Pertussis (DTaP). Four doses.
- H. influenzae type b (Hib). Four doses.
- Inactivated **Polio**. Three doses.
- Pneumococcal Conjugate. Three doses.
- Measles, **Mumps**, **Rubella** (MMR). One dose.
- Varicella (chickenpox). One dose.
- Hepatitis A. (In certain geographical areas and with certain high risk groups.)

Some immunizations may cause mild side effects, or more rarely, serious adverse reactions. However, the benefits of immunization greatly outweigh the incidence of health problems arising from them.

There are serious chronic diseases and health problems that are frequently diagnosed in childhood and cannot be vaccinated against. These include, but are not limited to, **asthma**, type I diabetes (juvenile diabetes), leukemia, **hemophilia**, and **cystic fibrosis**.

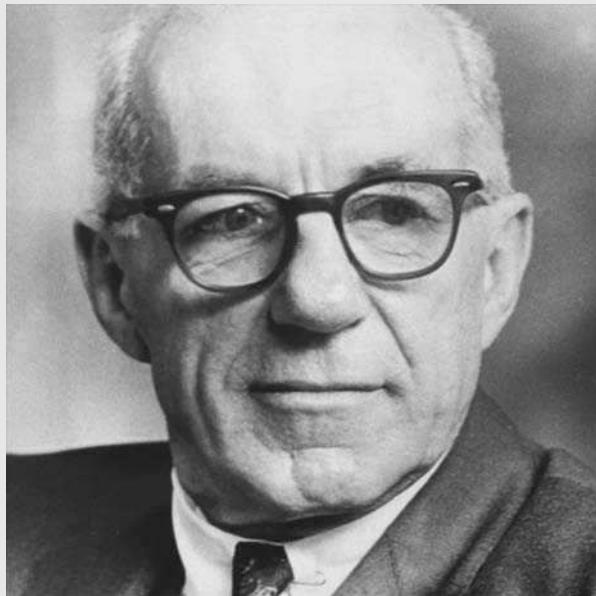
Mental health

Children who have difficulty in areas of language acquisition, cognitive development, and behavior control

may be suffering from mental illness. Mental health problems that may afflict children include:

- Attention Deficit Hyperactivity Disorder (**ADHD**). According to the AAP, 4–12% of school-aged children have ADHD, a condition characterized by poor impulse control and excessive motor activity.
- **Learning disorders**. Learning disabilities affect one in 10 school children.
- Depression, **anxiety**, and **bipolar disorder**. Affective, or mood, disorders can affect kids as well as adults.
- Eating disorders. **Anorexia nervosa**, **bulimia nervosa**, and binge eating disorder (BED) frequently occur in adolescent girls.
- **Schizophrenia**. A disorder characterized by bizarre thoughts and behaviors, **paranoia**, impaired sense of reality, and **psychosis** may be diagnosed in childhood.
- Obsessive-compulsive disorder. Also called OCD, this anxiety disorder afflicts one in 200 children.
- **Autism** and pervasive developmental disorder. Severe developmental disabilities that cause a child to become withdrawn and unresponsive.
- **Mental retardation**. Children under age 18 with an IQ of 70 or below and impairments in adaptive functioning are considered mentally retarded.

DR. BENJAMIN SPOCK (1903–1998)



(Library of Congress.)

Benjamin Spock, pediatrician and political activist, was most noted for his authorship of *Baby and Child Care*, which significantly changed predominant attitudes

toward the raising of infants and children. He began medical school at Yale University in 1925, and transferred to Columbia University's College of Physicians and Surgeons in 1927. Spock had decided well before starting his medical studies that he would "work with children, who have their whole lives ahead of them" and so, upon taking his M.D. degree in 1929 and serving his general internship at the prestigious Presbyterian Hospital, he specialized in pediatrics at a small hospital crowded with children in New York's Hell's Kitchen area.

On a summer vacation in 1943 he began to write his most famous book and he continued to work on it from 1944 to 1946 while serving as a medical officer in the Navy. The book sharply broke with the authoritarian tone and rigorous instructions found in earlier generations of baby-care books, most of which said to feed infants on a strict schedule and not to pick them up when they cried. Spock, who spent ten years trying to reconcile his psychoanalytic training with what mothers were telling him about their children, told his readers "You know more than you think you do. Don't be afraid to trust your own common sense. Take it easy, trust your own instincts, and follow the directions that your doctor gives you." The response was overwhelming. *Baby and Child Care* rapidly became America's all-time best-seller except for Shakespeare and the Bible; by 1976 it had also eclipsed Shakespeare.

Emotional and social health

Children take their first significant steps toward socialization and peer interaction when they begin to engage in cooperative play at around age four. Their social development will progress throughout childhood and adolescence as they develop friendships, start to be influenced by their peers, and begin to show interest in the opposite sex.

Factors which can have a negative impact on the emotional and social well-being of children include:

- **Violence.** Bullying can cause serious damage to a child's sense of self-esteem and personal safety, as can experiences with school violence.
- **Family turmoil.** Divorce, **death**, and other life-changing events that alter the family dynamic can have a serious impact on a child. Even a positive event such as the birth of a sibling or a move to a new city and school can put emotional strain on a child.
- **Stress.** The pressure to perform well academically and in extracurricular activities such as sports can be overwhelming to some children.

- **Peer pressure.** Although it can have a positive impact, peer pressure is often a source of significant stress for children. This is particularly true in adolescence when "fitting in" seems all-important.
- **Drugs and alcohol.** Curiosity is intrinsic to childhood, and over 30% of children have experimented with alcohol by age 13. Open communication with children that sets forth parental expectations about drug and alcohol use is essential.
- **Negative sexual experiences.** Sexual **abuse** and assault can emotionally scar a child and instill negative feelings about sexuality and relationships.

Causes and symptoms

Childhood health problems may be congenital (i.e., present at birth) or acquired through infection, immune system deficiency, or another disease process. They may also be caused by physical trauma (e.g., a car accident or a playground fall) or a toxic substance (e.g., an allergen, drug, or poisonous chemical), or triggered by genetic or environmental factors.

Physical and mental health problems in childhood can cause a wide spectrum of symptoms. However, the following behaviors frequently signify a larger emotional, social, or mental disturbance:

- signs of alcohol and drug use
- falling grades
- lack of interest in activities that were previously enjoyable to the child
- excessive anxiety
- persistent, prolonged depression
- withdrawal from friends and family
- violence
- temper tantrums or inappropriate displays of anger
- self-inflicted injury
- bizarre behavior and/or speech
- trouble with the police
- sexual promiscuity
- suicide attempts

The causes of developmental disorders and delays and learning disabilities are not always fully understood. Pervasive developmental disorder (PDD) and autistic spectrum disorder (more commonly known as autism) are characterized by unresponsiveness and severe impairments in one or more of the following areas:

- Social interaction. Autistic children are often unaware of acceptable social behavior and are withdrawn and socially isolated. They frequently do not like physical contact.
- Communication and language. A child with autism or PDD may not speak or may display limited or immature language skills.
- Behavior. Autistic or PDD children may have difficulty dealing with anger, can be self-injurious, and may display obsessive behavior.

Autism is associated with brain abnormalities, but the exact mechanisms that trigger the disorder are yet to be determined. It has been linked to certain congenital conditions such as **neurofibromatosis**, **fragile X syndrome**, and **phenylketonuria** (PKU).

Diagnosis

Physical, intellectual, emotional, and social maturation are all important markers of a child's overall health and well-being. When evaluating children, pediatricians and child-care specialists assess related skill sets, such as a child's acquisition and use of language, fine and gross motor skills, cognitive growth, and socialization, and

achievement of certain milestones in these areas. A developmental milestone is a task or skill set that a child is expected to reach at a certain age or stage of life. For example, by age one, most children have achieved the physical milestone of walking with the assistance of an adult. Developmental disorders may be identified and/or diagnosed by physicians, teachers, child psychologists, therapists, counselors, and other professionals who interact with children on a regular basis.

It is important to remember that all children are unique, and develop at different paces within this broad framework. Reaching a milestone early or late does not necessarily indicate a developmental problem. However, if a child is consistently lagging on achieving milestones, or has a significant deficit in one developmental area, he or she may be experiencing developmental delays.

Pediatricians and other medical professionals typically diagnose physical illness and disease in children. In cases of illness and injury, children will undergo a thorough **physical examination** and patient history. Diagnostic tests may be performed as appropriate. In cases of mental or emotional disorders, a psychologist or other mental healthcare professional will meet with the patient to conduct an interview and take a detailed social and medical history. 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

Medications may be prescribed to treat certain childhood illnesses. Proper dosage is particularly important with infants and children, as medications such as **acetaminophen** can be toxic in excessive amounts. Parents and caregivers should always follow the instructions for use that accompany medications, and inform the child's pediatrician if the child is taking any other drugs or **vitamins** to prevent potentially negative drug interactions. Any side effects or adverse reactions to medication should be reported to the child's physician. If **antibiotics** are prescribed, the full course should always be taken.

Other treatments for childhood illness and/or injuries include, but are not limited to, nutritional therapy, physical therapy, respiratory therapy, medical devices (e.g., **hearing aids**, glasses, braces), and in some cases, surgery.

Counseling is typically a front-line treatment for psychological disorders. Therapy approaches include psychotherapy, cognitive therapy, behavioral therapy, family counseling, and **group therapy**. Therapy or counseling may be administered by social workers, nurses, licensed counselors and therapists, psychologists, or psy-

Leading Causes Of Illness/Injury In Adolescents

Trauma (this could be anything from sports-related injuries to gunshot wounds; alcohol or other drug abuse is frequently a factor)
 Mental health issues (substance abuse, depression, etc.)
 Sexually transmitted infections
 Acquired immunodeficiency syndrome (AIDS)
 Eating disorders

chiatrists. Psychoactive medication may also be prescribed for symptom relief in children and adolescents with mental disorders.

Support groups may also provide emotional support for children with chronic illnesses or mental disorders. This approach, which allows individuals to seek advice and counsel from others in similar circumstances, can be extremely effective, especially in older children who look towards their peers for guidance and support.

Speech therapy may be helpful to children with developmental delays in language acquisition. Children with learning disorders can benefit from special education therapy.

Alternative treatment

Therapeutic approaches that encourage self-discovery and empowerment may be useful in treating some childhood emotional traumas and mental disorders. **Art therapy**, the use of the creative process to express and understand emotion, encompasses a broad range of humanistic disciplines, including visual arts, dance, drama, music, film, writing, literature, and other artistic genres. It can be particularly effective in children who may have difficulty gaining insight to emotions and thoughts they are otherwise incapable of expressing.

Certain mild herbal remedies may also be safely used with children, such as ginger (*Zingiber officinale*) tea for nausea and aloe vera salve for **burns**. Parents and caregivers should always consult their healthcare provider before administering herbs to children.

Prognosis

The prognosis for childhood health problems varies widely. In general, early detection and proper treatment can greatly improve the odds of recovery from many childhood ailments.

Some learning disabilities and mild developmental disorders can be overcome or greatly improved through the therapies discussed above. However, as of early 2001, there was no known medical treatment or pharmacological therapy that is capable of completely eliminating all

of the symptoms associated with pervasive developmental disorder (PDD), autism spectrum disorder, and mental retardation. Mental illnesses such as schizophrenia and bipolar disorder are also chronic, lifelong disorders, although their symptoms can often be well-controlled with medication.

Prevention

Parents can take some precautions to ensure the safety of their children. Childproofing the home, following a recommended immunization schedule, educating kids on safety, learning **CPR**, and taking kids for regular well-child check-ups can help to protect against physical harm. In addition, encouraging open communication with children can help them grow both emotionally and socially. Providing a loving and supportive home environment can help to nurture an emotionally healthy child who is independent, self-confident, socially skilled, insightful, and empathetic towards others.

Because they are still developing motor skills, kids can be particularly accident prone. Observe the following safety rules to protect children from injury:

- Helmets and padding. Children should always wear a properly fitted helmet and appropriate protective gear when riding a bike, scooter, or similar equipment or participating in sports. They should also ride on designated bike paths whenever possible, and learn bicycle safety rules (i.e., ride with traffic, use hand signals).
- Playground safety. Swing sets and other outdoor play equipment should be well-maintained have at least 12 in (30 cm) of loose fill materials (e.g., sand, wood chips) underneath to cushion falls, and children should always be properly supervised at play.
- Stay apprised of recalls. Children's toys, play equipment, and care products are frequently involved in product recalls. The U.S. Consumer Safety Products Commission (CSPC) is the agency responsible for tracking these recalls (see *Resources* below).
- Stay safe in the car. Up to 85% of children's car seats are improperly installed and/or used. Infants should always be in a rear-facing car seat until they are over 12 months of age and weigh more than 20 lb (9 kg). Never

Leading Causes Of Death In Adolescents

- Motor vehicle crashes
- Suicide (numbers 2 and 3 are approximately equal)
- Homicide
- Poisoning (which includes accidental poisonings due to alcohol or other drug overdose)
- Drowning

put an infant or car seat in a front passenger seat that has an air bag. Once they outgrow their forward facing car seats, children between the ages of four and eight who weigh between 40–80 lb (18–36 kg) should ride in a booster seat. Every child who rides in a car over this age and weight should buckle up with a properly fitted lap and shoulder belt.

- Teach children pedestrian safety. Younger children should never be allowed to cross the street by themselves, and older kids should know to follow traffic signs and signals, cross the street at the corner, and look both ways before stepping off the curb.
- Teach children about personal safety. Kids should know what to do in case they get lost or are approached by a stranger. It is also imperative that parents talk openly with their children about their body and sexuality, and what behavior is inappropriate, to protect them against sexual predators.

Child-proofing the household is also an important step towards keeping kids healthy. To make a house a safe home:

- Ban guns. Accidental shootings in the home injure an estimated 1,500 children under age 14 each year. If a gun must be in the home, it should be securely locked in a tamper proof box or safe.
- Keep all matches, lighters, and flammable materials properly stored and out of the reach of children.
- Make sure hot water heaters are set to 120 degrees or below to prevent scalding injuries.
- Equip the home with working fire extinguishers and smoke alarms, and teach children what to do in case of fire.
- Secure all medications (including vitamins, herbs, and supplements), hazardous chemicals, and poisonous substances (including alcohol and tobacco).
- Don't smoke. Aside from causing **cancer** and other health problems in smokers, second-hand smoke is hazardous to a child's health.
- Keep small children away from poisonous plants outdoors, and remove any indoor plants that are toxic.

- Post the phone numbers of poison control and the pediatrician near the phone, and teach children about dialing 9-1-1 for emergencies.
- Children under age five should never be left alone in the bathtub, wading pool, or near any standing water source (including an open toilet). Drowning is the leading cause of death by injury for children between the ages of one and four.
- Remove lead paint. Lead is a serious health hazard for children, and houses built before 1978 should be tested for lead paint. If lead is found, the paint should be removed using the appropriate safety precautions.

These safety guidelines are not all-inclusive, and there are many age-specific safety precautions that parents and guardians of children should observe. For example, infants should never be left with a propped-up bottle in their mouths or given small play items because of the **choking** hazards involved.

Resources

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ORGANIZATIONS

- National Institute of Mental Health. 6001 Executive Boulevard, Rm. 8184, MSC 9663, Bethesda, MD 20892-9663. (301) 443-4513.
 National SAFE KIDS Campaign. Children's National Medical Center. (202) 662-0600. <<http://www.safekids.org>>.
 U.S. Consumer Products Safety Commission (CPSC). 4330 East-West Highway, Bethesda, MD 20814-4408. (800) 638-2772. <<http://www.cpsc.gov>>.

Paula Ford-Martin

Chinese traditional herbal medicine see
Traditional Chinese herbalism

Chinese traditional medicine see **Traditional Chinese medicine**

Chiropractic

Definition

Chiropractic is from Greek words meaning done by hand. It is grounded in the principle that the body can heal itself when the skeletal system is correctly aligned and the nervous system is functioning properly. To achieve this, the practitioner uses his or her hands or an adjusting tool to perform specific manipulations of the vertebrae. When these bones of the spine are not correctly articulated, resulting in a condition known as subluxation, the theory is that nerve transmission is disrupted and causes **pain** and illness manifested in the back as well as other areas of the body.

Chiropractic is one of the most popular alternative therapies currently available. Some would say it now qualifies as mainstream treatment as opposed to complementary medicine. Chiropractic treatment is covered by many insurance plans. It has become well-accepted treatment for acute pain and problems of the spine, including lower back pain and **whiplash**. Applications beyond that scope are not supported by current evidence, although there are ongoing studies into the usefulness of chiropractic for such problems as ear infections, **dysmenorrhea**, infant **colic**, migraine headaches, and other conditions.

Purpose

Most people will experience back pain at some time in their lives. Injuries due to overexertion and poor posture are among the most common. Depending on the cause and severity of the condition, options for treatment may include physical therapy, rest, medications, surgery, or chiropractic care. Chiropractic treatment carries none of the risks of surgical or pharmacologic treatment. Practitioners use a holistic approach to health, which is appreciated by most patients. The goal is not merely to relieve the present ailment, but to analyze the cause and recommend appropriate changes of lifestyle to prevent the problem from recurring again. They believe in a risk/benefit analysis before use of any intervention. The odds of an adverse outcome are extremely low. Chiropractic has proven in several studies to be less expensive than many more traditional routes such as outpatient physical therapy. Relief from some neuromuscular problems is immediate, although a series of treatments is likely to be required to maintain the improvement. Spinal

manipulation is an excellent option for acute lower back pain, and may also relieve neck pain as well as other musculoskeletal pain. Although most back pain will subside eventually with no treatment at all, chiropractic treatment can significantly shorten the time it takes to get relief. Some types of **headache** can also be successfully treated by chiropractic.

Description

Origins

Spinal manipulation has a long history in many cultures but Daniel D. Palmer is the founder of modern chiropractic theory, dating back to the 1890s. A grocer and magnetic healer, he applied his knowledge of the nervous system and manual therapies in an unusual situation. One renowned story concerns Harvey Lillard, a janitor in the office where Palmer worked. The man had been deaf for 17 years, ever since he had sustained an injury to his upper spine. Palmer performed an adjustment on a painful vertebra in the region of the injury and Lillard's hearing was reputedly restored. Palmer theorized that all communication from the brain to the rest of the body passes through the spinal canal, and areas that are poorly aligned or under **stress** can cause physical symptoms both in the spine and in other areas of the body. Thus the body has the innate intelligence to heal itself when unencumbered by spinal irregularities causing nerve interference. After his success with Lillard, other patients began coming to him for care, and responded well to adjustments. This resulted in Palmer's further study of the relationship between an optimally functional spine and normal health.

Palmer founded the first chiropractic college in 1897. His son, B. J. Palmer, continued to develop chiropractic philosophy and practice after his father's **death**. B. J. and other faculty members were divided over the role of subluxation in disease. B. J. saw it as the cause of all disease. The others disagreed and sought a more rational way of thinking, thus broadening the base of chiropractic education. From 1910–1920, many other chiropractic colleges were established. Other innovators, including John Howard, Carl Cleveland, Earl Homewood, Joseph Janse, Herbert Lee, and Claude Watkins, also helped to advance the profession.

The theories of the Palmers receive somewhat broader interpretation today. Many chiropractors believe that back pain can be relieved and health restored through chiropractic treatment even in patients who do not have demonstrable subluxations. Scientific development and research of chiropractic is gaining momentum. The twenty-first century will likely see the metaphysical concepts such as innate intelligence give way to more scientific proofs and reform.



An example of a McTimoney chiropractic technique on patient's lumbar vertebra. The McTimoney chiropractic is a system of adjustment by hand of displacements of the spinal column and bones. It can also be applied to animals. (Photograph by Françoise Sauze, Custom Medical Stock Photo. Reproduced by permission.)

Many people besides the Palmers have contributed to the development of chiropractic theory and technique. Some have gone on to create a variety of procedures and related types of therapy that have their roots in chiropractic, including McTimoney-Corley chiropractic, craniosacral manipulation, naprapathy, and **applied kinesiology**. **Osteopathy** is another related holistic discipline that utilizes spinal and musculoskeletal manipulation as a part of treatment, but osteopathic training is more similar in scope to that of an M.D.

Initial visit

An initial chiropractic exam will most often include a history and a physical. The patient should be asked about what the current complaint is, whether there are chronic health problems, family history of disease, dietary habits,

medical care received, and any medications currently being taken. Further, the current complaint should be described in terms of how long it has been a problem, how it has progressed, and whether it is the result of an injury or occurred spontaneously. Details of how an injury occurred should be given. The physical exam should evaluate by observation and palpation whether the painful area has evidence of inflammation or poor alignment. Range of motion may also be assessed. In the spine, either hypomobility (fixation) or hypermobility may be a problem. Laboratory analysis is helpful in some cases to rule out serious infection or other health issues that may require referral for another type of treatment. Many practitioners also insist on x rays during the initial evaluation

Manipulation

When spinal manipulation is employed, it is generally done with the hands, although some practitioners may use an adjusting tool. A classic adjustment involves a high velocity, low amplitude thrust that produces a usually painless popping noise, and improves the range of motion of the joint that was treated. The patient may lie on a specially designed, padded table that helps the practitioner to achieve the proper positions for treatment. Some adjustments involve manipulating the entire spine, or large portions of it, as a unit; others are small movements designed to affect a single joint. Stretching, **traction**, and slow manipulation are other techniques that can be employed to restore structural integrity and relieve nerve interference.

Length of treatment

The number of chiropractic treatments required will vary depending on several factors. Generally longer-term treatment is needed for conditions that are chronic, severe, or occur in conjunction with another health problem. Patients who are not in overall good health may also have longer healing times. Some injuries will inherently require more treatments than others in order to get relief. Care is given in three stages. Initially appointments are more frequent with the goal of relieving immediate pain. Next, the patient moves into a rehabilitative stage to continue the healing process and help to prevent a relapse. Finally, the patient may elect periodic maintenance, or wellness treatments, along with lifestyle changes if needed in order to stay in good health.

Follow-up care

Discharge and follow-up therapy are important. If an injury occurred as a result of poor fitness or health, a program of **exercise** or **nutrition** should be prescribed. Home therapy may also be recommended, involving such things as anti-inflammatory medication and appli-

cations of heat or ice packs. Conscious attention to posture may help some patients avoid sustaining a similar injury in the future, and the chiropractor should be able to discern what poor postural habits require correction. A sedentary lifestyle, particularly with a lot of time spent sitting, is likely to contribute to poor posture and may predispose a person to back pain and injury.

Types of practitioners

Some practitioners use spinal manipulation to the exclusion of all other modalities, and are known as straight chiropractors. Others integrate various types of therapy such as massage, nutritional intervention, or treatment with **vitamins**, herbs, or homeopathic remedies. They also embrace ideas from other health care traditions. This group is known as mixers. The vast majority of chiropractors, perhaps 85%, fall in this latter category.

Preparations

Patients should enter the chiropractic clinic with an open mind. This will help to achieve maximum results.

Precautions

Chiropractic is not an appropriate therapy for diseases that are severely degenerative and may require medication or surgery. Many conditions of the spine are amenable to manipulative treatment, but that does not include **fractures**. The practitioner should be informed in advance if the patient is on anticoagulants, or has **osteoporosis** or any other condition that may weaken the bones. There are other circumstances that would contraindicate chiropractic care, and these should be detected in the history or physical exam. In addition to fractures, **Down syndrome**, some congenital defects, and some types of **cancer** are a few of the things that may preclude spinal manipulation. On rare occasions, a fracture or dislocation may occur. There is also a very slim possibility of experiencing a **stroke** as a result of spinal manipulation, but estimates are that it is no more frequent than 2.5 occurrences per one million treatments.

Be wary of chiropractors who insist on costly x rays and repeated visits with no end in sight. Extensive use is not scientifically justifiable, especially in most cases of lower back pain. There are some circumstances when x rays are indicated, including acute or possibly severe injuries such as those that might result from a car accident.

Side effects

It is not uncommon to have local discomfort in the form of aches, pains, or spasms for a few days following

DANIEL PALMER (1845–1913)

Chiropractic inventor, Daniel David Palmer, was born on March 7, 1845, in Toronto, Ontario. He was one of five siblings, the children of a shoemaker and his wife, Thomas and Katherine Palmer. Daniel Palmer and his older brother fell victim to wanderlust and left Canada with a tiny cash reserve in April 1865. They immigrated to the United States on foot, walking for 30 days before arriving in Buffalo, New York. They traveled by boat through the St. Lawrence Seaway to Detroit, Michigan. There they survived by working odd jobs and sleeping on the dock. Daniel Palmer settled in What Cheer, Iowa, where he supported himself and his first wife as a grocer and fish peddler in the early 1880s. He later moved to Davenport, Iowa, where he raised three daughters and one son.

Palmer was a man of high curiosity. He investigated a variety of disciplines of medical science during his lifetime, many of which were in their infancy. He was intrigued by phrenology and assorted spiritual cults, and for nine years he investigated the relationship between magnetism and disease. Palmer felt that there was one thing that caused disease. He was intent upon discovering this one thing, or as he called it: the great secret.

In September 1895, Palmer purported to have cured a deaf man by placing pressure on the man's displaced vertebra. Shortly afterward Palmer claimed to cure another patient of heart trouble, again by adjusting a displaced vertebra. The double coincidence led Palmer to theorize that human disease might be the result of dislocated or luxated bones, as Palmer called them. That same year he established the Palmer School of Chiropractic where he taught a three-month course in the simple fundamentals of medicine and spinal adjustment.

Palmer, who was married six times during his life, died in California in 1913; he was destitute. His son, Bartlett Joshua Palmer, successfully commercialized the practice of chiropractic.

a chiropractic treatment. Some patients may also experience mild headache or **fatigue** that resolves quickly.

Research and general acceptance

As recently as the 1970s, the American Medical Association (a national group of medical doctors) was quite hostile to chiropractic, which it deemed a cult. AMA members were advised that it was unethical to be associated with chiropractors. Fortunately that has changed, and as of 2000, many allopathic or traditionally trained physicians enjoy cordial referral relationships with chiropractors. The public is certainly strongly in

KEY TERMS

Adjustment—A very specific type of manipulation of the spine designed to return it to proper structural and functional form.

Allopathic—Conventional practice of medicine generally associated with M.D. physicians.

Dysmenorrhea—Painful menstruation.

Osteoporosis—A condition of decreased bone density, causing increased bone fragility, that is most common in elderly women.

Subluxation—Misalignment between vertebrae that structurally and functionally impairs nerve function.

favor of chiropractic treatment. An estimated 15% of people in the United States used chiropractic care in 1997. Chiropractors see the lion's share of all patients who seek medical help for back problems.

Research has also supported the use of spinal manipulation for acute low back pain. There is some anecdotal evidence recommending chiropractic treatment for ailments unrelated to musculoskeletal problems, but there is not enough research-based data to support this. On the other hand, a chiropractor may be able to treat problems and diseases unrelated to the skeletal structure by employing therapies other than spinal manipulation.

Although many chiropractors limit their practice to spine and joint problems, others claim to treat disorders that are not closely related to the back or musculoskeletal system. These include **asthma**, **bed-wetting**, **bronchitis**, coughs, **dizziness**, dysmenorrhea, earache, **fainting**, headache, hyperactivity, **indigestion**, **infertility**, migraine, **pneumonia**, and issues related to **pregnancy**. There are at least three explanations for possible efficacy for these conditions. One is that the problem could be linked to a nerve impingement, as may be possible with bed-wetting, dizziness, fainting, and headache. In a second group, chiropractic treatment may offer some relief from complicating pain and spasms caused by the disease process, as with asthma, bronchitis, coughs, and pneumonia. The discomforts of pregnancy may also be relieved with gentle chiropractic therapy. A third possibility is that manipulation or use of soft-tissue techniques may directly promote improvement of some conditions. One particular procedure, known as the endonasal technique, is thought to help the eustachian tube to open and thus improve drainage of the middle ear. The tube is sometimes blocked off due to exudates or inflammatory

processes. This can offer significant relief from earaches. Some headaches also fall in this category, as skilled use of soft tissue techniques and adjustment may relieve the muscle tension that may initiate some headaches.

Dysmenorrhea, hyperactivity, indigestion, and infertility are said to be relieved as a result of improved flow of blood and nerve energy following treatment. Evidence for this is anecdotal at best, but manipulation is unlikely to be harmful if causes treatable by other modalities have been ruled out.

For conditions such as cancer, fractures, infectious diseases, neurologic disease processes, and anything that may cause increased orthopedic fragility, chiropractic treatment alone is not an effective therapy, and may even be harmful in some cases. Those who have known circulatory problems, especially with a history of thrombosis, should not have spinal manipulation.

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ORGANIZATIONS

American Chiropractic Association. 1701 Clarendon Blvd., Arlington, VA 22209. (800)986-4636. <<http://www.amerchiro.org/>>.

Judith Turner

Chlamydial infections see **Chlamydial pneumonia; Epididymitis; Nongonococcal urethritis; Sexually transmitted diseases**

Chlamydial pneumonia

Definition

Chlamydial pneumonia refers to one of several types of pneumonia that can be caused by various types of the bacteria known as *Chlamydia*.

Description

Pneumonia is an infection of the lungs. The air sacs (alveoli) and/or the tissues of the lungs become swollen, and the alveoli may fill with pus or fluid. This prevents the lungs from taking in sufficient oxygen, which deprives the blood and the rest of the body's tissues of oxygen.

There are three major types of *Chlamydia*: *Chlamydia psittaci*, *Chlamydia pneumoniae*, and *Chlamydia trachomatis*. Each of these has the potential to cause a type of pneumonia.

Causes and symptoms

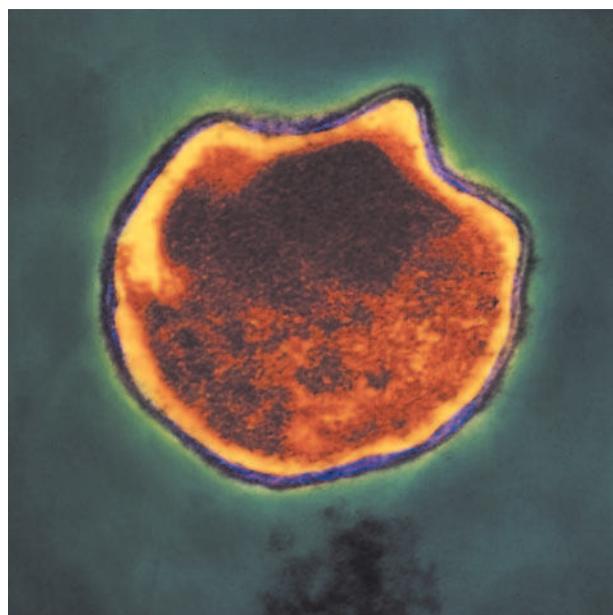
Chlamydia trachomatis is a major cause of **sexually transmitted diseases** (called **nongonococcal urethritis** and **pelvic inflammatory disease**). When a woman with an active chlamydial infection gives birth to a baby, the baby may aspirate (suck into his or her lungs) some of the mother's bacteria-laden secretions while passing through the birth canal. This can cause a form of relatively mild pneumonia in the newborn, occurring about two to six weeks after delivery.

Chlamydia psittaci is a bacteria carried by many types of birds, including pigeons, canaries, parakeets, parrots, and some gulls. Humans acquire the bacteria through contact with dust from bird feathers, bird droppings, or from the bite of a bird carrying the bacteria. People who keep birds as pets or who work where birds are kept have the highest risk for this type of pneumonia. This pneumonia, called **psittacosis**, causes **fever**, **cough**, and the production of sputum containing pus. This type of pneumonia may be quite severe, and is usually more serious in older patients. The illness can last several weeks.

Chlamydia pneumoniae usually causes a type of relatively mild "walking pneumonia." Patients experience fever and cough. This type of pneumonia is called a "community-acquired pneumonia" because it is easily passed from one member of the community to another.

Diagnosis

Laboratory tests indicating the presence of one of the strains of *Chlamydia* are sophisticated, expensive, and performed in only a few laboratories across the country. For this reason, doctors diagnose most cases of chlamydial pneumonia by performing a **physical examination** of the patient, and noting the presence of certain factors. For instance, if the mother of a baby sick with pneumonia is positive for a sexually transmitted disease caused by *Chlamydia trachomatis*, the diagnosis is obvious. History of exposure to birds in a patient sick with pneumonia suggests that *Chlamydia psittaci* may be the



A transmission electron microscopy (TEM) of a sectioned *Chlamydia pneumoniae* bacterium. (Photograph by Dr. Kari Lounatmaa, Custom Medical Stock Photo. Reproduced by permission.)

culprit. A mild pneumonia in an otherwise healthy person is likely to be a community-acquired walking pneumonia, such as that caused by *Chlamydia pneumoniae*.

Treatment

Treatment varies depending on the specific type of *Chlamydia* causing the infection. A newborn with *Chlamydia trachomatis* improves rapidly with erythromycin. *Chlamydia psittaci* infection is treated with tetracycline, bed rest, oxygen supplementation, and codeine-containing cough preparations. *Chlamydia pneumoniae* infection is treated with erythromycin.

Prognosis

The prognosis is generally excellent for the newborn with *Chlamydia trachomatis* pneumonia. *Chlamydia psittaci* may linger, and severe cases have a **death** rate of as high as 30%. The elderly are hardest hit by this type of pneumonia. A young, healthy person with *Chlamydia pneumoniae* has an excellent prognosis. In the elderly, however, there is a 5–10% death rate from this infection.

Prevention

Prevention of *Chlamydia trachomatis* pneumonia involves recognizing the symptoms of genital infection in the mother and treating her prior to delivery of her baby.

KEY TERMS

Alveoli—The small air sacs clustered at the ends of the bronchioles in the lungs, in which oxygen-carbon dioxide exchange takes place.

Aspiration—When solids or liquids that should be swallowed into the stomach are instead breathed into the respiratory system, or when substances from the outside environment are accidentally breathed into the lungs.

Sputum—Material produced within the alveoli in response to an infectious or inflammatory process.

Chlamydia psittaci can be prevented by warning people who have birds as pets, or who work around birds, to be careful to avoid contact with the dust and droppings of these birds. Sick birds can be treated with an antibiotic in their feed. Because people can contract psittacosis from each other, a person sick with this infection should be kept in **isolation**, so as not to infect other people.

Chlamydia pneumoniae is difficult to prevent because it is spread by respiratory droplets from other sick people. Because people with this type of pneumonia do not always feel very sick, they often continue to attend school, go to work, and go to other public places. They then spread the bacteria in the tiny droplets that are released into the air during coughing. Therefore, this pneumonia is very difficult to prevent and often occurs in outbreaks within communities.

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Rosalyn Carson-DeWitt, MD

Chlorhexidine see **Antibiotics, topical**

Chloroquine see **Antimalarial drugs**

Chlorzoxazone see **Muscle relaxants**

Choking

Definition

Choking is the inability to breathe because the trachea is blocked, constricted, or swollen shut.

Description

Choking is a medical emergency. When a person is choking, air cannot reach the lungs. If the airways cannot be cleared, **death** follows rapidly.

Anyone can choke, but choking is more common in children than in adults. Choking is a common cause of accidental death in young children who are apt to put toys or coins in their mouths, then unintentionally inhale them. About 3,000 adults die each year from choking on food.

People also choke because infection causes the throat tissue to swell shut. It is believed that this is what caused George Washington's death. Allergic reactions can also cause the throat to swell shut. Acute allergic reactions are called anaphylactic reactions and may be fatal. Strangulation puts external pressure on the trachea causing another form of choking.

Finally, people can choke from obstructive **sleep apnea**. This is a condition where tissues of the body obstruct the airways during sleep. Sleep apnea is most common in obese men who sleep on their backs. **Smoking**, heavy alcohol use, lung diseases such as **emphysema**, and an inherited tendency toward a narrowed airway and throat all increase the risk of choking during sleep.

Causes and symptoms

There are three reasons why people choke. These are:

- mechanical obstruction
- tissue swelling
- crushing of the trachea

Regardless of the cause, choking cuts off the air supply to the lungs. Indications that a person's airway is blocked include:

- the person cannot speak or cry out
- the person's face turns blue from lack of oxygen
- the person desperately grabs at his or her throat

- the person has a weak **cough** and labored breathing that produces a high-pitched noise
- the person has all of the above symptoms, then becomes unconscious
- during sleep, the person has episodes of gasping, pauses in breathing, and sudden awakenings.

Diagnosis

Diagnosing choking due to mechanical obstruction is straightforward, since the symptoms are obvious even to an untrained person. In choking due to infection, the person, usually a child, will have a **fever** and signs of illness before labored breathing begins. If choking is due to an allergic reaction to medication or insect bites, the person's earlobes and face will swell, giving an external sign that internal swelling is also occurring.

Choking due to sleep apnea is usually diagnosed on reports of symptoms by the person's sleep partner. There are also alarm devices to detect the occurrence of sleep apnea. Eventually sleep may be interrupted so frequently that daytime drowsiness becomes a problem.

Treatment

Choking, except during sleep apnea, is a medical emergency. If choking is due to allergic reaction or infection, people should summon emergency help or go immediately to an emergency room. If choking is due to obstructed airways, the **Heimlich maneuver** (an emergency procedure in which a person is grasped from behind in order to forcefully expel the obstruction) should be performed immediately. In severe cases a **tracheotomy** (an incision into the trachea through the neck below the larynx) must be performed.

Patients who suffer airway obstruction during sleep can be treated with a device similar to an oxygen mask that creates positive airway pressure and delivers a mixture of oxygen and air.

Prognosis

Many people are treated successfully for choking with no permanent effects. However, if treatment is unsuccessful, the person dies from lack of oxygen. In cases where the airway is restored after the critical period passes, there may be permanent brain damage.

Prevention

Watching children carefully to keep them from putting **foreign objects** in their mouth and avoiding giving young children food like raisins, round slices of hot

KEY TERMS

Trachea—The windpipe. A tube extending from below the voice box into the chest where it splits into two branches, the bronchi, that go to each lung.

Tracheotomy—The surgical creation of an opening in the trachea that functions as an alternative airway so that the patient may breathe.

dogs, and grapes can reduce the chance of choking in children. Adults should avoid heavy alcohol consumption when eating and avoid talking and laughing with food in their mouths. The risk of obstructive sleep apnea choking can be reduced by avoiding alcohol, tobacco smoking, tranquilizers, and sedatives before bed.

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Tish Davidson

Cholangitis

Definition

The term cholangitis means inflammation of the bile ducts. The term applies to inflammation of any portion of the bile ducts, which carry bile from the liver to the gallbladder and intestine. The inflammation is produced by bacterial infection or sometimes other causes.

Description

Bile, which is needed for digestion, is produced in the liver and then enters the common bile duct (CBD) through the hepatic ducts. Bile enters the gallbladder between meals, when the muscle or sphincter that controls flow of bile between the CBD and intestine is closed. During this period, bile accumulates in the CBD;

the pressure in the CBD rises, as would a pipe closed off at one end. The increase in pressure eventually causes the bile to flow into the gallbladder. During meals, the gallbladder contracts and the sphincter between the gallbladder and intestine relaxes, permitting bile to flow into the intestine and take part in digestion.

Bile that has just been produced by the liver is sterile (free of bacteria). This is partly due to its antibacterial properties; these are produced by the immunoglobulins (antibodies) secreted in bile, the bile acids which inhibit bacterial growth themselves, and mucus.

A small number of bacteria may be present in the bile ducts and gallbladder, getting there by moving backward from the intestine, which unlike the bile ducts, contains large numbers of bacteria. The normal flow of bile out of the ducts and into the intestine also helps keep too many organisms from multiplying. Bacteria also reach the bile ducts from the lymph tissue or from the blood stream.

When the passage of bile out of the ducts is blocked, the few bacteria that are there rapidly reproduce. A partial blockage to the flow of bile can occur when a stone from the gallbladder blocks the duct, and also allows bacteria to flow back into the CBD, and creates ideal conditions for their growth. Tumors, on the other hand, cause a more complete blockage of bile flow, both in and out, so fewer infections occur. The reproducing organisms are often able to enter the bloodstream and infect multiple organs such as the liver and heart valves.

Another source of inflammation of the bile ducts occurs in diseases of altered immunity, known as "autoimmune diseases." In these diseases, the body fails to recognize certain cells as part of its normal composition. The body thinks these cells are foreign and produces antibodies to fight them off, just as it fights against bacteria and viruses. Primary sclerosing cholangitis is a typical example of an autoimmune disease involving the bile ducts.

Causes and symptoms

As noted above, the two things that are needed for cholangitis to occur are: 1) obstruction to bile flow, and 2) presence of bacteria within the bile ducts. The most common cause of cholangitis is infection of the bile ducts due to blockage by a gallstone. Strictures (portions of ducts that have become narrow) also function in the same way. Strictures may be due to congenital (birth) abnormalities of the bile ducts, form as a result of injury to the bile duct (such as surgery, trauma), or result from inflammation that leads to scar tissue and narrowing.

The bacterium most commonly associated with infection of the bile ducts is *Escherichia coli* (*E. coli*) which is a normal inhabitant of the intestine. In some cases, more

than one type of bacteria is involved. Patients with **AIDS** can develop infection of narrowed bile ducts with unusual organisms such as *Cryptosporidium* and others.

The three symptoms present in about 70% of patients with cholangitis are abdominal **pain**, **fever**, and **jaundice**. Some patients only have chills and fever with minimal abdominal symptoms. Jaundice or yellow discoloration of the skin and eyes occurs in about 80% of patients. The color change is due to bile pigments that accumulate in the blood and eventually in the skin and eyes.

Inflammation due to the autoimmune disease primary sclerosing cholangitis leads to multiple areas of narrowing and eventual infection. Tumors can block the bile duct and also cause cholangitis, but as noted, infection is relatively infrequent; in fact cholangitis occurs in only about one in six patients with tumors.

Another type of bile duct infection occurs mainly in Southeast Asia and is known as recurrent pyogenic cholangitis or Oriental cholangitis. It has also been identified in Asians immigrating to North America. Most patients have stones in the bile ducts and/or gallbladder, and many cases are associated with the presence of parasites within the ducts. The role of parasites in causing infection is not clear. Many researchers believe that they are just coincidental, and have nothing to do with the stones or infection.

Diagnosis

The above symptoms alone are very suggestive of cholangitis; however, it is important to determine the exact cause and site of possible obstruction. This is because attacks are likely to recur, and different causes require different treatments. For example, the treatment of cholangitis due to a stone in the CBD is different from that due to bile duct strictures. An elevated white **blood count** suggests infection, but may be normal in 20% of patients. Abnormal or elevated tests of liver function, such as bilirubin and others are also frequently present. The specific bacteria is sometimes identified from blood cultures.

X-ray techniques

A number of x-ray techniques can make the diagnosis of bile duct obstruction; these include ultrasound and **computed tomography scans** (CT scans). However, ultrasound often cannot tell if an obstruction is due to a stricture or stone, missing a stone in about half the cases. CT scans have an even poorer record of stone detection.

Another method of diagnosing and sometimes treating the cause of bile duct obstruction or narrowing is called **percutaneous transhepatic cholangiography**. In this procedure, dye is injected into the ducts by means of

KEY TERMS

Antibiotic—A medication that is designed to kill or weaken bacteria.

Bilirubin—A pigment produced by the liver that is excreted in bile which causes a yellow discoloration of the skin and eyes when it accumulates in those organs. Bilirubin levels can be measured by blood tests, and are most often elevated in patients with liver disease or a blockage to bile flow.

Computed tomography scan (CT scan)—A specialized x-ray procedure in which cross-sections of the area in question can be examined in detail. In evaluating the bile ducts, iodine-based dye is often injected intravenously. The procedure is of greatest value in diagnosing the complications of gallstones (such as abscesses, pancreatitis) rather than documenting the presence of a stone.

Endoscope—An endoscope as used in the field of gastroenterology is a thin flexible tube which uses a lens or miniature camera to view various areas of the gastrointestinal tract. When the procedure is performed to examine certain organs such as the bile ducts or pancreas, the organs are not viewed directly, but rather indirectly through the injection of x-ray dye into the bile duct.

Endoscopy—The performance of an exam using an endoscope is referred by the general term endoscopy. Diagnosis through biopsies or other means and therapeutic procedures can be done with these instruments.

a needle placed into the liver. It is also used to drain bile and relieve an obstruction.

ENDOSCOPIC TECHNIQUES. An endoscope is a thin flexible tube that uses a lens or mirror to look at various parts of the gastrointestinal tract. **Endoscopic retrograde cholangiopancreatography (ERCP)** can accurately determine the cause and site of blockage. It also has the advantage of being able to treat the cause of obstruction, by removing stones and dilating (stretching) strictures. ERCP involves the injection of x-ray dye into the bile ducts through an endoscope. Endoscopic ultrasound is another endoscopic alternative, but is not as available as ERCP and is not therapeutic.

Treatment

The first aim is to control the bacterial infection. Broad-spectrum **antibiotics** are usually used. If the

Extracorporeal shock-wave lithotripsy (ESWL)—This is a technique that uses high-pressure waves similar to sound waves that can be “focused” on a very small area, thereby fracturing small solid objects such as gallstones, kidney stones, etc. The small fragments can pass more easily and harmlessly into the intestine or can be dissolved with medications.

Primary sclerosing cholangitis—A chronic disease in which it is believed that the immune system fails to recognize the cells that compose the bile ducts as part of the same body, and attempts to destroy them. It is not clear what exactly causes the disease, but it is frequently associated with another inflammatory disease of the digestive tract, ulcerative colitis. The inflammation of the ducts eventually produces formation of scar tissue, causing multiple areas of narrowing (strictures) that block bile flow and lead to bacterial infection. Liver transplant gives the best chance for long-term survival.

Ultrasound—A non-invasive procedure based on changes in sound waves of a frequency that cannot be heard, but respond to changes in tissue composition. It requires no preparation and no radiation occurs. It has become the “gold standard” for diagnosis of stones in the gallbladder, but is less accurate in diagnosing stones in the bile ducts. Gallstones as small as 2 mm can be identified. The procedure can now also be done through an endoscope, greatly improving investigation of the bile ducts.

infection does not come under control promptly, as noted by decrease in fever and pain, then other methods to relieve the obstruction and infection will be needed. Either way, definitive treatment of the cause of bile duct infection is the next step, and this has undergone revolutionary changes in the past decade. Endoscopic, radiographic and other techniques have made it possible to successfully remove stones and dilate strictures that previously required surgical intervention, often with high morbidity and mortality.

Radiologic and endoscopic techniques

Just as with diagnosis, treatment of cholangitis involves a number of similar procedures that differ mainly in the way the bile ducts are entered. The aims of these techniques are immediate relief of obstruction and infection as well as correction of any abnormalities that have

caused them. It is important to realize that even with endoscopy, x-ray dye is injected into the ducts and therefore the radiologist plays a role in both types of procedures. When endoscopy is used, the muscle between the intestine and bile duct is widened, to allow stones to pass. This is called a sphincterotomy and is often enough to relieve any obstruction and help clear infection. The widening of the muscle is needed if other procedures involving the bile duct are going to be performed.

The above techniques can be summarized as follows:

- Insertion of a catheter or thin flexible tube to drain bile and relieve obstruction. When performed by insertion of a needle into the liver the technique is called percutaneous transhepatic biliary drainage (PTBD); when performed endoscopically the catheter exits through the nose and is called a nasobiliary drain.
- Balloons can be inserted into the ducts with either method to dilate strictures.
- Insertion of a prosthesis which is a rigid or flexible tube designed to keep a narrowed area open; it is usually placed after a stricture is dilated with a balloon.
- Removal of stones can be accomplished most often by endoscopic techniques. A number of methods have been developed to perform this including laser and contact **lithotripsy** in which stones are fragmented by high-energy waves.

Surgical treatment

Fortunately, with recent advances in the above methods, this is a last option. Nonetheless, about 5–10% of patients will need to undergo surgical exploration of the bile ducts.

In some instances, the bile duct is so narrowed due to prior inflammation or tumor, that it needs connection to a different area of the intestinal tract to drain. This is rather complicated surgery and carries a mortality rate of 2%.

Other treatment

Extracorporeal shock-wave lithotripsy (ESWL) was first used to break up **kidney stones**. The technique has been extended to the treatment of **gallstones**, in both the gallbladder and bile ducts. It is often combined with endoscopic procedures to ease the passage of fragmented stones, or oral medications that can dissolve the fragments. Rarely, stones are also dissolved by instilling various chemicals such as ether directly into the bile ducts.

Prognosis

The outlook for those with cholangitis has markedly improved in the last several years due in large part to the

development of the techniques described above. For those patients whose episode of infection is caused by something other than a simple stone, the future is not as bright, but still often responsive to treatment. Some patients with autoimmune disease will need **liver transplantation**.

Prevention

This involves eliminating those factors that increase the risk of infection of the bile ducts, mainly stones and strictures. If it is medically possible, patients who have their gallbladder and suffer a bout of cholangitis should undergo surgical removal of the gallbladder and removal of any stones.

For other patients, a variety of therapies as outlined above, including dissolving small stones with bile acids, are also available. A combination of several of these methods is needed in some patients. Patients should discuss the risks and alternatives of these treatments with their physicians.

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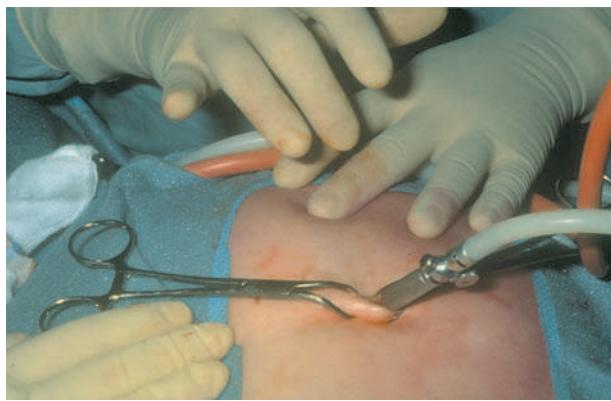
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David Kaminstein, MD



A surgeon performs a laparoscopic cholecystectomy on a patient. (Custom Medical Stock Photo. Reproduced by permission.)

Cholecystectomy

Definition

A cholecystectomy is the surgical removal of the gallbladder. The two basic types of this procedure are open cholecystectomy and the laparoscopic approach. It is estimated that the laparoscopic procedure is currently used for approximately 80% of cases.

Purpose

A cholecystectomy is performed to treat cholelithiasis and **cholecystitis**. In cholelithiasis, **gallstones** of varying shapes and sizes form from the solid components of bile. The presence of stones, often referred to as gallbladder disease, may produce symptoms of excruciating right upper abdominal **pain** radiating to the right shoulder. The gallbladder may become the site of acute infection and inflammation, resulting in symptoms of upper right abdominal pain, **nausea and vomiting**. This condition is referred to as cholecystitis. The surgical removal of the gallbladder can provide relief of these symptoms.

Precautions

Although the laparoscopic procedure requires general anesthesia for about the same length of time as the open procedure, **laparoscopy** generally produces less postoperative pain, and a shorter recovery period. The laparoscopic procedure would not be preferred in cases where the gallbladder is so inflamed that it could rupture, or when adhesions (additional fibrous bands of tissue) are present.

Description

The laparoscopic cholecystectomy involves the insertion of a long narrow cylindrical tube with a camera on the end, through an approximately 1 cm incision in the

abdomen, which allows visualization of the internal organs and projection of this image onto a video monitor. Three smaller incisions allow for insertion of other instruments to perform the surgical procedure. A laser may be used for the incision and cautery (burning unwanted tissue to stop bleeding), in which case the procedure may be called laser laparoscopic cholecystectomy.

In a conventional or open cholecystectomy, the gallbladder is removed through a surgical incision high in the right abdomen, just beneath the ribs. A drain may be inserted to prevent accumulation of fluid at the surgical site.

Preparation

As with any surgical procedure, the patient will be required to sign a consent form after the procedure is explained thoroughly. Food and fluids will be prohibited after midnight before the procedure. **Enemas** may be ordered to clean out the bowel. If nausea or vomiting are present, a suction tube to empty the stomach may be used, and for laparoscopic procedures, a urinary drainage catheter will also be used to decrease the risk of accidental puncture of the stomach or bladder with insertion of the trocar (a sharp-pointed instrument).

Aftercare

Post-operative care for the patient who has had an open cholecystectomy, as with 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 due to the pain caused by the proximity of the incision to the muscles used for respiration. The patient is shown how to support the operative site when breathing deeply and coughing, and given pain medication as necessary. Fluid intake and output is measured, and the

KEY TERMS

Cholecystitis—Infection and inflammation of the gallbladder, causing severe pain and rigidity in the upper right abdomen.

Cholelithiasis—Also known as gallstones, these hard masses are formed in the gallbladder or passages, and can cause severe upper right abdominal pain radiating to the right shoulder, as a result of blocked bile flow.

Gallbladder—A hollow pear-shaped sac on the under surface of the right lobe of the liver. Bile comes to it from the liver, and passes from it to the intestine to aid in digestion.

operative site is observed for color and amount of 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 encouraged to walk 8 hours after surgery and is discharged from the hospital within three to five days, with return to work approximately four to six weeks after the procedure.

Care received immediately after laparoscopic cholecystectomy is similar to that of any patient undergoing surgery with general anesthesia. A unique post-operative pain may be experienced in the right shoulder related to pressure from carbon dioxide used through the laparoscopic tubes. This pain may be relieved by laying on the left side with right knee and thigh drawn up to the chest. Walking will also help increase the body's reabsorption of the gas. The patient is usually discharged the day after surgery, and allowed to shower on the second postoperative day. The patient is advised to gradually resume normal activities over a three day period, while avoiding heavy lifting for about 10 days.

Risks

Potential problems associated with open cholecystectomy include respiratory problems related to location of the incision, wound infection, or **abscess** formation. Possible complications of laparoscopic cholecystectomy include accidental puncture of the bowel or bladder and uncontrolled bleeding. Incomplete reabsorption of the carbon dioxide gas could irritate the muscles used in respiration and cause respiratory distress.

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Cholecystitis

Definition

Cholecystitis refers to a painful inflammation of the gallbladder's wall. The disorder can occur a single time (acute), or can recur multiple times (chronic).

Description

The gallbladder is a small, pear-shaped organ in the upper right hand corner of the abdomen. It is connected by a series of ducts (tube-like channels) to the liver, pancreas, and duodenum (first part of the small intestine). To aid in digestion, the liver produces a substance called bile, which is passed into the gallbladder. The gallbladder concentrates this bile, meaning that it reabsorbs some of the fluid from the bile to make it more potent. After a meal, bile is squeezed out of the gallbladder by strong muscular contractions, and passes through a duct into the duodenum. Due to the chemical makeup of bile, the contents of the duodenum are kept at an optimal pH level for digestion. The bile also plays an important part in allowing fats within the small intestine to be absorbed.

Causes and symptoms

In about 95% of all cases of cholecystitis, the gallbladder contains **gallstones**. Gallstones are solid accumulations of the components of bile, particularly cholesterol, bile pigments, and calcium. These solids may occur when the components of bile are not in the correct proportion to each other. If the bile becomes overly concentrated, or if too much of one component is present, stones may form. When these stones block the duct leaving the gallbladder, bile accumulates within the gallbladder. The gallbladder continues to contract, but the bile

cannot pass out of the gallbladder in the normal way. Back pressure on the gallbladder, chemical changes from the stagnating bile trapped within the gallbladder, and occasionally bacterial infection, result in damage to the gallbladder wall. As the gallbladder becomes swollen, some areas of the wall do not receive adequate blood flow, and lack of oxygen causes cells to die.

When the stone blocks the flow of bile from the liver, certain normal byproducts of the liver's processing of red blood cells (called bilirubin) build up. The bilirubin is reabsorbed into the bloodstream, and over time this bilirubin is deposited in the skin and in the whites of the eyes. Because bilirubin contains a yellowish color, it causes a yellowish cast to the skin and eyes that is called **jaundice**.

Gallstone formation is seen in twice as many women as men, particularly those between the ages of 20 and 60. Pregnant women, or those on birth control pills or estrogen replacement therapy have a greater risk of gallstones, as do Native Americans and Mexican Americans. People who are overweight, or who lose a large amount of weight quickly are also at greater risk for developing gallstones. Not all individuals with gallstones will go on to have cholecystitis, since many people never have any symptoms from their gallstones and never know they exist. However, the vast majority of people with cholecystitis will be found to have gallstones. Rare causes of cholecystitis include severe **burns** or injury, massive systemic infection, severe illness, diabetes, obstruction by a tumor of the duct leaving the gallbladder, and certain uncommon infections of the gallbladder (including bacteria and worms).

Although there are rare reports of patients with chronic cholecystitis who never experience any **pain**, nearly 100% of the time cholecystitis will be diagnosed after a patient has experienced a bout of severe pain in the region of the gallbladder and liver. The pain may be crampy and episodic, or it may be constant. The pain is often described as pushing through to the right upper back and shoulder. Because deep breathing increases the pain, breathing becomes shallow. **Fever** is often present, and **nausea and vomiting** are nearly universal. Jaundice occurs when the duct leaving the liver is also obstructed, although it may take a number of days for it to become apparent. When bacterial infection sets in, the patient may begin to experience higher fever and shaking chills.

Diagnosis

Diagnosis of cholecystitis involves a careful abdominal examination. The enlarged, tender gallbladder may be felt through the abdominal wall. Pressure in the upper right corner of the abdomen may cause the patient to stop breathing in, due to an increase in pain. This is called



A close-up view of an inflamed gallbladder. (Custom Medical Stock Photo. Reproduced by permission.)

Murphy's sign. **Physical examination** may also reveal an increased heart rate and an increased rate of breathing.

Blood tests will show an increase in the white **blood count**, as well as an increase in bilirubin. Ultrasound is used to look for gallstones and to measure the thickness of the gallbladder wall (a marker of inflammation and scarring). A scan of the liver and gallbladder, with careful attention to the system of ducts throughout (called the biliary tree) is also used to demonstrate obstruction of ducts.

Rare complications of cholecystitis include:

- massive infection of the gallbladder, in which the gallbladder becomes filled with pus (called **empyema**)
- perforation of the gallbladder, in which the build-up of material within the gallbladder becomes so great that the wall of the organ bursts, with a resulting abdominal infection called **peritonitis**
- formation of abnormal connections between the gallbladder and other organs (the duodenum, large intestine, stomach), called **fistulas**
- obstruction of the intestine by a very large gallstone (called **gallstone ileus**)
- emphysema of the gallbladder, in which certain bacteria that produce gas infect the gallbladder, resulting in stretching of the gallbladder and disruption of its wall by gas

Treatment

Initial treatment of cholecystitis usually requires hospitalization. The patient is given fluids, salts, and sugars through a needle placed in a vein (intravenous or IV). No food or drink is given by mouth, and often a tube, called a nasogastric or NG tube, will need to be passed through the nose and down into the stomach to drain out

KEY TERMS

Bile—A substance produced by the liver, and concentrated and stored in the gallbladder. Bile contains many different substances, including bile salts, cholesterol, and bilirubin. After a meal, the gallbladder pumps bile into the duodenum (the first part of the small intestine) to keep the intestine's contents at the appropriate pH for digestion, and to help break down fats.

Bilirubin—Produced when red blood cells break down. It is a yellowish color and when levels are abnormally high, it causes the yellowish tint to eyes and skin known as jaundice.

Cholecystectomy—An operation to remove the gallbladder.

Cholecystotomy—An operation during which the gallbladder is opened, gallstones are removed, and excess bile is drained. The gallbladder is not removed.

Duct—A tube through which various substances can pass. These substances can travel through ducts to another organ or into the bloodstream.

the excess fluids. If infection is suspected, **antibiotics** are given.

Ultimately, treatment almost always involves removal of the gallbladder, a surgery called **cholecystectomy**. While this is not usually recommended while the patient is acutely ill, patients with complications usually do require emergency surgery (immediately following diagnosis) because the **death** rate increases in these cases. Similarly, those patients who have cholecystitis with no gallstones have about a 50% chance of death if the gallbladder is not quickly removed. Most patients, however, do best if surgery is performed after they have been stabilized with fluids, an NG tube, and antibiotics as necessary. When this is possible, gallbladder removal is done within five to six days of diagnosis. In patients who have other serious medical problems that may increase the risks of gallbladder removal surgery, the surgeon may decide to leave the gallbladder in place. In this case, the operation may involve removing obstructing gallstones and draining infected bile (called cholecystotomy).

Both cholecystectomy and cholecystotomy may be performed via the classical open abdominal operation (laparotomy). Tiny, "keyhole" incisions, a flexible scope, and a laser device that shatters the stones (a laparoscopic

laser) can be used to destroy the gallstones. The laparoscopic procedure can also be used to remove the gallbladder through one of the small incisions. Because of the smaller incisions, laparoscopic cholecystectomy is a procedure that is less painful and promotes faster healing.

Prognosis

Hospital management of cholecystitis ends the symptoms for about 75% of all patients. Of these patients, however, 25% will go on to have another attack of cholecystitis within a year, and 60% will have another attack within six years. Each attack of cholecystitis increases a patient's risk of developing life-threatening complications, requiring risky emergency surgery. Therefore, early removal of the gallbladder, rather than a "wait-and-see" approach, is usually recommended. Cure is complete in those patients who undergo cholecystectomy.

Prevention

Prevention of cholecystitis is probably best attempted by maintaining a reasonably ideal weight. Some studies have suggested that eating a diet high in fiber, vegetables, and fruit is also protective.

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 National Digestive Diseases Information Clearinghouse. 2 Information Way, Bethesda, MD 20892-3570. (800) 891-5389. <<http://www.niddk.nih.gov/health/digest/nddic.htm>>.

Rosalyn Carson-DeWitt, MD

Cholecystography see **Gallbladder x rays**

Choledocholithiasis see **Gallstones**

Cholelithiasis see **Gallstones**

Cholelithotomy see **Gallstone removal**

Cholera

Definition

Cholera is an acute illness characterized by watery **diarrhea** that is caused by the bacterium *Vibrio cholerae*. Cholera is spread by eating food or drinking water contaminated with the bacteria. Although cholera was a public health problem in the United States and Europe a hundred years ago, modern sanitation and the treatment of drinking water have virtually eliminated the disease in developed countries. In third world countries, however, cholera is still common.

Description

Cholera is spread by eating food or drinking water that has been contaminated with cholera bacteria. Contamination usually occurs when human feces from a person who has the disease seeps into a community water supply. Fruits and vegetables can also be contaminated in areas where crops are fertilized with human feces. Cholera bacteria also live in warm, brackish water and can infect persons who eat raw or undercooked seafood obtained from such waters. Cholera is rarely transmitted directly from one person to another.

Cholera often occurs in outbreaks or epidemics. The World Health Organization (WHO) estimates that during any cholera epidemic, approximately 0.2–1% of the local population will contract the disease. Anyone can get cholera, but infants, children, and the elderly are more likely to die from the disease because they become dehydrated faster than adults. There is no particular season in which cholera is more likely to occur.

Because of an extensive system of sewage and water treatment in the United States, Canada, Europe, Japan, and Australia, cholera is generally not a concern for visitors and residents of these countries. People visiting or living in other parts of the world, particularly on the Indian subcontinent and in parts of Africa and South America, should be aware of the potential for contracting cholera and practice prevention. Fortunately, the disease is both preventable and treatable.

Causes and symptoms

Because *V. cholerae* bacteria are sensitive to acid, most cholera-causing bacteria die in the acidic environment of the stomach. However, when a person has ingested food or water containing large amounts of cholera bacteria, some will survive to infect the intestines. As would be expected, antacid usage or the use of any medication that blocks acid production in the stomach would allow more bacteria to survive and cause infection.



A false color transmission electron micrograph (TEM) of *Vibrio cholerae* bacterium magnified 6,000 times its original size. (Photography by T. McCarthy, Custom Medical Stock Photo. Reproduced by permission.)

In the small intestine, the rapidly multiplying bacteria produce a toxin that causes a large volume of water and electrolytes to be secreted into the bowels and then to be abruptly eliminated as watery diarrhea. Vomiting may also occur. Symptoms begin to appear between one and three days after the contaminated food or water has been ingested.

In the small intestine, the rapidly multiplying bacteria produce a toxin that causes a large volume of water and electrolytes to be secreted into the bowels and then to be abruptly eliminated as watery diarrhea. Vomiting may also occur. Symptoms begin to appear between one and three days after the contaminated food or water has been ingested.

Most cases of cholera are mild, but about one in 20 patients experience severe, potentially life-threatening symptoms. In severe cases, fluids can be lost through diarrhea and vomiting at the rate of one quart per hour. This can produce a dangerous state of **dehydration** unless the lost fluids and electrolytes are rapidly replaced.

Signs of dehydration include intense thirst, little or no urine output, dry skin and mouth, an absence of tears, glassy or sunken eyes, muscle cramps, weakness, and rapid heart rate. The soft spot on an infant's head will appear to be sunken or drawn in. Dehydration occurs most rapidly in the very young and the very old because they have fewer fluid reserves. A doctor should be consulted immediately any time signs of severe dehydration occur. Immediate replacement of the lost fluids and electrolytes is necessary to prevent kidney failure, **coma**, and **death**.

Diagnosis

Rapid diagnosis of cholera can be made by examining a fresh stool sample under the microscope for the

KEY TERMS

Antibody—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

Electrolytes—Salts and minerals that ionize in body fluids. Common human electrolytes are sodium, chloride, potassium, and calcium. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Toxin—A poison. In the case of cholera, a poison secreted as a byproduct of the growth of the cholera bacteria in the small intestine.

presence of *V. cholerae* bacteria. Cholera can also be diagnosed by culturing a stool sample in the laboratory to isolate the cholera-causing bacteria. In addition, a blood test may reveal the presence of antibodies against the cholera bacteria. In areas where cholera occurs often, however, patients are usually treated for diarrhea and vomiting symptoms as if they had cholera without laboratory confirmation.

Treatment

The key to treating cholera lies in preventing dehydration by replacing the fluids and electrolytes lost through diarrhea and vomiting. The discovery that rehydration can be accomplished orally revolutionized the treatment of cholera and other, similar diseases by making this simple, cost-effective treatment widely available throughout the world. The World Health Organization has developed an inexpensive oral replacement fluid containing appropriate amounts of water, sugar, and salts that is used worldwide. In cases of severe dehydration, replacement fluids must be given intravenously. Patients should be encouraged to drink when they can keep liquids down and eat when their appetite returns. Recovery generally takes three to six days.

Adults may be given the antibiotic tetracycline to shorten the duration of the illness and reduce fluid loss. The World Health Organization recommends this antibiotic treatment only in cases of severe dehydration. If **antibiotics** are overused, the cholera bacteria organism may become resistant to the drug, making the antibiotic ineffective in treating even severe cases of cholera. Tetracycline is not given to children whose permanent teeth

have not come in because it can cause the teeth to become permanently discolored.

Prognosis

Today, cholera is a very treatable disease. Patients with milder cases of cholera usually recover on their own in three to six days without additional complications. They may eliminate the bacteria in their feces for up to two weeks. Chronic carriers of the disease are rare. With prompt fluid and electrolyte replacement, the death rate in patients with severe cholera is less than 1%. Untreated, the death rate can be greater than 50%. The difficulty in treating severe cholera is not in knowing how to treat it, but in getting medical care to ill people in underdeveloped areas of the world where medical resources are limited.

Prevention

The best form of cholera prevention is to establish good sanitation and waste treatment systems. In the absence of adequate sewage treatment, the following guidelines should be followed to reduce the possibility of infection:

- Boil it. Drink and brush teeth only with water that has been boiled or treated with chlorine or iodine tablets. Safe drinks include coffee and tea made with boiling water or carbonated bottled water and carbonated soft drinks.
- Cook it. Eat only thoroughly cooked foods, and eat them while they are still hot. Avoid eating food from street vendors.
- Peel it. Eat only fruit or nuts with a thick, intact skin or shell that is removed immediately before eating.
- Forget it. Do not eat raw foods such as oysters or ceviche. Avoid salads and raw vegetables. Do not use untreated ice cubes in otherwise safe drinks.
- Stay out of it. Do not swim or fish in polluted water.

A cholera vaccine exists that can be given to travelers and residents of areas where cholera is known to be active, but the vaccine is not highly effective. It provides only 25–50% immunity, and then only for a period of about six months. The vaccine is never given to infants under six months of age. The United States Centers for Disease Control and Prevention do not currently recommend cholera **vaccination** for travelers. Residents of cholera-plagued areas should discuss the value of the vaccine with their doctor.

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Tish Davidson

Cholestasis

Definition

Cholestasis is a condition caused by rapidly developing (acute) or long-term (chronic) interruption in the excretion of bile (a digestive fluid that helps the body process fat). The term is taken from the Greek *chole*, bile, and *stasis*, standing still.

Description

Cholestasis is caused by obstruction within the liver (intrahepatic) or outside the liver (extrahepatic). The obstruction causes bile salts, the bile pigment bilirubin, and fats (lipids) to accumulate in the blood stream instead of being eliminated normally.

Intrahepatic cholestasis is characterized by widespread blockage of small ducts or by disorders, such as hepatitis, that impair the body's ability to eliminate bile. Extrahepatic cholestasis can occur as a side effect of many medications. It can also occur as a complication of surgery, serious injury, tissue-destroying infection, or intravenous feeding. Extrahepatic cholestasis can be caused by conditions such as tumors and **gallstones** that block the flow of bile from the gallbladder to the first part of the small intestine (duodenum).

Pregnancy increases the sensitivity of the bile ducts to estrogen, and cholestasis often develops during the second and third trimesters of pregnancy. This condition is the second most common cause of **jaundice** during pregnancy, but generalized **itching** (*pruritus gravidarum*) is the only symptom most women experience. Cholestasis of pregnancy tends to run in families. Symptoms usually disappear within two to four weeks after the baby's birth but may reappear if the woman becomes pregnant again.

A similar condition affects some women who take birth-control pills. Symptoms disappear after the woman

stops using **oral contraceptives**. This condition does not lead to chronic liver disease. A woman who develops cholestasis from either of these causes (pregnancy or birth control hormones) has an increased risk of developing cholestasis from the other.

Benign familial recurrent cholestasis is a rare condition characterized by brief, repeated episodes of itching and jaundice. Symptoms often disappear. This condition does not cause **cirrhosis**.

Drug-induced cholestasis may be a complication of **chemotherapy** or other medications. The two major types of drug-induced cholestasis are direct toxic injury and reactions unique to an individual (idiosyncratic reactions). In direct toxic injury, the severity of symptoms parallels the amount of medication involved. This condition:

- develops a short time after treatment begins
- follows a predictable pattern
- usually causes liver damage

Direct toxic reactions develop in 1% of all patients who take chlorpromazine (Thorazine), a tranquilizer and antinausea drug. Idiosyncratic reactions may occur at the onset of treatment or at a later time. Allergic responses are varied and are not related to the amount of medication being taken.

Causes and symptoms

Intrahepatic cholestasis is usually caused by hepatitis or by medications that can produce symptoms resembling hepatitis. Phenothiazine-derivative drugs, including chlorpromazine, can cause sudden **fever** and inflammation. Symptoms usually disappear after use of the drug(s) is stopped. In rare cases, a condition resembling chronic biliary cirrhosis (a progressive disease characterized by destruction of small bile ducts) persists even after the medication is stopped. Some patients experience a similar reaction in response to tricyclic antidepressants (amitriptyline, imipramine), phenylbutazone (Butazolidin), erythromycin estolate (Estomycin, Purmycin), and other drugs. Intrahepatic cholestasis may also be caused by alcoholic liver disease, **primary biliary cirrhosis**, **cancer** that has spread (metastasized) from another part of the body, and a number of rare disorders.

Extrahepatic cholestasis is most often caused by a stone obstructing the passage through which bile travels from the gallbladder to the small intestine (common bile duct) or by pancreatic cancer. Less often, the condition occurs as a result of non-cancerous narrowing of the common duct (strictures), ductal carcinoma, or disorders of the pancreas.

Cholestasis caused by the use of steroids causes little, if any, inflammation. Symptoms develop gradually

KEY TERMS

Bile—A bitter yellow-green substance produced by the liver. Bile breaks down fats in the small intestine so that they can be used by the body. It is stored in the gallbladder and passes from the gallbladder through the common bile duct to the top of the small intestine (duodenum) as needed to digest fat.

Biliary—Of bile or of the gallbladder and bile ducts that transport bile and make up the biliary system or tract.

Endoscopic retrograde cholangiopancreatography—A diagnostic procedure for mapping the pancreatic and common bile ducts. A flexible tube with a light transmitter (fiberoptics) is placed in the duct. A contrast dye is instilled directly into the duct and a series of x-ray images are taken.

Computed tomography scans (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.

Hepatic—Of the liver, from the Greek *hepar*.

Liver function tests—Tests used to evaluate liver metabolism, storage, filtration, and excretion. The tests include alkaline phosphatase and serum alanine aminotransferase and aspartate aminotransferase.

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.

Percutaneous transhepatic cholangiography—An x-ray examination of the bile ducts. A needle is passed through the skin (percutaneous) across or over the liver (transhepatic) and directly into a bile duct to inject a contrast dye. The dye enhances the x-ray image mapping the system of bile ducts (cholangiography).

Phenothiazine-derivative drugs—A large family of drugs derived from phenothiazine, a compound that in itself is too poisonous for human consumption. Phenothiazine derivatives include tranquilizers, medications that prevent vomiting, antihistamines, and drugs used to enhance the effectiveness of anesthesia.

Ultrasonography—A test using sound waves to measure blood flow. Gel is applied to a hand-held transducer that is pressed against the patient's body. Images are displayed on a monitor.

and usually disappear after the drug is discontinued. Other drugs that can cause cholestasis include:

- allopurinol (Zyloprim)
- amitriptyline (Elavil)
- azathioprine (Imuran)
- benoxaprofen (Oraflex)
- capotril (Capoten)
- carbamazepine (Tegretol)
- cimetidine (Tagamet)
- hydralazine hydrochloride (Apresoline Hydrochloride)
- imipramine (Tofranil)
- penicillin
- quinidine sulfate (Quinidex)
- ranitidine (Zantac)
- sulfonamides (Apo-Sulfatrim, sulfamethoxazole)
- sulindac (Clinoril, Saldac)

Symptoms of both intrahepatic and extrahepatic cholestasis include a yellow discoloration of the skin (jaundice), dark urine, and pale stools. Itching over the skin may be severe if the condition is advanced.

Symptoms of chronic cholestasis include:

- skin discoloration
- scars or skin injuries caused by scratching
- bone pain
- yellowish fat deposits beneath the surface of the skin (xanthoma) or around the eyes (xanthelasma)

Patients with advanced cholestasis feel ill, tire easily, and are often nauseated. Abdominal pain and such systemic symptoms as anorexia, vomiting, and fever are usually due to the underlying condition that causes cholestasis.

Diagnosis

Determining whether obstruction exists inside or outside the liver is the essential part of diagnosis. A his-

tory of hepatitis or heavy drinking, recent use of certain drugs, and symptoms like **ascites** (abnormal abdominal swelling) and splenomegaly (enlarged spleen) suggest intrahepatic cholestasis. Pain or rigidity in the gallbladder or pancreas suggest an extrahepatic form.

Blood tests and **liver function tests** can reveal the pattern and extent of liver injury, indicate functional abnormalities, and establish the cause of the condition. However, most misdiagnoses occur when physicians rely more on laboratory analysis than on detailed medical history and the results of a thorough **physical examination**. Special attention should be paid to three liver function tests. Levels of alkaline phosphatase (ALP), alanine aminotransferase (ALT), and aspartate aminotransferase (AST) can indicate whether the patient's condition is caused by an obstructive condition like cholestasis or a disease of the liver cells (hepatocellular disease) like viral hepatitis or cancer. ALP levels more than three times greater than normal indicate cholestasis. High levels of AST and particularly of ALT, which is found predominantly in liver cells, indicate hepatocellular disease.

Once the disease pattern has been established, ultrasound may be performed to determine whether obstruction of the large duct has caused widening of small ducts located close to it. **Computed tomography scans** (CT) and **magnetic resonance imaging** (MRI) can provide more detailed information about the source of the obstruction. If these procedures that do not enter the patient's body (non-invasive procedures) do not provide the information a family physician, internist, or gastroenterologist needs to make a diagnosis of cholestasis, one of these procedures may be performed:

- direct cholangiography, an x-ray map of the bile ducts, enhanced by the use of contrast dye
- percutaneous transhepatic cholangiography, used to identify obstructions that impede the flow of bile from the liver to the digestive system, takes x-ray images of the bile ducts after a contrast dye has been injected by a needle passed directly into a hepatic duct
- endoscopic retrograde cholangiopancreatography (ERCP), which uses a special dye to outline the pancreatic and common bile ducts and highlight the position of any obstruction; a special tube with a light transmitter is inserted into the duct and a series of x-ray images is taken

A doctor who thinks a physical obstruction is responsible for progressive deterioration of a patient's condition may consider an exploratory surgical procedure (diagnostic laparotomy). **Liver biopsy** is sometimes performed if imaging tests do not indicate why a duct is enlarged, but results of a single biopsy may not represent the status of the entire organ.

Treatment

The goal of treatment is to eliminate or control the patient's symptoms. Discontinuing the use of certain drugs can restore normal liver function, but surgery may be needed to drain or remove obstructions or to widen affected ducts.

Rifampin (Rifadin, Rimactane), an antibacterial drug; phenobarbital, a barbiturate anticonvulsant; and other drugs are sometimes prescribed to cleanse the system and eliminate bile salts and other toxic compounds.

Patients who have chronic cholestasis and have trouble digesting fat may have to restrict the amount of fat in their diet and take calcium and water-soluble vitamin supplements. A liver transplant may become necessary if complications occur.

Prognosis

Symptoms almost always disappear after the underlying condition is controlled.

Some patients who have cholestasis experience symptoms only after infection develops, but chronic bile-duct obstruction always leads to cirrhosis. It may also cause **osteoporosis** (fragile bones) or osteomalacia (soft bones).

Emergency care is not required unless inflammation of the bile ducts (**cholangitis**) develops. Cancer should be considered when an adult suddenly develops cholestasis after the age of 50.

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American Liver Foundation. 1425 Pompton Ave., Cedar Grove, NJ 07009. (800) 223-0179. <<http://www.liverfoundation.org>>.

National Institute of Diabetes, Digestive, and Kidney Diseases of the National Institutes of Health. 31 Center Drive, Bethesda, MD 20892-2560. (301) 496-3583. <<http://www.niddk.nih.gov>>.

National Organization for Rare Disorders. P.O. Box 8923, New Fairfield, CT 06812-8923. (800) 999-6673. <<http://www.rarediseases.org>>.

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Maureen Haggerty

Cholesterol, high

Definition

Cholesterol is a fatty substance found in animal tissue and is an important component to the human body. It is manufactured in the liver and carried throughout the body in the bloodstream. Problems can occur when too much cholesterol forms an accumulation of plaque on blood vessel walls, which impedes blood flow to the heart and other organs. The highest cholesterol content is found in meat, poultry, shellfish, and dairy products.

Description

Cholesterol is the Dr. Jekyll and Mr. Hyde of medicine, since it has both a good side and bad side. It is necessary to digest fats from food, make hormones, build cell walls, and participate in other processes for maintaining a healthy body. When people talk about cholesterol as a medical problem, they are usually referring to high cholesterol. This can be somewhat misleading, since there are four components to cholesterol. These are:

- LDL, the so-called bad cholesterol
- HDL, the so-called good cholesterol
- triglycerides, a blood fat lipid that increases the risk for heart disease
- total cholesterol

High LDL (low-density lipoprotein) is a major contributing factor of heart disease. The cholesterol forms plaque in the heart's blood vessels, which restricts or blocks the supply of blood to the heart, and causes a condition called **atherosclerosis**. This can lead to a "heart attack," resulting in damage to the heart and possibly **death**. The U.S. Food and Drug Administration (FDA) estimates that 90 million American adults, roughly half the adult population, have elevated cholesterol levels.

The population as a whole is at some risk of developing high LDL cholesterol in their lifetimes. Specific risk factors include a family history of high cholesterol, **obesity**, **heart attack or stroke**, **alcoholism**, and lack of regular **exercise**. The chances of developing high cholesterol increase after the age of 45. One of the primary causes of high LDL cholesterol is too much fat or sugar in the diet, a problem especially true in the United States. Cholesterol is also produced naturally in the liver and overproduction may occur even in people who limit their intake of high cholesterol food. Low HDL and high triglyceride levels are also risk factors for atherosclerosis.

Types Of Cholesterol

Types	Levels
Total cholesterol:	
Desirable	<200
Borderline	200 to 240
Undesirable	>240
HDL cholesterol:	
Desirable	>45
Borderline	35 to 45
Undesirable	<35
LDL cholesterol:	
Desirable	<130
Borderline	130 to 160
Undesirable	>160
Ratio of total cholesterol to HDL cholesterol:	
Desirable	<3
Borderline	3 to 4
Undesirable	>4

Causes and symptoms

There are no readily apparent symptoms that indicate high LDL or triglycerides, or low HDL. The only way to diagnose the problems is through a simple blood test. However, one general indication of high cholesterol is obesity. Another is a high-fat diet.

Diagnosis

High cholesterol is often diagnosed and treated by general practitioners or family practice physicians. In some cases, the condition is treated by an endocrinologist or cardiologist. Total cholesterol, LDL, HDL, and triglyceride levels as well as the cholesterol to HDL ratio are measured by a blood test called a lipid panel. The cost of a lipid panel is generally \$40–100 and is covered by most health insurance and HMO plans, including Medicare, providing there is an appropriate reason for the test. Home cholesterol testing kits are available over the counter but test only for total cholesterol. The results should only be used as a guide and if the total cholesterol level is high or low, a lipid panel should be performed by a physician. In most adults the recommended levels, measured by milligrams per deciliter (mg/dL) of blood, are: total cholesterol, less than 200; LDL, less than 130; HDL, more than 35; triglycerides, 30–200; and cholesterol to HDL ratio, four to one. However, the recommended cholesterol levels may vary, depending on other risk factors such as **hypertension**, a family history of heart disease, diabetes, age, alcoholism, and **smoking**.

Doctors have always been puzzled by why some people develop heart disease while others with identical

HDL and LDL levels do not. New studies indicate it may be due to the size of the cholesterol particles in the bloodstream. A test called a nuclear magnetic resonance (NMR) LipoProfile exposes a blood sample to a magnetic field to determine the size of the cholesterol particles. Particle size can also be determined by a centrifugation test, where blood samples are spun very quickly to allow particles to separate and move at different distances. The smaller the particles, the greater the chance of developing heart disease. It allows physicians to treat patients who have normal or close to normal results from a lipid panel but abnormal particle size.

Treatment

A wide variety of prescription medicines are available to treat cholesterol problems. These include statins such as Mevacor (lovastatin), Lescol (fluvastatin), Pravachol (pravastatin), Zocor (simvastatin), Baycol (cerivastatin), and Lipitor (atorvastatin) to lower LDL. A group of drugs called fibric acid derivatives are used to lower triglycerides and raise HDL. These include Lopid (gemfibrozil), Atromid-S (clofibrate), and Tricor (fenofibrate). Doctors decide which drug to use based on the severity of the cholesterol problem, side effects, and cost.

Alternative treatment

The primary goal of cholesterol treatment is to lower LDL to under 160 mg/dL in people without heart disease and who are at lower risk of developing it. The goal in people with higher risk factors for heart disease is less than 130 mg/dL. In patients who already have heart disease, the goal is under 100 mg/dL, according to FDA guidelines. Also, since low HDL levels increase the risks of heart disease, the goal of all patients is more than 35 mg/dL.

In both alternative and conventional treatment of high cholesterol, the first-line treatment options are exercise, diet, weight loss, and stopping smoking. Other alternative treatments include high doses of niacin, soy protein, garlic, algae, and the Chinese medicine supplement Cholestin (a red yeast fermented with rice).

Diet and exercise

Since a large number of people with high cholesterol are overweight, a healthy diet and regular exercise are probably the most beneficial natural ways to control cholesterol levels. In general, the goal is to substantially reduce or eliminate foods high in animal fat. These include meat, shellfish, eggs, and dairy products. Several specific diet options are beneficial. One is the vegetarian diet. Vegetarians typically get up to 100% more fiber and up to 50% less cholesterol from food than non-vegetari-

KEY TERMS

Atherosclerosis—A build-up of fatty substances in the inner layers of the arteries.

Estrogen—A hormone that stimulates development of female secondary sex characteristics.

Glycemic—The presence of glucose in the blood.

Hypertension—Abnormally high blood pressure in the arteries.

Legumes—A family of plants that bear edible seeds in pods, including beans and peas.

Lipid—Any of a variety of substances that, along with proteins and carbohydrates, make up the main structural components of living cells.

Polyunsaturated fats—A non-animal oil or fatty acid rich in unsaturated chemical bonds not associated with the formation of cholesterol in the blood.

ans. The vegetarian low-cholesterol diet consists of at least six servings of whole grain foods, three or more servings of green leafy vegetables, two to four servings of fruit, two to four servings of legumes, and one or two servings of non-fat dairy products daily.

A second diet is the Asian diet, with brown rice being the staple. Other allowable foods include fish, vegetables such as bok choy, bean sprouts, and black beans. It allows for one weekly serving of meat and very few dairy products. The food is flavored with traditional Asian spices and condiments, such as ginger, chilies, turmeric, and soy sauce.

Another regimen is the low glycemic or diabetic diet, which can raise the HDL (good cholesterol) level by as much as 20% in three weeks. Low glycemic foods promote a slow but steady rise in blood sugar levels following a meal, which increases the level of HDL. They also lower total cholesterol and triglycerides. Low glycemic foods include certain fruits, vegetables, beans, and whole grains. Processed and refined foods and sugars should be avoided.

Exercise is an extremely important part of lowering bad cholesterol and raising good cholesterol. It should consist of 20–30 minutes of vigorous aerobic exercise at least three times a week. Exercises that cause the heart to beat faster include fast walking, bicycling, jogging, roller skating, swimming, and walking up stairs. There are also a wide selection of aerobic programs available at gyms or on videotape.

Garlic

A number of clinical studies have indicated that garlic can offer modest reductions in cholesterol. A 1997 study by **nutrition** researchers at Pennsylvania State University found men who took garlic capsules for five months reduced their total cholesterol by 7% and LDL by 12%. Another study showed that seven cloves of fresh garlic a day significantly reduced LDL, as did a daily dose of four garlic extract pills. Other studies in 1997 and 1998 back up these results. However, two more recent studies have questioned the effectiveness of garlic in lowering "bad cholesterol."

Cholestin

Cholestin hit the over-the-counter market in 1997 as a cholesterol-lowering dietary supplement. It is a processed form of red yeast fermented with rice, a traditional herbal remedy used for centuries by the Chinese. Two studies released in 1998 showed Cholestin lowered LDL cholesterol by 20–30%. It also appeared to raise HDL and lower triglyceride levels. Although the supplement contains hundreds of compounds, the major active LDL-lowering ingredient is lovastatin, a chemical also found in the prescription drug Mevacor. The FDA banned Cholestin in early 1998 but a federal district court judge lifted the ban a year later, ruling the product was a dietary supplement, not a drug. It is not fully understood how the substance works and patients may want to consult with their physician before taking Cholestin. No serious side effects have been reported, but minor side effects, including bloating and **heartburn**, have been reported.

Other treatments

A study released in 1999 indicated that blue-green algae contains polyunsaturated fatty acids that lower cholesterol. The algae, known as alga *Aphanizomenon flos-aquae* (AFA) is available as an over-the-counter dietary supplement. Niacin, also known as nicotinic acid or vitamin B₃, has been shown to reduce LDL levels by 10–20%, and raise HDL levels by 15–35%. It also can reduce triglycerides. But because an extremely high dose of niacin (2–3 g) is needed to treat cholesterol problems, it should only be taken under a doctor's supervision to monitor possible toxic side effects. Niacin can also cause flushing when taken in high doses. Soy protein with high levels of isoflavones also have been shown to reduce bad cholesterol by up to 10%. A daily diet that contains 62 mg of isoflavones in soy protein is recommended, and can be incorporated into other diet regimens, including vegetarian, Asian, and low glycemic.

Prognosis

High cholesterol is one of the key risk factors for heart disease. Left untreated, too much bad cholesterol

can clog the blood vessels, leading to chest **pain (angina)**, blood clots, and heart attacks. Heart disease is the number one killer of men and women in the United States. By reducing LDL, people with heart disease may prevent further heart attacks and strokes, prolong and improve the quality of their lives, and slow or reverse cholesterol build-up in the arteries. In people without heart disease, lowering LDL can decrease the risk of a first heart attack or stroke.

Prevention

The best way to prevent cholesterol problems is through a combination of healthy lifestyle activities, a primarily low-fat and high-fiber diet, regular aerobic exercise, not smoking, and maintaining an optimal weight. But for people with high risk factors for heart disease, such as a family history of heart disease, diabetes, and being over the age of 45, these measures may not be enough to prevent the onset of high cholesterol. There are studies being done on the effectiveness of some existing anti-cholesterol drugs for controlling cholesterol levels in patients who do not meet the criteria for high cholesterol but no definitive results are available.

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<http://www.nhlbi.nih.gov>.

Ken R. Wells

Cholesterol-reducing drugs

Definition

Cholesterol-reducing drugs are medicines that lower the amount of cholesterol (a fat-like substance) in the blood.

Purpose

Cholesterol is a chemical that can both benefit and harm the body. On the good side, cholesterol plays important roles in the structure of cells and in the production of hormones. But too much cholesterol in the blood can lead to heart and blood vessel disease. To complicate matters, not all cholesterol contributes to heart and blood vessel problems. One type, called high-density lipoprotein (HDL) cholesterol, or “good cholesterol,” actually lowers the risk of these problems. The other type, low-density lipoprotein (LDL) cholesterol, or “bad cholesterol,” is the type that threatens people’s health. The names reflect the way cholesterol moves through the body. To travel through the bloodstream, cholesterol must attach itself to a protein. The combination of a protein and a fatty substance like cholesterol is called a lipoprotein.

Many factors may contribute to the fact that some people have higher cholesterol levels than others. A diet high in certain types of fats is one factor. Medical problems such as poorly controlled diabetes, an underactive thyroid gland, an overactive pituitary gland, liver disease or kidney failure also may cause **high cholesterol** levels. And some people have inherited disorders that prevent their bodies from properly using and eliminating fats. This allows cholesterol to build up in the blood.

Treatment for high cholesterol levels usually begins with changes in daily habits. By losing weight, stopping **smoking**, exercising more and reducing the amount of fat and cholesterol in the diet, many people can bring their cholesterol levels down to acceptable levels. However, some may need to use cholesterol-reducing drugs to reduce their risk of health problems.

Description

There are four different classes of cholesterol lowering drugs:

Bile acid sequestrants are drugs that act by binding with the bile produced by the liver. Bile helps the digestion and absorption of fats in the intestine. By blocking the digestion of fats, bile acid sequestrants prevent the formation of cholesterol. Drugs in this class include: cholestyramine (Questran); colestipol (Colestid); and colesevelam (Welchol).

HMG-CoA inhibitors, often called “statins,” are drugs that block an enzyme called “3-hydroxy-3-methyl-glutaryl-coenzyme A reductase.” This blocks one of the steps in converting fat to cholesterol. These are the most effective cholesterol lowering agents available. Drugs in this group include: atorvastatin (Lipitor); cerivastatin (Baycol); fluvastatin (Lescol); lovastatin (Mevacor); pravastatin (Pravachol); and simvastatin (Zocor).

Fibric acid derivatives include clofibrate (Atromid-S); gemfibrozil (Lopid); and fenofibrate (Tricor). Although these drugs are less effective than the statins at lowering total cholesterol, they may be able to lower the low-density lipoprotein (LDL) cholesterol while raising the high-density lipoprotein (HDL) cholesterol. Their exact mechanism of action is believed to be associated with inhibition of lipoprotein lipase activity.

Niacin, vitamin B-3, is also effective in lowering cholesterol levels. Although the normal vitamin dose of niacin is only 20 mg, the dose required to reduce cholesterol levels is at least 500 mg each day. The mechanism of action of niacin in cholesterol reduction is associated with the inhibition of VLDL secretion in the bloodstream.

Recommended dosage

The recommended dosage depends on the type of cholesterol-reducing drug used. The prescribing physician or the pharmacist who filled the prescription can advise about the correct dosage.

Cholesterol-reducing drugs should be taken exactly as directed and doses should not be missed. Double doses should not be taken to make up for a missed dose.

Physicians may prescribe a combination of cholesterol-reducing drugs, such as pravastatin and colestipol. Following the directions for how and when to take the drugs is very important. The medicine may not work properly if both drugs are taken at the same time of day.

Niacin should not be taken at the same time as an HMG-CoA inhibitor, as this combination may cause severe muscle problems. If niacin is taken in an over-the-counter form, both the prescribing physician and pharmacist should be informed. There are no problems when the niacin is taken in normal doses as a vitamin.

The prescription should not be stopped without first checking with the physician who prescribed it. Cholesterol levels may increase when the medicine is stopped, and the physician may prescribe a special diet to make this less likely.

Precautions

Seeing a physician regularly while taking cholesterol-reducing drugs is important. The physician will check to

make sure the medicine is working as it should and will decide whether it is still needed. Blood tests and other medical tests may be ordered to help the physician monitor the drug's effectiveness and check for side effects.

For most people, cholesterol-reducing drugs are just one part of a whole program for lowering cholesterol levels. Other important elements of the program may include weight loss, **exercise**, special **diets** and changes in other habits. The medication should never be viewed as a substitute for other measures ordered by the physician. Cholesterol-reducing drugs will not cure problems that cause high cholesterol; they will only help control cholesterol levels.

People over 60 years of age may be unusually sensitive to the effects of some cholesterol-reducing drugs. This may increase the chance of side effects.

Anyone who is taking an HMG-CoA reductase inhibitor should notify the health care professional in charge before having any surgical or dental procedures or receiving emergency treatment.

Special conditions

People who have certain medical conditions or who are taking certain other medications may have problems if they take cholesterol-reducing drugs. Before taking these drugs, the prescribing physician should be informed of any of the following conditions:

ALLERGIES. Anyone who has had unusual reactions to cholesterol-reducing drugs in the past should inform the prescribing physician before taking the drugs again. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

PREGNANCY. Studies of laboratory animals have shown that giving high doses of gemfibrozil during **pregnancy** increases the risk of **birth defects** and other problems, including **death** of the unborn baby. The effects of this drug have not been studied in pregnant women. Women who are pregnant or who may become pregnant should check with their physicians before using gemfibrozil.

Cholesterol-reducing drugs in the group known as HMG-CoA reductase inhibitors (such as lovastatin, fluvastatin, pravastatin and simvastatin) should not be taken by women who are pregnant or who plan to become pregnant soon. By blocking the production of cholesterol, these drugs prevent a fetus from developing properly. Women who are able to bear children should use an effective birth control method while taking these drugs. Any woman who becomes pregnant while taking these drugs should check with her physician immediately.

Cholestyramine and colestipol will not directly harm an unborn baby, because these drugs are not taken into the body. However, the drugs may keep the mother's body from absorbing **vitamins** that she and the baby need. Pregnant women who take these drugs should ask their physicians whether they need to take extra vitamins.

BREASTFEEDING. Because cholestyramine and colestipol interfere with the absorption of vitamins, women who use these drugs while breastfeeding should ask their physicians if they need to take extra vitamins.

Women who are breastfeeding should talk to their physicians before using gemfibrozil. Whether this drug passes into breast milk is not known. But because animal studies suggest that it may increase the risk of some types of **cancer**, women should carefully consider the safety of using it while breastfeeding.

HMG-CoA reductase inhibitors (such as lovastatin, pravastatin, fluvastatin and simvastatin) should not be used by women who are breastfeeding their babies.

OTHER MEDICAL CONDITIONS. Cholesterol-reducing drugs may make some medical problems worse. Before using these drugs, people with any of these medical conditions should make sure their physicians are aware of their conditions:

- stomach problems, including stomach ulcer
- **constipation**
- hemorrhoids
- **gallstones** or gallbladder disease
- bleeding problems
- underactive thyroid
- heart or blood vessel disease

In addition, people with kidney or liver disease may be more likely to have blood problems or other side effects when they take certain cholesterol-reducing drugs. And some drugs of this type may actually raise cholesterol levels in people with liver disease.

Patients with any of the following medical conditions may develop problems that could lead to kidney failure if they take HMG-CoA reductase inhibitors:

- treatments to prevent rejection after an organ transplant
- recent major surgery
- seizures (convulsions) that are not well controlled

People with **phenylketonuria** (PKU) should be aware that sugar-free formulations of some cholesterol-reducing drugs contain phenylalanine in aspartame. This ingredient can cause problems in people who have phenylketonuria.

USE OF CERTAIN MEDICINES. Cholesterol-reducing drugs may change the effects of other medicines. Patients should not take any other medicine that has not been prescribed or approved by a physician who knows they are taking cholesterol-reducing drugs.

Side effects

Gemfibrozil

Studies in animals and humans suggest that gemfibrozil increases the risk of some types of cancer. The drug may also cause gallstones or muscle problems. Patients who need to take this medicine should ask their physicians for the latest information on its benefits and risks.

Patients taking gemfibrozil should check with a physician immediately if any of these side effects occur:

- fever or chills
- severe stomach **pain** with nausea and vomiting
- pain in the lower back or side
- pain or difficulty when urinating
- cough or hoarseness

HMG-CoA reductase inhibitors

These drugs may damage the liver or muscles. Patients who take the drugs should have blood tests to check for liver damage as often as their physician recommends. Any unexplained pain, tenderness or weakness in the muscles should be reported to the physician at once.

All cholesterol-reducing drugs

Minor side effects such as **heartburn, indigestion**, belching, bloating, gas, nausea or vomiting, stomach pain, **dizziness** and **headache** usually go away as the body adjusts to the drug and do not require medical treatment unless they continue or they interfere with normal activities.

Patients who have constipation while taking cholesterol-reducing drugs should bring the problem to a physician's attention as soon as possible.

Additional side effects are possible. Anyone who has unusual symptoms while taking cholesterol-reducing drugs should get in touch with his or her physician.

Interactions

Cholesterol-reducing drugs 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 cholesterol-reducing drugs should let the physician know all other medicines he or

KEY TERMS

Cell—The basic unit that makes up all living tissue.

Cholesterol—Fatty substance found in tissue. Necessary to maintain a healthy body.

Enzyme—A type of protein, produced in the body, that brings about or speeds up chemical reactions.

Hormone—A substance that is produced in one part of the body, then travels through the bloodstream to another part of the body where it has its effect.

Phenylketonuria—(PKU) A genetic disorder in which the body lacks an important enzyme. If untreated, the disorder can lead to brain damage and mental retardation.

Pituitary gland—A pea-sized gland at the base of the brain that produces many hormones that affect growth and body functions.

she is taking and should ask whether the possible interactions can interfere with drug therapy. Examples of possible interactions are listed below.

Some cholesterol-reducing drugs may prevent the following medicines from working properly:

- thyroid hormones
- water pills (diuretics)
- certain **antibiotics** taken by mouth, such as **tetracyclines**, penicillin G and vancomycin
- the beta-blocker Inderal, used to treat high blood pressure
- digitalis heart medicines
- phenylbutazone, a nonsteroidal anti-inflammatory drug

Taking some cholesterol-reducing drugs with blood thinners (anticoagulants) may increase the chance of bleeding.

Combining HMG-CoA reductase inhibitors with gemfibrozil, cyclosporine (Sandimmune) or niacin may cause or worsen problems with the kidneys or muscles.

Resources

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Nancy Ross-Flanigan

Cholesterol test

Definition

The cholesterol test is a quantitative analysis of the cholesterol levels in a sample of the patient's blood. Total serum cholesterol (TC) is the measurement routinely taken. Doctors sometimes order a complete lipoprotein profile to better evaluate the risk for **atherosclerosis (coronary artery disease, or CAD)**. The full lipoprotein profile also includes measurements of triglyceride levels (a chemical compound that forms 95% of the fats and oils stored in animal or vegetable cells) and lipoproteins (high density and low density). Blood fats are also called "lipids."

The type of cholesterol in the blood is as important as the total quantity. Cholesterol is a fatty substance and cannot be dissolved in water. It must combine with a protein molecule called a lipoprotein in order to be transported in the blood. There are five major types of lipoproteins in the human body; they differ in the amount of cholesterol that they carry in comparison to other fats and fatty acids, and in their functions in the body. Lipoproteins are classified, as follows, according to their density:

- Chylomicrons. These are normally found in the blood only after a person has eaten foods containing fats. They contain about 7% cholesterol. Chylomicrons transport fats and cholesterol from the intestine into the liver and then into the bloodstream. They are metabolized in the process of carrying food energy to muscle and fat cells.
- Very low-density lipoproteins (VLDL). These lipoproteins carry mostly triglycerides, but they also contain 16–22% cholesterol. VLDLs are made in the liver and eventually become IDL particles after they have lost their triglyceride content.
- Intermediate-density lipoproteins (IDL). IDLs are short-lived lipoproteins containing about 30% cholesterol that are converted in the liver to low-density lipoproteins (LDLs).
- Low-density lipoproteins (LDL). LDL molecules carry cholesterol from the liver to other body tissues. They contain about 50% cholesterol. Extra LDLs are absorbed by the liver and their cholesterol is excreted into the bile. LDL particles are involved in the formation of plaques (abnormal deposits of cholesterol) in the walls of the coronary arteries. LDL is known as "bad cholesterol."
- High-density lipoproteins (HDL). HDL molecules are made in the intestines and the liver. HDLs are about 50% protein and 19% cholesterol. They help to remove cholesterol from artery walls. Lifestyle changes, including exercising, keeping weight within recom-

mended limits, and giving up **smoking** can increase the body's levels of HDL cholesterol. HDL is known as "good cholesterol."

Because of the difference in density and cholesterol content of lipoproteins, two patients with the same total cholesterol level can have very different lipid profiles and different risk for CAD. The critical factor is the level of HDL cholesterol in the blood serum. Some doctors use the ratio of the total cholesterol level to HDL cholesterol when assessing the patient's degree of risk. A low TC/HDL ratio is associated with a lower degree of risk.

Purpose

The purpose of the TC test is to measure the levels of cholesterol in the patient's blood. The patient's cholesterol can also be fractionated (separated into different portions) in order to determine the TC/HDL ratio. The results help the doctor to assess the patient's risk for coronary artery disease (CAD). High LDL levels are associated with increased risk of CAD whereas high HDL levels are associated with relatively lower risk.

In addition, the results of the cholesterol test can assist the doctor in evaluating the patient's metabolism of fat, or in diagnosing inflammation of the pancreas, liver disease, or disorders of the thyroid gland.

The frequency of cholesterol testing depends on the patient's degree of risk for CAD. People with low cholesterol levels may need to be tested once every five years. People with high levels of blood cholesterol should be tested more frequently, according to their doctor's advice. The doctor may recommend a detailed evaluation of the different types of lipids in the patient's blood. It is ideal to check the HDL and triglycerides as well as the cholesterol and LDL. In addition, the National Cholesterol Education Program (NCEP) suggests further evaluation if the patient has any of the symptoms of CAD or if she or he has two or more of the following risk factors for CAD:

- male sex
- high blood pressure
- smoking
- diabetes
- low HDL levels
- family history of CAD before age 55

Precautions

Patients who are seriously ill or hospitalized for surgery should not be given cholesterol tests because the results will not indicate the patient's normal cholesterol level. Acute illness, high **fever, starvation**, or recent surgery lowers blood cholesterol levels.

KEY TERMS

Atherosclerosis—A disease of the coronary arteries in which cholesterol is deposited in plaques on the arterial walls. The plaque narrows or blocks blood flow to the heart. Atherosclerosis is sometimes called coronary artery disease, or CAD.

Fractionation—A laboratory test or process in which blood or another fluid is broken down into its components. Fractionation can be used to assess the proportions of the different types of cholesterol in a blood sample.

High-density lipoprotein (HDL)—A type of lipoprotein that protects against CAD by removing cholesterol deposits from arteries or preventing their formation.

Hypercholesterolemia—The presence of excessively high levels of cholesterol in the blood.

Lipid—Any organic compound that is greasy, insol-

uble in water, but soluble in alcohol. Fats, waxes, and oils are examples of lipids.

Lipoprotein—A complex molecule that consists of a protein membrane surrounding a core of lipids. Lipoproteins carry cholesterol and other lipids from the digestive tract to the liver and other body tissues. There are five major types of lipoproteins.

Low-density lipoprotein (LDL)—A type of lipoprotein that consists of about 50% cholesterol and is associated with an increased risk of CAD.

Plaque—An abnormal deposit of hardened cholesterol on the wall of an artery.

Triglyceride—A chemical compound that forms about 95% of the fats and oils stored in animal and vegetable cells. Triglyceride levels are sometimes measured as well as cholesterol when a patient is screened for heart disease.

Description

The cholesterol test requires a sample of the patient's blood. **Fasting** before the test is required to get an accurate triglyceride and LDL level. The blood is withdrawn by the usual vacuum tube technique from one of the patient's veins. The blood test takes between three and five minutes.

Preparation

Patients who are scheduled for a lipid profile test should fast (except for water) for 12–14 hours before the blood sample is drawn. If the patient's cholesterol is to be fractionated, he or she should also avoid alcohol for 24 hours before the test.

Patients should also stop taking any medications that may affect the accuracy of the test results. These include **corticosteroids**, estrogen or androgens, **oral contraceptives**, some **diuretics**, haloperidol, some **antibiotics**, and niacin. Antilipemics are drugs that lower the concentration of fatty substances in the blood. When these are taken by the patient, blood testing may be done frequently to evaluate the liver function as well as lipids. The patient's doctor will give the patient a list of specific medications to be discontinued before the test.

Aftercare

Aftercare includes routine care of the skin around the needle puncture. Most patients have no aftereffects, but

some may have a small bruise or swelling. A washcloth soaked in warm water usually relieves any discomfort. In addition, the patient should resume taking any prescription medications that were discontinued before the test.

Risks

The primary risk to the patient is a mild stinging or burning sensation during the venipuncture, with minor swelling or bruising afterward.

Normal results

The "normal" values for serum lipids depend on the patient's age, sex, and race. Normal values for people in Western countries are usually given as 140–220 mg/dL in adults, although as many as 5% of the population has TC higher than 300 mg/dL. Among Asians, the figures are about 20% lower. As a rule, both TC and LDL levels rise as people get older.

Some doctors prefer to speak of "desired" rather than "normal" cholesterol values, on the grounds that "normal" refers to statistically average levels that may still be too high for good health. Desirable values are as follows:

- Total cholesterol (TC): less than 200 mg/dL
- HDL cholesterol: 40–70 mg/dL in males, 40–80 mg/dL in females
- LDL cholesterol: less than 130 mg/dL
- TC/HDL ratio: under 4.0 in males, 3.8 in females.

Abnormal results

It is possible for blood cholesterol levels to be too low as well as too high.

Abnormally low levels

TC levels less than 160 mg/dL are associated with higher mortality rates from **cancer**, liver disease, respiratory disorders, and injuries. The connection between unusually low cholesterol and increased mortality is not clear, although some researchers think that the low level is a secondary sign of the underlying disease and not the cause of disease or **death**.

Low levels of serum cholesterol are also associated with **malnutrition** or **hyperthyroidism**. Further diagnostic testing may be necessary in order to locate the cause.

Abnormally high levels

Prior to 1980, **hypercholesterolemia** (an abnormally high TC level) was defined as any value above the 95th percentile for the population. These figures ranged from 210 mg/dL in persons younger than 20 to more than 280 mg/dL in persons older than 60. It is now known, however, that TC levels over 200 mg/dL are associated with significantly higher risk of CAD. Levels of 280 mg/dL or more are considered elevated. Treatment with diet and medication has proven to successfully lower risk of **heart attack** and **stroke**.

Elevated cholesterol levels may also result from hepatitis, blockage of the bile ducts, disorders of lipid metabolism, **nephrotic syndrome**, inflammation of the pancreas, or **hypothyroidism**.

Resources

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Rebecca J. Frey

Cholinergic drugs

Definition

Cholinergic drugs are medications that produce the same effects as the parasympathetic nervous system.

Purpose

Cholinergic drugs produce the same effects as acetylcholine. Acetylcholine is the most common neurohormone of the parasympathetic nervous system, the part of the peripheral nervous system responsible for the everyday work of the body. While the sympathetic nervous system acts during times of excitation, the parasympathetic system deals with everyday activities such as salivation, digestion, and muscle relaxation.

The cholinergic drugs may be used in several ways. The cholinergic muscle stimulants are used to diagnose and treat myasthenia gravis, a disease that causes severe muscle weakness. This class of drugs includes ambenonium chloride (Mytelase), edrophonium chloride (Tensilon), neostigmine (Prostigmine), and pyridostigmine (Mestinón). These drugs are also widely used in surgery, both to reduce the risk of urinary retention, and to reverse the effects of the muscle relaxant drugs that are used in surgery.

Cholinergic drugs are also used in control of **glaucoma**, a disease that is caused by increased pressure inside the eye. The most common drugs used for this purpose are demecarium (Humorsol) and echthiophate (Phospholine iodide).

Description

Cholinergic drugs usually act in one of two ways. Some directly mimic the effect of acetylcholine, while others block the effects of acetylcholinesterase. Acetylcholinesterase is an enzyme that destroys naturally occurring acetylcholine. By blocking the enzyme, the naturally occurring acetylcholine has a longer action.

Recommended dosage

Cholinergic drugs are available only by prescription. They may be available as eye drops, capsules, tablets, or injections.

Precautions

Cholinergic drugs should be avoided when the patient has any sort of obstruction in the urinary or digestive tracts, such as a tumor, or severe inflammation which is causing blockage.

They should be used with caution in patients with **asthma**, epilepsy, slow heart beat, **hyperthyroidism**, or gastric ulcers.

The effects of the cholinergic drugs are to produce the same effects as stimulation of the parasympathetic nervous system. These effects include slowing of the heartbeat, increases in normal secretions including the

KEY TERMS

Cholinergic—Nerves that are stimulated by acetylcholine.

Glaucoma—a disease of the eye marked by increased pressure within the eyeball that can result in damage to the optic disk and gradual loss of vision.

Myasthenia gravis—a disease characterized by progressive weakness and exhaustibility of voluntary muscles without atrophy or sensory disturbance and caused by an autoimmune attack on acetylcholine receptors at neuromuscular junctions.

Parasympathetic nervous system—the part of the nervous system that contains chiefly cholinergic fibers, that tends to induce secretion, to increase the tone and contractility of smooth muscle, and to slow the heart rate.

digestive acids of the stomach, saliva and tears. For this reason, patients who already have a problem in one of these areas, such as a slow heartbeat or stomach ulcers should use these drugs with great caution, since the medication will make their conditions worse.

Side effects

When used properly, cholinergic drugs will increase muscle strength in patients with **myasthenia gravis**. In eye drop form, they can reduce the intraocular pressure in glaucoma.

The possible adverse effects of cholinergic drugs are:

- slow heart beat, possibly leading to cardiac arrest
- muscle weakness, muscle cramps, and muscle pain
- convulsions
- weak breathing, inability to breath
- increased stomach acid and saliva
- nausea and vomiting
- dizziness, drowsiness, and headache

Resources

BOOKS

Beyond the Limits: A Self Portrait of Myasthenia Gravis.
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Samuel Uretsky, PharmD

Chondromalacia patellae

Definition

Chondromalacia patellae refers to the progressive erosion of the articular cartilage of the knee joint, that is the cartilage underlying the kneecap (patella) that articulates with the knee joint.

Description

Chondromalacia patellae (CMP), also known as patello-femoral **pain** syndrome or patello-femoral **stress** syndrome, is a syndrome that causes pain/discomfort at the front of the knee. It is associated with irritation or wear on the underside of the kneecap, or patella. In a normal knee, the articular cartilage is smooth and elastic and glides smoothly over the surface of the thighbone, or femur when the knee is bent. Erosion of the cartilage roughens the surface and prevents this smooth action.

CMP is most common in adolescent females, although older people may also develop it. An average of two out of 10,000 people develop this condition, many of them runners or other athletes.

Causes and symptoms

CMP is the result of the normal **aging** process, overuse, injury, or uneven pressures exerted on the knee joint. In teens, CMP may be caused by uneven growth or uneven strength in the thigh muscles. Growth spurts, common in teens, may result in a mildly abnormal alignment of the patella, which increases the angle formed by the thigh and the patellar tendon (Q-angle). This condition adds to the damage. Symptoms include pain, normally around the kneecap, and a grinding sensation felt when extending the leg. The pain may radiate to the back of the knee, or it may be intermittent and brought on by squatting, kneeling, going up or down stairs, especially down, or by repeated bending of the joint.

Diagnosis

Diagnosis is established during a **physical examination** performed by a general practitioner or an orthopedist, and is based on frequency of symptoms and confirmed by x rays of the knee. The CMP erosion can also

KEY TERMS

Arthroscopic knee surgery—Surgery performed to examine or repair tissues inside the knee joint through a special scope (arthroscope).

Femur—The thigh bone.

Isometric exercises—Exercises which strengthen through muscle resistance.

Osteoarthritis—Degenerative joint disease.

Quadriceps, hip flexors, hamstrings—Major muscles in the thigh area which affect knee mechanics.

be seen on an MRI, although this type of scan is not routinely performed for this purpose. The patient should inform the doctor about any previous injuries to the joint.

Treatment

Initial treatment may consist of resting the knee using crutches, along with **aspirin**, Tylenol, or a non-steroidal anti-inflammatory drug (NSAID) such as Motrin for seven to 10 days. The person should limit sports activity until the joint is healed and may use ice followed by heat to decrease inflammation. When the doctor allows the patient to resume sports, a knee brace may be prescribed in the form of a stabilizer with a hole at the kneecap.

Treatment also includes low impact exercises to strengthen the quadriceps muscles which help stabilize the knee joint. Physical therapy may be suggested at the start of this program so as to help the patient learn the correct method of performing the exercises.

Approximately 85% of people do well with conservative CMP treatment. The remainder still have severe pain and may require **arthroscopic surgery** to repair the tissues inside the knee joint. In more severe cases, open surgery may be required to realign the kneecap and perhaps other corrections.

Alternative treatments

Physical therapy offers treatments that may help CMP patients. Aqua therapy has the benefit of exercising the knee without putting stress on it and it also strengthens the thigh muscles. **Biofeedback** can be used to learn tensing and relaxing specific muscles to relieve pain. These techniques have the benefit of no side effects. **Massage therapy** might be beneficial as well. Calcium, minerals, and vitamins as part of a balanced diet will aid healing and help prevent further problems.

Prognosis

In most teens with CMP, the prognosis is excellent since the damage is reversible when treatment starts before the cartilage begins to break down. With proper treatment and preventive techniques, teenagers will complete their growth without permanent damage to the joint. Only about 15% of patients require surgical intervention. Older people may go on to develop **osteoarthritis** in the knee.

Prevention

Proper exercises are the best preventive measure. Since tightness of thigh muscles is a risk factor, warming up before athletic activities is recommended, as well as participating in a variety of sports rather than just one. Stretching exercises increase flexibility of the quadriceps, hip flexors, and hamstrings. Strengthening exercises such as short arc leg extensions, straight leg raises, quadriceps isometric exercises, and stationary bicycling are also recommended.

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Barbara J. Mitchell

Chorea see **Movement disorders**

Choriocarcinoma

Definition

A choriocarcinoma is type of **cancer** germ cell containing trophoblast cells.

Description

Choriocarcinomas are cancers that develop from germ cells, cells that ordinarily turn into sperm or eggs. Choriocarcinomas resemble the cells that surround an

KEY TERMS

Biopsy—A sample of an organ taken to look for abnormalities. Also, the technique used to take such samples.

Chemotherapy—The treatment of cancer with drugs.

Computed tomography (CT)—A special x ray technique that produces a cross sectional image of the organs inside the body.

Extragonadal—In a location other than the reproductive organs.

Germ cell—One of the cells that ordinarily develop into eggs or sperm (also sperm and eggs).

Gonads—The ovaries or testes.

Klinefelter syndrome—A condition caused by extra X chromosome(s) in a male, that results in small testes and infertility together with increased height, decreased facial hair, and sometimes breast enlargement.

Magnetic resonance imaging—A type of study that uses changes induced by magnets to see cells and tissues inside the body.

Mole—A mass of abnormal, partially developed tissues inside the uterus (womb). Moles develop during a pregnancy that begins with an abnormal fertilization.

Ovaries—The female sex organs that make eggs and female hormones.

Remission—The disappearance of the symptoms of cancer, although all of the cancer cells may not be gone.

Reproductive organs—The group of organs (including the testes, ovaries, and uterus) whose purpose is to produce a new individual and continue the species.

Testes—The male sex organs that make sperm and male hormones.

Testicular cancer—A cancer that originates in the testes.

Trophoblast—The tissues that surround an embryo and attach it to the uterus.

Tumor—A lump made up of abnormal cells.

Uterus—The organ where a child develops (womb).

embryo in the uterus. Most of these cancers form inside the reproductive organs. Some originate in the testes or ovaries, especially in young adults. Others develop in the uterus after a **pregnancy** or miscarriage—particularly often after a mole. A few choriocarcinomas arise in sites outside the reproductive organs. Such “extragonadal” tumors are usually found in young adults and are more common in males.

Choriocarcinomas are one of the most dangerous germ cell cancers. Choriocarcinomas usually grow quickly and spread widely. Occasionally, this cancer grows so fast that the original tumor outgrows its blood supply and dies, leaving behind only a small scar.

Causes and symptoms

Choriocarcinomas result from genetic damage to a germ cell. Males with **Klinefelter syndrome** are especially likely to develop extragonadal germ cell tumors.

The symptoms of a choriocarcinoma vary, depending on where the tumor originates and where it spreads. In the uterus, the most common symptom is bleeding. Cancers in the ovary often have only subtle signs such as

widening of the waistline or **pain**. In the testes, choriocarcinomas can often be felt as small painless lumps. Choriocarcinomas that spread to other organs may reveal their presence by bleeding. In the brain, this bleeding can cause a **stroke**.

Diagnosis

Choriocarcinomas are usually referred to an oncologist, a doctor who specializes in cancer treatment. To diagnose this tumor, the doctor will do a **physical examination** and examine the internal organs with x rays or ultrasound studies. Choriocarcinomas are not always biopsied before being treated, because they tend to bleed heavily. Spreading of the cancer is detected with x rays, ultrasound studies, computed tomography (CT), or **magnetic resonance imaging** (MRI) scans.

Most choriocarcinomas make human chorionic gonadotropin (hCG), a hormone normally found only during pregnancy. The presence of hCG in the blood can help diagnose this cancer and monitor the success of treatment.

Treatment

Choriocarcinomas are usually treated by surgical removal of the tumor and **chemotherapy**. Radiation is occasionally used, particularly for tumors in the brain.

Alternative treatment

Complementary treatments can decrease **stress**, reduce the side effects of cancer treatment, and help patients feel more in control. For instance, some people find activities such as **yoga**, massage, **music therapy**, **meditation**, prayer, or mild physical exercise helpful.

Prognosis

The prognosis for choriocarcinomas in the uterus is very good. Although these tumors have often spread throughout the body, chemotherapy results in a cure or remission in at least 80–90% of cases. Women who have had choriocarcinomas often go on to have normal pregnancies and deliveries.

Choriocarcinomas in other sites have a poorer prognosis. These tumors tend to spread quickly and don't always respond well to chemotherapy. Although treatment can be effective, the outcome usually depends on how widely the cancer is dispersed. Generally, the prognosis is worse if the cancer can be found in the liver or brain, if hCG levels are high, or if the original tumor developed outside the gonads. Five-year survival with testicular cancers can range from 92% for tumors that have spread only to the lungs to 48% for tumors that have spread to other internal organs.

Prevention

There is no known means of prevention. However, early detection of the symptoms and prompt medical treatment can improve the odds of survival.

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Anna Rovid Spickler, D.V.M., Ph.D.

Chorionic gonadotropin test see **Human chorionic gonadotropin pregnancy test**

Chorionic villus sampling

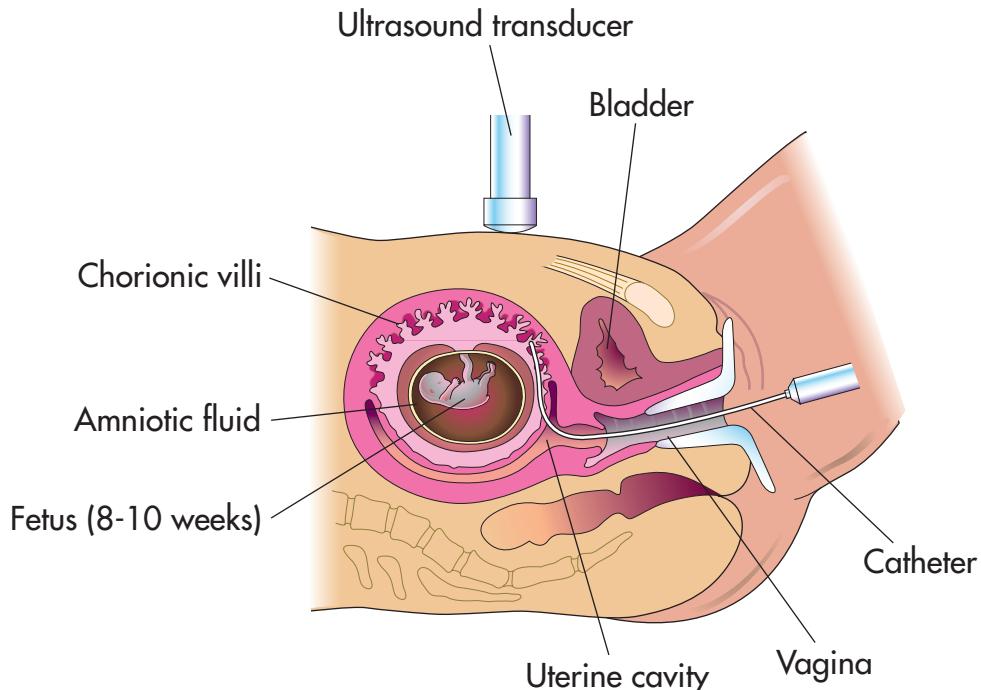
Definition

Chorionic villus sampling (CVS), also known as chorionic villus biopsy, is a prenatal test that can detect genetic and chromosomal abnormalities of an unborn baby.

Purpose

Chorionic villus sampling is performed on pregnant women who are at risk for carrying a fetus with a genetic or chromosomal defect. Although it carries a slightly higher risk, CVS may be used in place of **amniocentesis** for women who have one or more of the following risk factors:

- Women age 35 and older. The chance of having a child with **Down syndrome** increases with maternal age. For instance, the chance of having a baby with Down syndrome is one in 378 for a 35-year-old woman and increases to one in 30 for a 45-year-old woman.
- A history of miscarriages or children born with **birth defects**.
- A family history of genetic disease. Prenatal **genetic testing** is recommended if either the mother or father of



Chorionic villus sampling is performed on pregnant women who are at risk for carrying a fetus with a genetic or chromosomal defect. This procedure can be performed through the vagina and the cervix (transcervically) or through the abdomen (transabdominally). In the transcervical procedure, as depicted above, the physician uses ultrasound to help guide a catheter through the cervix into the uterus. By applying suction from the syringe attached to the other end of the catheter, a small sample of the chorionic villi are obtained. (Illustration by Electronic Illustrators Group.)

the unborn baby has a family history of genetic disease or is known to be a carrier of a genetic disease.

Precautions

Chorionic villus sampling is not recommended for women who have vaginal bleeding or spotting during the **pregnancy**. It is not typically recommended for women who have Rh sensitization from a previous pregnancy.

Description

Chorionic villus sampling has been in use since the 1980s. This prenatal testing procedure involves taking a sample of the chorion frondosum—that part of the chorionic membrane containing the villi—for laboratory analysis. The chorionic membrane is the outer sac which surrounds the developing fetus. Chorionic villi are microscopic, finger-like projections that emerge from the chorionic membrane and eventually form the placenta. The cells that make up the chorionic villi are of fetal origin so laboratory analysis can identify any genetic, chromosomal, or biochemical diseases of the fetus.

Chorionic villus sampling is best performed between 10 and 12 weeks of pregnancy. The procedure is performed either through the vagina and the cervix (transcervically) or through the abdomen (transabdominally) depending upon the preferences of the patient or the doctor. In some cases, the location of the placenta dictates which method the doctor uses. Both methods are equally safe and effective. Following the preparation time, both procedures take only about five minutes. Women undergoing chorionic villus sampling may experience no **pain** at all or feel cramping or pinching. Occasionally, a second sampling procedure must be performed if insufficient villus material was obtained.

For the transcervical procedure, the woman lies on an examining table on her back with her feet in stirrups. The woman's vaginal area is thoroughly cleansed with an antiseptic, a sterile speculum is inserted into her vagina and opened, and the cervix is cleansed with an antiseptic. Using ultrasound (a device which uses sound waves to visualize internal organs) as a guide, the doctor inserts a thin, plastic tube called a catheter through the cervix and into the uterus. The passage of the catheter through the

KEY TERMS

Chorionic villi—Microscopic, finger-like projections that emerge from the outer sac which surrounds the developing baby. Chorionic villi are of fetal origin and eventually form the placenta.

Chromosomes—Human cells carry DNA in tightly compressed rod-like structures called chromosomes. Humans have 23 pairs of chromosomes including the sex chromosomes.

Down syndrome—A chromosomal disorder caused by an extra copy or a rearrangement of chromosome 21. Children with Down syndrome have varying degrees of mental retardation and may have heart defects.

Fetus—Term for an unborn baby after the eighth week of pregnancy. Prior to seven weeks, it is called an embryo.

Rh sensitization—A woman with a negative blood type (Rh negative) who has produced antibodies against her fetus with a positive blood type (Rh positive). The mother's body considered the fetal blood cells a foreign object and mounted an immune attack on it.

Ultrasound—A safe, painless procedure which uses sound waves to visualize internal organs. A wand that transmits and receives the sound waves is moved over the woman's abdomen and internal organs can be seen on a video screen.

cervix may cause cramping. The doctor carefully watches the image produced by the ultrasound and advances the catheter to the chorionic villi. By applying suction from the syringe attached to the other end of the catheter, a small sample of the chorionic villi are obtained. A cramping or pinching feeling may be felt as the sample is being taken. The catheter is then easily withdrawn.

For the transabdominal method, the woman lies on her back on an examining table. Ultrasound enables the doctor to locate the placenta. The specific area on the woman's abdomen is cleansed thoroughly with an anti-septic and a local anesthetic may be injected to numb the area. With ultrasound guidance, a long needle is inserted through the woman's abdominal wall, through the uterine wall and to the chorionic villi. The sample is obtained by applying suction from the syringe.

The chorionic villus sample is immediately placed into nutrient medium and sent to the laboratory. At the lab-

oratory, the sample is examined under the microscope and any contaminating cells or material is carefully removed. The villi can be analyzed immediately, or incubated for a day or more to allow for cell division. The cells are stopped in the midst of cell division and spread onto a microscope slide. Cells with clearly separated chromosomes are photographed so that the type and number of chromosomes can be analyzed. Chromosomes are strings of DNA which have been tightly compressed. Humans have 23 pairs of chromosomes including the sex chromosomes. Rearrangements of the chromosomes or the presence of additional or fewer chromosomes can be identified by examination of the photograph. Down syndrome, for instance, is caused by an extra copy of chromosome 21. In addition to the chromosomal analysis, specialized tests can be performed as needed to look for specific diseases such as **Tay-Sachs disease**. Depending upon which tests are performed, results may be available as early as two days or up to eight days after the procedure.

Chorionic villus sampling costs between \$1,200 and \$1,800. Insurance coverage for this test may vary.

Alternate procedures

There are alternate procedures for diagnosing genetic and chromosomal disorders of the fetus. Amniocentesis is commonly used and involves inserting a needle through the pregnant woman's abdomen to obtain a sample of amniotic fluid. Amniocentesis is usually performed in the second trimester at approximately 16 weeks gestation and the laboratory analysis may take two to three weeks. The two advantages of chorionic villus sampling are that it is performed during the first trimester and the results are available in about one week. However, as of 1997, amniocentesis is being performed in the first trimester, but this is still very rare. The risk of **miscarriage** after amniocentesis is 0.5–1% (one to two women out of 200) which is lower than that for chorionic villus sampling (1–3%).

A noninvasive alternative is the maternal blood test called triple marker screening or multiple marker screening. A sample of the pregnant woman's blood is analyzed for three different markers: alphafetoprotein (AFP), human chorionic gonadotropin, and unconjugated estriol. The levels of these three markers in the mother's blood can identify unborn babies who are at risk for certain genetic or chromosomal defects. This is a screening test which determines the chance that the fetus has the defect, but it can not diagnose defects. A negative test result does not necessarily mean the unborn baby does not have a birth defect. For instance, this screening test can only predict 60–70% of the fetuses with Down syndrome. Pregnant women who have a positive triple marker

screen are encouraged to undergo a diagnostic test, such as amniocentesis (by the time an AFP is done, it is too late to perform a CVS).

Preparation

Prior to the chorionic villus sampling procedure the woman needs to drink fluids and refrain from urinating to ensure her bladder is full. These preparations create a better ultrasound picture.

Aftercare

It is generally recommended that women undergoing chorionic villus sampling have someone drive them home and have no plans for the rest of the day. Women with Rh negative blood must receive a Rho (D) immune globulin injection following the procedure. Women should call their doctor if they experience excessive bleeding, vaginal discharge, **fever**, or abdominal pain after the procedure.

Risks

Of women who undergo transcervical chorionic villus sampling, one third experience minimal vaginal spotting and 7–10% experience vaginal bleeding. One out of five women experience cramping following the procedure. Two to three women out of 100 (or 2–3%) will miscarry following chorionic villus sampling. The risk of infection is very low. Rupture of the amniotic membranes is a rare complication. Women with Rh negative blood may be at an increased risk for developing Rh incompatibility following chorionic villus sampling.

There have been reports of limb defects in babies following chorionic villus sampling. However, in 1996 the World Health Organization reported that the incidence of babies born with limb defects from 138,966 women who had undergone chorionic villus sampling was the same as for women who had not. Therefore, this study found no connection between chorionic villus sampling and limb defects.

Normal results

No genetic, chromosomal, or biochemical abnormalities were found in the fetal cells. The gender of the fetus will be identified but will be made known to the parents only with their approval.

Abnormal results

Analysis of the cells from the chorionic villus enables the detection of over 200 diseases and disorders such as Down Syndrome, Tay-Sachs disease, and **cystic**

fibrosis. Gross rearrangements of the chromosomes and chromosome additions or losses are detected.

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Belinda Rowland, PhD

Choroiditis see **Uveitis**

Choroiretinitis see **Uveitis**

Chromosome studies see **Genetic testing**

Chronic arthritis of childhood see **Juvenile arthritis**

Chronic constrictive pericarditis see
Pericarditis

Chronic Epstein-Barr virus see **Chronic fatigue syndrome**

Chronic fatigue syndrome

Definition

Chronic **fatigue** syndrome (CFS) is a condition that causes extreme tiredness. People with CFS have debilitating fatigue that lasts for six months or longer. They also have many other symptoms. Some of these are **pain** in the joints and muscles, **headache**, and **sore throat**. CFS does not have a known cause, but appears to result from a combination of factors.

Description

CFS is the most common name for this disorder, but it also has been called chronic fatigue and immune disor-

der (CFIDS), myalgic encephalomyelitis, low natural killer cell disease, post-viral syndrome, Epstein-Barr disease, and Yuppie flu. CFS has so many names because researchers have been unable to find out exactly what causes it and because there are many similar, overlapping conditions. Reports of a CFS-like syndrome called neurasthenia date back to 1869. Later, people with similar symptoms were said to have **fibromyalgia** because one of the main symptoms is myalgia, or muscle pain. Because of the similarity of symptoms, fibromyalgia and CFS are considered to be overlapping syndromes.

In the early to mid-1980s, there were outbreaks of CFS in some areas of the United States. Doctors found that many people with CFS had high levels of antibodies to the Epstein-Barr virus (EBV), which causes mononucleosis, in their blood. For a while they thought they had found the culprit, but it turned out that many healthy people also had high EBV antibodies. Scientists have also found high levels of other viral antibodies in the blood of people with CFS. These findings have led many scientists to believe that a virus or combination of viruses may trigger CFS.

CFS was sometimes referred to as Yuppie flu because it seemed to often affect young, middle-class professionals. In fact, CFS can affect people of any gender, age, race, or socioeconomic group. Although anyone can get CFS, most patients diagnosed with CFS are 25–45 years old, and about 80% of cases are in women. Estimates of how many people are afflicted with CFS vary due to the similarity of CFS symptoms to other diseases and the difficulty in identifying it. The Centers for Disease Control and Prevention (CDC) has estimated that four to 10 people per 100,000 in the United States have CFS. According to the CFIDS Foundation, about 500,000 adults in the United States (0.3% of the population) have CFS. This probably is a low estimate since these figures do not include children and are based on the CDC definition of CFS, which is very strict for research purposes.

Causes and symptoms

There is no single known cause for CFS. Studies have pointed to several different conditions that might be responsible. These include:

- viral infections
- chemical toxins
- **allergies**
- immune abnormalities
- psychological disorders

Although the cause is still controversial, many doctors and researchers now think that CFS may not be a single illness. Instead, they think CFS may be a group of

symptoms caused by several conditions. One theory is that a microorganism, such as a virus, or a chemical injures the body and damages the immune system, allowing dormant viruses to become active. About 90% of all people have a virus in the herpes family dormant (not actively growing or reproducing) in their bodies since childhood. When these viruses start growing again, the immune system may overreact and produce chemicals called cytokines that can cause flu-like symptoms. Immune abnormalities have been found in studies of people with CFS, although the same abnormalities are also found in people with allergies, autoimmune diseases, **cancer**, and other disorders.

The role of psychological problems in CFS is very controversial. Because many people with CFS are diagnosed with depression and other psychiatric disorders, some experts conclude that the symptoms of CFS are psychological. However, many people with CFS did not have psychological disorders before getting the illness. Many doctors think that patients become depressed or anxious because of the effects of the symptoms of their CFS. One recent study concluded that depression was the result of CFS and was not its cause.

Having CFS is not just a matter of being tired. People with CFS have severe fatigue that keeps them from performing their normal daily activities. They find it difficult or impossible to work, attend school, or even to take part in social activities. They may have sleep disturbances that keep them from getting enough rest or they may sleep too much. Many people with CFS feel just as tired after a full night's sleep as before they went to bed. When they **exercise** or try to be active in spite of their fatigue, people with CFS experience what some patients call "payback"—debilitating exhaustion that can confine them to bed for days.

Other symptoms of CFS include:

- muscle pain (myalgia)
- joint pain (arthralgia)
- sore throat
- headache
- **fever** and chills
- tender lymph nodes
- trouble concentrating
- memory loss

A recent study at Johns Hopkins University found an abnormality in blood pressure regulation in 22 of 23 patients with CFS. This abnormality, called neurally mediated **hypotension**, causes a sudden drop in blood pressure when a person has been standing, exercising or exposed to heat for a while. When this occurs, patients

feel lightheaded and may faint. They often are exhausted for hours to days after one of these episodes. When treated with salt and medications to stabilize blood pressure, many patients in the study had marked improvements in their CFS symptoms.

Diagnosis

CFS is diagnosed by evaluating symptoms and eliminating other causes of fatigue. Doctors carefully question patients about their symptoms, any other illnesses they have had, and medications they are taking. They also conduct a **physical examination**, neurological examination, and laboratory tests to identify any underlying disorders or other diseases that cause fatigue. In the United States, many doctors use the CDC case definition to determine if a patient has CFS.

To be diagnosed with CFS, patients must meet both of the following criteria:

- Unexplained continuing or recurring chronic fatigue for at least six months that is of new or definite onset, is not the result of ongoing exertion, and is not mainly relieved by rest, and causes occupational, educational, social, or personal activities to be greatly reduced.
- Four or more of the following symptoms: loss of short-term memory or ability to concentrate; sore throat; tender lymph nodes; muscle pain; multi-joint pain without swelling or redness; headaches of a new type, pattern, or severity; unrefreshing sleep; and post-exertional malaise (a vague feeling of discomfort or tiredness following exercise or other physical or mental activity) lasting more than 24 hours. These symptoms must have continued or recurred during six or more consecutive months of illness and must not have started before the fatigue began.

Treatment

There is no cure for CFS, but many treatments are available to help relieve the symptoms. Treatments usually are individualized to each person's particular symptoms and needs. The first treatment most doctors recommend is a combination of rest, exercise, and a balanced diet. Prioritizing activities, avoiding overexertion, and resting when needed are key to maintaining existing energy reserves. A program of moderate exercise helps to keep patients from losing physical conditioning, but too much exercise can worsen fatigue and other CFS symptoms. Counseling and **stress reduction** techniques also may help some people with CFS.

Many medications, nutritional supplements, and herbal preparations have been used to treat CFS. While many of these are unproven, others seem to provide some people with relief. People with CFS should discuss their treatment

KEY TERMS

Arthralgia—Joint pain.

Cytokines—Proteins produced by certain types of lymphocytes. They are important controllers of immune functions.

Depression—A psychological condition, with feelings of sadness, sleep disturbance, fatigue, and inability to concentrate.

Epstein-Barr virus (EBV)—A virus in the herpes family that causes mononucleosis.

Fibromyalgia—A disorder closely related to CFS. Symptoms include pain, tenderness, and muscle stiffness.

Lymph node—Small immune organs containing lymphocytes. They are found in the neck, armpits, groin, and other locations in the body.

Lymphocytes—White blood cells that are responsible for the actions of the immune system.

Mononucleosis—A flu-like illness caused by the Epstein-Barr virus.

Myalgia—Muscle pain.

Myalgic encephalomyelitis—An older name for chronic fatigue syndrome; encephalomyelitis refers to inflammation of the brain and spinal cord.

Natural killer (NK) cell—A lymphocyte that acts as a primary immune defense against infection.

Neurally mediated hypotension—A rapid fall in blood pressure that causes dizziness, blurred vision, and fainting, and is often followed by prolonged fatigue.

Neurasthenia—Nervous exhaustion—a disorder with symptoms of irritability and weakness, commonly diagnosed in the late 1800s.

plan with their doctors, and carefully weigh the benefits and risks of each therapy before making a decision.

Drugs

Nonsteroidal anti-inflammatory drugs (NSAIDs), such as ibuprofen and naproxen, may be used to relieve pain and reduce fever. Another medication that is prescribed to relieve pain and muscle spasms is cyclobenzaprine (sold as Flexeril).

Many doctors prescribe low dosages of antidepressants for their sedative effects and to relieve symptoms of

depression. **Antianxiety drugs**, such as **benzodiazepines** or buspirone may be prescribed for excessive **anxiety** that has lasted for at least six months.

Other medications that have been tested or are being tested for treatment of CFS are:

- Fludrocortisone (Florinef), a synthetic steroid, which is currently being tested for treatment of people with CFS. It causes the body to retain salt, thereby increasing blood pressure. It has helped some people with CFS who have neurally mediated hypotension.
- Beta-adrenergic blocking drugs, often prescribed for high blood pressure. Such drugs, including atenolol (Tenoretic, Tenormin) and propranolol (Inderal), are sometimes prescribed for neurally mediated hypotension.
- Gamma globulin, which contains human antibodies to a variety of organisms that cause infection. It has been used experimentally to boost immune function in people with CFS.
- Ampligen, a drug which stimulates the immune system and has antiviral activity. In one small study, ampligen improved mental function in people with CFS.

Alternative treatment

A variety of nutritional supplements are used for treatment of CFS. Among these are vitamin C, vitamin B₁₂, vitamin A, vitamin E, and various dietary **minerals**. These supplements may help improve immune and mental functions. Several herbs have been shown to improve immune function and have other beneficial effects. Some that are used for CFS are astragalus (*Astragalus membranaceus*), **echinacea** (*Echinacea spp.*), garlic (*Allium sativum*), ginseng (*Panax ginseng*), gingko (*Gingko biloba*), evening primrose oil (*Oenothera biennis*), shiitake mushroom extract (*Lentinus edodes*), borage seed oil, and quercetin.

Many people have enhanced their healing process for CFS with the use of a treatment program inclusive of one or more alternative therapies. **Stress** reduction techniques such as **biofeedback**, **meditation**, **acupuncture**, and **yoga** may help people with sleep disturbances relax and get more rest. They also help some people reduce depression and anxiety caused by CFS.

Prognosis

The course of CFS varies widely for different people. Some people get progressively worse over time, while others gradually improve. Some individuals have periods of illness that alternate with periods of good health. While many people with CFS never fully regain their health, they find relief from symptoms and adapt to

the demands of the disorder by carefully following a treatment plan combining adequate rest, **nutrition**, exercise, and other therapies.

Prevention

Because the cause of CFS is not known, there currently are no recommendations for preventing the disorder.

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- The CFIDS Association. Community Health Services, P.O. Box 220398, Charlotte, NC 28222-0398. (704) 362-2343.
- The National CFS Association. 919 Scott Ave., Kansas City, KS 66105. (913) 321-2278.
- The National CFIDS Foundation. 103 Aletha Road, Needham, MA 02192. (781) 449-3535. <<http://www.cfidsfoundation.org>>.

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<http://www.nih.gov>.

Toni Rizzo

Chronic granulomatous disease

Definition

Chronic granulomatous disease (CGD) is an inherited disorder in which white blood cells lose their ability to destroy certain bacteria and fungi.

Description

CGD is an X-linked genetic disease, meaning the defective gene is carried on the X chromosome (one of the sex chromosomes). Females have two copies of the X chromosome, whereas males have one X and one Y. CGD also is a recessive defect meaning that both copies of the chromosome must have the defect before it can be expressed. Females who have one X chromosome without the defect do not get this disease. Males, since they only have one X chromosome, get the disease if the defect is present. Thus, CGD affects mostly males.

CGD is an **immunodeficiency** disorder. Patients with immunodeficiency disorders suffer frequent infections. This happens because part of their immune system isn't working properly and the infectious microorganisms are not killed as rapidly as is normal. In CGD there is a defect in the ability of the white blood cells to kill bacteria and fungi. The white blood cells affected are phagocytic cells. They are part of the non-specific immune system and move via the blood to all parts of the body where they ingest and destroy microbes. Phagocytic cells are the first line of defense against microorganisms. In this disease, the decreased ability to kill microbes that they have ingested leads to a failure to effectively combat infectious diseases. Patients with CGD are subject to certain types of recurring infection, especially those of the skin, lungs, mouth, nose, intestines, and lymph nodes. With the exception of the lymph nodes, all of these areas are considered external tissues that come into contact with microorganisms from the environment. The lymph system drains all areas of the body to eliminate destroyed microorganisms and to assist the immune system in attacking microorganisms. Infections occur in the lymph nodes as a consequence of the normal draining function.

KEY TERMS

Immunodeficiency—A weakening of the body's immune system.

Phagocytic cells—A cell that ingests microorganisms and foreign particles.

Causes and symptoms

The genetic defect that causes CGD reduces the amount of hydrogen peroxide and superoxide that white blood cells can make. These chemicals are important for killing bacteria and fungi. Without them the white blood cells ingest the microorganisms, but can't kill them. In some cases, the microbes then replicate inside the white blood cell eventually causing its **death**.

Symptoms of the disease usually appear by age two. Frequent, recurrent infections of the skin, lungs (e.g. **pneumonia**), mouth (e.g. gingivitis), nose, intestines and lymph nodes are a hallmark of this disease. Patients may also develop multiple, recurrent liver abscesses and bone infections (**osteomyelitis**).

Diagnosis

Diagnosis is made based on the observation of a pattern of recurrent infections. Blood tests of lymphocyte and antibody functions will be normal. Tests of phagocytic cells will show normal ingestion, but a greatly decreased ability to kill bacteria.

Treatment

Early, aggressive treatment of all infections is critical to the successful management of CGD. Patients are treated with **antibiotics** and immune serum. Antibiotics are used at the first sign of infection. Immune serum is a source of antibodies that help fight infections. Interferon gamma is an experimental treatment for CGD that has shown promising results. There is no cure for the underlying cause of chronic granulomatous disease.

Prognosis

Although antibiotics can treat most infections and may help prevent others, premature death may result, typically due to repeated lung infections.

Prevention

Since CGD is a hereditary disorder, it cannot currently be prevented. Patients and their families may ben-

efit from **genetic counseling**. Preventive (prophylactic) antibiotics may help keep some infections from occurring, and good hygiene, especially rigorous skin and mouth care, can help prevent infections in these areas. Avoiding crowds or other people who have infections are also effective preventive measures.

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ORGANIZATIONS

- Chronic Granulomatous Disease Association. 2616 Monterey Road, San Marino, CA 91108-1646. (818) 441-4118.
National Organization for Rare Disorders. P.O. Box 8923, New Fairfield, CT 06812-8923. (800) 999-6673. <<http://www.rarediseases.org>>.

John T. Lohr, PhD

Chronic kidney failure

Definition

Chronic kidney failure occurs when disease or disorder damages the kidneys so that they are no longer capable of adequately removing fluids and wastes from the body or of maintaining the proper level of certain kidney-regulated chemicals in the bloodstream.

Description

Chronic kidney failure, also known as chronic renal failure, affects over 250,000 Americans annually. It is caused by a number of diseases and inherited disorders, but the progression of chronic kidney failure is always the same. The kidneys, which serve as the body's natural filtration system, gradually lose their ability to remove fluids and waste products (urea) from the bloodstream. They also fail to regulate certain chemicals in the bloodstream, and deposit protein into the urine. Chronic kidney failure is irreversible, and will eventually lead to total kidney failure, also known as end-stage renal disease (ESRD). Without proper treatment intervention to remove wastes and fluids from the bloodstream, ESRD is fatal.

Causes and symptoms

Kidney failure is triggered by disease or a hereditary disorder in the kidneys. Both kidneys are typically affect-

ed. The four most common causes of chronic kidney failure include:

- **Diabetes.** **Diabetes mellitus** (DM), both insulin dependent (IDDM) and non-insulin dependant (NIDDM), occurs when the body cannot produce and/or use insulin, the hormone necessary for the body to process glucose. Long-term diabetes may cause the glomeruli, the filtering units located in the nephrons of the kidneys, to gradually lose functioning.
- **Glomerulonephritis.** Glomerulonephritis is a chronic inflammation of the glomeruli, or filtering units of the kidney. Certain types of glomerulonephritis are treatable, and may only cause a temporary disruption of kidney functioning.
- **Hypertension.** High blood pressure is unique in that it is both a cause and a major symptom of kidney failure. The kidneys can become stressed and ultimately sustain permanent damage from blood pushing through them at an excessive level of pressure over a long period of time.
- **Polycystic kidney disease.** Polycystic kidney disease is an inherited disorder that causes cysts to be formed on the nephrons, or functioning units, of the kidneys. The cysts hamper the regular functioning of the kidney.

Other possible causes of chronic kidney failure include **kidney cancer**, obstructions such as **kidney stones**, **pyelonephritis**, reflux nephropathy, **systemic lupus erythematosus**, **amyloidosis**, sickle cell anemia, **Alport syndrome**, and oxalosis.

Initially, symptoms of chronic kidney failure develop slowly. Even individuals with mild to moderate kidney failure may show few symptoms in spite of increased urea in their blood. Among the symptoms that may be present at this point are frequent urination during the night and high blood pressure.

Most symptoms of chronic kidney failure are not apparent until kidney disease has progressed significantly. Common symptoms include:

- **Anemia.** The kidneys are responsible for the production of erythropoietin (EPO), a hormone which stimulates red blood cell production. If kidney disease causes shrinking of the kidney, this red cell production is hampered.
- **Bad breath or a bad taste in mouth.** Urea, or waste products, in the saliva may cause an ammonia-like taste in the mouth.
- **Bone and joint problems.** The kidneys produce vitamin D, which aids in the absorption of calcium and keeps bones strong. For patients with kidney failure, bones may become brittle, and in the case of children, normal growth may be stunted. Joint **pain** may also occur as a result of unchecked phosphate levels in the blood.

- Edema. Puffiness or swelling around the eyes, arms, hands, and feet.
- Frequent urination.
- Foamy or bloody urine. Protein in the urine may cause it to foam significantly. Blood in the urine may indicate bleeding from diseased or obstructed kidneys, bladder, or ureters.
- Headaches. High blood pressure may trigger headaches.
- Hypertension, or high blood pressure. The retention of fluids and wastes causes blood volume to increase, which in turn, causes blood pressure to rise.
- Increased **fatigue**. Toxic substances in the blood and the presence of anemia may cause feelings of exhaustion.
- **Itching**. Phosphorus, which is typically eliminated in the urine, accumulates in the blood of patients with kidney failure. This heightened phosphorus level may cause itching of the skin.
- Lower back pain. Pain where the kidneys are located, in the small of the back below the ribs.
- Nausea, loss of appetite, and vomiting. Urea in the gastric juices may cause upset stomach. This can lead to **malnutrition** and weight loss.

Diagnosis

Kidney failure is typically diagnosed and treated by a nephrologist, a doctor that specializes in treating the kidneys. The patient that is suspected of having chronic kidney failure will undergo an extensive blood work-up. A blood test will assess the levels of creatinine, blood urea nitrogen (BUN), uric acid, phosphate, sodium, and potassium in the blood. Urine samples will also be collected, usually over a 24-hour period, to assess protein loss.

Uncovering the cause of kidney failure is critical to proper treatment. A full assessment of the kidneys is necessary to determine if the underlying disease is treatable and if the kidney failure is chronic or acute. An x ray, MRI, computed tomography scan, ultrasound, renal biopsy, and/or arteriogram of the kidneys may be employed to determine the cause of kidney failure and level of remaining kidney function. X rays and ultrasound of the bladder and/or ureters may also be taken.

Treatment

Chronic kidney failure is an irreversible condition. Hemodialysis, peritoneal dialysis, or **kidney transplantation** must be employed to replace the lost function of the kidneys. In addition, dietary changes and treatment to relieve specific symptoms such as anemia and high blood pressure are critical to the treatment process.

KEY TERMS

End-stage renal disease (ESRD)—Total kidney failure; chronic kidney failure is diagnosed as ESRD when kidney function falls to 5–10% of capacity.

Nephrotic syndrome—Characterized by protein loss in the urine, low protein levels in the blood, and fluid retention.

Ureters—The two ducts that pass urine from each kidney to the bladder.

Hemodialysis

Hemodialysis is the most frequently prescribed type of dialysis treatment in the United States. Most hemodialysis patients require treatment three times a week, for an average of three to four hours per dialysis “run” depending on the type of dialyzer used and their current physical condition. The treatment involves circulating the patient’s blood outside of the body through an extracorporeal circuit (ECC), or dialysis circuit. The dialysis circuit consists of plastic blood tubing, a two-compartment filter known as a dialyzer, or artificial kidney, and a dialysis machine that monitors and maintains blood flow and administers dialysate, a chemical bath used to draw waste products out of the blood. The patient’s blood leaves and enters the body through two needles inserted into the patient’s vein, called an access site, and is pushed through the blood compartment of the dialyzer. Once inside of the dialyzer, excess fluids and toxins are pulled out of the bloodstream and into the dialysate compartment, where they are carried out of the body. At the same time, electrolytes and other chemicals in the dialysate solution move from the dialysate into the bloodstream. The purified, chemically-balanced blood is then returned to the body.

Peritoneal dialysis

In peritoneal dialysis (PD), the patient’s peritoneum, or lining of the abdomen, acts as a blood filter. A catheter is surgically inserted into the patient’s abdomen. During treatment, the catheter is used to fill the abdominal cavity with dialysate. Waste products and excess fluids move from the patient’s bloodstream into the dialysate solution. After a waiting period of six to 24 hours, depending on the treatment method used, the waste-filled dialysate is drained from the abdomen, and replaced with clean dialysate. There are three types of peritoneal dialysis, which vary by treatment time and administration method: Continuous Ambulatory Peritoneal Dialysis

(CAPD), Continuous Cyclic Peritoneal Dialysis (CCPD), and Intermittent Peritoneal Dialysis (IPD).

Kidney transplantation

Kidney transplantation involves surgically attaching a functioning kidney, or graft, from a brain dead organ donor (a cadaver transplant), or from a living donor, to a patient with ESRD. Patients with chronic renal disease who need a transplant and don't have a living donor register with UNOS (United Network for Organ Sharing), the federal organ procurement agency, to be placed on a waiting list for a cadaver kidney transplant. Kidney availability is based on the patient's health status. When the new kidney is transplanted, the patient's existing, diseased kidneys may or may not be removed, depending on the circumstances surrounding the kidney failure. A regimen of immunosuppressive, or anti-rejection medication, is required after transplantation surgery.

Dietary management

A diet low in sodium, potassium, and phosphorous, three substances that the kidneys regulate, is critical in managing kidney disease. Other dietary restrictions, such as a reduction in protein, may be prescribed depending on the cause of kidney failure and the type of dialysis treatment employed. Patients with chronic kidney failure also need to limit their fluid intake.

Medications and dietary supplements

Kidney failure patients with hypertension typically take medication to control their high blood pressure. Epoetin alfa, or EPO (Epogen), a hormone therapy, and intravenous or oral iron supplements are used to manage anemia. A multivitamin may be prescribed to replace **vitamins** lost during dialysis treatments. Vitamin D, which promotes the absorption of calcium, along with calcium supplements, may also be prescribed.

Since 1973, Medicare has picked up 80% of ESRD treatment costs, including the costs of dialysis and transplantation and of some medications. To qualify for benefits, a patient must be insured or eligible for benefits under Social Security, or be a spouse or child of an eligible American. Private insurance and state Medicaid programs often cover the remaining 20% of treatment costs.

Prognosis

Early diagnosis and treatment of kidney failure is critical to improving length and quality of life in chronic kidney failure patients. Patient outcome varies by the cause of chronic kidney failure and the method chosen to treat it. Overall, patients with chronic kidney disease leading to

ESRD have a shortened lifespan. According to the United States Renal Data System (USRDS), the lifespan of an ESRD patient is 18–47% of the lifespan of the age-sex-race matched general population. ESRD patients on dialysis have a lifespan that is 16–37% of the general population.

The demand for kidneys to transplant continues to exceed supply. In 1996, over 34,000 Americans were on the UNOS waiting list for a kidney transplant, but only 11,330 living donor and cadaver transplants were actually performed. Cadaver kidney transplants have a 50% chance of functioning nine years, and living donor kidneys that have two matching antigen pairs have a 50% chance of functioning for 24 years. However, some transplant grafts have functioned for over 30 years.

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ORGANIZATIONS

- American Association of Kidney Patients (AAKP). 100 S. Ashley Drive, Suite 280, Tampa, FL 33602. (800) 749-2257. <<http://www.aakp.org>>.
- American Kidney Fund (AKF). Suite 1010, 6110 Executive Boulevard, Rockville, MD 20852. (800) 638-8299. <<http://www.arbon.com/kidney>>.
- National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). Building 31, Room 9A04, 31 Center Drive, MSC 2560, Bethesda, MD 208792-2560. (301) 496-3583. <<http://www.niddk.nih.gov>>.
- National Kidney Foundation. 30 East 33rd St., New York, NY 10016. (800) 622-9010. <<http://www.kidney.org>>.
- United States Renal Data System (USRDS). The University of Michigan, 315 W. Huron, Suite 240, Ann Arbor, MI 48103. (734) 998-6611. <<http://www.med.umich.edu/usrds>>.

Paula Anne Ford-Martin

Chronic leukemias see **Leukemias, chronic**

Chronic obstructive lung disease

Definition

Chronic obstructive lung disease, also known as chronic obstructive pulmonary disease (COPD), is a general term for a group of conditions in which there is persistent difficulty in expelling (or exhaling) air from the lungs. COPD commonly refers to two related, progressive diseases of the respiratory system, chronic **bronchitis** and **emphysema**. Because **smoking** is the major cause of both diseases, chronic bronchitis and emphysema often occur together in the same patient.

Description

COPD is one of the fastest-growing health problems. Nearly 16 million people in the United States, 14 million with chronic bronchitis and two million with emphysema, suffer from COPD. COPD is responsible for more than 96,000 deaths annually, making it the fourth leading cause of **death**. Although COPD is more common in men than women, the increase in incidence of smoking among women since World War II has produced an increase in deaths from COPD in women. COPD has a large economic impact on the healthcare system and a destructive impact on the lives of patients and their families. Quality of life for a person with COPD decreases as the disease progresses.

Chronic bronchitis

In chronic bronchitis, chronic inflammation caused by cigarette smoking results in a narrowing of the openings in the bronchi, the large air tubes of the respiratory system, and interferes with the flow of air. Inflammation also causes the glands that line the bronchi to produce excessive amounts of mucus, further narrowing the airways and blocking airflow. The result is often a chronic **cough** that produces sputum (mainly mucus) and **shortness of breath**. Cigarette smoke also damages the cilia, small hair-like projections that move bacteria and foreign particles out of the lungs, increasing the risk of infections.

Emphysema

Emphysema is a disease in which cigarette smoke causes an overproduction of the enzyme elastase, one of the immune system's infection-fighting biochemicals. This results in irreversible destruction of a protein in the

lung called elastin which is important for maintaining the structure of the walls of the alveoli, the terminal small air sacs of the respiratory system. As the walls of the alveoli rupture, the number of alveoli is reduced and many of those remaining are enlarged, making the lungs of the patient with emphysema less elastic and overinflated. Due to the higher pressure inside the chest that must be developed to force air out of the less-elastic lungs, the bronchioles, small air tubes of the respiratory system, tend to collapse during exhalation. Stale air gets trapped in the air sacs and fresh air cannot be brought in.

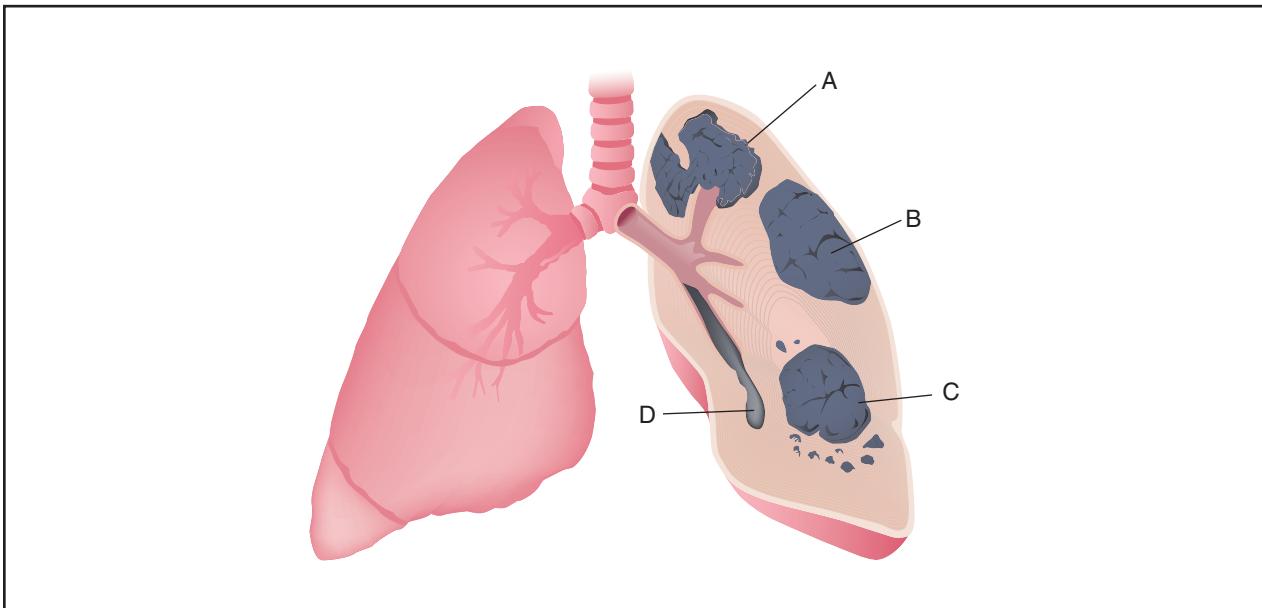
Causes and symptoms

There are several important risk factors for COPD:

- Lifestyle. Cigarette smoking is by far the most important risk factor for COPD (80% of all cases). Cigar and pipe smoking can also cause COPD. Air pollution and industrial dusts and fumes are other important risk factors.
- Age. Chronic bronchitis is more common in people over 40 years old; emphysema occurs more often in people 65 years of age and older.
- Socioeconomic class. COPD-related deaths are about twice as high among unskilled and semi-skilled laborers as among professionals.
- Family clustering. It is thought that heredity predisposes people in certain families to the development of COPD when other causes, such as smoking and air pollution, are present.
- Lung infections. Lung infections make all forms of COPD worse.

In the general population, emphysema usually develops in older individuals with a long smoking history. However, there is also a form of emphysema that runs in families. People with this type of emphysema have a hereditary deficiency of a blood component, an enzyme inhibitor called alpha-1-antitrypsin (AAT). This type of emphysema is sometimes called "early onset emphysema" because it can appear when a person is as young as 30 or 40 years old. It is estimated that there are between 75,000 and 150,000 Americans who were born with AAT-deficiency. Of this group, emphysema afflicts an estimated 20,000–40,000 people (1–3% of all cases of emphysema). The risk of developing emphysema for an AAT-deficient individual who also smokes is much greater than for others.

The first symptoms of chronic bronchitis are cough and mucus production. These symptoms resemble a chest cold that lingers on for weeks. Later, shortness of breath develops. Cough, sputum production, and shortness of breath may become worse if a person develops a lung infection. A person with chronic bronchitis may later



A. lung cancer. B. pneumonia. C. emphysema. D. phlegm from chronic bronchitis. (Illustration by Argosy, Inc.)

develop emphysema as well. In emphysema, shortness of breath on exertion is the predominant early symptom. Coughing is usually minor and there is little sputum. As the disease progresses, the shortness of breath occurs with less exertion, and eventually may be present even when at rest. At this point, a sputum-producing cough may also occur. Either chronic bronchitis or emphysema may lead to respiratory failure—a condition in which there occurs a dangerously low level of oxygen or a serious excess of carbon dioxide in the blood.

Diagnosis

The first step in diagnosing COPD is a good medical evaluation, including a medical history and a **physical examination** of the chest using a stethoscope. In addition, the doctor may request one or more of the following tests:

Pulmonary function test

Using a spirometer, an instrument that measures the air taken into and exhaled from the lungs, the doctor will determine two important values: (1) vital capacity (VC), the largest amount of air expelled after the deepest inhalation, and (2) forced expiratory volume (FEV1), the maximum amount of air expired in one second. The **pulmonary function test** can be performed in the doctor's office, but is expensive.

Chest x ray

Chest x rays can detect only about half of the cases of emphysema. Chest x rays are rarely useful for diagnosing chronic bronchitis.

Blood gas levels

Blood may be drawn from an artery (more painful than drawing blood from a vein) to determine the amount of oxygen and carbon dioxide present. Low oxygen and high carbon dioxide levels are often indicative of chronic bronchitis, but not always of emphysema.

Tests for cause of infection

If infection is present, blood and sputum tests may be done to determine the cause of infection.

Electrocardiogram (ECG)

Many patients with lung disease also develop heart problems. The ECG identifies signs of heart disease.

Treatment

The precise nature of the patient's condition will determine the type of treatment prescribed for COPD. With a program of complete respiratory care, disability can be minimized, acute episodes prevented, hospitalizations reduced, and some early deaths avoided. On the other hand, no treatment has been shown to slow the progress of the disease, and only oxygen therapy increases survival rate.

Drugs

Medications frequently prescribed for COPD patients include:

- **Bronchodilators.** These agents open narrowed airways and offer significant symptomatic relief for many, but not all, people with COPD. There are three types of bronchodilators: Beta₂ agonists, anticholinergic agents, and theophylline and its derivatives. Depending on the specific drug, a bronchodilator may be inhaled, injected, or taken orally.
- **Corticosteroids.** Corticosteroids, usually inhaled, block inflammation and are most useful for patients with chronic bronchitis with or without emphysema. Steroids are generally not useful in patients who have emphysema.
- Oxygen replacement. Eventually, patients with low blood oxygen levels may need to rely on supplemental oxygen from portable or stationary tanks.
- **Antibiotics.** Antibiotics are frequently given at the first sign of a respiratory infection, such as increased sputum production or a change in color of sputum from clear to yellow or green.
- Vaccines. To prevent pulmonary infection from viruses and bacteria, people with COPD should be vaccinated against **influenza** each year at least six weeks before flu season and have a one-time pneumococcal (**pneumonia**) vaccine.
- Expectorants. These agents help loosen and expel mucus secretions from the airways.
- Diuretics. These drugs are given to prevent excess water retention in patients with associated right **heart failure**.
- Augmentation therapy (for emphysema due to AAT-deficiency only). Replacement AAT (Prolastin), derived from human blood which has been screened for viruses, is injected weekly or bimonthly for life.

Surgery

Surgical procedures for emphysema are very rare. They are expensive and often not covered by insurance. The great majority of patients cannot be helped by surgery, and no single procedure is ideal for those who can be helped. In January of 1996, the government temporarily suspended Medicare payments for lung reduction surgery.

- **Lung transplantation.** Lung transplantation has been successfully employed in some patients with end-stage COPD. In the hands of an experienced team, the one-year survival rate is over 70%.
- Lung volume reduction. These procedures remove 20–30% of severely diseased lung tissue; the remaining parts of the lung are joined together. Mortality rates can be as high as 15% and complication rates are even

KEY TERMS

Alpha-1-antitrypsin (AAT)—A blood component that breaks down infection-fighting enzymes such as elastase.

Alveoli—Terminal air sacs of the respiratory system, where gas (oxygen and carbon dioxide) exchange occurs.

Bronchi—Large air tubes of the respiratory system.

Bronchioles—Small air tubes of the respiratory system.

Bronchodilators—Drugs that open wider the bronchial tubes of the respiratory system.

Corticosteroids—A group of hormones that are used as drugs to block inflammation.

Forced expiratory volume (FEV1)—The maximum amount of air expired in one second.

Spirometer—An instrument used by a doctor to perform a breathing test.

Vital capacity (VC)—The largest amount of air expelled after one's deepest inhalation.

higher. When the operation is successful, patients report significant improvement in symptoms.

Pulmonary rehabilitation

A structured, outpatient pulmonary **rehabilitation** program improves functional capacity in certain patients with COPD. Services may include general **exercise** training, administration of oxygen and nutritional supplements, intermittent mechanical ventilatory support, continuous positive airway pressure, relaxation techniques, breathing exercises and techniques (such as pursed lip breathing), and methods for mobilizing and removing secretions.

Alternative treatment

For both chronic bronchitis and emphysema, alternative practitioners recommend diet and nutritional supplements, a variety of herbal medicines, **hydrotherapy**, **acupressure** and **acupuncture**, **aromatherapy**, **homeopathy**, and **yoga**.

Prognosis

COPD is a disease that can be treated and controlled, but not cured. Survival of patients with COPD is clearly related to the degree of their lung function when they are

diagnosed and the rate at which they lose this function. Overall, the median survival is about 10 years for patients with COPD who have lost approximately two-thirds of their lung function at diagnosis.

Prevention

Lifestyle modifications that can help prevent COPD, or improve function in COPD patients, include: quitting smoking, avoiding respiratory irritants and infections, avoiding allergens, maintaining good **nutrition**, drinking lots of fluids, avoiding excessively low or high temperatures and very high altitudes, maintaining proper weight, and exercising to increase muscle tone.

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ORGANIZATIONS

- American Association for Respiratory Care. 11030 Ables Lane, Dallas, TX 75229. (214) 243-2272. <<http://www.aarc.org>>.
 American Lung Association. 1740 Broadway, New York, NY 10019. (800) 586-4872. <<http://www.lungusa.org>>.
 National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.
 National Jewish Medical and Research Center. 1400 Jackson St., Denver, CO 80206. (800) 222-LUNG (Lung Line). <<http://www.njc.org>>.

Harry W. Golden

Chronic obstructive pulmonary disease see
Emphysema; Chronic obstructive lung disease

Churg-Strauss syndrome see **Vasculitis**

Cingulotomy see **Psychosurgery**

Ciprofloxacin see **Fluoroquinolones**

Circadian rhythm sleep disorders see **Jet lag**

Circumcision

Definition

The surgical removal of the foreskin of the penis or prepuce.

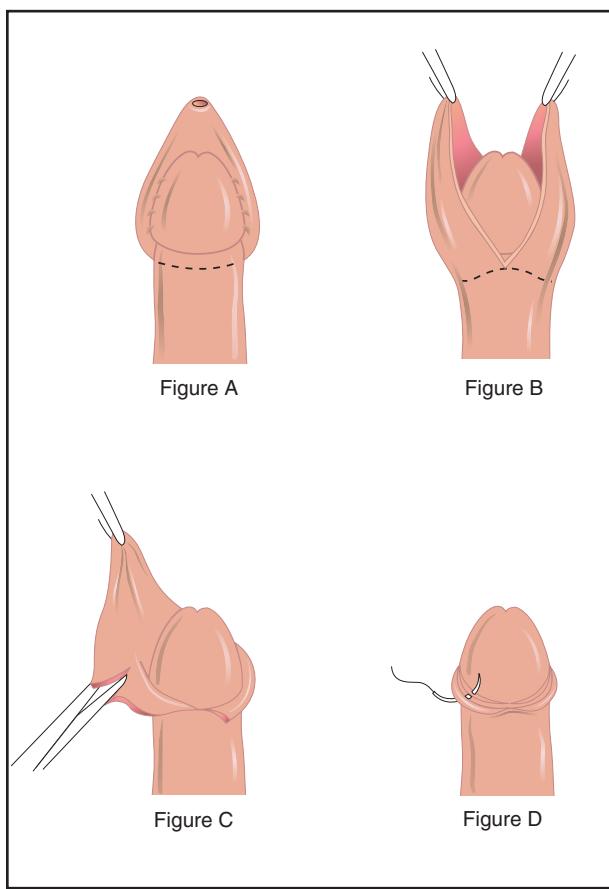
Purpose

In the United States, circumcision in infant boys is performed for social, medical, or cultural/religious reasons. Once a routine operation urged by pediatricians and obstetricians for newborns in the middle of the twentieth century, circumcision has become an elective option that parents make for their sons on an individual basis. Families who practice Judaism or Islam may select to have their sons circumcised as a religious practice. Others choose circumcision for medical benefits.

Female circumcision (also known as **female genital mutilation**) is usually performed for cultural and social reasons by family members and others who are not members of the medical profession, with no anesthesia. Not only is the prepuce removed but often the vaginal opening is sewn to make it smaller. This practice is supposed to ensure the virginity of a bride on her wedding day. It also prevents the woman from achieving sexual pleasure during coitus. This practice is not universally approved by the medical profession and is considered by some as a human rights violation.

Though the incidence of male circumcision has decreased from 90% in 1979 to 60% in 1996, it is still the most common surgical operation in the United States. Circumcision rates are much lower for the rest of the industrialized world. In Britain, it is only done for religious practices or to correct a specific medical condition of the penis.

Some of the medical reasons parents choose circumcision are to protect against infections of the urinary tract and the foreskin, prevent **cancer**, lower the risk of getting **sexually transmitted diseases**, and prevent phimosis (a tightening of the foreskin that may close the opening of the penis). Though studies indicate that uncircumcised boys under the age of five are 20 times more likely than circumcised boys to have



A typical circumcision procedure involves the following steps: Figure A: The surgeon makes an incision around the foreskin. Figure B: The foreskin is then freed from the skin covering the penile shaft. Figure C: The surgeon cuts the foreskin to the initial incision, lifting the foreskin from the mucous membrane. Figure D: The surgeon sutures the top edge of the skin that covers the penile shaft and the mucous membrane. (Illustration by Electronic Illustrators Group.)

urinary tract infections (UTIs), the rate of incidence of UTIs is quite low. There are also indications that circumcised men are less likely to suffer from **penile cancer**, inflammation of the penis, or have many sexually transmitted diseases. Here again, the rate of incidence is low. Good hygiene usually prevents most infections of the penis. Phimosis and penile cancer are very rare, even in men who have not been circumcised. Education and good safe sex practices can prevent sexually transmitted diseases in ways that a surgical procedure cannot because these are diseases acquired through risky behaviors.

With these factors in mind, the American Academy of Pediatrics has issued a policy statement that states though there is existing scientific evidence that indicates the medical benefits of circumcision, the benefits aren't

KEY TERMS

Foreskin—A covering fold of skin over the tip of the penis.

Glans—The cone-shaped tip of the penis.

Hernia—Bulging of abdominal structures through an abnormal opening in the muscular wall.

Hydrocele—Collection of fluid in the scrotum.

Hypospadias—A congenital deformity of the penis where the urinary tract opening is not at the tip of the glans.

Phimosis—A tightening of the foreskin that may close the opening of the penis.

Prepuce—A fold like the foreskin that covers the clitoris; another name for foreskin.

strong enough to recommended circumcision as a routine practice.

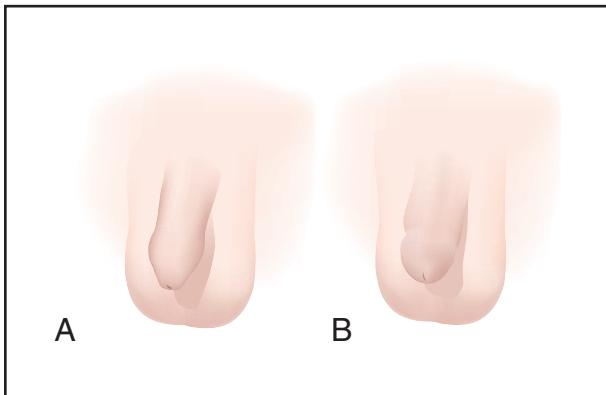
Precautions

Circumcision should not be performed on infants with certain deformities of the penis that may require a portion of the foreskin for repair. The most common condition for surgery using the foreskin is hypospadias, a congenital deformity of the penis where the urinary tract opening is not at the tip of the glans. Also, infants with a large hydrocoele or **hernia** may suffer important complications through circumcision. Premature infants and infants with serious infections are also poor candidates to be circumcised, as are infants with **hemophilia**, other bleeding disorders, or whose mothers had taken anticoagulant drugs. In older boys or men, circumcision is a minor procedure. Therefore, it can be performed in virtually anyone without a serious illness or unusual deformity.

Description

The foreskin of the penis protects the sensitivity of the glans and shields it from irritation by urine, feces, and foreign materials. It also protects the urinary opening against infection and incidental injury.

In circumcision of infants, the foreskin is pulled tightly into a specially designed clamp, and the foreskin pulls away from the broadened tip of the penis. Pressure from the clamp stops bleeding from blood vessels that supplied the foreskin. In older boys or adults, an incision is made around the base of the foreskin, the foreskin is



A. uncircumcised penis. B. circumcised penis. (Illustration by Argosy Inc.)

pulled back, and then it is cut away from the tip of the penis. Stitches are usually used to close the skin edges.

Preparation

Despite a long-standing belief that infants do not experience serious **pain** from circumcision, most authorities now believe that some form of local anesthesia is necessary. The physician injects local anesthesia at the base of the penis or under the skin around the penis (subcutaneous ring block). Both anesthetics block key nerves. EMLA cream, a topical formula of several anesthetics can also be used.

Aftercare

After circumcision, the wound should be washed daily. An antibiotic ointment or petroleum jelly may be applied to the site. If there is an incision, a wound dressing will be present and should be changed each time the diaper is changed. Sometimes a plastic ring is used instead of a bandage. The ring will usually fall off in five to eight days. The penis will heal in seven to 10 days.

Infants who undergo circumcision may be fussy for some hours afterward, so parents should be prepared for crying, feeding problems, and sleep problems. Generally these go away within a day. In older boys, the penis may be painful, but this will go away gradually. A topical anesthetic ointment or spray may be used to relieve this temporary discomfort. There may also be a "bruise" on the penis, which typically goes away with no particular attention.

Risks

Complications following newborn circumcision appear in one out of every 500 procedures. Most complications are minor. Bleeding occurs in half of the compli-

cations and is usually easy to control. Infections are rare and present with **fever** and signs of inflammation.

There may be injuries to the penis itself, and these may be difficult to repair. In 2000, there were reports that the surgical clamps used in circumcision were at fault in over 100 injuries reported between July 1996 and January 2000. In nearly all cases, the clamps were assumed to be in working order but had been repaired with replacement parts that were not of the manufacturer's specifications. Physicians were urged to inspect the clamps before use and ensure that their dimensions fit their infant patients.

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Janie F. Franz

Cirrhosis

Definition

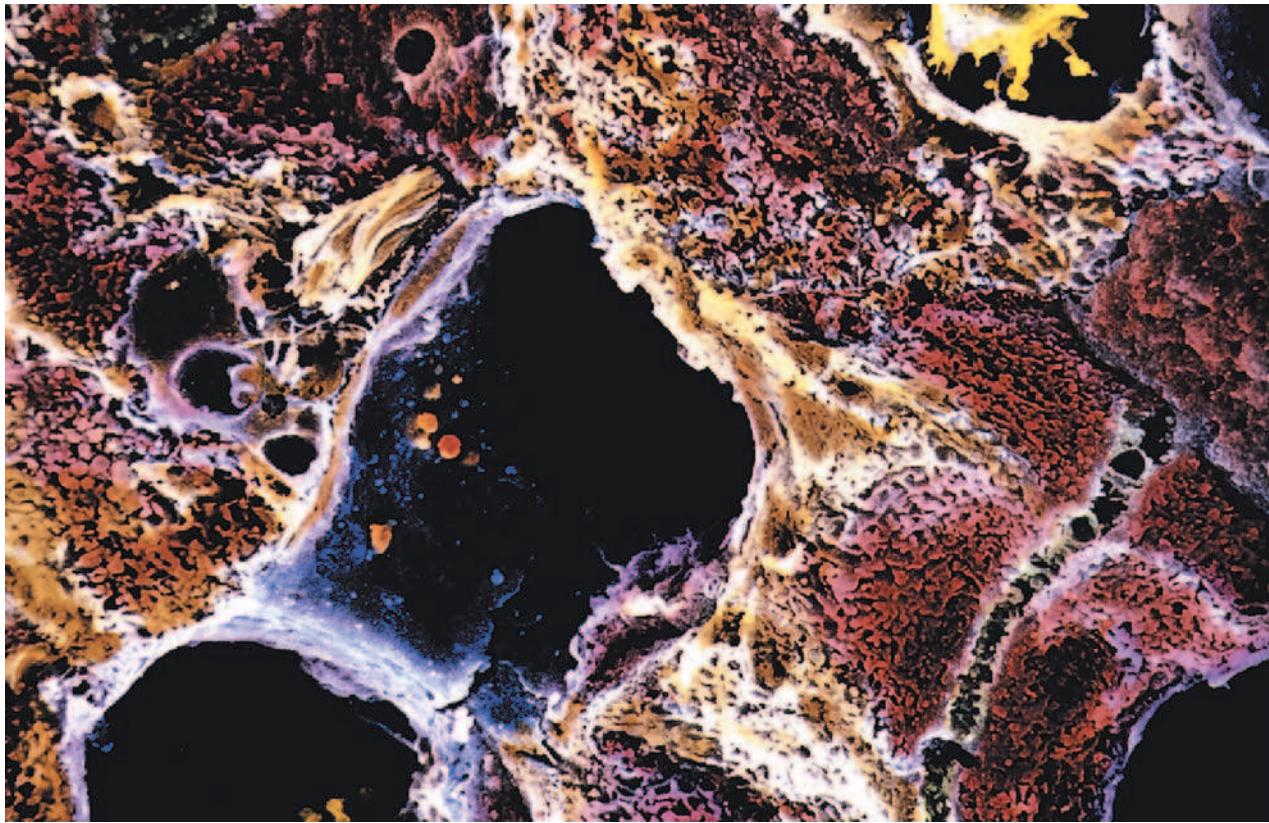
Cirrhosis is a chronic, degenerative disease in which normal liver cells are damaged and are then replaced by scar tissue.

Description

Cirrhosis changes the structure of the liver and the blood vessels that nourish it. The disease reduces the liver's ability to manufacture proteins and process hormones, nutrients, medications, and poisons.

Cirrhosis gets worse over time and can become potentially life threatening. This disease can cause:

- excessive bleeding (hemorrhage)
- impotence
- liver **cancer**
- coma due to accumulated ammonia and body wastes (liver failure)



A micrograph of a human liver showing tissue damaged by cirrhosis. (Photograph by Professor P. Motta, Photo Researchers, Inc. Reproduced by permission.)

• death

Cirrhosis is the seventh leading cause of disease-related death in the United States. It is twice as common in men as in women. The disease occurs in more than half of all malnourished chronic alcoholics and kills about 25,000 people a year. It is the third most common cause of death in adults between the ages of 45 and 65.

Types of cirrhosis

Portal or nutritional cirrhosis is the form of the disease most common in the United States. About 30–50% of all cases of cirrhosis are this type. Nine out of every 10 people who have nutritional cirrhosis have a history of **alcoholism**. Portal or nutritional cirrhosis is also called Laënnec's cirrhosis.

Biliary cirrhosis is caused by intrahepatic bile-duct diseases that impede bile flow. Bile is formed in the liver and is carried by ducts to the intestines. Bile then helps digest fats in the intestines. Biliary cirrhosis can scar or block these ducts. It represents 15–20% of all cirrhosis.

Various types of chronic hepatitis, especially **hepatitis B** and **hepatitis C**, can cause postnecrotic cirrhosis.

This form of the disease affects up to 40% of all patients who have cirrhosis.

Disorders like the inability to metabolize iron and similar disorders may cause pigment cirrhosis (**hemochromatosis**), which accounts for 5–10% of all instances of the disease.

Causes and symptoms

Long-term alcoholism is the primary cause of cirrhosis in the United States. Men and women respond differently to alcohol. Although most men can safely consume two to five drinks a day, one or two drinks a day can cause liver damage in women. Individual tolerance to alcohol varies, but people who drink more and drink more often have a higher risk of developing cirrhosis. In some people, one drink a day can cause liver scarring.

Chronic liver infections like hepatitis B and particularly hepatitis C are commonly linked to cirrhosis. People at high risk of contracting hepatitis B include those exposed to the virus through contact with blood and body fluids. This includes healthcare workers and intravenous

(IV) drug users. People in the past have contracted hepatitis C through blood transfusions.

Liver injury, reactions to prescription medications, exposure to toxic substances, and repeated episodes of **heart failure** with liver congestion can cause cirrhosis. The disorder can also be a result of diseases that run in families (inherited diseases) like:

- a lack of a specific liver enzyme (α_1 -antitrypsin deficiency)
- the absence of a milk-digesting enzyme (galactosemia)
- an inability to convert sugars to energy (glycogen storage disease)
- an absorption deficit in which excess iron is deposited in the liver, pancreas, heart, and other organs (hemochromatosis)
- a disorder characterized by accumulations of copper in the liver, brain, kidneys, and corneas (Wilson's disease)

Poor **nutrition** increases a person's risk of developing cirrhosis. In about 10 out of every 100 patients, the cause of cirrhosis cannot be determined. Many people who have cirrhosis do not have any symptoms (often called compensated cirrhosis). Their disease is detected during a routine physical or when tests for an unrelated medical problem are performed. This type of cirrhosis can also be detected when complications occur (decompensated cirrhosis).

Symptoms of cirrhosis are usually caused by the loss of functioning liver cells or organ swelling due to scarring. The liver enlarges during the early stages of illness. The palms of the hands turn red and patients may experience:

- constipation
- diarrhea
- dull abdominal **pain**
- fatigue
- indigestion
- loss of appetite
- nausea
- vomiting
- weakness
- weight loss

As the disease progresses, the spleen enlarges and fluid collects in the abdomen (**ascites**) and legs (**edema**). Spider-like blood vessels appear on the chest and shoulders, and bruising becomes common. Men sometimes lose chest hair. Their breasts may grow and their testicles may shrink. Women may have menstrual irregularities.

Cirrhosis can cause extremely dry skin and intense **itching**. The whites of the eyes and the skin may turn yellow (**jaundice**), and urine may be dark yellow or brown. Stools may be black or bloody. Sometimes the patient develops persistent high blood pressure due to the scarring (**portal hypertension**). This type of hypertension can be life threatening. It can cause veins to enlarge in the stomach and in the tube leading from the mouth to the stomach (esophagus). These enlarged veins are called varices, and they can rupture and bleed massively.

Other symptoms of cirrhosis include:

- anemia
- bleeding gums
- decreased interest in sex
- fever
- fluid in the lungs
- hallucinations
- lethargy
- lightheadedness
- muscle weakness
- musty breath
- painful nerve inflammation (neuritis)
- slurred speech
- tremors

If the liver loses its ability to remove toxins from the brain, the patient may have additional symptoms. The patient may become forgetful and unresponsive, neglect personal care, have trouble concentrating, and acquire new sleeping habits. These symptoms are related to ammonia intoxication and the failure of the liver to convert ammonia to urea. High protein intake in these patients can also lead to these symptoms.

Diagnosis

A patient's medical history can reveal illnesses or lifestyles likely to lead to cirrhosis. Liver changes can be seen during a **physical examination**. A doctor who suspects cirrhosis may order blood and urine tests to measure liver function. Because only a small number of healthy cells are needed to carry out essential liver functions, test results may be normal even when cirrhosis is present.

Computed tomography scans (CT), ultrasound, and other imaging techniques can be used during diagnosis. They can help determine the size of the liver, indicate healthy and scarred areas of the organ, and detect **gallstones**. Cirrhosis is sometimes diagnosed during surgery or by examining the liver with a laparoscope. This view-

ing device is inserted into the patient's body through a tiny incision in the abdomen.

Liver biopsy is usually needed to confirm a diagnosis of cirrhosis. In this procedure, a tissue sample is removed from the liver and is examined under a microscope in order to learn more about the organ.

Treatment

The goal of treatment is to cure or reduce the condition causing cirrhosis, prevent or delay disease progression, and prevent or treat complications.

Salt and fluid intake are often limited, and activity is encouraged. A diet high in calories and moderately high in protein can benefit some patients. **Tube feedings** or vitamin supplements may be prescribed if the liver continues to deteriorate. Patients are asked not to consume alcohol.

Medication

Iron supplements, **diuretics**, and **antibiotics** may be used for anemia, fluid retention, and ammonia accumulation associated with cirrhosis. Vasoconstrictors are sometimes needed to stop internal bleeding and antiemetics may be prescribed to control nausea.

Laxatives help the body absorb toxins and accelerate their removal from the digestive tract. **Beta blockers** may be prescribed to control cirrhosis-induced portal hypertension. Because the diseased liver can no longer efficiently neutralize harmful substances, medications must be given with caution. Interferon medicines may be used by patients with chronic hepatitis B and hepatitis C to prevent post-hepatitis cirrhosis.

Surgery

Medication that causes scarring can be injected directly into veins to control bleeding from varices in the stomach or esophagus. Varices may require a special surgical procedure called balloon tamponade ligation to stop the bleeding. Surgery may be required to repair disease-related throat damage. It is sometimes necessary to remove diseased portions of the spleen and other organs.

Liver transplants can benefit patients with advanced cirrhosis. However, the new liver will eventually become diseased unless the underlying cause of cirrhosis is removed. Patients with alcoholic cirrhosis must demonstrate a willingness to stop drinking before being considered suitable transplant candidates.

Supportive measures

A balanced diet promotes regeneration of healthy liver cells. Eating five or six small meals throughout the

day should prevent the sick or bloated feeling patients with cirrhosis often have after eating. Alcohol and **caffeine**, which destroy liver cells, should be avoided. So should any foods that upset the stomach. Patients with brain disease associated with cirrhosis should avoid excessive amounts of protein in the diet.

A patient can keep a food diary that describes what was eaten, when it was eaten, and how the patient felt afterwards. This diary can be useful in identifying foods that are hard to digest and in scheduling meals to coincide with the times the patient is most hungry.

Patients who have cirrhosis should weigh themselves every day and notify their doctor of a sudden gain of five pounds or more. A doctor should also be notified if symptoms of cirrhosis appear in anyone who has not been diagnosed with the disease. A doctor should also be notified if a patient diagnosed with cirrhosis:

- vomits blood
- passes black stools
- seems confused or unresponsive
- shows signs of infection (redness, swelling, tenderness, pain)

Alternative treatment

Alternative treatments for cirrhosis are aimed at promoting the function of healthy liver cells and relieving the symptoms associated with the disease. Several herbal remedies may be helpful to cirrhosis patients. Dandelion (*Taraxacum officinale*) and rock-poppy (*Chelidonium majus*) may help improve the efficiency of liver cells. Milk thistle extract (*Silybum marianum*) may slow disease progression and significantly improve survival rates in alcoholics and other cirrhosis patients. Practitioners of **homeopathy** and **traditional Chinese medicine** can also prescribe treatments that support healthy liver function.

Prognosis

Cirrhosis-related liver damage cannot be reversed, but further damage can be prevented by patients who:

- eat properly
- get enough rest
- do not consume alcohol
- remain free of infection

If the underlying cause of cirrhosis cannot be corrected or removed, scarring will continue. The liver will fail, and the patient will probably die within five years. Patients who stop drinking after being diagnosed with cirrhosis can increase their likelihood of living more than a few years from 40% to 60–70%.

Prevention

Eliminating alcohol abuse could prevent 75–80% of all cases of cirrhosis.

Other preventive measures include:

- obtaining counseling or other treatment for alcoholism
- taking precautions (practicing safe sex, avoiding dirty needles) to prevent hepatitis
- getting immunizations against hepatitis if a person is in a high-risk group
- receiving appropriate medical treatment quickly when diagnosed with hepatitis B or hepatitis C
- having blood drawn at regular intervals to rid the body of excess iron from hemochromatosis
- using medicines (chelating agents) to rid the body of excess copper from Wilson's disease
- wearing protective clothing and following product directions when using toxic chemicals at work, at home, or in the garden

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Maureen Haggerty

Cisapride see Antigastroesophageal reflux drugs

CK test see Creatine kinase test

Clap see Gonorrhea

Clarithromycin see Erythromycins

Cleft lip and palate

Definition

A cleft is a birth defect that occurs when the tissues of the lip and/or palate of the fetus do not fuse very early in **pregnancy**. A cleft lip, sometimes referred to as a harelip, is an opening in the upper lip that can extend into the base of the nostril. A cleft palate is an opening in the roof of the mouth.

Description

Babies born with cleft lips will have an opening involving the upper lip. The length of the opening ranges from a small notch, to a cleft that extends into the base of the nostril. Cleft lips may involve one or both sides of the lip.

Babies born with cleft palates have openings in the palate, which is the roof of the mouth. The size and position of the opening varies. The cleft may be only in the hard palate, the bony portion of the roof of the mouth, opening into the floor of the nose. It may be only in the soft palate, the soft portion of the roof of the mouth. The cleft palate may involve both the hard and soft palate and may occur on both sides of the center of the palate.

Babies may have cleft lips with or without cleft palates. Cleft palates may also occur without cleft lips.

The incidence of cleft lip and palate not associated with a syndrome is one in 700 newborns. Native Americans have an incidence of 3.6 in 1,000 newborns. The incidence among Japanese newborns is 2.1 in 1,000. The incidence among whites is one in 1,000 newborns. African Americans have an incidence of 0.3 in 1,000 newborns.

Causes and symptoms

Cleft lips and palates not associated with a syndrome are caused by a combination of genetic and environmental factors. Inheritance caused by such a combination is called multifactorial. The embryo inherits genes that increase the risk for cleft lip and/or palate. When an embryo with such genes is exposed to certain environmental factors the embryo develops a cleft.

The risk of a baby being born with a cleft lip or palate increases with the number of affected relatives and increases with relatives that have more severe clefts.

Environmental factors that increase the risk of cleft lip and palate include cigarette and alcohol use during

pregnancy. Some drugs also increase the incidence of clefting, such as phenytoin, sodium valproate, and methotrexate. The pregnant mother's **nutrition** may affect the incidence of clefting as well.

Babies born with a cleft lip will be seen to have an elongated opening in the upper lip. The size of this opening may range from a small notch in the upper lip to an opening that extends into the base of the nostril. The cleft lip may be below the right or left nostril or below both nostrils.

Babies born with a cleft palate will be seen to have an opening into the roof of the mouth. The size and position of the cleft varies and it may involve only the hard palate, or only the soft palate and may occur on both sides of the center of the palate.

In some cases the cleft palate will be covered with the normal lining of the mouth and can only be felt by the examiner.

Babies with cleft lips and palates have feeding difficulties, which are more severe in babies with cleft palates. The difficulty in feeding is due to the baby being unable to achieve complete suction. In the case of clefts of the hard palate, liquids enter the nose from the mouth through the opening in the hard palate.

A cleft palate also affects a child's speech, since the palate is necessary for speech formation. The child's speech pattern may still be affected despite surgical repair.

Ear infections are more common in babies born with cleft palates. The infections occur because the muscles of the palate do not open the Eustachian tubes which drain the middle ear. This allows fluid to collect and increases the risk of infection and **hearing loss**.

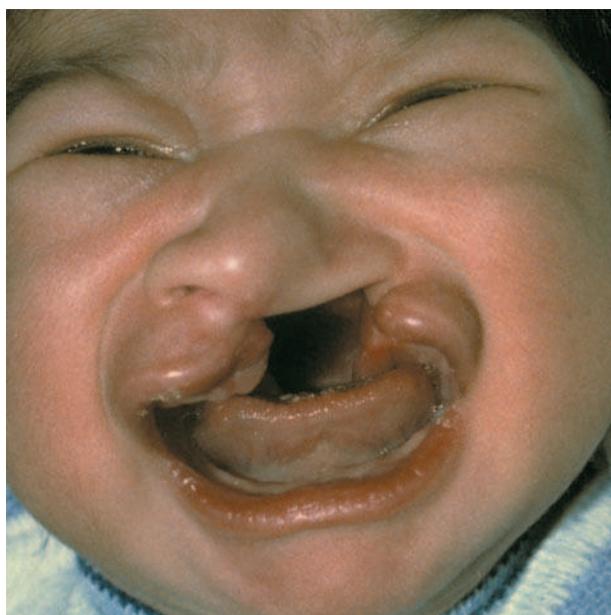
Teeth may also erupt misaligned.

Diagnosis

Cleft lip and palate can be diagnosed before birth by ultrasound. After birth, cleft lip and palate are diagnosed by physical exam.

Treatment

If cleft lip and/or palate are diagnosed by ultrasound before birth, further testing may be required to diagnose associated abnormalities if present. Referral to a cleft team is essential. A cleft team consists of specialists in the management of babies with clefts and includes surgeons as well as nurses and speech therapists. Members of the team inform the parents of all aspects of management. Feeding methods are also discussed, since feeding is the first problem that must be dealt with. It may be possible to breastfeed a baby born with only a cleft lip, but babies born with cleft palates usually have more



This infant has an unilateral cleft lip and palate. (Custom Medical Stock Photo. Reproduced by permission.)

problems with feeding and frequently require special bottles and teats. A palatal obturator is a device that fits into the roof of the mouth, thus blocking the cleft opening and allowing easier suckling.

Surgery to repair cleft lips is sometimes performed after orthodontic treatment to narrow the gap in the upper lip. The orthodontic treatment can involve acrylic splints with or without screws or may involve the use of adhesive tape placed across the gap in the lip. The orthodontic treatment for cleft lip should be started within the first three weeks of life and continue until the cleft lip is repaired.

The timing of surgical cleft lip repair depends on the judgment of the surgeon who will perform the operation. The procedure is usually performed between one and three months of age. The goals of the operation are to close the gap in the upper lip, place scars in the natural skin curves and to repair muscle so that the lip appears normal during movement. The closure is done in the three layers (skin, muscle, and mucosa) that line the inside of the lip. At the time of the procedure, if the nose is shaped abnormally due to the cleft lip, it is also corrected. Sometimes further surgery may be needed on the lip and/or nose to refine the result.

The goals of the surgeon repairing a cleft palate are normal speech, normal facial growth, and hearing for the affected infant. The repair of the cleft palate is usually performed between three and 18 months of age. The timing may extend beyond this and varies with the type of cleft palate and center where the procedure is being performed.

Depending on the type of cleft palate, more than one operation may be needed to close the cleft and improve speech.

Nonsurgical treatment of a cleft palate is available for patients who are at high risk for surgery and consists of a prosthetic appliance worn to block the opening in the palate.

Babies born with cleft palates are vulnerable to ear infections. Their Eustachian tubes do not effectively drain fluid from the middle ear so fluid accumulates and infection sets in. This may lead to hearing loss. These children require drainage tubes to be inserted to prevent fluid accumulation.

Babies born with clefts usually require orthodontic treatment between 13 and 18 years of age. They also require speech therapy.

Prognosis

Babies born with cleft lip and palate have a good prognosis, and approximately 80% will develop normal speech. There is no known means of preventing clefting. Good prenatal care is essential and avoiding harmful substances appear to reduce the risk.

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ORGANIZATIONS

- Cleft Palate Foundation. (800) 24-CLEFT. <<http://www.cleftline.org>>.

Farris F. Gulli, MD

Cleft palate see **Cleft lip and palate**

Climacteric see **Menopause**

Clenched fist injury

Definition

A clenched fist injury (CFI) is a bite wound on the hand, caused when a person's closed fist strikes the teeth of another person, usually in the course of a fight. CFIs are sometimes referred to as closed fist injuries or fight bites.

Description

Clenched fist injuries are most common over the metacarpophalangeal joint. Their appearance is deceptive because they do not bleed heavily and the underlying injury is hidden by soft tissue when the patient opens his hand and straightens the injured finger. CFIs can, however, have serious consequences, including infection, cellulitis, inflammation of the bone or bone marrow (**osteomyelitis**), septic arthritis, and inflammation of the sheaths covering the tendons of the hand (tenosynovitis). These may lead to permanent loss of function or **amputation**.

Most CFIs result in tissue injury due to the force of impact, ragged-edged tears in the skin resulting from contact with the teeth, and contamination of the wound by the bacteria in human saliva. As the patient opens his hand, the skin of the finger is pulled backward over the deeper part of the wound, thus sealing bacteria within the injured tissue. This sealing of the wound by normal motions of the finger is the reason why clenched fist injuries have the highest rate of infection of any human bite. The rate of infection of clenched-fist injuries varies from 15–50%.

Causes and symptoms

The causes of CFIs include fighting and other forms of aggressive behavior, often combined with drug or alcohol consumption.

The symptoms of clenched fist injury include **pain** in the affected part of the hand and some stiffness of the injured finger with limitation of movement. If the patient has delayed getting medical treatment, there may be evidence of infection, including swelling, redness, and suppuration (a discharge of pus). The skin around the wound will be warm to the touch and **fever** may be present.

Diagnosis

Diagnosis of clenched fist injuries is usually made on the basis of the location of the injury and x-ray findings. The most common finding in CFI x rays is soft tissue swelling, but the x rays may also reveal air pockets in deep tissues or the joint spaces, fragments of teeth, frac-

ture lines in the bones, or small loose bone chips. Diagnosis is often complicated by the fact that the patient will be reluctant to admit how the injury happened. The treating physician must maintain a high level of suspicion and often ask directly.

Treatment

Treatment of clenched fist injuries is complicated by several factors. One factor is the anatomical structure of the human hand, which contains many small closed spaces that make it easy for infection to spread and persist. Another is the number of disease-causing bacteria transmitted by human bites; at least 42 different species have been identified. In addition, CFIs typically do not receive immediate treatment because the patient is concerned about legal consequences. The longer the delay, the higher the chances of infection and permanent damage to the hand. Patients who wait longer than 24 hours to seek treatment or have signs of infection or damage to the tendon, joint capsule, or bones are usually referred immediately to a doctor who specializes in hand surgery.

The first step in treatment of clenched fist injury is irrigation, a procedure by which the wound is flushed with a stream of water under high pressure or with an antiseptic solution. Incision and drainage of the wound (I&D) may be required as well as **debridement**, the surgical removal of dead tissue and **foreign objects** from a wound. Careful examination of the depth of the wound is essential to proper treatment. The surgeon may need to enlarge the sides of the wound in order to make an accurate evaluation. The patient will be asked to move the affected joint through its full range of motion so that the surgeon can determine whether the tendon or joint capsule has been damaged. Following these procedures, the surgeon will pack the wound and put the hand in a splint. Bite **wounds** are never sutured (sewn shut) because of the possibility of enclosing bacteria inside the injury. After 24 hours, the packing will be removed and the hand reexamined for signs of infection.

If the wound has become infected, the patient is usually hospitalized and given parenteral (injectable) **antibiotics**. The wound is irrigated and examined to determine the extent of the injury. Cultures are taken for both aerobic (requiring air or oxygen to live) and anaerobic (not requiring air or oxygen) species of bacteria. The cultures should be taken from areas deep in the wound rather than from the surface for greater accuracy. **Tetanus** toxoid should be given if the patient has not been immunized within the last 10 years. The patient should also receive treatment and follow-up for the rare possibility of HIV and hepatitis transmission. Although no well-documented cases of HIV transmission by human bites exist as of 2001, the potential for transmission by this route is still present.

KEY TERMS

Antibiotic—A chemical substance produced by a microorganism which can inhibit the growth of or kill other microorganisms.

Debridement—Surgical removal of damaged tissue and foreign objects from a wound.

I&D—Incision and drainage of a wound.

Irrigation—Cleansing a wound with large amounts of water and/or an antiseptic solution.

PARENTERAL—Administered inside the body but outside the digestive tract.

Tetanus toxoid—Tetanus toxoid is a vaccine used to prevent tetanus (also known as lockjaw).

Infected clenched fist injuries usually contain several disease-causing bacteria, the most common being *Streptococcus pyogenes*, *Staphylococcus aureus*, *Bacteroides sp.*, *Peptostreptococcus sp.*, and *Eikenella corrodens*. Broad-spectrum antibiotics are usually given. Uninfected and relatively superficial CFIs may be treated with oral penicillin plus dicloxacillin or Augmentin. For infected CFIs, parenteral penicillin G is usually given together with nafcillin or cefuroxime. CFIs infected by drug-resistant strains of *S. aureus* may require treatment with vancomycin.

Prognosis

The prognosis depends on the patient's underlying state of health and compliance with treatment; depth of the wound; the involvement of the joint capsule or tendon; and the length of time before the wound is treated. The more superficial the wound and the faster the treatment, the better the prognosis.

Prevention

The best way to prevent clenched fist injuries is to avoid fist fights, intoxication, and association with people who practice these forms of behavior. If involved in a fistfight, people should avoid directing punches at their opponent's mouth. The next best preventive measure is to get medical treatment at once for a clenched-fist injury.

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ORGANIZATIONS

- Massachusetts College of Emergency Physicians (MACEP). P. O. Box 296, Swansea, MA 02777. (508) 643-0117. Fax: (508) 643-0141.

Rebecca J. Frey, PhD

Clomiphene see **Infertility drugs**

Clonazepam see **Benzodiazepines**

Closed fracture reduction see **Fracture repair**

Clostridium difficile colitis see **Antibiotic-associated colitis**

Clotrimazole see **Antifungal drugs, topical**

Clotting disorders see **Coagulation disorders**

Clubfoot can affect one foot or both. Sometimes an infant's feet appear abnormal at birth because of the intrauterine position of the fetus birth. If there is no anatomic abnormality of the bone, this is not true clubfoot, and the problem can usually be corrected by applying special braces or casts to straighten the foot.

The ratio of males to females with clubfoot is 2.5 to 1. The incidence of clubfoot varies only slightly. In the United States, the incidence is approximately 1 in every 1,000 live births. A 1980 Danish study reported an overall incidence of 1.20 in every 1,000 children; by 1994, that number had doubled to 2.41 in every 1,000 live births. No reason was offered for the increase.

Causes and symptoms

Experts do not agree on the precise cause of clubfoot. The exact genetic mechanism of inheritance has been extensively investigated using family studies and other epidemiological methods. As of 1999, no definitive conclusions had been reached, although a Mendelian pattern of inheritance is suspected. This may be due to the interaction of several different inheritance patterns, different patterns of development appearing as the same condition, or a complex interaction between genetic and environmental factors. The MSX1 gene has been associated with clubfoot in animal studies. But, as of 2001, these findings have not been replicated in humans.

A family history of clubfoot has been reported in 24.4% of families in a single study. These findings suggest the potential role of one or more genes being responsible for clubfoot.

Several environmental causes have been proposed for clubfoot. Obstetricians feel that intrauterine crowding causes clubfoot. This theory is supported by a significantly higher incidence of clubfoot among twins compared to singleton births. Intrauterine exposure to the drug misoprostol has been linked with clubfoot. Misoprostol is commonly used when trying, usually unsuccessfully, to induce abortion in Brazil and in other countries in South and Central America. Researchers in Norway have reported that males who are in the printing trades have significantly more offspring with clubfoot than men in other occupations. For unknown reasons, **amniocentesis**, a prenatal test, has also been associated with clubfoot. The infants of mothers who smoke during **pregnancy** have a greater chance of being born with clubfoot than are offspring of women who do not smoke.

True clubfoot is usually obvious at birth. The four most common varieties have been described. A clubfoot has a typical appearance of pointing downward and

Clubfoot

Definition

Clubfoot is a condition in which one or both feet are twisted into an abnormal position at birth. The condition is also known as talipes.

Description

True clubfoot is characterized by abnormal bone formation in the foot. There are four variations of clubfoot, including talipes varus, talipes valgus, talipes equinus, and talipes calcaneus. In talipes varus, the most common form of clubfoot, the foot generally turns inward so that the leg and foot look somewhat like the letter J. In talipes valgus, the foot rotates outward like the letter L. In talipes equinus, the foot points downward, similar to that of a toe dancer. In talipes calcaneus, the foot points upward, with the heel pointing down.

being twisted inwards. Since the condition starts in the first trimester of pregnancy, the abnormality is quite well established at birth, and the foot is often very rigid. Uncorrected clubfoot in an adult causes only part of the foot, usually the outer edge, or the heel or the toes, to touch the ground. For a person with clubfoot, walking becomes difficult or impossible.

Diagnosis

True clubfoot is usually recognizable and obvious on **physical examination**. A routine x ray of the foot that shows the bones to be malformed or misaligned supplies a confirmed diagnosis of clubfoot. Ultrasonography is not always useful in diagnosing the presence of clubfoot prior to the birth of a child.

Treatment

Most orthopedic surgeons agree that the initial treatment of congenital (present at birth) clubfoot should be non-operative. Non-surgical treatment should begin in the first days of life to take advantage of the favorable fibro-elastic properties of the foot's connective tissues, those forming the ligaments, joint capsules, and tendons. In a common treatment, a series of casts is applied over a period of months to reposition the foot into a normal alignment. In mild cases, splinting and wearing braces at night may correct the abnormality.

When clubfoot is severe enough to require surgery, the condition is usually not completely correctable, although significant improvement is possible. In the most severe cases, surgery may be required, especially when the Achilles tendon, which joins the muscles in the calf to the bone of the heel, needs to be lengthened. Because an early operation induces fibrosis, a scarring and stiffness of the tissue, surgery should be delayed until an affected child is at least three months old.

Much of a clubfoot abnormality can be corrected by the use of manipulation and casting during the first three months of life. Proper manipulative techniques must be followed by applications of appropriately molded plaster casts to provide effective and safe correction of most varieties of clubfoot. Long-term care by an orthopedist is required after initial treatment to ensure that the correction of the abnormality is maintained. Exercises, corrective shoes, or nighttime splints may be needed until the child stops growing.

Prognosis

With prompt, expert treatment, clubfoot is usually correctable. Most individuals are able to wear regular



Person suffering from clubfoot. About one of every 400 newborns has some form of this birth defect. (Photo Researchers, Inc. Reproduced by permission.)

shoes and lead active lives. If clubfoot is not appropriately treated, the abnormality becomes fixed. This has an effect on the growth of the leg and foot, and some degree of permanent disability usually results.

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KEY TERMS

Enterovirus—Any of a group of viruses that primarily affect the gastrointestinal tract.

Intrauterine—Situated or occurring in the uterus.

Orthopedist—A doctor specializing in treatment of the skeletal system and its associated muscles and joints.

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- March of Dimes/Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (888) 663-4637. resource-center@modimes.org. <<http://www.modimes.org>>.
- National Easter Seal Society. 230 W. Monroe St., Suite 1800, Chicago, IL 60606-4802. (312) 726-6200 or (800) 221-6827. <<http://www.easter-seals.org>>.
- National Organization for Rare Disorders (NORD). PO Box 8923, New Fairfield, CT 06812-8923. (203) 746-6518 or (800) 999-6673. Fax: (203) 746-6481. <<http://www.rare-diseases.org>>.

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L. Fleming Fallon, Jr., MD, DrPH

Cluster headache

Definition

Cluster headaches are characterized by an intense one-sided **pain** centered by the eye or temple. The pain lasts for one to two hours on average and may recur several times in a day.

Description

Cluster headaches have been known as histamine headaches, red migraines, and Horton's disease, among others. The constant factor is the pain, which transcends by far the distress of the more common tension-type **headache** or even that of a **migraine headache**.

Cluster headaches afflict less than 0.5% of the population and predominantly affect men; approximately 80% of sufferers are male. Onset typically occurs in the late 20s, but there is no absolute age restriction. Approximately 80% of cluster headaches are classified as episodic; the remaining 20% are considered chronic. Both display the same symptoms. However, episodic cluster headaches occur during one- to five-month periods followed by six- to 24-month attack-free, or remission, periods. There is no such reprieve for chronic cluster headache sufferers.

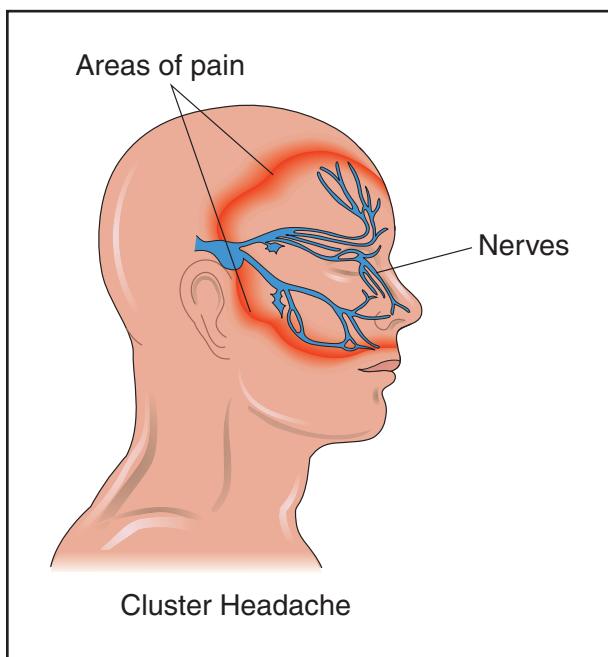
Causes and symptoms

Biochemical, hormonal, and vascular changes induce cluster headaches, but why these changes occur remains unclear. Episodic cluster headaches seem to be linked to changes in day length, possibly signaling a connection to the so-called biological clock. Alcohol, tobacco, histamine, or **stress** can trigger cluster headaches. Decreased blood oxygen levels (hypoxemia) can also act as a trigger, particularly during the night when an individual is sleeping. Interestingly, the triggers do not cause cluster headaches during remission periods.

The primary cluster headache symptom is excruciating one-sided head pain centered behind an eye or near the temple. This pain may radiate outward from the initial focus and encompass the mouth and teeth. For this reason, some cluster headache sufferers may mistakenly attribute their pain to a dental problem. Secondary symptoms, occurring on the same side as the pain, include eye tearing, nasal congestion followed by a runny nose, pupil contraction, and facial drooping or flushing.

Diagnosis

Cluster headache symptoms guide the diagnosis. A medical examination includes recording headache



The primary cluster headache symptom is excruciating one-sided head pain located behind an eye or near the temple. Secondary symptoms include eye tearing, nasal congestion, and a runny nose. (Illustration by Electronic Illustrators Group.)

details, such as frequency and duration, when it occurs, pain intensity and location, possible triggers, and any prior symptoms. This history allows other potential problems to be discounted.

Treatment

Treatment for cluster headaches is composed of induction, maintenance, and symptomatic therapies. The first two therapies are prophylactic treatments, geared toward preventing headaches. Symptomatic therapy is meant to stop or shorten a headache.

Induction and maintenance therapies begin together. Induction therapy is intended to break the headache cycle with drugs such as **corticosteroids** (for example, prednisone) or dihydroergotamine. These drugs are not meant for long-term therapy, but rather as a jump-start for maintenance therapy. Maintenance therapy drugs include verapamil, lithium carbonate, ergotamine, and methysergide. These drugs have long-term effectiveness, but must be taken for at least a week before a response is observed. With long-term treatment, methysergide must be stopped for one month each year to avoid dangerous side effects (formation of fibrous tissue inside the abdominal artery, lungs, and heart valves).

Despite prophylactic treatment, headaches may still occur. Symptomatic therapy includes oxygen inhalation,

KEY TERMS

Biological clock—A synonym for the body's circadian rhythm, the natural biological variations that occur over the course of a day.

Migraine headache—An intense throbbing pain that occurs on one or both sides of the head. The headache is usually accompanied by other symptoms, such as nausea, vomiting, and aversion to light.

Prophylactic—Referring to treatment that prevents symptoms from occurring.

Tension-type headache—A dull pain that seems to exert pressure on the head; the most common form of headache.

sumatriptan injection, and application of local anesthetics inside the nose. Surgery is a last resort for chronic cluster headaches that fail to respond to therapy.

Alternative treatment

Since some cluster headaches are triggered by stress, **stress reduction** techniques, such as **yoga**, **meditation**, and regular **exercise**, may be effective. Some cluster headaches may be an allergic response triggered by food or environmental substances, therefore identifying and removing the allergen(s) may be key to resolution of the problem. Histamine is another suspected trigger of cluster headaches, and this response may be controlled with vitamin C and the bioflavonoids quercetin and bromelain (pineapple enzyme). Supplementation with essential fatty acids (EFA) will help decrease any inflammatory response.

Physical medicine therapies such as adjustments of the spine, craniosacral treatment, and massage at the temporomandibular joint (TMJ) can clear blockages, as can traditional Chinese medical therapies including **acupuncture**. Homeopathic treatment can also be beneficial. Nervous system relaxant herbs, used singly or in combination, can allow the central nervous system to relax as well as assist in peripheral nerve response. A few herbs to consider for relaxation are valerian (*Valeriana officinalis*), chamomile (*Matricaria recutita*), rosemary (*Rosemarinus officinalis*), and skullcap (*Scutellaria baicalensis*).

Prognosis

In general, drug therapy offers effective treatment.

Prevention

Avoiding triggers, adhering to medical treatment, and controlling stress can help ward off some cluster headaches.

Resources

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ORGANIZATIONS

- American Council for Headache Education (ACHE). 19 Man-tua Road, Mt. Royal, NJ 08061. (800) 255-2243. <<http://www.achenet.org>>.
 National Headache Foundation. 428 W. St. James Place, Chicago, IL 60614. (800) 843-2256. <<http://www.headaches.org>>.

Julia Barrett

CMV see **Cytomegalovirus infection**

CNS depressants see **Central nervous system depressants**

CNS stimulants see **Central nervous system stimulants**

order affecting platelet production or one of the many steps in the entire process can disrupt clotting.

Coagulation disorders arise from different causes and produce different complications. Some common coagulation disorders are:

- Hemophilia, or hemophilia A (Factor VIII deficiency), an inherited coagulation disorder, affects about 20,000 Americans. This genetic disorder is carried by females but most often affects males.
- Christmas disease, also known as hemophilia B or Factor IX deficiency, is less common than hemophilia A with similar symptoms.
- Disseminated intravascular coagulation disorder, also known as consumption coagulopathy, occurs as a result of other diseases and conditions. This disease accelerates clotting, which can actually cause hemorrhage.
- **Thrombocytopenia** is the most common cause of coagulation disorder. It is characterized by a lack of circulating platelets in the blood. This disease also includes idiopathic thrombocytopenia.
- Von Willebrand's disease is a hereditary disorder with prolonged **bleeding time** due to a clotting factor deficiency and impaired platelet function. It is the most common hereditary coagulation disorder.
- Hypoprothrombinemia is a congenital deficiency of clotting factors that can lead to hemorrhage.
- Other coagulation disorders include Factor XI deficiency, also known as hemophilia C, and Factor VII deficiency. Hemophilia C afflicts one in 100,000 people and is the second most common bleeding disorder among women. Factor VII is also called serum prothrombin conversion accelerator (SPCA) deficiency. One in 500,000 people may be afflicted with this disorder that is often diagnosed in newborns because of bleeding into the brain as a result of traumatic delivery.

Coagulation disorders

Definition

Coagulation disorders deal with disruption of the body's ability to control blood clotting. The most commonly known coagulation disorder is **hemophilia**, a condition in which patients bleed for long periods of time before clotting. There are other coagulation disorders with a variety of causes.

Description

Coagulation, or clotting, occurs as a complex process involving several components of the blood. Plasma, the fluid component of the blood, carries a number of proteins and coagulation factors that regulate bleeding. Platelets, small colorless fragments in the blood, initiate contraction of damaged blood vessels so that less blood is lost. They also help plug damaged blood vessels and work with plasma to accelerate blood clotting. A dis-

Causes and symptoms

Some coagulation disorders present symptoms such as severe bruising. Others will show no apparent symptoms, but carry the threat of severe internal bleeding.

Hemophilia

Because of its hereditary nature, hemophilia A may be suspected before symptoms occur. Some signs of hemophilia A are numerous large, deep **bruises** and **pain** and swelling of joints caused by internal bleeding. Patients with hemophilia do not bleed faster, just longer. A person with mild hemophilia may first discover the disorder with prolonged bleeding following a surgical procedure. If there is bleeding into the neck, head, or

digestive tract, or bleeding from an injury, emergency measures may be required.

Mild and severe hemophilia A are inherited through a complex genetic system that passes a recessive gene on the female chromosome. Women usually do not show signs of hemophilia but are carriers of the disease. Each male child of the carrier has a 50% chance of having hemophilia, and each female child has a 50% chance of passing the gene on.

Christmas disease

Christmas disease, or hemophilia B, is also hereditary but less common than hemophilia A. The severity of Christmas disease varies from mild to severe, although mild cases are more common. The severity depends on the degree of deficiency of the Factor IX (clotting factor). Hemophilia B symptoms are similar to those of hemophilia A, including numerous, large, and deep bruises and prolonged bleeding. The more dangerous symptoms are those that represent possible internal bleeding, such as swelling of joints, or bleeding into internal organs upon trauma. Hemophilia most often occurs in families with a known history of the disease, but occasionally, new cases will occur in families with no apparent history.

Disseminated intravascular coagulation

The name of this disorder arises from the fact that malfunction of clotting factors cause platelets to clot in small blood vessels throughout the body. This action leads to a lack of clotting factors and platelets at a site of injury that requires clotting. Patients with disseminated intravascular coagulation (DIC) will bleed abnormally even though there is no history of coagulation abnormality. Symptoms may include minute spots of hemorrhage on the skin, and purple patches or hematomas caused by bleeding in the skin. A patient may bleed from surgery or intravenous injection (IV) sites. Related symptoms include vomiting, seizures, **coma**, **shortness of breath**, **shock**, severe pain in the back, muscles, abdomen, or chest.

DIC is not a hereditary disorder or a common one. It is most commonly caused by complications during **pregnancy** or delivery, overwhelming infections, acute leukemia, metastatic **cancer**, extensive **burns** and trauma, and even snakebites. There are a number of other causes of DIC, and it is not commonly understood why or how these various disorders can lead to the coagulation problem. What the underlying causes of DIC have in common is some factor that affects proteins, platelets, or other clotting factors and processes. For example, uterine tissue can enter the mother's circulation during prolonged labor, introducing foreign proteins into the blood, or the venom of some exotic snakes can activate one of the clotting fac-

KEY TERMS

Clotting factor—Also known as coagulation factors. Proteins in the plasma which serve to activate various parts of the blood clotting process by being transformed from inactive to active form.

Enzyme—A substance that causes a chemical reaction, usually a protein. Enzymes are secreted by cells.

Hemorrhage—Abnormal bleeding from the blood vessels.

Heparin—An anticoagulant, or blood clot "dis-solver."

Idiopathic—Refers to a disease of unknown cause, and sometime to a primary disease.

Metastatic—The term used to describe a secondary cancer, or one that has spread from one area of the body to another.

Serum reagents—Serum is fluid, or the fluid portion of the blood retained after removal of the blood cells and fibrin clot. Reagents are substances added to the serum to produce a chemical reaction.

Thrombosis—Formation of a clot in the blood that either blocks, or partially blocks a blood vessel. The thrombus may lead to infarction, or death of tissue, due to a blocked blood supply.

tors. Severe head trauma can expose blood to brain tissue. No matter the cause of DIC, the results are a malfunction of thrombin (an enzyme) and prothrombin (a glycoprotein), which activate the fibrinolytic system, releasing clotting factors in the blood. DIC can alternate from hemorrhage to thrombosis, and both can exist, which further complicates diagnosis and treatment.

Thrombocytopenia

Thrombocytopenia may be acquired or congenital. It represents a defective or decreased production of platelets. Symptoms include sudden onset of small spots of hemorrhage on the skin, or bleeding into mucous membranes (such as nosebleeds). The disorder may also be evident as blood in vomit or stools, bleeding during surgery, or heavy menstrual flow in women. Some patients show none of these symptoms, but complain of **fatigue** and general weakness. There are several causes of thrombocytopenia, which is more commonly acquired as a result of another disorder. Common underlying disorders include leukemia, drug toxicity, or **aplastic ane-**

mia, all of which lead to decreased or defective production of platelets in the bone marrow. Other diseases may destroy platelets outside the marrow. These include severe infection, disseminated intravascular coagulation, and **cirrhosis** of the liver. The idiopathic form most commonly occurs in children, and is most likely the result of production of antibodies that cause destruction of platelets in the spleen and to a lesser extent the liver.

Von Willebrand's disease is caused by a defect in the Von Willebrand clotting factor, often accompanied by a deficiency of Factor VIII as well. It is a hereditary disorder that affects both males and females. In rare cases, it may be acquired. Symptoms include easy bruising, bleeding in small cuts that stops and starts, abnormal bleeding after surgery, and abnormally heavy menstrual bleeding. Nosebleeds and blood in the stool with a black, tarlike appearance are also signs of Von Willebrand's disease.

Hypoprothrombinemia

This disorder is a deficiency in prothrombin, or Factor II, a glycoprotein formed and stored in the liver. Prothrombin, under the right conditions, is converted to thrombin, which activates fibrin and begins the process of coagulation. Some patients may show no symptoms, and others will suffer severe hemorrhaging. Patients may experience easy bruising, profuse nosebleeds, postpartum hemorrhage, excessively prolonged or heavy menstrual bleeding, and postsurgical hemorrhage. Hypoprothrombinemia may also be acquired rather than inherited, and usually results from a **Vitamin K deficiency** caused by liver diseases, newborn hemorrhagic disease, or a number of other factors.

Other coagulation disorders

Factor XI deficiency, or hemophilia C, occurs more frequently among certain ethnic groups, with an incidence of about one in 10,000 among Ashkenazi Jews. Nearly 50% of patients with this disorder experience no symptoms, but others may notice blood in their urine, nosebleeds, or bruising. Although joint bleeding seldom occurs, some factor XI patients will experience bleeding long after an injury occurs. Some women will experience prolonged bleeding after **childbirth**. Patients with factor VII deficiency vary greatly in their bleeding severity. Women may experience heavy menstrual bleeding, bleeding from the gums or nose, bleeding deep within the skin, and episodes of bleeding into the stomach, intestine, and urinary tract. Factor VII patients may also suffer bleeding into joints.

Diagnosis

Several blood tests can be used to detect various coagulation disorders. There are hundreds of different tests a doctor can order to look for indications of specific

diseases. In addition to blood tests, physicians will complete a medical history and **physical examination**. In the case of acquired coagulation disorders, information such as prior or current diseases and medications will be important in determining the cause of the blood disorder.

- Hemophilia A will be diagnosed with laboratory tests detecting presence of clotting factor VIII, factor IX, and others, as well as the presence or absence of clotting factor inhibitors.
- Christmas disease will be checked against normal bleeding and clotting time, as well as for abnormal serum reagents in factor IX deficiency. Other tests of **prothrombin time** and thromboplastic generation may also be ordered.
- There is no one test or group of tests that can always make (or exclude) a diagnosis of DIC. DIC can be diagnosed through a number of laboratory tests which measure concentration of platelets and fibrinogen in the blood with normal counts and prolonged prothrombin time. Other supportive data include diminished levels of factors V, fibrinogen, and VIII, decreased hemoglobin, and others. Since many of the test results also indicate other disorders, the physician may have to put together several results to reach a diagnosis of DIC. Serial tests may also be recommended, because a single examine at one moment in time may not reveal the process that is occurring.
- Tests for thrombocytopenia include coagulation tests revealing a decreased **platelet count**, prolonged bleeding time, and other measurements. If these tests indicate that platelet destruction is causing the disorder, the physician may order bone marrow examination.
- Von Willebrand's disease will be diagnosed with the assistance of laboratory tests which show prolonged bleeding time, absent or reduced levels of factor VIII, normal platelet count, and others.
- Hypothrombinemia is diagnosed with history information and the use of tests that measure vitamin K deficiency, deficiency of prothrombin, and clotting factors V, VII, IX, and X.
- Factor XI deficiency is diagnosed most often after injury-related bleeding. Blood tests can help pinpoint factor VII deficiency.

Treatment

In mild cases, treatment may involve the use of drugs that stimulate the release of deficient clotting factors. In severe cases, bleeding may only stop if the clotting factor that is missing is replaced through infusion of donated human blood in the form of fresh frozen plasma or cryoprecipitate.

- Hemophilia A in mild episodes may require infusion of a drug called desmopressin or DDAVP. Severe bleeding episodes will require transfusions of human blood clotting factors. Hemophiliacs are encouraged to receive physical therapy to help damaged joints and to exercise in non-contact sports such as swimming, bicycle riding, or walking.
- Christmas disease patients are treated similarly to hemophilia A patients. There are commercial products and human blood products available to provide coagulation. Cryoprecipitate was invented in 1965 to replace the need for whole plasma transfusions, which introduced more volume than needed. By the 1970s, people were able to infuse themselves with freeze-dried clotting factor. Superficial **wounds** can be cleaned and bandaged. Parents of hemophiliac children receiving immunizations should inform the **vaccination** provider in advance to decrease the possibility of bleeding problems. These children should probably not receive injections which go into the muscle.
- Treatment for disseminated intravascular coagulation patients is complicated by the large variety of underlying causes of the disorder. If at all possible, the physician will first treat this underlying disorder. If the patient is not already bleeding, this supportive treatment may eliminate the DIC. However, if bleeding is occurring, the patient may need blood, platelets, fresh frozen plasma, or other blood products. Heparin has been controversial in treating DIC, but it is often used as a last resort to stop hemorrhage. Heparin has not proven useful in treating patients with DIC resulting from heat **stroke**, exotic snakebites, trauma, mismatched transfusions, and acute problems resulting from obstetrical complications.
- Secondary acquired thrombocytopenia is best alleviated by treating the underlying cause or disorder. The specific treatment may depend on the underlying cause. Sometimes, corticosteroids or immune globulin may be given to improve platelet production.
- Von Willebrand's disease is treated by several methods to reduce bleeding time and to replace factor VIII, which consequently will replace the Von Willebrand factor. This may include infusion of cryoprecipitate or fresh frozen plasma. Desmopressin may also help raise levels of the Von Willebrand factor.
- Hypoprothrombinemia may be treated with concentrates of prothrombin. Vitamin K may also be produced, and in bleeding episodes, the patient may receive fresh plasma products.
- Factor XI (hemophilia C) is most often treated with plasma, since there are no commercially available concentrates of factor XI in the United States. Factor VII

patients may be treated with prothrombin complex concentrates. As of early 1998, factor VII concentrate was not licensed in the United States and could only be used with special permission.

Alternative treatment

This can be a very severe condition and should be managed by a practitioner of alternative medicine in conjunction with a medical doctor; this condition should not be self managed. For patients known to suffer from hemophilia A or B and other bleeding disorders, avoidance of activities that can cause severe injury should be practiced. Comprehensive care addresses the whole person by helping to deal with the psychosocial aspects of the disease.

Prognosis

The prognosis for patients with mild forms of coagulation disorders is normally good. Many people can lead a normal life and maintain a normal life expectancy. Without treatment of bleeding episodes, severe muscle and joint pain, and eventually, damage, can occur. Any incident that causes blood to collect in the head, neck, or digestive system can be very serious and requires immediate attention. DIC can be severe enough to cause clots to form and a stroke could occur. DIC is also serious enough to cause **gangrene** in the fingers, nose, or genitals. The prognosis depends on early intervention and treatment of the underlying condition. Hemorrhage from a coagulation disorder, particularly into the brain or digestive track, can prove fatal. In the past, patients who received regular transfusions of human blood products were subject to increased risk of **AIDS** and other diseases. However, efforts have been made since the early 1990s to ensure the safety of the blood supply.

Prevention

Prevention of coagulation disorders varies. Acquired disorders may only be prevented by preventing onset of the underlying disorder (such as cirrhosis). Hereditary disorders can be predicted with prenatal testing and **genetic counseling**. Prevention of severe bleeding episodes may be accomplished by refraining from activities that could cause injury, such as contact sports. Open communication with healthcare providers prior to procedures or tests that could cause bleeding may prevent a severe bleeding incident.

Resources

BOOKS

Bellenir, Karen. *Genetic Disorders Sourcebook*. Omnigraphics, Inc., 1995.

PERIODICALS

Community Alert. New York: National Hemophilia Foundation.

ORGANIZATIONS

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

National Hemophilia Foundation. 116 West 32nd St., 11th Floor, New York, NY 10001. 800-424-2634. <<http://www.hemophilia.org/home.htm>>.

Teresa Norris, RN

Coagulopathies see **Coagulation disorders**

Coal miner's disease see **Black lung disease**

Coal worker's pneumoconiosis see **Black lung disease**

Among the consequences of coarctation of the aorta is ventricular hypertrophy, an enlarging of the left ventricle in response to the increased back pressure of the blood and the demand for more blood by the body. Symptoms in infants include **shortness of breath** (dyspnea), difficulty in feeding, and poor weight gain. Older children usually don't have symptoms, but may display **fatigue**, shortness of breath, or a feeling of lameness in their legs.

Diagnosis

Infants usually have an abnormal "gallop" heart rhythm and may also have **heart murmurs**. Sometimes excessive arterial pulses can be seen in the carotid and suprasternal notch arteries, indicating increased pressure in these arteries, while the femoral pulse is weak or can't be detected. The systolic pressure is higher in the arms than in the legs. Enlargement of the heart can be seen in x rays. Similar symptoms are seen in older children and adults. A 10 mm Hg (mercury) pressure difference between the upper and lower extremities is diagnostic for coarctation of the aorta. For some patients, the systolic pressure difference is observed only during **exercise**. Infants frequently have an abnormal electrocardiogram (ECG) that indicates that the right or both ventricles are enlarged, while in older children the ECG may be normal or show that the left ventricle is enlarged. The coarctation may be detected in echocardiographic examination.

Coarctation of the aorta

Definition

A defect that develops in the fetus in which there is a narrowing of the aortic arch, the main blood artery that delivers blood from the left ventricle of the heart to the rest of the body. Coarctation of the aorta is diagnosed in both newborns and adults. Approximately 10% of newborns with **congenital heart disease** have coarctation of the aorta.

Description

Blood leaves the heart by way of the left ventricle and is distributed to the body by arteries. The aortic arch is the first artery to carry blood as it leaves the heart. Other arteries to the head and arms branch off the aortic arch. A narrowing of the aorta at any spot produces resistance to the flow of blood. This causes high blood pressure before the narrowing and low pressure below the narrowing (downstream). Parts of the body supplied by arteries that branch off the aortic arch before the narrowing have high blood pressure, while most of the lower body doesn't receive enough blood supply. To compensate for this, the heart works harder, and the blood pressure rises.

Approximately half of all infants with coarctation of the aorta are diagnosed within the first two months of life. Frequently, there are other congenital cardiac complications present. Infants with **Turner syndrome** have a 45% rate of also having coarctation. There is evidence that some cases of coarctation may be inherited.

Causes and symptoms

In newborns with congenital heart disease, coarctation of the aorta develops while the baby is in the womb.

Treatment

Drugs can be used to treat the **hypertension** and **heart failure**. Surgery is recommended for infants with other, associated cardiac defects and for those infants not responding to drug therapy. Surgery is indicated for infants that don't require immediate surgery, but who develop severe hypertension during the first several months of life. Patients are advised to avoid vigorous exercise prior to surgical correction of the coarctation. Recoarctation can occur in some patients, even if they have had surgery.

Prognosis

Approximately half of all infants diagnosed with coarctation of the aorta have no other cardiac defects and will respond well to medical management. Most of these children will eventually outgrow the condition after several years of life. Although their hypertension may increase for several months early in life, it will eventually decrease as the circulatory system develops. Surgery is required for infants that have severe coarctation of the aorta or have associated cardiac defects. The average life span of children who have coarctation of the aorta is 34

KEY TERMS

Dyspnea—Difficulty in breathing. Usually associated with heart or lung diseases.

Electrocardiogram—A graph of the heart's beating produced by an instrument that detects the electrical signals made by the heart.

years of age. The most common complications for children who have not had surgery are hypertension, aortic rupture, intracranial bleeding, and congestive heart failure. Women who have an uncorrected coarctation of the aorta have a mortality rate of 10% during pregnancy and a 90% rate of complications.

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John T. Lohr, PhD

Cocaine

Definition

Cocaine is a highly addictive central nervous system stimulant extracted from the leaves of the coca plant, *Erythroxylon coca*.

Description

In its most common form, cocaine is a whitish crystalline powder that produces feelings of euphoria when ingested.

Now classified as a Schedule II drug, cocaine has legitimate medical uses as well as a long history of recreational abuse. Administered by a licensed physician, the drug can be used as a local anesthetic for certain eye and ear problems and in some kinds of surgery.

Forms of the drug

In powder form, cocaine is known by such street names as "coke," "blow," "C," "flake," "snow" and

"toot." It is most commonly inhaled or "snorted." It may also be dissolved in water and injected.

Crack is a smokable form of cocaine that produces an immediate and more intense high. It comes in off-white chunks or chips called "rocks." Little crumbs of crack are sometimes called "kibbles & bits."

In addition to their stand-alone use, both cocaine and crack are often mixed with other substances. Cocaine may be mixed with methcathinone (a more recent drug of abuse, known as "cat," that is similar to methamphetamine) to create a "wildcat." A hollowed-out cigar filled with a mixture of crack and marijuana is known as a "woolah." And either cocaine or crack used in conjunction with heroin is called a "speedball." Cocaine used together with alcohol represents the most common fatal two-drug combination.

History

Cocaine is one of the oldest known psychoactive drugs. Coca leaves, the source of cocaine, were used by the Incas and other inhabitants of the Andean region of South America for thousands of years, both as a stimulant and to depress appetite and combat apoxia (**altitude sickness**).

Despite the long history of coca leaf use, it was not until the latter part of the nineteenth century that the active ingredient of the plant, cocaine hydrochloride, was first extracted from those leaves. The new drug soon became a common ingredient in patent medicines and other popular products (including the original formula for Coca-Cola). This widespread use quickly raised concerns about the drug's negative effects. In the early 1900s, several legislative steps were taken to address those concerns; the Harrison Act of 1914 banned the use of cocaine and other substances in non-prescription products. In the wake of those actions, cocaine use declined substantially.

The drug culture of the 1960s sparked renewed interest in cocaine. With the advent of crack in the 1980s, use of the drug had once again become a national problem. Cocaine use declined significantly during the early 1990s, but it remains a significant problem and is on the increase in certain geographic areas and among certain age groups.

Causes and symptoms

As with other forms of **addiction**, cocaine abuse is the result of a complex combination of internal and external factors. Genetic predisposition, family history, and immediate environment can all affect a person's probability of becoming addicted.

As many as three to four million people are estimated to be chronic cocaine users. The 1997 National Household Survey on Drug Abuse reported an estimated 600,000 current crack users, showing no significant change since the late 1980s.

How cocaine affects the brain

Extensive research has been conducted to determine how cocaine works on the brain and why it is so addictive. Cocaine has been found to affect an area of the brain known as the ventral tegmental area (VTA), which connects with the nucleus accumbens, a major pleasure center. Like other commonly abused addictive drugs, cocaine's effects are related to the action of the neurotransmitter dopamine, which carries information between neurons. Cocaine interferes with the normal functioning of neurons by blocking the re-uptake of dopamine, which builds up in the synapses and is believed to cause the pleasurable feelings reported by cocaine users.

Short-term effects of use

The short-term effects of cocaine can include:

- rapid heartbeat
- constricted blood vessels
- dilated pupils
- increased temperature
- increased energy
- reduced appetite
- increased sense of alertness
- euphoria
- **death** due to overdose

Long-term effects of use

The long-term effects of cocaine and crack use include:

- dependence, addiction
- irritability
- mood swings
- restlessness
- weight loss
- auditory hallucinations
- paranoia

Cocaine use and pregnancy

The rise in cocaine use as well as the appearance of crack cocaine in the late 1980s spurred fears about its

effects on the developing fetus and, since then, several research reports have suggested that prenatal cocaine use could be associated to a wide range of fetal, newborn, and child development problems. According to the The Lindesmith Center-Drug Policy Foundation, many of these early reports had methodological flaws, and most researchers nowadays propose more cautious conclusions concerning prenatal cocaine effects. Much evidence would seem to point to the lack of quality prenatal care and the use of alcohol and tobacco as primary factors in poor fetal development among pregnant cocaine users. Research sponsored by the National Institute on Drug Abuse (NIDA) and the Albert Einstein Medical Center in Philadelphia corroborate the Lindensmith Center findings in reporting that the lack of quality prenatal care is associated with undesirable effects often attributed to cocaine exposure such as **prematurity**, low birth weight, and fetal or infant death. The Center for Disease Control and Prevention (CDC) however, reports that mothers who use cocaine early in **pregnancy** are five times as likely to have a baby with a malformation of the urinary tract as mothers who do not use the drug. Thus, cocaine use during pregnancy is assuredly most inadvisable, especially since it is also often associated with the use of alcohol known to cause long-term developmental problems. Supporting the cocaine-exposed expecting mother so as to discourage cocaine use remains an important task for all health caregivers.

Diagnosis

Diagnosing cocaine addiction can be difficult. Many of the signs of short-term cocaine use are not obvious. Since cocaine users often also use other drugs, it may not be easy to distinguish the effects of one drug from another.

Cocaine use has been documented in significant numbers of eighth graders as well as older teens. Over all age groups, more men than women use the drug. The highest rate of cocaine use is found among adults 18 to 25 years old.

Medical complications

Cocaine has been linked to several serious health problems, including:

- arrhythmia
- heart attacks
- chest pain
- respiratory failure
- strokes
- seizures

Other complications may vary depending on how the drug is administered. Prolonged snorting, for example, can irritate the nasal septum, producing nosebleeds, chronic runny nose, and other problems. Intravenous users face an increased risk of infectious diseases such as HIV/AIDS and hepatitis.

Testing

Drug testing can be useful in diagnosing and treating cocaine abuse. Urine testing can detect cocaine; besides providing an objective alternative to reliance on what a patient says, such tests can also be used as a follow-up to treatment to confirm that the patient has remained drug-free.

Treatment

The last two decades have seen a dramatic rise in the number of cocaine addicts seeking treatment. But like all forms of drug abuse, cocaine abuse/addiction is a multi-faceted phenomenon involving environmental, social, and familial as well as physiological factors. This greatly complicates the challenge of effectively treating cocaine addiction.

Pharmacological treatments

To date, no medications have been approved specifically for treating cocaine addiction. But several were under development at this writing. Selegeline, delivered either via a time-release pill or a transdermal patch, shows promise as a possible anti-cocaine medication. Clinical studies have shown the drug disulfiram (also used to treat alcoholics) to be effective in treating cocaine abusers. In addition, antidepressant medications are sometimes used to control the mood swings associated with the early stages of cocaine withdrawal.

Behavioral approaches

A wide range of behavioral interventions have been successfully used to treat cocaine addiction. The approach used must be tailored to the specific needs of each individual patient, however.

Contingency management rewards drug abstinence (confirmed by urine testing) with points or vouchers which patients can exchange for such things as an evening out or membership in a gym. **Cognitive-behavioral therapy** helps users learn to recognize and avoid situations most likely to lead to cocaine use and to develop healthier ways to cope with stressful situations. Residential programs/therapeutic communities may also be helpful, particularly in more severe cases. Patients typically spend six to 12 months in such programs, which may also include vocational training and other features.

KEY TERMS

Apoxia—Apoxia refers to altitude sickness.

Arrhythmia—Irregular heartbeat.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Nasal septum—The membrane that separates the nostrils.

Neurotransmitter—A chemical that carries nerve impulses across a synapse.

Synapse—The gap between two nerve cells.

Alternative treatment

Various alternative or complementary approaches have been used in treating cocaine addiction, often in combination with more conventional therapies. In Japan, the herb acorus has been traditionally used both to assist early-stage cocaine withdrawal and in later recovery stages. Other herbs sometimes used to treat drug addictions of various kinds include kola nut, guarana seed and yohimbe (to boost short-term energy), and valerian root, hops leaf, scullcap leaf, and chamomile (to calm the patient). The amino acids phenylalanine and tyrosine have been used to reduce cocaine addicts' craving for the drug, and vitamin therapy may be used to help strengthen the patient. Gentle massage has been used to help infants born with congenital cocaine addiction. Other techniques, such as **acupuncture**, **EEG biofeedback**, and visualization, may also be useful in treating addiction.

Prognosis

Because addiction involves so many different factors, prospects for individual addicts vary widely. However, research has consistently shown that treatment can significantly reduce both drug abuse and subsequent criminal activity. The comprehensive Services Research Outcomes Study (1998) found a 45% drop in cocaine use five years after treatment, compared to use during the five years before treatment. The study also found that females generally respond better to treatment than males, and older patients tend to reduce their drug use more than younger patients.

Some research also supports the idea that 12-step programs used in conjunction with other approaches can significantly enhance the prospects for a positive outcome. One study of people in outpatient drug-treatment programs found that participation in a 12-step program nearly doubled their chances of remaining drug-free.

Prevention

Despite significant variation over time, cocaine addiction has proven to be a persistent public health problem. Interdiction and source control are expensive and have failed to eliminate the problem, and some law enforcement officials are now recommending more emphasis on demand reduction through education and other measures to address the causes of cocaine addiction.

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ORGANIZATIONS

- Cocaine Anonymous. 6125 Washington Blvd. Suite 202, Culver City, CA 90232. (800) 347-8998.
- Nar-Anon Family Group Headquarters, Inc. P.O. Box 2562, Palos Verdes Peninsula, CA 90274. (310) 547-5800.

Peter Gregutt

Coccidioidomycosis

Definition

Coccidioidomycosis is an infection caused by inhaling the microscopic spores of the fungus *Coccidioides immitis*. Spores are the tiny, thick-walled structures that fungi use to reproduce. Coccidioidomycosis exists in three forms. The acute form produces flu-like symptoms. The chronic form can develop as many as 20 years after initial infection and, in the lungs, can produce inflamed,

injured areas that can fill with pus (abscesses). Disseminated coccidioidomycosis describes the type of coccidioidomycosis that spreads throughout the body affecting many organ systems and is often fatal.

Description

Coccidioidomycosis is an airborne infection. The fungus that causes the disease is found in the dry desert soil of the southwestern United States, Mexico, and Central and South America. Coccidioidomycosis is sometimes called San Joaquin fever, valley fever, or desert fever because of its prevalence in the farming valleys of California. Although commonly acquired, overt coccidioidomycosis is a rare disease. Chronic infections occur in only one out of every 100,000 people.

Although anyone can get coccidioidomycosis, farm laborers, construction workers, and archaeologists who work where it is dusty are at greater risk to become infected. People of any age can get coccidioidomycosis, but the disease most commonly occurs in the 25–55 age group. In its acute form, coccidioidomycosis infects men and women equally.

Chronic and disseminated forms of coccidioidomycosis occur more frequently in men and pregnant women. Although it is not clear why, people of color are 10–20 times more likely to develop the disseminated form of the disease than caucasians. People who have a weakened immune system (immunocompromised), either from diseases such as AIDS or leukemia, or as the result of medications that suppressed the immune system (corticosteroids, chemotherapy), are more likely to develop disseminated coccidioidomycosis.

Causes and symptoms

When the spores of *C. immitis* are inhaled, they can become lodged in the lungs, divide, and cause localized inflammation. This is known as acute or primary coccidioidomycosis. The disease is not spread from one person to another. Approximately 60% of people who are infected exhibit no symptoms (asymptomatic). In the other 40%, symptoms appear 10–30 days after exposure. These symptoms include a fever which can reach 104°F (39.5°C), dry cough, chest pains, joint and muscle aches, headache, and weight loss. About two weeks after the start of the fever, some people develop a painful red rash or lumps on the lower legs. Symptoms usually disappear without treatment in about one month. People who have been infected gain partial immunity to reinfection.

The chronic form of coccidioidomycosis normally occurs after a long latent period of 20 or more years during which the patient experiences no symptoms of the disease. In the chronic phase, coccidioidomycosis causes

lung abscesses that rupture, spilling pus and fluid into the lungs, and causing serious damage to the lungs. The patient experiences difficulty breathing and has a fever, chest **pain**, and other signs of **pneumonia**. Medical treatment is essential for recovery.

In its disseminated form, coccidioidomycosis spreads to other parts of the body including the liver, bones, skin, brain, heart, and lining around the heart (pericardium). Symptoms include fever, joint pain, loss of appetite, weight loss, night sweats, **skin lesions**, and difficulty breathing. Also, in 30–50% of patients with disseminated coccidioidomycosis, the tissue coverings of the brain and spinal cord become inflamed (**meningitis**).

Diagnosis

Many cases of coccidioidomycosis go undiagnosed because the symptoms resemble those of common viral diseases. However, a skin test similar to that for **tuberculosis** will determine whether a person has been infected. The test is simple and accurate, but it does not indicate whether the disease was limited to its acute form or if it has progressed to its chronic form.

Diagnosis of chronic or disseminated coccidioidomycosis is made by culturing a sample of sputum or other body fluids in the laboratory to isolate the fungus. A blood serum test is used to detect the presence of an antibody produced in response to *C. immitis* infection. Chest x rays are often used to assess lung damage, but alone cannot lead to a definitive diagnosis of coccidioidomycosis because other diseases can produce similar results on the x ray.

Treatment

In most cases of acute coccidioidomycosis, the body's own immune system is adequate to bring about recovery without medical intervention. Fever and pain can be treated with non-prescription drugs.

Chronic and disseminated coccidioidomycosis, however, are serious diseases that require treatment with prescription drugs. Patients with intact immune systems who develop chronic coccidioidomycosis are treated with the drug ketoconazole (Nizoral) or amphotericin B (Fungizone). Patients with suppressed immune systems are treated with amphotericin B (Fungizone). Amphotericin B is a powerful fungistatic drug with potentially toxic side effects. As a result, hospitalization is required in order to monitor patients. The patient may also receive other drugs to minimize the side effects of the amphotericin B.

Patients with AIDS must continue to take itraconazole (Sporonox) or fluconazole (Diflucan) orally or receive weekly intravenous doses of amphotericin B for the rest of their lives in order to prevent a relapse. Because of the high cost of fluconazole, Pfizer, the man-

KEY TERMS

Abscess—An area of inflamed and injured body tissue that fills with pus.

Acidophilus—The bacteria *Lactobacillus acidophilus* that usually found in yogurt.

Antibody—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

Antigen—A foreign protein to which the body reacts by making antibodies.

Asymptomatic—Persons who carry a disease but who do not exhibit symptoms of the disease are said to be asymptomatic.

Bifidobacteria—A group of bacteria normally present in the intestine. Commercial supplements containing these bacteria 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.

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

Meningitis—An inflammation of the membranes surrounding the brain or spinal cord.

Pericardium—The tissue sac around the heart.

ufacturer of the drug, has established a financial assistance plan to make the drug available at lower cost to those who meet certain criteria. Patients needing this drug should ask their doctors about this program.

Alternative treatment

Alternative treatment for fungal infections focuses on creating an internal environment where the fungus cannot survive. This is accomplished by eating a diet low in dairy products, sugars, including honey and fruit juice, and foods like beer that contain yeast. This is complemented by a diet consisting, in large part, of uncooked and unprocessed foods. Supplements of **vitamins C, E, A-plus, and B complex** may also be useful. *Lactobacillus acidophilus* and *Bifidobacterium* will replenish the good bacteria in the intestines. Antifungal herbs, like garlic (*Allium sativum*), can be consumed in relatively large doses and for an extended period of time in order to increase effectiveness.

Prognosis

Most people who are infected with coccidioidomycosis only suffer from the mild, acute form of the disease and recover without further complications. Patients who suffer from chronic coccidioidomycosis and who have no underlying lung or immune system diseases also stand a good chance of recovery, although they must be alert to a relapse.

The picture for patients with the disseminated form of the disease, many of whom have AIDS, is less positive. Untreated disseminated coccidioidomycosis is almost always fatal within a short time. With treatment, chance of survival increases, but the **death** rate remains high when meningitis or diffuse lung (pulmonary) disease is present. AIDS patients must constantly guard against relapse.

Prevention

Because the fungus that causes coccidioidomycosis is airborne and microscopic, the only method of prevention is to avoid visiting areas where it is found in the soil. Unfortunately, for many people this is impractical. Maintaining general good health and avoiding HIV infection will limit coccidioidomycosis to the acute and relatively mild form in most people.

Resources

ORGANIZATIONS

American Lung Association. 1740 Broadway, New York, NY 10019. (800) 586-4872. <<http://www.lungusa.org>>.
 Canadian HIV/AIDS Clearinghouse. 1565 Carling Avenue, Suite 400, Ottawa, ON K1Z 8R1. (877) 999-7740. <http://www.clearinghouse.cpha.ca/clearinghouse_e.htm>.
 Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. (800) 311-3435, (404) 639-3311. <<http://www.cdc.gov>>.
 National Aids Hotline. (800) 342-2437.
 Project Inform. 205 13th Street, #2001, San Francisco, CA 94103. (800) 822-7422. <<http://www.projinf.org>>.

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Tish Davidson

Coccyx injuries

Definition

The coccyx—or tailbone—is the last bone of the vertebral column, and usually consists of three to five

fused vertebrae that connect with the sacrum, a part of the pelvis.

Description

The coccyx consists of fused vertebrae, which are not flexible like the other vertebrae of the vertebral column which are all interspaced by intervertebral disks and joined together by elastic ligaments. Since the spinal cord ends just before the coccyx begins, coccygeal vertebrae also lack a central foramen (hole). In the coccyx, the vertebrae generally fuse together in early adulthood and may also fuse with the sacrum, the bone located between the 5th lumbar vertebra and the coccyx, as a person ages. In males, the coccyx curves downward, and in females, it is straighter to allow a baby to pass through the birth canal without impediment.

Pain in or around the coccyx is called coccydynia or coccygodynia. Coccydynia presents a range of symptoms associated to a variety of underlying causes and conditions.

Causes and symptoms

Causes

Coccydynia can be caused by a number of factors. Usually, patients report pain after a fall onto their buttocks, as occurs when going down stairs or while skating. Others have pain during **pregnancy** or after **childbirth**. Some experience repetitive strain from rowing or cycling, and some cite anal intercourse as the cause of pain. In many cases, pain derives from a malformation of the coccyx itself. Sometimes bony spurs appear on the coccyx, but only seem to be painful in thin patients who do not have the padding to protect the region from the spur.

Other causes of coccydynia include **cancer** or damage to the sacrum that generates referred pain, meaning pain that appears in one region but originates from another. Muscle strain or tension, pinched nerves or damaged nerves, or dislocation of the coccyx due to gross **obesity** are other causes.

Symptoms

The most common symptom of coccydynia, irrespective of the cause of the condition, is pain when sitting, or when rising from a sitting position. If the condition lasts long enough, the patient may even experience pain when standing or lying down. Sometimes, numbness occurs in the lower part of the spine. Some patients will experience pain during bowel movements, sexual intercourse, or menstruation.

Secondary symptoms include back pain from sitting in odd positions in order to relieve pain, and painful feet from standing too much, because patients avoid sitting.

KEY TERMS

Coccyx—The last bone of the spinal column, consisting of three to five fused vertebrae that connect with the sacrum, a part of the pelvis.

Coccydynia—Also called coccygodynia. Pain in or around the coccyx.

Foramen—A small opening, perforation, or orifice.

Magnetic resonance imaging (MRI)—An imaging technique that produces pictures of the inside of the body.

Sacrum—The triangle-shaped bone located between the fifth lumbar vertebra and the coccyx that consists of five vertebrae fused together. The sacrum joins on each side with the bones of the pelvis.

Spinal cord—Elongated nerve bundles that lie in the vertebral canal and from which the spinal nerves emerge.

Vertebrae—Bones in the cervical, thoracic, and lumbar regions of the body that make up the vertebral column. Vertebrae have a central foramen (hole), and their superposition makes up the vertebral canal that encloses the spinal cord.

Vertebral column—The vertebral column, also called the spinal column or spine, consists of a series of vertebrae connected by ligaments. It provides a supporting axis for the body and protects the spinal cord. The vertebral column consists of seven cervical vertebrae in the neck, followed by 12 thoracic vertebrae that connect to the ribs, five lumbar vertebrae in the lower back, the sacrum, and the coccyx.

Sometimes the entire buttocks experience pain. Rarely, exhaustion, depression, and lack of sleep may occur.

Diagnosis

Diagnosis of fracture is usually made by inserting a gloved finger in the rectum and pressing on the coccyx. X rays and **magnetic resonance imaging (MRI)** are also often used. Since coccyx pain may be the result of other factors like cancer, these must be ruled out through a variety of tests before treatment can begin.

Treatment

Treatment exists to either control the pain or eliminate the cause. Pain control may be dangerous if an

underlying condition exists of which the pain is a warning sign. Nerve blocks and a variety of drugs are other options to control pain.

Elimination of the root cause of the pain is ideal. This is done through careful diagnosis and the application of manual treatments, corticosteroid injections into the coccyx vertebrae, or surgery. Injections into the fourth and fifth sacral nerves and coccygeal nerves often bring relief, but are considered more as a pain control measure than as curative treatment. Manual treatments have not been found to be effective. Surgery is a radical procedure whose indications are inconsistent and dependent on the subjectivity of the physician.

Prognosis

With current treatment, prognosis is good and patients usually are able to live pain free.

Prevention

There probably is no real prevention, except weight control. Some women may choose to give birth through cesarian section instead of vaginally after an episode of coccyx pain from a previous delivery.

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Janie Franz

Cochlear implants

Definition

A cochlear implant is a surgical treatment for **hearing loss** that works like an artificial human cochlea in the inner ear, helping to send sound from the ear to the brain. It is different from a hearing aid, which simply amplifies sound.

Purpose

A cochlear implant bypasses damaged hair cells and helps establish some degree of hearing by stimulating the hearing (auditory) nerve directly.



A close-up view of a cochlear implant. (Photograph by L. Steinmark, Custom Medical Stock Photo. Reproduced by permission.)

Precautions

Because the implants are controversial, very expensive, and have uncertain results, the U.S. Food and Drug Administration (FDA) has limited the implants to people:

- who get no significant benefit from **hearing aids**
- who are at least 2 years old (the age at which specialists can verify severity of deafness)
- with severe to profound hearing loss.

Description

Hearing loss is caused by a number of different problems that occur either in the hearing nerve or parts of the middle or inner ear. The most common type of deafness is caused by damaged hair cells in the cochlea, the hearing part of the inner ear. Normally, hair cells stimulate the hearing nerve, which transmits sound signals to the brain. When hair cells stop functioning, the hearing nerve remains unstimulated, and the person can't hear. Hair cells can be destroyed by many things, including infection, trauma, loud noise, **aging**, or **birth defects**.

All cochlear implants consist of a microphone worn behind the ear that picks up sound and sends it along a wire to a speech processor, which is worn in a small shoulder pouch, pocket, or belt. The processor boosts the sound, filters out background noise, and turns sound into digital signals before sending it to a transmitter worn behind the ear. A magnet holds the transmitter in place through its attraction to the receiver-stimulator, a part of the device that is surgically attached beneath the skin in the skull. The receiver picks up digital signals forwarded by the transmitter, and converts them into electrical impulses. These electrical impulses flow through electrodes contained in a narrow, flexible tube that has been threaded into the cochlea.

As many as 24 electrodes (depending on the type of implant) carry the impulses that stimulate the hearing nerve. The brain then interprets the signals as specific sounds.

Despite the benefits that the implant appears to offer, some hearing specialists and members of the deaf community still believe that the benefits may not outweigh the risks and limitations of the device. Because the device must be surgically implanted, it carries some surgical risk. Also, manufacturers can't promise how well a person will hear with an implant. Moreover, after getting an implant, some people say they feel alienated from the Deaf community, while at the same time not feeling fully a part of the hearing world.

The sounds heard through an implant are different from the normal hearing sounds, and have been described as artificial or "robotlike." This is because the implant's handful of electrodes cannot hope to match the complexity of a person's 15,000 hair cells.

Surgical procedure

During the procedure, the surgeon makes an incision behind the ear and opens the mastoid bone (the ridge on the skull behind the ear) leading into the middle ear. The surgeon then places the receiver-stimulator in the bone, and gently threads the electrodes into the cochlea. This operation takes between one and one-half to five hours.

Preparation

Before a person gets an implant, specialists at an implant clinic conduct a careful evaluation, including extensive hearing tests to determine how well the candidate can hear.

Unfortunately, it is not possible to predict who will benefit from an implant. In general, the later in life a person becomes deaf, and the shorter the duration of deafness, the better the person is likely to understand speech with an implant. Likewise, someone with a healthy hearing nerve will do better than someone with a damaged nerve.

First, candidates undergo a trial with a powerful hearing aid. If the aid can't improve hearing enough, a physician then performs a physical exam and orders a scan of the inner ear (some patients with a scarred cochlea aren't good candidates). A doctor may also order a psychological exam to better understand the person's expectations. Patients need to be highly motivated, and have a realistic understanding of what an implant can and cannot do.

Aftercare

The patient remains in the hospital for a day or two after the surgery. After a month, the surgical **wounds** will

have healed and the patient returns to the implant clinic to be fitted with the external parts of the device (the speech processor, microphone, and transmitter). A clinician tunes the speech processor and sets levels of stimulation for each electrode, from soft to loud.

The patient is then trained in how to interpret the sounds heard through the device. The length of the training varies from days to years, depending on how well the person can interpret the sounds heard through the device.

Risks

As with all operations, there are a few risks of surgery. These include:

- dizziness
- facial **paralysis** (rarely)
- infection at the incision site

Scientists aren't sure about the long-term effects of electrical stimulation on the nervous system. It is also possible to damage the implant's internal components by a blow to the head, which will render the device unworkable.

Normal results

Most profoundly deaf patients who receive an implant are able to discern medium and loud sounds, including speech, at comfortable listening levels. Many use sound clues from the implant, together with speech reading and other facial cues. Almost all adults improve their communication skills when combining the implant with speech reading (lip reading), and some can understand spoken words without speech reading. More than half of adults who lost hearing after they learned to speak can understand some speech without speech reading. About 30% can understand spoken sounds well enough to use the phone.

Children who were born deaf or who lost their hearing before they could speak have the most difficulty in learning to use the implant. Research suggests, however, that most of these children are able to learn spoken language and understand speech using the implant.

Resources

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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.

Hair cells—Sensory receptors in the inner ear that transform sound vibrations into messages that travel to the brain.

Inner ear—The interior section of the ear, where sound vibrations and information about balance are translated into nerve impulses.

Middle ear—The small cavity between the eardrum and the oval window that houses the three tiny bones of hearing.

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ORGANIZATIONS

- Alexander Graham Bell Association for the Deaf. 3417 Volta Place NW, Washington, DC 20007. (202) 337-5220. <<http://www.agbell.org>>.
- American Speech-Language-Hearing Association. 10801 Rockville Pike, Rockville, MD 20852. (800) 638-8255. <<http://www.asha.org>>.
- Cochlear Implant Club International. 5335 Wisconsin Ave. NW, Suite 440, Washington, D.C. 20015-2052. (202) 895-2781. <<http://www.cici.org>>.
- Hearing Loss Link. 2600 W. Peterson Ave., Ste. 202, Chicago, IL 60659. (312) 743-1032, (312) 743-1007 (TDD).
- National Association for the Deaf. 814 Thayer Ave., Silver Spring, MD 20910. (301) 587-1788, (301) 587-1789 (TDD). <<http://www.nad.org>>.

Carol A. Turkington

Cognitive-behavioral therapy

Definition

Cognitive-behavioral therapy is an action-oriented form of psychosocial therapy that assumes that maladap-

tive, or faulty, thinking patterns cause maladaptive behavior and “negative” emotions. (Maladaptive behavior is behavior that is counter-productive or interferes with everyday living.) The treatment focuses on changing an individual’s thoughts (cognitive patterns) in order to change his or her behavior and emotional state.

Purpose

Theoretically, cognitive-behavioral therapy can be employed in any situation in which there is a pattern of unwanted behavior accompanied by distress and impairment. It is a recommended treatment option for a number of mental disorders, including affective (mood) disorders, **personality disorders**, social phobia, **obsessive-compulsive disorder** (OCD), eating disorders, substance abuse, **anxiety or panic disorder**, **agoraphobia**, **post-traumatic stress disorder** (PTSD), and **attention-deficit/hyperactivity disorder** (ADHD). It is also frequently used as a tool to deal with chronic **pain** for patients with illnesses such as **rheumatoid arthritis**, back problems, and **cancer**. Patients with **sleep disorders** may also find cognitive-behavioral therapy a useful treatment for **insomnia**.

Precautions

Cognitive-behavioral therapy may not be suitable for some patients. Those who don’t have a specific behavioral issue they wish to address and whose goals for therapy are to gain insight into the past may be better served by psychodynamic therapy. Patients must also be willing to take a very active role in the treatment process.

Cognitive-behavioral intervention may be inappropriate for some severely psychotic patients and for cognitively impaired patients (for example, patients with organic brain disease or a traumatic brain injury), depending on their level of functioning.

Description

Cognitive-behavioral therapy combines the individual goals of cognitive therapy and behavioral therapy.

Pioneered by psychologists Aaron Beck and Albert Ellis in the 1960s, cognitive therapy assumes that maladaptive behaviors and disturbed mood or emotions are the result of inappropriate or irrational thinking patterns, called *automatic thoughts*. Instead of reacting to the reality of a situation, an individual reacts to his or her own distorted viewpoint of the situation. For example, a person may conclude that he is “worthless” simply because he failed an exam or didn’t get a date. Cognitive therapists attempt to make their patients aware of these dis-

torted thinking patterns, or cognitive distortions, and change them (a process termed cognitive restructuring).

Behavioral therapy, or behavior modification, trains individuals to replace undesirable behaviors with healthier behavioral patterns. Unlike psychodynamic therapies, it does not focus on uncovering or understanding the unconscious motivations that may be behind the maladaptive behavior. In other words, strictly behavioral therapists don’t try to find out why their patients behave the way they do, they just teach them to change the behavior.

Cognitive-behavioral therapy integrates the cognitive restructuring approach of cognitive therapy with the behavioral modification techniques of behavioral therapy. The therapist works with the patient to identify both the thoughts and the behaviors that are causing distress, and to change those thoughts in order to readjust the behavior. In some cases, the patient may have certain fundamental core beliefs, called schemas, which are flawed and require modification. For example, a patient suffering from depression may be avoiding social contact with others, and suffering considerable emotional distress because of his **isolation**. When questioned why, the patient reveals to his therapist that he is afraid of rejection, of what others may do or say to him. Upon further exploration with his therapist, they discover that his real fear is not rejection, but the belief that he is hopelessly uninteresting and unlovable. His therapist then tests the reality of that assertion by having the patient name friends and family who love him and enjoy his company. By showing the patient that others value him, the therapist both exposes the irrationality of the patient’s belief and provides him with a new model of thought to change his old behavior pattern. In this case, the person learns to think, “I am an interesting and lovable person; therefore I should not have difficulty making new friends in social situations.” If enough “irrational cognitions” are changed, this patient may experience considerable relief from his depression.

A number of different techniques may be employed in cognitive-behavioral therapy to help patients uncover and examine their thoughts and change their behaviors. They include:

- Behavioral homework assignments. Cognitive-behavioral therapists frequently request that their patients complete homework assignments between therapy sessions. These may consist of real-life “behavioral experiments” where patients are encouraged to try out new responses to situations discussed in therapy sessions.
- Cognitive rehearsal. The patient imagines a difficult situation and the therapist guides him through the step-by-step process of facing and successfully dealing with it. The patient then works on practicing, or rehearsing,

these steps mentally. Ideally, when the situation arises in real life, the patient will draw on the rehearsed behavior to address it.

- **Journal.** Patients are asked to keep a detailed diary recounting their thoughts, feelings, and actions when specific situations arise. The journal helps to make the patient aware of his or her maladaptive thoughts and to show their consequences on behavior. In later stages of therapy, it may serve to demonstrate and reinforce positive behaviors.
- **Modeling.** The therapist and patient engage in role-playing exercises in which the therapist acts out appropriate behaviors or responses to situations.
- **Conditioning.** The therapist uses reinforcement to encourage a particular behavior. For example, a child with ADHD gets a gold star every time he stays focused on tasks and accomplishes certain daily chores. The gold star reinforces and increases the desired behavior by identifying it with something positive. Reinforcement can also be used to extinguish unwanted behaviors by imposing negative consequences.
- **Systematic desensitization.** Patients imagine a situation they fear, while the therapist employs techniques to help the patient relax, helping the person cope with their fear reaction and eventually eliminate the anxiety altogether. For example, a patient in treatment for agoraphobia, or fear of open or public places, will relax and then picture herself on the sidewalk outside of her house. In her next session, she may relax herself and then imagine a visit to a crowded shopping mall. The imagery of the anxiety-producing situations gets progressively more intense until, eventually, the therapist and patient approach the anxiety-causing situation in real-life (a “graded exposure”), perhaps by visiting a mall. Exposure may be increased to the point of “flooding,” providing maximum exposure to the real situation. By repeatedly pairing a desired response (relaxation) with a fear-producing situation (open, public spaces), the patient gradually becomes desensitized to the old response of fear and learns to react with feelings of relaxation.
- **Validity testing.** Patients are asked to test the validity of the automatic thoughts and schemas they encounter. The therapist may ask the patient to defend or produce evidence that a schema is true. If the patient is unable to meet the challenge, the faulty nature of the schema is exposed.

Initial treatment sessions are typically spent explaining the basic tenets of cognitive-behavioral therapy to the patient and establishing a positive working relationship between therapist and patient. Cognitive-behavioral therapy is a collaborative, action-oriented therapy effort. As such, it empowers the patient by giving him an active role

KEY TERMS

Automatic thoughts—Thoughts that automatically come to mind when a particular situation occurs. Cognitive-behavioral therapy seeks to challenge automatic thoughts.

Cognitive restructuring—The process of replacing maladaptive thought patterns with constructive thoughts and beliefs.

Maladaptive—Unsuitable or counterproductive; for example, maladaptive behavior is behavior that is inappropriate to a given situation.

Psychodynamic therapy—A therapeutic approach that assumes dysfunctional or unwanted behavior is caused by unconscious, internal conflicts and focuses on gaining insight into these motivations.

Relaxation technique—A technique used to relieve stress. Exercise, biofeedback, hypnosis, and meditation are all effective relaxation tools. Relaxation techniques are used in cognitive-behavioral therapy to teach patients new ways of coping with stressful situations.

Schemas—Fundamental core beliefs or assumptions that are part of the perceptual filter people use to view the world. Cognitive-behavioral therapy seeks to change maladaptive schemas.

in the therapy process and discourages any overdependence on the therapist that may occur in other therapeutic relationships. Therapy is typically administered in an outpatient setting in either an individual or group session. Therapists include psychologists (Ph.D., Psy.D., Ed.D. or M.A. degree), clinical social workers (M.S.W., D.S.W., or L.S.W. degree), counselors (M.A. or M.S. degree), or psychiatrists (M.D. with specialization in psychiatry) and should be trained in cognitive-behavioral techniques, although some brief cognitive-behavioral interventions may be suggested by a primary physician/caregiver. Treatment is relatively short in comparison to some other forms of psychotherapy, usually lasting no longer than 16 weeks. Many insurance plans provide reimbursement for cognitive-behavioral therapy services. Because coverage is dependent on the disorder or illness the therapy is treating, patients should check with their individual plans.

Rational-emotive behavior therapy

Rational-emotive behavior therapy (REBT) is a popular variation of cognitive-behavioral therapy developed

in 1955 by psychologist Albert Ellis. REBT is based on the belief that a person's past experiences shape their belief system and thinking patterns. People form illogical, irrational thinking patterns that become the cause of both their negative emotions and of further irrational ideas. REBT focuses on helping patients discover these irrational beliefs that guide their behavior and replace them with rational beliefs and thoughts in order to relieve their emotional distress.

There are 10 basic irrational assumptions that trigger maladaptive emotions and behaviors:

- It is a necessity for an adult to be loved and approved of by almost everyone for virtually everything.
- A person must be thoroughly competent, adequate, and successful in all respects.
- Certain people are bad, wicked, or villainous and should be punished for their sins.
- It is catastrophic when things are not going the way one would like.
- Human unhappiness is externally caused. People have little or no ability to control their sorrows or to rid themselves of negative feelings.
- It is right to be terribly preoccupied with and upset about something that may be dangerous or fearsome.
- It is easier to avoid facing many of life's difficulties and responsibilities than it is to undertake more rewarding forms of self-discipline.
- The past is all-important. Because something once strongly affected someone's life, it should continue to do so indefinitely.
- People and things should be different from the way they are. It is catastrophic if perfect solutions to the grim realities of life are not immediately found.
- Maximal human happiness can be achieved by inertia and inaction or by passively and without commitment.

Meichenbaum's self-instructional approach

Psychologist Donald Meichenbaum pioneered the self-instructional, or "self-talk," approach to cognitive-behavioral therapy in the 1970s. This approach focuses on changing what people say to themselves, both internally and out loud. It is based on the belief that an individual's actions follow directly from this self-talk. This type of therapy emphasizes teaching patients coping skills that they can use in a variety of situations to help themselves. The technique used to accomplish this is self-instructional inner dialogue, a method of talking through a problem or situation as it occurs.

Preparation

Patients may seek therapy independently, or be referred for treatment by a primary physician, psychologist, or psychiatrist. Because the patient and therapist work closely together to achieve specific therapeutic objectives, it is important that their working relationship is comfortable and their goals are compatible. Prior to beginning treatment, the patient and therapist should meet for a consultation session, or mutual interview. The consultation gives the therapist the opportunity to make an initial assessment of the patient and recommend a course of treatment and goals for therapy. It also gives the patient an opportunity to find out important details about the therapist's approach to treatment, professional credentials, and any other issues of interest.

In some managed-care clinical settings, an intake interview or evaluation is required before a patient begins therapy. The intake interview is used to evaluate the patient and assign him or her to a therapist. It may be conducted by a psychiatric nurse, counselor, or social worker.

Normal results

Many patients who undergo cognitive-behavioral therapy successfully learn how to replace their maladaptive thoughts and behaviors with positive ones that facilitate individual growth and happiness. Cognitive-behavioral therapy may be used in conjunction with pharmaceutical and other treatment interventions, so overall success rates are difficult to gauge. However, success rates of 65% or more have been reported with cognitive-behavioral therapy alone as a treatment for panic attacks and agoraphobia. Relapse has been reported in some patient populations, perhaps due to the brief nature of the therapy, but follow-up sessions can put patients back on track.

Resources

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ORGANIZATIONS

Albert Ellis Institute. 45 East 65th St., New York, NY 10021. (800) 323-4738. <<http://www.rebt.org>>.

Beck Institute. GSB Building, City Line and Belmont Avenues, Suite 700, Bala Cynwyd, PA 19004-1610. (610) 664-3020. <<http://www.beckinstitute.org>>.

The National Association of Cognitive-Behavioral Therapists. P.O. Box 2195, Weirton, WV 26062. (800) 853-1135. <<http://www.nacbt.org>>.

Paula Anne Ford-Martin

Colchicine see **Gout drugs**

COLD see **Chronic obstructive lung disease**

Cold agglutinins test

Definition

The cold agglutinins test is performed to detect the presence of antibodies in blood that are sensitive to temperature changes. Antibodies are proteins produced by the immune system in response to specific disease agents; autoantibodies are antibodies that the body produces against one of its own substances. Cold agglutinins are autoantibodies that cause red blood cells to clump, but only when the blood is cooled below the normal body temperature of 98.6°F (37°C). The clumping is most pronounced at temperatures below 78°F (25.6°C).

Purpose

The cold agglutinins test is used to confirm the diagnosis of certain diseases that stimulate the body to produce cold agglutinins. The disease most commonly diagnosed by this test is mycoplasmal **pneumonia**, but mononucleosis, **mumps**, **measles**, **scarlet fever**, some parasitic infections, **cirrhosis** of the liver, and some types of **hemolytic anemia** can also cause the formation of cold agglutinins. Hemolytic **anemias** are conditions in which the blood is low in oxygen because the red blood cells are breaking down at a faster rate than their normal life expectancy of 120 days. In addition to these illnesses, some people have a benign condition called chronic cold agglutinin disease, in which exposure to cold causes temporary clumping of red blood cells and consequent numbness in ears, fingers, and toes.

Description

Since cold agglutinins cause red blood cells to clump only at temperatures lower than 98.6°F (37°C), the test consists of chilling a sample of the patient's blood. There is a bedside version of the test in which the doctor collects four or five drops of blood in a small tube,

KEY TERMS

Agglutinin—An antibody that causes red blood cells to stick or clump together.

Antibody—A protein molecule produced by the immune system that is specific to a disease agent, such as *Mycoplasma pneumoniae*. The antibody combines with the organism and disables it.

Autoantibody—An antibody produced by the body in reaction to any of its own cells or cell products.

Cold agglutinins—Antibodies that cause clumping of red blood cells when the blood temperature falls below normal body temperature (98.6°F/37°C).

Hemolytic anemia—Oxygen deficiency in the blood, caused by shortened survival of red blood cells.

Mycoplasma—A type of free-living microorganism that has no cell wall. Mycoplasmas cause some varieties of pneumonia and urinary tract infections that stimulate the body to produce cold agglutinins.

Titer—The concentration of a substance in a given sample of blood or other tissue fluid.

cools the tube in ice water for 30–60 seconds, and looks for clumping of red blood cells. If the cells clump after chilling and unclump as they rewarm, a cold agglutinin titer (concentration) greater than 1:64 is present. Bedside test results, however, should be confirmed by a laboratory. The laboratory test measures the clumping of red blood cells in different dilutions of the patient's blood serum at 39.2°F (4°C).

Normal results

The results of the cold agglutinins test require a doctor's interpretation. In general, however, a normal value is lower than 1:32.

Abnormal results

Any value higher than 1:32 suggests a diagnosis of mycoplasmal pneumonia or one of the other viral infections or disease conditions indicated by this test.

Resources

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Rebecca J. Frey

Cold sensitivity antibodies test see
Cryoglobulin test

Cold sore

Definition

A cold sore is a fluid-filled blister which usually appears at the edge of the lips. Cold sores are caused by a herpes simplex virus infection.

Description

A cold sore is a fluid-filled, painful blister that is usually on or around the lips. Other names for a cold sore are **fever blister**, oral herpes, labial herpes, herpes labialis, and herpes febrilis. Cold sores most often occur on the lips which distinguishes them from the common canker sore, which is usually inside the mouth. Cold sores do not usually occur inside the mouth except during the initial episode. **Canker sores** usually form either on the tongue or inside the cheeks.

Cold sores are caused by a herpes virus. There are eight different kinds of human herpes viruses. Only two of these, herpes simplex types 1 and 2, can cause cold sores. It is commonly believed that herpes simplex virus type 1 infects above the waist and herpes simplex virus type 2 infects below the waist. This is not completely true. Both herpes virus type 1 and type 2 can cause herpes lesions on the lips or genitals, but recurrent cold sores are almost always type 1.

Oral herpes is very common. More than 60% of Americans have had a cold sore, and almost 25% of those infected experience recurrent outbreaks. Most of these persons became infected before age 10. Anyone can become infected by herpes virus and, once infected, the virus remains latent for life. Herpes viruses are spread from person to person by direct skin-to-skin contact. The highest risk for spreading the virus is the time period beginning with the appearance of blisters and ending with scab formation. However, infected persons need not have visible blisters to spread the infection to others since the virus may be present in the saliva without obvious oral lesions.

Viruses are different from bacteria. While bacteria are independent and can reproduce on their own, viruses enter human cells and force them to make more virus. The infected human cell is usually killed and releases thousands of new viruses. The cell **death** and resulting tissue damage causes the actual cold sores. In addition, the herpes virus can infect a cell and instead of making the cell produce new viruses, it hides inside the cell and waits. The herpes virus hides in the nervous system. This is called "latency." A latent virus can wait inside the nervous system for days, months, or even years. At some future time, the virus "awakens" and causes the cell to produce thousands of new viruses that cause an active infection.

This process of latency and active infection is best understood by considering the cold sore cycle. An active infection is obvious because cold sores are present. The first infection is called the "primary" infection. This active infection is then controlled by the body's immune system and the sores heal. In between active infections, the virus is latent. At some point in the future, latent viruses become activated and once again cause sores. These are called "recurrent" infections. Although it is unknown what triggers latent virus to activate, several conditions seem to bring on infections. These include **stress**, illness, tiredness, exposure to sunlight, menstruation, fever, and diet.

Causes and symptoms

While anyone can be infected by herpes virus, not everyone will show symptoms. The first symptoms of herpes occur within two-20 days after contact with an infected person. Symptoms of the primary infection are usually more severe than those of recurrent infections. The primary infection can cause symptoms like other viral infections including tiredness, **headache**, fever, and swollen lymph nodes in the neck.

Typically, 50-80% of persons with oral herpes experience a prodrome (symptoms of oncoming disease) of **pain**, burning, **itching**, or tingling at the site where blisters will form. This prodrome stage may last anywhere from a few hours to one to two days. The herpes infection prodrome occurs in both the primary infection and recurrent infections.

In 95% of the patients with cold sores, the blisters occur at the outer edge of the lips which is called the "vermilion border." Less often, blisters form on the nose, chin, or cheek. Following the prodrome, the disease process is rapid. First, small red bumps appear that quickly form fluid-filled blisters. The painful blisters may either burst and form a scab or dry up and form a scab. Within two days of the first red bumps, all the blisters have formed scabs. The skin heals completely and without scarring within six to ten days.

Some children have a very serious primary (first episode) herpes infection called “gingivostomatitis.” This causes fever, swollen lymph glands, and numerous blisters inside the mouth and on the lips and tongue that may form large, open sores. These painful sores may last up to three weeks and can make eating and drinking difficult. Because of this, young children with gingivostomatitis are at risk for **dehydration** (excessive loss of water from the body).

Most people experience fewer than two recurrent outbreaks of cold sores each year. Some people never experience outbreaks, while some have very frequent outbreaks. In most people, the blisters form in the same area each time and are triggered by the same factors (such as stress, sun exposure, etc.).

Diagnosis

Because oral herpes is so common, it is diagnosed primarily by symptoms. It can be diagnosed and treated by the family doctor, dermatologists (doctors who specialize in skin diseases) and infectious disease specialists. Laboratory tests may be performed to look for the virus. Because healing sores do not shed much virus, a sample from an open sore would be taken for viral culture. A sterile cotton swab would be wiped over open sores and the sample used to infect human cells in culture. Cells that are killed by the herpes virus have a certain appearance under microscopic examination. The results of this test are available within two to 10 days.

Oral herpes may resemble a bacterial infection called impetigo. This skin infection is most commonly seen in children and causes herpes-like blisters around the mouth and nose. Also, because oral herpes can occur inside the mouth, the blisters could be mistaken for common canker sores. Therefore, the doctor would need to determine whether the blisters are oral herpes, canker sores, or **impetigo**. The diagnosis and treatment of herpes infections should be covered by most insurance providers.

Treatment

There is no cure for herpes virus infections. There are **antiviral drugs** available that have some effect on lessening the symptoms and decreasing the length of herpes outbreaks. There is evidence that some may also prevent future outbreaks. These antiviral drugs work by interfering with the replication of the viruses, and are most effective when taken as early in the infection process as possible. For the best results, drug treatment should begin during the prodrome stage before blisters are visible. Depending on the length of the outbreak, drug treatment could continue for up to 10 days.



A close-up view of a patient's mouth with gingivostomatitis cold sores. (Custom Medical Stock Photo. Reproduced by permission.)

Acyclovir (Zovirax) is the drug of choice for herpes infection and can be given intravenously or taken by mouth. It can be applied directly to sores as an ointment but is not very useful in this form. A liquid form for children is also available. Acyclovir is effective in treating both the primary infection and recurrent outbreaks. When taken by mouth to prevent an outbreak, acyclovir reduces the frequency of herpes outbreaks.

During an outbreak of cold sores, salty foods, citrus foods (oranges etc.), and other foods that irritate the sores should be avoided. Wash the sores once or twice a day with warm, soapy water and pat gently to dry. Over-the-counter lip products that contain the chemical phenol (such as Blistex Medicated Lip Ointment) and numbing ointments (Anbesol) help to relieve cold sores. A bandage may be placed over the sores to protect them and prevent spreading the virus to other sites on the lips or face. **Acetaminophen** (Tylenol) or ibuprofen (Motrin, Advil) may be taken if necessary to reduce pain and fever.

Alternative treatment

Vitamin and mineral supplements and diet may have an effect on the recurrence and duration of cold sores. In general, cold sore sufferers should eat a healthy diet of unprocessed foods such as vegetables, fruits, and whole grains. Alcohol, **caffeine**, and sugar should be avoided.

An imbalance in the amino acids lysine and arginine is thought to be one contributing factor in herpes virus outbreaks. A diet that is rich in the amino acid lysine may help prevent recurrences of cold sores. Foods which contain high levels of lysine include most vegetables, legumes, fish, turkey, and chicken. In one study, patients taking lysine supplements had milder symptoms during an outbreak, a shorter healing time, and had fewer outbreaks

KEY TERMS

Latent—A nonactive virus which is in a dormant state within a cell. The herpes virus is latent in the nervous system.

Prodrome—The symptoms that warn of the beginning of disease. The herpes prodrome consists of pain, burning, tingling, or itching at a site before blisters are visible.

Recurrence—The return of an active infection following a period of latency.

than patients who did not take lysine. Patients should take 1,000 mg of lysine three times a day during a cold sore outbreak and 500 mg daily on an ongoing basis to prevent recurrences. Intake of the amino acid arginine should be reduced. Foods rich in arginine that should be avoided are chocolate, peanuts, almonds, and other nuts and seeds.

Vitamin C and bioflavonoids (a substance in fruits that helps the body to absorb and use vitamin C) have been shown to reduce the duration of a cold sore outbreak and reduce the number of sores. The vitamin B complex includes important **vitamins** that support the nervous system where viruses can hide out. B complex vitamins can also help manage stress, an important contributing factor to the outbreak of herpes viruses. Applying the oil in vitamin E capsules directly to cold sores may provide relief. Zinc lozenges appear to affect the reproduction of viruses and also enhance the immune system. Ointments containing lemon balm (*Melissa officinalis*) or licorice (*Glycyrrhiza glabra*) and peppermint (*Mentha piperita*) have been shown to help cold sores heal.

Prognosis

Oral herpes can be painful and embarrassing, but it is not a serious infection. There is no cure for oral herpes, but outbreaks usually occur less frequently after age 35. The spread of the herpes virus to the eyes is very serious. The herpes virus can infect the cells in the cornea and cause scarring that may impair vision.

Prevention

The only way to prevent oral herpes is to avoid contact with infected persons. This is not an easy solution because many people aren't aware that they are infected and can easily infect others. Currently there are no herpes vaccines available, although herpes vaccines are being tested.

Several practices can reduce the occurrence of cold sores and the spread of virus to other body locations or people. These practices are:

- Avoidance of sun exposure to the face. Before getting prolonged exposure to the sun, apply sunscreen to the face and especially to the lips. Wearing a hat with a large brim is also helpful.
- Avoid touching cold sores. Squeezing, picking, or pinching blisters can allow the virus to spread to other parts of the lips or face and infect those sites.
- Wash hands frequently. Persons with oral herpes should wash their hands carefully before touching others. An infected person can spread the virus to others even when he or she has no obvious blisters.
- Avoid contact with others during active infection. Infected persons should avoid kissing or sexual contact with others until after the cold sores have healed.
- Wear gloves when applying ointment to a child's sore.
- Be especially careful with infants. Never kiss the eyes or lips of a baby who is under six months old.
- Be watchful of infected children. Do not allow infected children to share toys that may be put into the mouth. Toys that have been mouthing should be disinfected before other children play with them.
- Maintain good general health. A healthy diet, plenty of sleep, and **exercise** help to minimize the chance of getting a cold or the flu, which are known to bring on cold sores. Also, good general health keeps the immune system strong; this helps to keep the virus in check and prevents outbreaks.

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Belinda Rowland, PhD

Cold spot myocardial imaging see **Thallium heart scan**

Colds see **Common cold**

Colic

Definition

Colic is persistent, unexplained crying in a healthy baby between two weeks and five months of age.

Description

Colic, which is not a disease, affects 10–20% of all infants. It is more common in boys than in girls and most common in a family's first child. Symptoms of colic usually appear when a baby is 14–21 days old, reach a crescendo at the age of three months, and disappear within the next eight weeks. Episodes occur frequently but intermittently and usually begin with prolonged periods of crying in the late afternoon or evening. They can last for just a few minutes or continue for several hours. Some babies who have colic are simply fussy. Others cry so hard that their faces turn red, then pale.

Causes and symptoms

No one knows what causes colic. The condition may be the result of swallowing large amounts of air, which becomes trapped in the digestive tract and causes bloating and severe abdominal pain.

Other possible causes of colic include:

- digestive tract immaturity
- food intolerances
- hunger or overfeeding
- lack of sleep
- loneliness
- overheated milk or formula
- overstimulation resulting from noise, light, or activity
- tension

During a colicky episode, babies' bellies often look swollen, feel hard, and make a rumbling sound. Crying intensifies, tapers off, then gets louder. Many babies grow rigid, clench their fists, curl their toes, and draw their legs toward their body. A burp or a bowel movement can end an attack. Most babies who have colic don't seem to be in pain between attacks.

Diagnosis

Pediatricians and family physicians suspect colic in an infant who:

- has cried loudly for at least three hours a day at least three times a week for three weeks or longer

- is not hungry but cries for several hours between dinner time and midnight
- demonstrates the clenched fists, rigidity, and other physical traits associated with colic

The baby's medical history and a parent's description of eating, sleeping, and crying patterns are used to confirm a diagnosis of colic. **Physical examination** and laboratory tests are used to rule out infection, intestinal blockage, and other conditions that can cause abdominal pain and other colic-like symptoms.

Treatment

Medications do not cure colic. Doctors sometimes recommend simethicone (Mylicon Drops) to relieve gas pain, but generally advise parents to take a practical approach to the problem.

Gently massaging the baby's back can release a trapped gas bubble, and holding the baby in a sitting position can help prevent air from being swallowed during feedings. Bottle-fed babies can swallow air if nipple holes are either too large or too small.

Nipple-hole size can be checked by filling a bottle with cold formula, turning it upside down, and counting the number of drops released when it is shaken or squeezed. A nipple hole that is the right size will release about one drop of formula every second.

Babies should not be fed every time they cry, but feeding and burping a baby more often may alleviate symptoms of colic. A bottle-fed baby should be burped after every ounce, and a baby who is breastfeeding should be burped every five minutes.

When cow's milk is the source of the symptoms, bottle-fed babies should be switched to a soy milk hydrolyzed protein formula. A woman whose baby is breastfeeding should eliminate dairy products from her diet for seven days, then gradually reintroduce them unless the baby's symptoms reappear.

Since intolerance to foods other than cow's milk may also lead to symptoms of colic, breastfeeding women may also relieve their babies' colic by eliminating from their diet:

- coffee
- tea
- cocoa
- citrus
- peanuts
- wheat
- broccoli and other vegetables belonging to the cabbage family

Rocking a baby in a quiet, darkened room can prevent overstimulation, and a baby usually calms down when cuddled in a warm, soft blanket.

Colicky babies cry less when they are soothed by the motion of a wind-up swing, a car ride, or being carried in a parent's arms. Pacifiers can soothe babies who are upset, but a pacifier should never be attached to a string.

A doctor should be notified if a baby who has been diagnosed with colic:

- develops a rectal **fever** higher than 101°F (38.3°C)
- cries for more than four hours
- vomits
- has **diarrhea** or stools that are black or bloody
- loses weight
- eats less than normal

Alternative treatment

Applying gentle pressure to the webbed area between the thumb and index finger of either hand can calm a crying child. So can gently massaging the area directly above the child's navel and the corresponding spot on the spine. Applying warm compresses or holding your hand firmly over the child's abdomen can relieve cramping.

Teas made with chamomile (*Matricaria recutita*), lemon balm (*Melissa officinalis*), peppermint (*Mentha piperita*), or dill (*Anethum graveolens*) can lessen bowel inflammation and reduce gas. A homeopathic combination called "colic" may be effective, and constitutional homeopathic treatment can help strengthen the child's entire constitution.

Prognosis

Colic is distressing, but it is not dangerous. Symptoms almost always disappear before a child is six months old.

Prevention

Many doctors believe that colic cannot be prevented. Some alternative practitioners, however, feel that colic can be prevented by an awareness of food intolerances and their impact.

Resources

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Maureen Haggerty

Collapsed lung see **Pneumothorax**

Colloidal bath see **Therapeutic baths**

Colon cancer

Definition

Cancer of the colon is the disease characterized by the development of malignant cells in the lining or epithelium of the first and longest portion of the large intestine. Malignant cells have lost normal control mechanisms governing growth. These cells may invade surrounding local tissue, or they may spread throughout the body and invade other organ systems.

Synonyms for the colon include the large bowel or the large intestine. The rectum is the continuation of the large intestine into the pelvis that terminates in the anus.

Description

The colon is a tubular organ beginning in the right lower aspect of the abdomen. Anatomically, it ascends on the right side of the abdomen, traverses from right to left in the upper abdomen, descends vertically down the left side, takes an S-shaped curve in the lower left abdomen, and then flows into the rectum as it leaves the abdomen for the pelvis. These portions of the colon are named separately though they are part of the same organ:

- cecum, the beginning of the colon
- ascending colon, the right vertical ascent of the colon
- transverse colon, the portion traversing from right to left
- descending colon, the left vertical descent of the colon
- sigmoid colon, the s-shaped segment of colon above the pelvis

These portions of the colon are recognized anatomically based on the arterial blood supply and venous and lymphatic drainage of these segments of the colon. Lymph, a protein-rich fluid that bathes the cells of the body, is transported in small channels known as lymphatics that run along side the veins of the colon. Lymph nodes are small filters through which the lymph travels on its way back to the blood stream. Cancer can spread elsewhere in the body by invading the lymph and vascular systems. Therefore, these anatomic considerations become very important in the treatment of colon cancer.

The small intestine is the continuation of the upper gastrointestinal tract that is responsible for the transport of ingested nutrients into the body. The waste left after the small intestine has completed absorption of nutrients amounts to a few liters (about the same as quart) of material per day and is directly delivered to the colon (at the cecum) for processing. Physiologically, the colon is responsible for the preservation of fluid and electrolytes as it propels the increasingly solid waste towards the rectum and anus for excretion.

When cells lining the colon become malignant, they first grow locally and may invade partially or totally through the wall of the bowel and even into adjacent structures and organs. In the process, the tumor can penetrate and invade the lymphatics or the capillaries locally and it gains access to the circulation. 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 colon primary tumor, are then known as metastases. If metastases are found in the regional lymph nodes from the primary, they are known as regional metastases or regional nodal metastases. If they are distant from the primary tumor, they are known as distant metastases. The patient with distant metastases has systemic disease. Thus the cancer originating in the colon begins locally and, given time, can become systemic in its extent.

By the time the primary is originally detected, it is usually larger than 0.4 in (1 cm) in size and has over a million cells. This amount of growth itself 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 and screening becomes a very important endeavor to aid in earlier detection of this disease.

There are about 94,000 cases of colon 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. (Lung cancer is the first affecting both men and women, breast is the leader in women and prostate the leader in men.) Nearly 48,000 people died from colon cancer in the United States in 2000. 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 colon is thought to arise sporadically in about 80% of those who develop the disease. Twenty percent of cases are thought to have genetic predisposition that ranges from familial syndromes affecting 50% of the offspring of a mutation carrier, to a risk of 6% when there is just a family history of colon cancer occurring in a first degree relative. Development of colon cancer at an early age, or at multiple sites, or recurrent colon cancer suggests a genetically transmitted form of the disease as opposed to the sporadic form.

Causes and symptoms

Causes of colon cancer are probably environmental in the sporadic cases (80%) and genetic in the heredity predisposed cases (20%). Since malignant cells have a changed genetic makeup, this means that in 80% of cases, the environment spontaneously induces change, whereas in those born with a genetic predisposition, they 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 (cancer-causing agents). Specific carcinogens have been difficult to identify; however, dietary factors seem to be involved.

Colon cancer is more common in industrialized nations and **diets** high in fat, red meat, total calories, and alcohol seem to predispose. Diets high in fiber are associated with a decreased risk. The mechanism for protection by high-fiber diets may be related to less exposure of the colon lining to carcinogens from the environment, as the transit time through the bowel is faster with a high-fiber diet than it is with a low-fiber diet.

Age plays a definite role in the predisposition to colon cancer. Colon cancer is uncommon before age 40. This incidence increases substantially after age 50 and doubles with each succeeding decade.

There is also a slight increase risk for colon cancer in the individual who smokes.

Patients who suffer from inflammatory diseases of the colon known as **ulcerative colitis** and Crohn's colitis are also at increased risk.

As for genetic predisposition, on chromosome 5, there is a gene called the APC gene associated with the familial adenomatous polyposis syndrome. There are multiple different 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 genetic aberration is transmitted to 50% of offspring and each of those affected will develop colon cancer, usually at an early age. There is another syndrome, hereditary non-polyposis colon cancer (also known as Lynch syndrome), related to mutations in any of four genes responsible for DNA mismatch repair. In patients with colon 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. This allows for the damaged cell to perpetuate itself, and continued replication of the damaged DNA may lead to tumor development. Though these syndromes have a very high incidence of colon cancer, family history without the syndrome is also a substantial risk factor. When considering first-degree relatives, history of one with colon cancer raises the baseline risk of 2% to 6%. (Most physicians think that this baseline is about 4%.) The presence of a second raises the risk to 17%.

The development of polyps of the colon almost always precedes the development of colon cancer by five or more years. Polyps are benign growths of the colon lining. They can be unrelated to cancer, precancerous, or malignant. Polyps, when identified, are removed for diagnosis. If the polyps are benign, the patient should undergo careful surveillance for the development of more polyps or the development of colon cancer.

Colon cancer causes symptoms related to its local presence in the large bowel or by its effect on other organs if it has spread. These symptoms may occur alone or in combination:

- a change in bowel habit
- blood in the stool
- bloating, persistent abdominal distention
- constipation
- a feeling of fullness even after having a bowel movement
- narrowing of the stool—so-called ribbon stools
- persistent, chronic **fatigue**
- abdominal discomfort
- unexplained weight loss
- very rarely, nausea and vomiting

Most of these symptoms are caused by the physical presence of the tumor mass in the colon. Similar symptoms can be caused by other processes; these are not absolutely specific to colon cancer. The key is recognizing that the persistence of these types of symptoms without ready explanation should prompt the individual to seek medical evaluation.

Many of the symptoms are understood by remembering that the colon is a tubular conduit. If a tumor develops, as it reaches a certain size it will begin to cause symptoms related to the obstruction of that conduit. In addition, the tumor commonly oozes blood that is lost in the stool. (Often, this blood is not visible.) This phenomenon results in anemia and chronic fatigue. Weight loss is a late symptom, often implying substantial obstruction or the presence of systemic disease.

Diagnosis

Screening

Of all of the major cancers, only colorectal cancer can be prevented by screening. In all other cancers (breast and prostate, for example), screening tests look for small, malignant lesions. Screening for colorectal cancers, however, is the search for pre-malignant, benign polyps. This screening can be close to 100% effective in preventing cancer development, not just in detecting small cancers.

Screening involves physical exam, simple laboratory tests, and the visualization of the lining of the colon. The ways to visualize the colon epithelium are with x rays (indirect visualization), and endoscopy (direct visualization).

The **physical examination** involves the performance of a digital rectal exam (DRE). The DRE includes manual examination of the rectum, anus, and the prostate. During this examination, the physician examines the anus and the surrounding skin for **hemorrhoids**, abscesses, and other irregularities. After lubricating the gloved finger and anus, the examiner gently slides the finger into the anus and follows the contours of the rectum. The examiner notes the tone of the anus and feels the walls and the edges for texture, tenderness and masses as far as the examining finger can reach. 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 3 such specimens are on the card, the card is then easily chemically tested for occult blood also. (The stool analysis mentioned here is known as a **fecal occult blood test**, or FOBT, and, while it can be helpful, it is not 100% accurate—only about 50% of cancers are FOBT-positive.) 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 patient's blood. When this occurs, the protein produced is known as a tumor marker. There is a tumor marker for some cancers of the colon; it is known as carcinoembryonic antigen, or CEA. Unfortunately, this protein may be made by other adenocarcinomas as well, or it may not be produced by a particular colon cancer. Therefore, screening by chemical analysis for CEA has not been helpful. CEA has been helpful when used in a follow-up role for patients treated for colon cancer if their tumor makes the protein.

Indirect visualization of the colon may be accomplished by placing barium through the rectum and filling the colon with this compound. Barium produces a white contrast image of the lining of the colon on x ray and thus, the contour of the lining of the colon may be seen. Detail can be increased if the barium utilized is thinned and air also introduced. These studies are known as the **barium enema** (BE) and the double contrast barium enema (DCBE).

Direct visualization of the lining of the colon is accomplished using a scope or endoscope. The physician introduces the instrument through the rectum and passes it proximally, visualizing the colon epithelium in the process. Older, shorter scopes were rigid. Today, utilizing fiberoptic technology, the scopes are flexible and can reach much farther. If the left colon only is visualized, it is called **flexible sigmoidoscopy**. When the entire colon is visualized, the procedure is known as **colonoscopy**.

Unlike the indirect visualizations of the colon (the BE and the DCBE), the endoscopic screenings allow the physician to remove polyps and biopsy suspicious tissue. (A biopsy is a removal of tissue for examination by a pathologist.) For this reason, many physicians prefer endoscopic screening. All of the visualizations, the BE, DCBE, and each type of endoscopy require pre-procedure preparation (evacuation) of the colon.

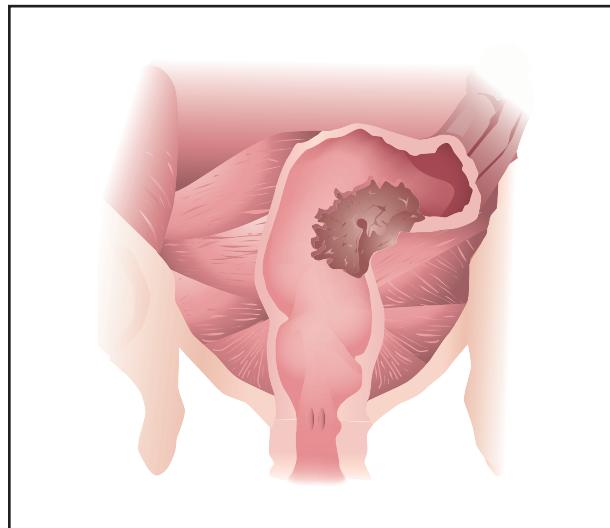
The American Cancer Society has recommended the following screening protocol for those of normal risk over 50 years of age:

- yearly DRE with occult blood in stool testing
- flexible sigmoidoscopy at age 50
- flexible sigmoidoscopy repeated every five years

Many physicians, however, recommend full colonoscopy every five to seven years. Screening evaluations should start sooner for patients who have predisposing factors, such as family history, history of polyps, or a familial syndrome.

Evaluation of patients with symptoms

For those whose symptoms prompt them to visit their physician, and if their symptoms could possibly be



A colon with a cancerous growth. (Illustration by Argosy Inc.)

related to colon cancer, the entire colon will be inspected. The combination of a flexible sigmoidoscopy and DCBE may be performed but the preferred evaluation of the entire colon and rectum is that of complete colonoscopy. Colonoscopy allows direct visualization, photography, and the opportunity to obtain a biopsy of any abnormality visualized. If, for technical reasons, the entire colon is not visualized endoscopically, a DCBE should complement the colonoscopy.

The diagnosis of colon cancer is actually made by the performance of a biopsy of any abnormal lesion in the colon. When a tumor growth is identified, it could be either a benign polyp (or lesion) or a cancer; the biopsy resolves the issue. The endoscopist may take many samples so as to exclude any sampling errors.

If the patient presents with advanced disease, or has advanced disease at the time of diagnosis, areas where the tumor has spread (such as the liver) may be amenable to biopsy. Such biopsies are usually obtained using a special needle under local anesthesia.

Once a diagnosis of colon cancer has been established by biopsy, in addition to the physical exam, studies will be performed to assess the extent of the disease. Blood studies include a complete **blood count**, **liver function tests**, and a CEA. Imaging studies will include a **chest x ray** and a CAT scan (computed tomography scan) of the abdomen. The chest x ray will determine if there is spread to the lung, and the CAT scan will evaluate potential spread to the liver as well as any local invasive characteristics of the primary tumor. If the patient has any neurologic symptoms, a CAT scan of the brain will be performed, and if the

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 reconnection 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 manifested in part by fatigue.

Carcinogens—Substances in the environment that cause cancer, presumably by inducing mutations, with prolonged exposure.

Electrolytes—Salts, such as sodium and chloride.

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 seen with the naked eye.

Polyps—Localized growths of the epithelium that can be benign, precancerous, or harbor malignancy.

Radical resection—Surgical resection that takes the blood supply and lymph system supplying the organ along with the organ.

Resect—To remove surgically.

Sacrum—Posterior bony wall of the pelvis.

Systemic—Referring to throughout the body.

patient is experiencing bone **pain**, a bone scan will also be performed.

Treatment

Once the diagnosis has been confirmed by biopsy, the clinical stage of the cancer is assigned. Using the characteristics of the primary tumor, its depth of penetration through the bowel, and the presence or absence of regional or distant metastases, the stage of the cancer is derived. Often, the depth of penetration through the bowel or the presence of regional lymph nodes can't be assigned before surgery.

Colon 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 bowel wall.
- Stage II: the tumor has penetrated through to the outer wall of the colon or has gone through it, possibly invading other local tissue.
- Stage III: any depth or size of tumor associated with regional lymph node involvement.

- Stage IV: any of previous criteria associated with distant metastasis.

With many cancers other than colon cancer, staging plays an important pre-treatment role to best determine treatment options. In colon cancer, almost all colon cancers are treated with surgery first, regardless of stage. Colon cancers through stage III, and even some stage IV colon cancers, are treated with surgery first before any other treatments are considered.

Surgery

Surgical removal of the involved anatomic segment of colon (colectomy) along with its blood supply and regional lymph nodes is the primary therapy for colon cancer. Usually, on the basis of the blood supply, the partial colectomies are separated into right, left, transverse, or sigmoid. The removal of the blood supply at its origin along with the regional lymph nodes that accompany it assures an adequate margin of normal colon on either side of the primary tumor. When the cancer lies in a position such that the blood supply and lymph drainage lies between two of the major vessels, both vessels are taken to assure complete radical resection or removal (extend-

ed radical right or left colectomy). If the primary tumor penetrates through the bowel wall, any tissue adjacent to the tumor extension is also taken if feasible.

Surgery is used as primary therapy for stages I through III colon cancer unless there are signs that local invasion will not permit complete removal of the tumor, as may occur in advanced stage III tumors. However, this circumstance is very rare, and occurs in less than 2% of all colon cancer cases.

After the resection is completed, the ends of the remaining colon are reconstructed; the hook-up is called an anastomosis. Once healing has occurred, there may be a slight increase in the frequency of bowel movements. This effect usually lasts only for several weeks. Most patients go on to develop completely normal bowel function.

Occasionally, the anastomosis would be risky and cannot be performed. (Most commonly, this occurs when the bowel could not be adequately evacuated in an emergency circumstance due to bowel obstruction.) When the anastomosis cannot be performed, a **colostomy** is performed instead. A colostomy is performed by bringing the end of the colon through the abdominal wall and sewing it to the skin. The patient will have to wear an appliance (a bag) to manage the stool. The colostomy may be temporary and the patient may undergo a hook-up at a later, safer date, or the colostomy may be permanent. In most cases, emergent colostomies are not reversed and are permanent.

Radiation

Radiation therapy is used as an adjunct to surgery if there is concern about potential for local recurrence post-operatively and the area of concern will tolerate the radiation. For instance, if the tumor invaded muscle of the abdominal wall but was not completely removed, this area would be considered for radiation. Radiation has significant dose limits when residual bowel is exposed to it because the small and large intestine do not tolerate radiation well.

Radiation is also used in the treatment of patients who present with or progress to having metastatic disease. It is particularly useful in shrinking metastatic colon cancer to the brain.

Chemotherapy

Chemotherapy is useful for patients who have had all identifiable tumor removed and are at risk for recurrence (adjuvant chemotherapy). Chemotherapy may also be used when the cancer is stage IV and is beyond the scope of regional therapy, but this use is rare.

Adjuvant therapy is considered in stage II disease with deep penetration or in stage III patients. Standard

therapy is treatment with 5-fluorouracil, (5FU) combined with leucovorin for a period of six to 12 months. 5FU is an antimetabolite and leucovorin improves the response rate. (A response is a temporary regression of the cancer in response to the chemotherapy.) 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 do have some toxicity, but usually are tolerated fairly well.

Similar chemotherapy may be administered for stage IV disease or if a patient progresses and develops metastases. Results show response rates of about 20%. Unfortunately, these patients eventually succumb to the disease, and this chemotherapy may not prolong survival or improve quality of life in Stage IV patients. 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 it improved response rates to 39%, added two to three months to disease-free survival, and prolonged overall survival by a little over two months.

Alternative treatment

Alternative therapies have not been studied in a large-scale, scientific way. Large doses of **vitamins**, fiber, and green tea are among therapies tried. Avoiding cigarettes and alcohol may be helpful. Before initiating any alternative therapies, the patient is wise to consult his/her physician to be sure that these therapies do not complicate or interfere with the established therapy.

Prognosis

Prognosis is the long-term outlook or survival after therapy. Overall, about 50% of patients treated for colon 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 a very worthwhile endeavor.

About 15% of patients present with stage I disease and 85–90% survive. Stage II represents 20–30% of cases and 65–75% survive. Thirt to forty percent comprise the stage III presentation of which 55% survive. The remaining 20–25% present with stage IV disease and are very rarely cured.

Prevention

There is not an absolute way of preventing colon cancer. Still, there are steps an individual can take to dramatically lessen the risk or to identify the precursors of colon cancer so that it does not manifest itself. 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. 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

Colon therapy see **Colonic irrigation**

Colonic irrigation

Definition

Colonic irrigation is also known as **hydrotherapy** of the colon, high colonic, entero-lavage, or simply colonic. It is the process of cleansing the colon by passing several gal-

lons of water through it with the use of special equipment. It is similar to an enema but treats the whole colon, not just the lower bowel. This has the effect of flushing out impacted fecal matter, toxins, mucous, and even parasites that often build up over the passage of time. It is a procedure that should only be undertaken by a qualified practitioner.

Purpose

Anyone suffering from gas, bloating, cramping pains, **acne** and other skin complaints, arthritis, and a list of bowel complaints such as diverticulitis and irritable bowel etc., may benefit from colonic irrigation. In particular, **cancer** patients are often advised to undertake a course of colonic irrigation sessions as an essential part of their treatment. When a biological cancer therapy begins to enable the body to breakdown a cancerous mass, it is essential that speedy and effective elimination of the resulting toxins is achieved.

Colon and bowel cancer is one of the leading causes of **death** in the United States, and alternative practitioners insist that it can be prevented by efficient hygiene procedures. Providing that care is taken to replace the natural organisms that flourish in the bowel, many health benefits can be expected from colonic irrigation. In general, alternative practitioners maintain that an ill-functioning bowel is the source of all disease, and therefore keeping it clean will be an effective protection against this.

Removing large amounts of toxic matter relieves the patient and can lead to the alleviation of symptoms such as arthritis, **chronic fatigue syndrome**, **candidiasis**, and a host of other illnesses. Properly executed, colonic irrigation can help restore normal peristaltic action to a sluggish bowel, thus reducing the need for more hydrotherapy treatments over time. In addition, removing the layer of fecal matter which coats the intestines in many individuals allows improved assimilation of the nutrients from foods and can alleviate symptoms of vitamin and other nutrient deficiencies. Many alternative health practitioners consider some form of hydrotherapy for the bowel to be essential in the treatment of degenerative diseases.

Description

Origins

Cleansing the colon with the use of hydrotherapy is not a new concept. Forms of colonic irrigation have been used successfully for decades to relieve chronic toxicity and even acute cases of toxemia.

Over time, many people develop a thick layer of fecal matter that coats their colon. It hardens and becomes impacted, reducing the efficiency of the bowel,

and in some cases, completely obstructing normal elimination of waste matter from the body. It is quite common for people to only have one bowel movement per day, and some as few as one or two per week.

Alternative practitioners advise that we probably should have one bowel movement for every meal that we eat. If not, then we are not eliminating wastes completely, and if input exceeds output, then we will surely suffer the consequences at some point.

Incomplete elimination of body wastes may result in the following, depending on where the deposits end up:

- sluggish system
- joint **pain** and arthritis
- irritable bowel syndrome
- diverticulitis
- **Crohn's disease**
- leaky gut syndrome
- heart problem
- migraine
- **allergies**
- bad breath
- acne and other skin problems such as psoriasis
- asthma
- early senility and **Alzheimer's disease**
- chronic **fatigue** syndrome
- cancer, particularly of the bowel
- multiple sclerosis

During colonic irrigation, a small speculum is passed into the patient's bowel through the rectum. This is attached to a tube, which leads to a machine that pumps temperature-controlled water into the colon at a controlled rate (to be controlled by either the practitioner or the patient). The temperature of the water should ideally be kept as close to body temperature as possible.

The patient will temporarily be filled with water up to the level of the entire colon. Patients say they can feel the water up under their ribs but that the process, although sometimes uncomfortable, is not painful. The amount of water will vary but will generally be in the region of between two and six liters (or quarts) at any one time. This triggers peristaltic action and the patient will begin to expel the water along with fecal matter back through the tube and into the machine.

The fecal matter is flushed out through a viewing tube, so that what is eliminated may be monitored. Quite often, unsuspected parasites are expelled, along with very old fecal material, very dark in color, which may

KEY TERMS

Dysbiosis—The condition that results when the natural flora of the gut are thrown out of balance, such as when antibiotics are taken.

Peristalsis—The natural wave-like action of a healthy bowel that transports matter from one end of the bowel to the other.

Probiotics—Supplements of beneficial microorganisms that normally colonize the gut.

Toxemia—Poisoning of the blood.

have been in the colon for years. Some therapists comment that it looks like aging rubber.

During the treatment, the therapist will gently massage the patient's abdomen to help dislodge impacted fecal matter. In addition to massage, sometimes **acupressure**, **reflexology**, or lymphatic drainage techniques may be used to loosen deposits and stimulate the bowel. It is important that the right amount of water is used, as too much will cause discomfort and too little will be ineffective. If correctly done, colonic irrigation is not painful at all and some patients claim to sleep through their treatment.

Sanitation is vital to this process. The tubes and speculums used are generally disposable, but other parts of the machine, such as the viewing tube, must be sterilized after each patient.

Normally, a series of treatments will be required to achieve desired results regarding the elimination of impacted, decaying matter, and restoration of bowel regularity. Initially, only gas and recent fecal matter may be expelled. The residue attached to the colon wall is usually the result of years of neglect, and therapists say that one cannot expect complete relief in only one session.

Impacted fecal matter can cause an imbalance of the natural organisms that normally populate the bowel, causing what is known as dysbiosis. Under ideal conditions, the bowel is populated by a variety of naturally occurring organisms. It seems that the enzymes occurring in fresh fruit and vegetables encourage these beneficial organisms. One of the results of eating processed denatured foods is that this natural balance is upset, and food may begin to rot in the bowel instead of being processed.

Decomposing matter can cause a toxic condition and may lead to many health problems, as **constipation** causes backed up pollution of the body cells. The process of repair and elimination of wastes enters a downward spiral which at best will cause fatigue, lack of energy and

premature aging, and, at worst, can cause degenerative diseases, among them allergies, and even cancer and Alzheimer's disease.

The cost of colonic irrigation treatments varies, but is generally between \$35–70 per session, which may last from 45 minutes to one hour. The cost of the machine itself ranges from \$4,000–12,000, but again, it should be noted that only qualified therapists should conduct sessions.

Preparations

Most practitioners prefer that distilled or purified water is used for colonic irrigation, but others use sterilized tap water.

Precautions

It may be advisable to use a probiotic pessary after colonic irrigation, to ensure replacement of desirable natural flora. There are certain conditions that either partly or completely preclude the use of colonic irrigation, such as an active attack of Crohn's disease, bleeding ulcers, and hyperacidosis. If in doubt, a qualified practitioner should be consulted. Anyone suffering from these conditions should always notify the practitioner when receiving colonic irrigation treatments.

Side effects

Some allopathic practitioners claim that colonic irrigation flushes out essential electrolytes and friendly bacteria from the bowel and that it can be dangerous. Practitioners counter that this can easily be remedied with the use of probiotics, and that in any case, these possible disadvantages are easily offset by the benefits of having large amounts of putrefying matter, harmful organisms, and parasites removed from the system.

Research and general acceptance

Although many alternative health care practitioners swear by colonic irrigation, there is a large allopathic lobby that claims that there are no benefits to be had, and that there are dangers involved. However, there are many decades of records and research from the alternative health care community that indicate that this therapy may have a valuable place in the treatment of degenerative diseases and toxic conditions.

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ORGANIZATIONS

California Colon Hygienist Society. 333 Miller Ave., Suite 1, Mill Valley, CA 94941. (415) 383-7224.

Intestinal Health Institute. 4427 East Fifth St., Tucson, AZ 85711. (520) 325-9686. info@sheilas.com. <<http://www.sheilas.com>>.

Patricia Skinner

Colonoscopy

Definition

Colonoscopy is a medical procedure where a long, flexible, tubular instrument called the colonoscope is used to view the entire inner lining of the colon (large intestine) and the rectum.

Purpose

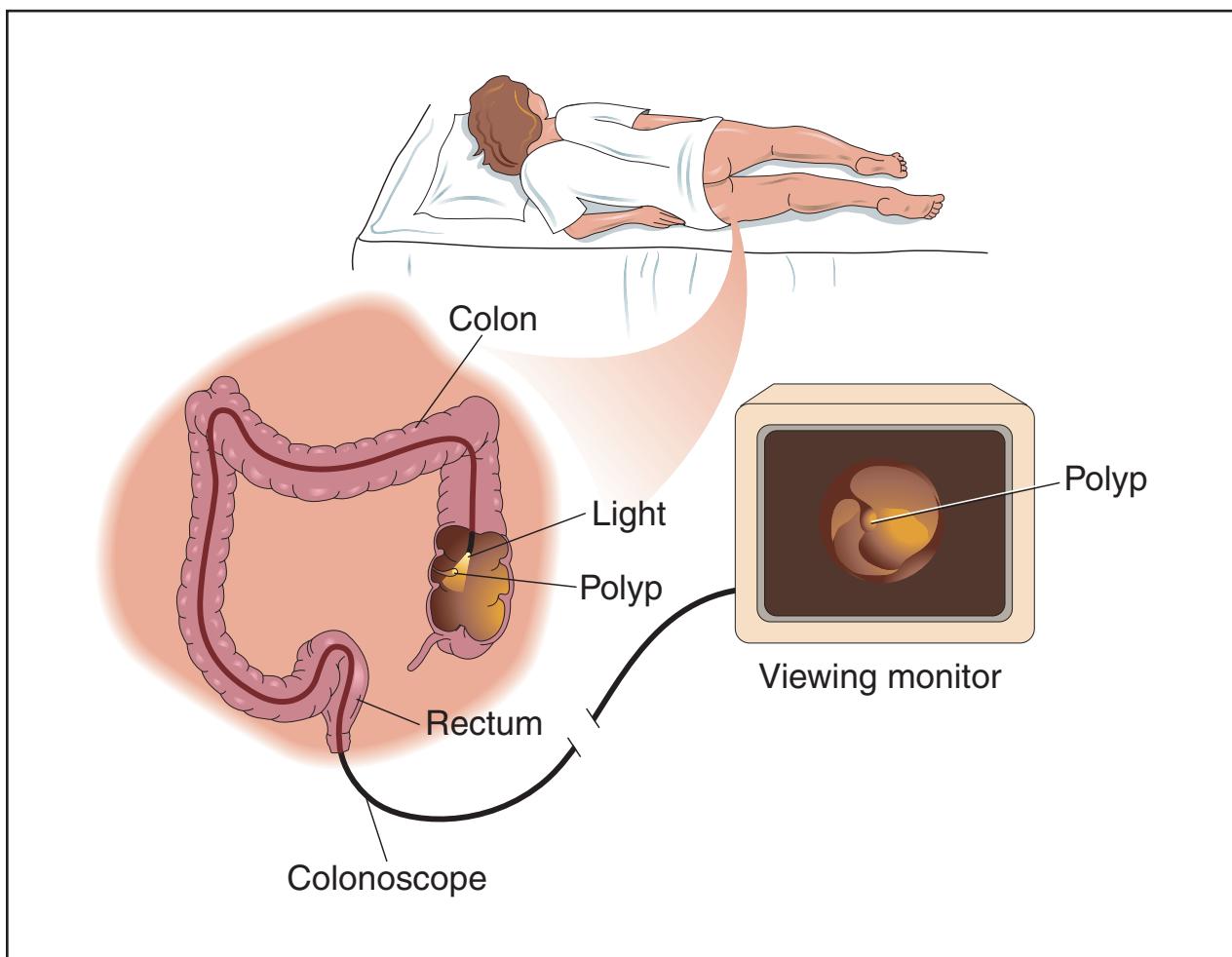
A colonoscopy is generally recommended when the patient complains of rectal bleeding or has a change in bowel habits and other unexplained abdominal symptoms. The test is frequently used to test for colorectal cancer, especially when polyps or tumor-like growths have been detected using the **barium enema** and other diagnostic tests. Polyps can be removed through the colonoscope and samples of tissue (biopsies) can be taken to test for the presence of cancerous cells.

The test also enables the physician to check for bowel diseases such as **ulcerative colitis** and **Crohn's disease**. It is a necessary tool in monitoring patients who have a past history of polyps or **colon cancer**.

Description

The procedure can be done either in the doctor's office or in a special procedure room of a local hospital. An intravenous (IV) line will be started in a vein in the arm. The patient is generally given a sedative and a pain-killer through the IV line.

During the colonoscopy, the patient will be asked to lie on his/her left side with his/her knees drawn up towards the abdomen. The doctor begins the procedure by inserting a lubricated, gloved finger into the anus to check for any abnormal masses or blockage. A thin, well-lubricated colonoscope will then be inserted into the anus and it will be gently advanced through the colon. The lining of the intestine will be examined through the scope. Occasionally air may be pumped through the colonoscope to help clear the path or open the colon. If there are



Colonoscopy is a procedure where a long and flexible tubular instrument called a colonoscope is inserted into the patient's anus in order to view the lining of the colon and rectum. It is performed to test for colorectal cancer and other bowel diseases, and enables the physician to collect tissue samples for laboratory analysis. (Illustration by Electronic Illustrators Group.)

excessive secretions, stool, or blood that obstruct the viewing, they will be suctioned out through the scope. The doctor may press on the abdomen or ask the patient to change his/her position in order to advance the scope through the colon.

The entire length of the large intestine can be examined in this manner. If suspicious growths are observed, tiny biopsy forceps or brushes can be inserted through the colon and tissue samples can be obtained. Small polyps can also be removed through the colonoscope. After the procedure, the colonoscope is slowly withdrawn and the instilled air is allowed to escape. The anal area is then cleansed with tissues.

The procedure may take anywhere from 30 minutes to two hours depending on how easy it is to advance the scope through the colon. Colonoscopy can be a long and uncomfortable procedure, and the bowel cleaning prepa-

ration may be tiring and can produce **diarrhea** and cramping. During the colonoscopy, the sedative and the **pain** medications will keep the patient very drowsy and relaxed. Most patients complain of minor discomfort and pressure from the colonoscope moving inside. However, the procedure is not painful.

Preparation

The doctor should be notified if the patient has **allergies** to any medications or anesthetics; any bleeding problems; or if the woman is pregnant. The doctor should also be informed of all the medications that the person is currently on and if he or she has had a barium x-ray examination recently. If the patient has had heart valves replaced, the doctor should be informed so that appropriate **antibiotics** can be administered to prevent any chance of infection. The risks of the procedure will be

KEY TERMS

Barium enema—An x-ray test of the bowel after giving the patient an enema of a white chalky substance that outlines the colon and the rectum.

Biopsy—Removal of a tissue sample for examination under the microscope to check for cancer cells.

Colonoscope—A thin, flexible, hollow, lighted tube that is inserted through the rectum into the colon to enable the doctor to view the entire lining of the colon.

Crohn's disease—A chronic inflammatory disease where the immune system starts attacking one's own body. The disease generally starts in the gastrointestinal tract.

Diverticulosis—A pouchlike section that bulges through the large intestine's muscular walls but is not inflamed. It may cause bleeding, stomach distress and excess gas.

Pathologist—A doctor who specializes in the diagnosis of disease by studying cells and tissues under a microscope.

Polyps—An abnormal growth that develops on the inside of a hollow organ such as the colon.

Ulcerative colitis—A chronic condition where recurrent ulcers are found in the colon. It is manifested clinically by abdominal cramping, and rectal bleeding.

explained to the patient before performing the procedure and the patient will be asked to sign a consent form.

It is important that the colon be thoroughly cleaned before performing the examination. Hence, before the examination, considerable preparation is necessary to clear the colon of all stool. The patient will be asked to refrain from eating any solid food for 24–48 hours before the test. Only clear liquids such as juices, broth, and Jello are recommended. The patient is advised to drink plenty of water to avoid **dehydration**. The evening before the test, the patient will have to take a strong laxative that the doctor has prescribed. Several 1 qt **enemas** of warm tap water may have to be taken on the morning of the exam. Commercial enemas (e.g., Fleet) may be used.

The patient will be given specific instructions on how to use the enema and how many such enemas are necessary. Generally, the procedure has to be repeated

until the return from the enema is clear of stool particles. On the morning of the examination, the patient is instructed not to eat or drink anything. The preparatory procedures are extremely important since, if the colon is not thoroughly clean, the exam cannot be done.

Aftercare

After the procedure, the patient is kept under observation until the effects of the medications wear off. The patient will have to be driven home by somebody and can generally resume a normal diet and usual activities unless otherwise instructed. The patient will be advised to drink lots of fluids to replace those lost by **laxatives** and **fasting**.

For a few hours after the procedure, the patient may feel groggy. There may be some abdominal cramping and considerable amount of gas may be passed. If a biopsy was performed or a polyp was removed, there may be small amounts of blood in the stool for a few days. If the patient experiences severe abdominal pain or has persistent and heavy bleeding, it should be brought to the doctor's attention immediately.

Risks

The procedure is virtually free of any complications and risks. Very rarely (two in 1000 cases) there may be a perforation (a hole) in the intestinal wall. Heavy bleeding due to the removal of the polyp or from the biopsy site occurs very infrequently (one in 1000 cases). Infections due to a colonoscopy are also extremely rare. Patients with artificial or abnormal heart valves are usually given antibiotics before and after the procedure to prevent an infection.

Normal results

The results are said to be normal if the lining of the colon is a pale reddish pink and there are no abnormal looking masses that are found in the lining of the colon.

Abnormal results

Abnormal results would imply that polyps or other suspicious-looking masses were detected in the lining of the intestine. Polyps can be removed during the procedure and tissue samples can be biopsied. If cancerous cells are detected in the tissue samples, then a diagnosis of colon cancer is made. The pathologist analyzes the tumor cells further to estimate the aggressiveness of the tumor and the extent of spread of the disease. This is crucial before deciding on the mode of treatment for the disease. Abnormal findings could also be due to inflam-

matory bowel diseases such as ulcerative colitis or Crohn's disease. A condition called diverticulosis, where many small fingerlike pouches protrude from the colon wall, may also contribute to an abnormal result in the colonoscopy.

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ORGANIZATIONS

- American Cancer Society. 1599 Clifton Rd., NE, Atlanta, GA 30329-4251. (800) 227-2345. <<http://www.cancer.org>>.
 Cancer Research Institute. 681 Fifth Ave., New York, N.Y. 10022. (800) 992-2623. <<http://www.cancerresearch.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>>.
 United Ostomy Association, Inc. (UOA). 19772 MacArthur Blvd., Suite 200, Irvine, CA 92612-2405. (800) 826-0826. <<http://www.oua.org>>.

Lata Cherath, PhD

Color blindness

Definition

Color blindness is an abnormal condition characterized by the inability to clearly distinguish different colors of the spectrum. The difficulties can be mild to severe. It is a misleading term because people with color blindness are not blind. Rather, they tend to see colors in a limited range of hues; a rare few may not see colors at all.

Description

Normal color vision requires the use of specialized receptor cells called cones, which are located in the retina of the eye. There are three types of cones, termed red, blue, and green, which enable people to see a wide spectrum of colors. An abnormality, or deficiency, of any of the types of cones will result in abnormal color vision.

There are three basic variants of color blindness. Red/green color blindness (deutanopia) is the most common deficiency, affecting 8% of Caucasian males and 0.5% of Caucasian females. The prevalence varies with culture.

Blue color blindness (protanopia) is an inability to distinguish both blue and yellow, which are seen as white or gray. Protanopia is quite rare and has equal prevalence in males and females. It is common for young children to have blue/green confusion that becomes less pronounced in adulthood. Blue color deficiency often appears in people who have physical disorders such as liver disease or **diabetes mellitus**.

A total inability to distinguish colors (achromatopsia) is exceedingly rare. These affected individuals view the world in shades of gray. They frequently have poor visual acuity and are extremely sensitive to light (photophobia), which causes them to squint in ordinary light.

Researchers studying red/green color blindness in the United Kingdom reported an average prevalence of only 4.7% in one group. Only 1% of Eskimo males are color blind. Approximately 2.9% of boys from Saudi Arabia and 3.7% from India were found to have deficient color vision. Red/green color blindness may slightly increase an affected person's chances of contracting **leprosy**. Pre-term infants exhibit an increased prevalence of blue color blindness. Achromatopsia has a prevalence of about 1 in 33,000 in the United States and affects males and females equally.

Causes and symptoms

Red/green and blue color blindness appear to be located on at least two different gene locations. The majority of affected individuals are males. Females are carriers, but are not normally affected. This indicates that the X chromosome is one of the locations for color blindness. Male offspring of females who carry the altered gene have a 50-50 chance of being color-blind. The rare female that has red/green color blindness, or rarer still, blue color blindness, indicates there is an involvement of another gene. As of 2001, the location of this gene has not been identified.

Achromatopsia, the complete inability to distinguish color, is an autosomal recessive disease of the retina. This means that both parents have one copy of the altered gene but do not have the disease. Each of their children has a 25% chance of not having the gene, a 50% chance of having one altered gene (and, like the parents, being unaffected), and a 25% risk of having both the altered gene and the condition. In 1997, the achromatopsia gene was located on chromosome 2.

The inability to correctly identify colors is the only sign of color blindness. It is important to note that people with red/green or blue varieties of color blindness use other cues such as color saturation and object shape or location to distinguish colors. They can often distinguish

KEY TERMS

Achromatopsia—The inability to distinguish any colors.

Cones—Receptor cells that allow the perception of colors.

Deuteranopia—The inability or difficulty in distinguishing red/green colors.

Photophobia—An extreme sensitivity to light.

Protanopia—The inability of difficulty in distinguishing blue and yellow colors.

Retina—The light-sensitive layer of tissue in the back of the eye that receives and transmits visual signals to the brain through the optic nerve.

Rod—Photoreceptor that is highly sensitive to low levels of light and transmits images in shades of gray.

red or green if they can visually compare the colors. However, most have difficulty accurately identifying colors without any other references. Most people with any impairment in color vision learn colors, as do other young children. These individuals often reach adolescence before their visual deficiency is identified.

Color blindness is sometimes acquired. Chronic illnesses that can lead to color blindness include **Alzheimer's disease**, diabetes mellitus, **glaucoma**, leukemia, liver disease, chronic alcoholism, **macular degeneration**, **multiple sclerosis**, **Parkinson's disease**, sickle cell anemia, and **retinitis pigmentosa**. Accidents or strokes that damage the retina or affect particular areas of the brain eye can lead to color blindness. Some medications such as **antibiotics**, **barbiturates**, anti-tuberculosis drugs, high blood pressure medications, and several medications used to treat nervous disorders and psychological problems may cause color blindness. Industrial or environmental chemicals such as carbon monoxide, carbon disulfide, fertilizers, styrene, and some containing lead can cause loss of color vision. Occasionally, changes can occur in the affected person's capacity to see colors after age 60.

Diagnosis

There are several tests available to identify problems associated with color vision. The most commonly used is the American Optical/Hardy, Rand, and Ritter Pseudoisochromatic test. It is composed of several discs filled with colored dots of different sizes and colors. A person with normal color vision looking at a test item sees a

number that is clearly located somewhere in the center of a circle of variously colored dots. A color-blind person is not able to distinguish the number.

The Ishihara test is comprised of eight plates that are similar to the American Optical Pseudoisochromatic test plates. The individual being tested looks for numbers among the various colored dots on each test plate. Some plates distinguish between red/green and blue color blindness. Individuals with normal color vision perceive one number. Those with red/green color deficiency see a different number. Those with blue color vision see yet a different number.

A third analytical tool is the Titmus II Vision Tester Color Perception test. The subject looks into a stereoscopic machine. The test stimulus most often used in professional offices contains six different designs or numbers on a black background, framed in a yellow border. Titmus II can test one eye at a time. However, its value is limited because it can only identify red/green deficiencies and is not highly accurate.

Treatment

There is no treatment or cure for color blindness. Most color vision deficient persons compensate well for their abnormality and usually rely on color cues and details that are not consciously evident to persons with typical color vision.

Inherited color blindness cannot be prevented. In the case of some types of acquired color deficiency, if the cause of the problem is removed, the condition may improve with time. But for most people with acquired color blindness, the damage is usually permanent.

Prognosis

Color blindness that is inherited is present in both eyes and remains constant over an individual's entire life. Some cases of acquired color vision loss are not severe, may appear in only one eye, and last for only a short time. Other cases tend to be progressive, becoming worse with time.

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ORGANIZATIONS

- Achromatopsia Network. C/O Frances Futterman, PO Box 214, Berkeley, CA 94701-0214. <http://www.achromat.org/how_to_join.html>.
- American Academy of Ophthalmology. PO Box 7424, San Francisco, CA 94120-7424. (415) 561-8500. <<http://www.eyenet.org>>.
- International Colour Vision Society: Forschungsstelle fuer Experimentelle Ophthalmologie. Roentgenweg 11, Tuebingen, D-72076. Germany <<http://orlab.optom.unsw.edu.au/ICVS>>.
- National Society to Prevent Blindness. 500 East Remington Rd., Schaumburg, IL 60173. (708) 843-2020 or (800) 331-2020. <<http://www.preventblindness.org>>.

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L. Fleming Fallon, Jr., MD, MPH

Colorectal cancer see **Colon cancer; Rectal cancer**

Colostomy

Definition

Ostomy is a surgical procedure used to create an opening for urine and feces to be released from the body.

Colostomy refers to a surgical procedure where a portion of the large intestine is brought through the abdominal wall to carry stool out of the body.

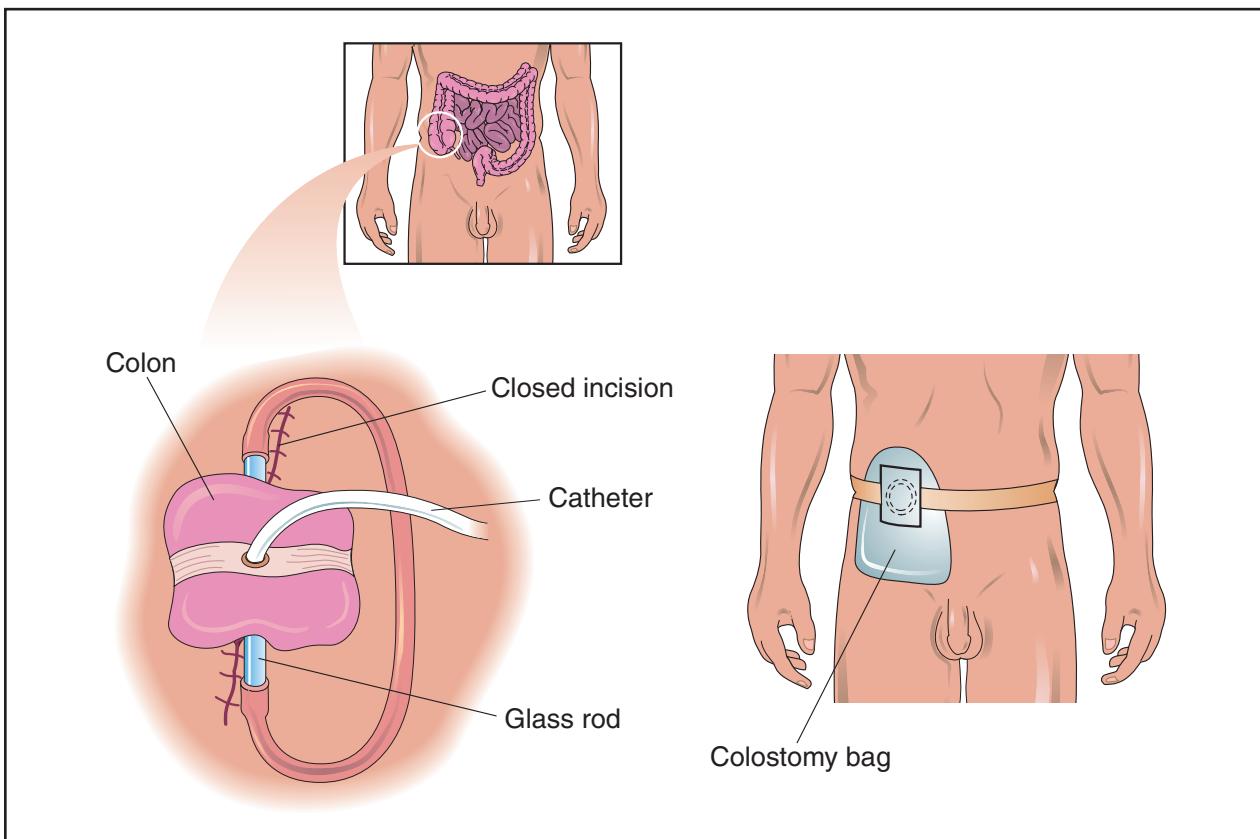
Purpose

A colostomy is created as a means to treat various disorders of the large intestine, including **cancer**, obstruction, inflammatory bowel disease, ruptured diverticulum, **ischemia** (compromised blood supply), or traumatic injury. Temporary colostomies are created to divert stool from injured or diseased portions of the large intestine, allowing rest and healing. Permanent colostomies are performed when the distal bowel (bowel at the farthest distance) must be removed or is blocked and inoperable. Although colorectal cancer is the most common indication for a permanent colostomy, only about 10–15% of patients with this diagnosis require a colostomy.

Description

Surgery will result in one of three types of colostomies:

- End colostomy. The functioning end of the intestine (the section of bowel that remains connected to the upper gastrointestinal tract) is brought out onto the surface of the abdomen, forming the stoma by cuffing the intestine back on itself and suturing the end to the skin. A stoma is an artificial opening created to the surface of the body. The surface of the stoma is actually the lining of the intestine, usually appearing moist and pink. The distal portion of bowel (now connected only to the rectum) may be removed, or sutured closed and left in the abdomen. An end colostomy is usually a permanent ostomy, resulting from trauma, cancer or another pathological condition.
- Double-barrel colostomy. This colostomy involves the creation of two separate stomas on the abdominal wall. The proximal (nearest) stoma is the functional end that is connected to the upper gastrointestinal tract and will drain stool. The distal stoma, connected to the rectum and also called a mucous fistula, drains small amounts of mucus material. This is most often a temporary colostomy performed to rest an area of bowel, and to be later closed.
- Loop colostomy. This colostomy is created by bringing a loop of bowel through an incision in the abdominal wall. The loop is held in place outside the abdomen by a plastic rod slipped beneath it. An incision is made in the bowel to allow the passage of stool through the loop colostomy. The supporting rod is removed approximately 7–10 days after surgery, when healing has occurred that will prevent the loop of bowel from



A colostomy is a surgical procedure in which a portion of the large intestine, or colon, is brought through the abdominal wall to carry feces out of the body. There are three types of colostomies: end colostomy, double-barrel colostomy, and loop colostomy. The loop colostomy is featured in the illustration above. (Illustration by Electronic Illustrators Group.)

retracting into the abdomen. A loop colostomy is most often performed for creation of a temporary stoma to divert stool away from an area of intestine that has been blocked or ruptured.

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 and an electrocardiograph (EKG), may be ordered as the doctor deems necessary. If possible, the patient should visit an enterostomal therapist, who will mark an appropriate place on the abdomen for the stoma, and offer pre-operative education on ostomy management.

In order to empty and cleanse the bowel, the patient may be placed on a low residue diet for several days prior to surgery. A liquid diet may be ordered for at least the day before surgery, with nothing by mouth after midnight. A series of **enemas** and/or oral preparations (GoLyteLy or Colyte) may be ordered to empty the bowel

of stool. Oral anti-infectives (neomycin, erythromycin, or kanamycin sulfate) may be ordered to decrease bacteria in the intestine and help prevent post-operative infection. A nasogastric tube is inserted from the nose to the stomach on the day of surgery or during surgery to remove gastric secretions and prevent **nausea and vomiting**. A urinary catheter (a thin plastic tube) may also be inserted to keep the bladder empty during surgery, giving more space in the surgical field and decreasing chances of accidental injury.

Aftercare

Post-operative care for the patient with a new colostomy, as with those who have had any major surgery, involves monitoring of blood pressure, pulse, respirations, 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 instructed how to support the operative site during deep breathing and coughing, and given pain medication as necessary. Fluid intake and output is measured, and the operative site is observed

for color and amount of wound drainage. The nasogastric tube will remain in place, attached to low intermittent suction until bowel activity resumes. For the first 24–48 hours after surgery, the colostomy will drain bloody mucus. Fluids and electrolytes are infused intravenously until the patient's diet is can gradually be resumed, beginning with liquids. Usually within 72 hours, passage of gas and stool through the stoma begins. Initially the stool is liquid, gradually thickening as the patient begins to take solid foods. The patient is usually out of bed in 8–24 hours after surgery and discharged in 2–4 days.

A colostomy pouch will generally have been placed on the patient's abdomen, around the stoma, during surgery. During the hospital stay, the patient and his or her caregivers will be educated on how to care for the colostomy. Determination of appropriate pouching supplies and a schedule of how often to change the pouch should be established. Regular assessment and meticulous care of the skin surrounding the stoma is important to maintain an adequate surface on which to apply the pouch. Some patients with colostomies are able to routinely irrigate the stoma, resulting in regulation of bowel function; rather than needing to wear a pouch, these patients may need only a dressing or cap over their stoma. Often, an enterostomal therapist will visit the patient at home after discharge to help with the patient's resumption of normal daily activities.

Risks

Potential complications of colostomy surgery include:

- excessive bleeding
- surgical wound infection
- thrombophlebitis (inflammation and blood clot to veins in the legs)
- pneumonia
- pulmonary **embolism** (blood clot or air bubble in the lungs' blood supply)

Normal results

Complete healing is expected without complications. The period of time required for recovery from the surgery may vary depending of the patient's overall health prior to surgery. The colostomy patient without other medical complications should be able to resume all daily activities once recovered 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

KEY TERMS

Diverticulum—Pouches that project off the wall of the intestine, visible as opaque on an x ray after the patient has swallowed a contrast (dye) substance.

Embolism—Blockage of a blood vessel by any small piece of material traveling in the blood. The emboli may be caused by germs, air, blood clots, or fat.

Enema—Insertion of a tube into the rectum to infuse fluid into the bowel and encourage a bowel movement. Ordinary enemas contain tap water, mixtures of soap and water, glycerine and water, or other materials.

Intestine—Commonly called the bowels, divided into the small and large intestine. They extend from the stomach to the anus. The small intestine is about 20 ft (6 m) long. The large intestine is about 5 ft (1.5 m) long.

Ischemia—A compromise in blood supply delivered to body tissues that causes tissue damage or death.

Ostomy—A surgically created opening in the abdomen for elimination of waste products (urine or stool).

- headache, muscle aches, **dizziness**, or **fever**
- increased abdominal pain or swelling, **constipation**, nausea or vomiting or black, tarry stools

Stomal complications to be monitored include:

- Death (necrosis) of stomal tissue. Caused by inadequate blood supply, this complication is usually visible 12–24 hours after the operation and may require additional surgery.
- Retraction (stoma is flush with the abdomen surface or has moved below it). Caused by insufficient stomal length, this complication may be managed by use of special pouching supplies. Elective revision of the stoma is also an option.
- Prolapse (stoma increases length above the surface of the abdomen). Most often results from an overly large opening in the abdominal wall or inadequate fixation of the bowel to the abdominal wall. Surgical correction is required when blood supply is compromised.
- Stenosis (narrowing at the opening of the stoma). Often associated with infection around the stoma or scarring. Mild stenosis can be removed under local anesthesia.

Severe stenosis may require surgery for reshaping the stoma.

- Parastomal **hernia** (bowel causing bulge in the abdominal wall next to the stoma). This is due to placement of the stoma where the abdominal wall is weak or creation of an overly large opening in the abdominal wall. The use of an ostomy support belt and special pouching supplies may be adequate. If severe, the defect in the abdominal wall should be repaired and the stoma moved to another location.

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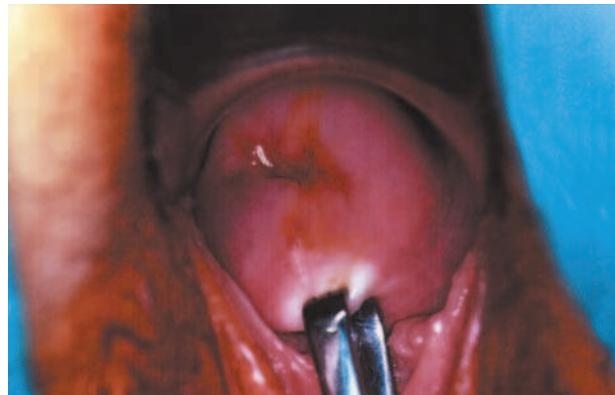
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Kathleen D. Wright, RN



A colposcopy makes it possible for a physician to view this healthy cervix without surgery. (Photograph by Dr. P. Marazzi, Custom Medical Stock Photo. Reproduced by permission.)

sue. If a **PAP test** shows abnormal cell growth, further testing, such as colposcopy, often is required. A PAP test is a screening test that involves scraping cells from the outside of the cervix. If abnormal cells are found, the physician will attempt to find the area that produced the abnormal cells and remove it for further study (biopsy). Only then can a diagnosis be made.

Colposcopy may also be performed if the cervix looks abnormal during a routine examination. It may also be suggested for women with **genital warts** and for diethylstilbestrol (DES) daughters (women whose mothers took DES when pregnant with them).

Precautions

Women who are pregnant, or who suspect that they are pregnant, must tell their doctor before the procedure begins. Pregnant women can, and should, have a colposcopy if they have an abnormal PAP test. However, special precautions must be taken during biopsy of the cervix.

Description

A colposcopy is performed in a physician's office and is similar to a regular gynecologic exam. An instrument called a speculum is used to hold the vagina open, and the gynecologist looks at the cervix and vagina through the colposcope instead simply by eye, as in a routine examination.

The colposcope is placed outside the patient's body and never touches the skin. The cervix and vagina are swabbed with dilute acetic acid (vinegar). The solution highlights abnormal areas by turning them white (instead of a normal pink color). Abnormal areas can also be identified by looking for a characteristic pattern made by abnormal blood vessels.

Colposcopy

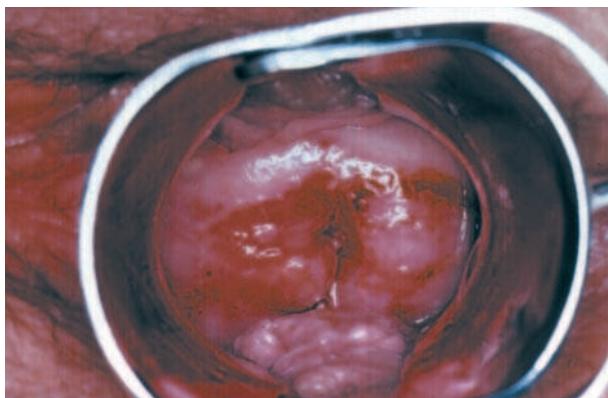
Definition

Colposcopy is a procedure that allows a physician to take a closer look at a woman's cervix and vagina using a special instrument called a colposcope. It is used to check for precancerous or abnormal areas. The colposcope can magnify the area between 10 and 40 times; some devices also can take photographs.

Purpose

The colposcope helps to identify abnormal areas of the cervix or vagina so that small pieces of tissue (biopsies) can be taken for further analysis.

Colposcopy is used to identify or rule out the existence of any precancerous conditions in the cervical tis-



This colposcopic view of the cervix reveals CIN 2 dysplasia, or abnormal growth of cells. This is the second stage in the development of cervical cancer. (Custom Medical Stock Photo. Reproduced by permission.)

If any abnormal areas are seen, the doctor will take a biopsy of the tissue, a common procedure that takes about 15 minutes. Several samples might be taken, depending on the size of the abnormal area. A biopsy may cause temporary discomfort and cramping, which usually go away within a few minutes. If the abnormal area appears to extend inside the cervical canal, a scraping of the canal may be done. The biopsy results are usually available within a week.

If the tissue sample indicates abnormal growth (dysplasia) or precancer, and if the entire abnormal area can be seen, the doctor can destroy the tissue using one of several procedures, including ones that use high heat (diathermy), extreme cold (cryosurgery), or lasers. Another procedure, called a loop electrosurgical excision (LEEP), uses low-voltage high-frequency radio waves to excise tissue. If any of the abnormal tissue is within the cervical canal, a cone biopsy (removal of a conical section of the cervix for inspection) will be needed.

Preparation

Colposcopy is a painless procedure that does not require any anesthetic medication. If a biopsy is done, there may be mild cramps or a sharp pinching when the tissue is removed. To lessen this **pain**, your doctor may recommend 800 mg of ibuprofen (Motrin) taken the night before and the morning of the procedure (no later than 30 minutes before the appointment). Patients who are pregnant or allergic to **aspirin** or ibuprofen can take two tablets of **acetaminophen** (Tylenol) instead.

Aftercare

If a biopsy was done, there may be a dark vaginal discharge afterwards. After the sample is removed, the

KEY TERMS

Biopsy—Removal of sample of abnormal tissue for more extensive examination under a microscope.

Cervix—The neck of the uterus.

Cryosurgery—Freezing and destroying abnormal cells.

DES—The abbreviation for diethylstilbestrol, a synthetic form of estrogen that was widely prescribed to women from 1940 to 1970 to prevent complications. It was linked to several serious birth defects and disorders of the reproductive system in daughters of women who took DES. In 1971, the FDA suggested it not be used during pregnancy and banned its use in 1979 as a growth promoter in livestock.

Diathermy—Also called electrocautery, this is a procedure that heats and destroys abnormal cells. It is gradually being replaced by cryosurgery, lasers, or LEEP.

Human papilloma virus—A virus that causes common warts of the hands and feet, as well as lesions in the genital and vaginal area. More than 50 types of HPV have been identified, some of which are linked to cancerous and precancerous conditions, including cancer of the cervix.

Loop electrosurgical excision (LEEP)—A procedure that can help diagnose and treat cervical abnormalities, using a thin wire loop that emits a low-voltage high-frequency radio wave that can excise tissue. It is considered better than either lasers or electrocautery because it can both diagnose and treat precancerous cells or early stage cancer at the same time.

PAP test—The common term for the Papanicolaou test, a simple smear method of examining stained cells to detect cancer of the cervix.

Speculum—A retractor used to separate the walls of the vagina to make visual examination easier.

doctor applies Monsel's solution to the area to stop the bleeding. When this mixes with blood it creates a black fluid that looks like coffee grounds for a couple of days after the procedure. It is also normal to have some spotting after a colposcopy.

Patients should not use tampons or put anything else in the vagina for at least a week after the procedure, or until the doctor says it's safe. In addition, women should

not have sex or douche for at least a week after the procedure because of the risk of infection.

Risks

Occasionally, patients may have bleeding or infection after biopsy. Bleeding is usually controlled with a topical medication.

A patient should call her doctor right away if she notices any of the following symptoms:

- heavy vaginal bleeding (more than one sanitary pad an hour)
- fever, chills, or an unpleasant vaginal odor
- lower abdominal pain.

Normal results

If visual inspection shows that the surface of the cervix is smooth and pink, this is considered normal. If abnormal areas are found and biopsied and the results show no indication of **cancer**, a precancerous condition, or other disease, this also is considered normal.

Abnormal results

Abnormal conditions that can be detected using colposcopy and biopsy include precancerous tissue changes (cervical dysplasia), cancer, and cervical **warts** (human papilloma virus).

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ORGANIZATIONS

American Society for Colposcopy and Cervical Pathology. 20 W. Washington St., Ste. #1, Hagerstown, MD 21740. (800) 787-7227. <<http://www.asccp.org>>.

Carol A. Turkington

Coma

Definition

Coma, from the Greek word "koma," meaning deep sleep, is a state of extreme unresponsiveness, in which an individual exhibits no voluntary movement or behavior. Furthermore, in a deep coma, even painful stimuli

(actions which, when performed on a healthy individual, result in reactions) are unable to affect any response, and normal reflexes may be lost.

Description

Coma lies on a spectrum with other alterations in consciousness. The level of consciousness required by, for example, someone reading this passage lies at one extreme end of the spectrum, while complete brain **death** lies at the other end of the spectrum. In between are such states as obtundation, drowsiness, and stupor. All of these are conditions which, unlike coma, still allow the individual to respond to stimuli, although such a response may be brief and require stimulus of greater than normal intensity.

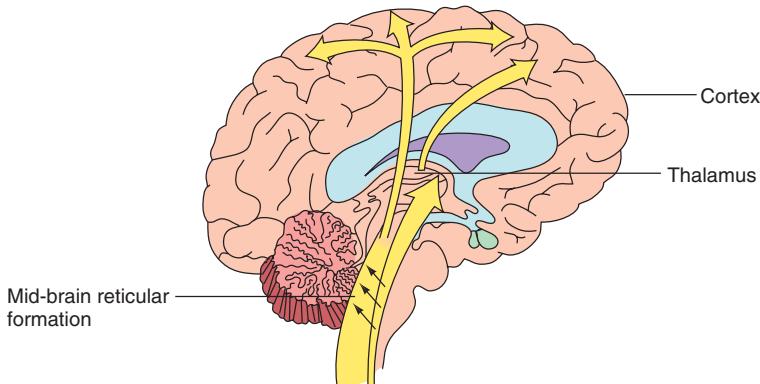
In order to understand the loss of function suffered by a comatose individual, it is necessary to first understand the important characteristics of the conscious state. Consciousness is defined by two fundamental elements: awareness and arousal.

Awareness allows one to receive and process all the information communicated by the five senses, and thus relate to oneself and to the outside world. Awareness has both psychological and physiological components. The psychological component is governed by an individual's mind and mental processes. The physiological component refers to the functioning of an individual's brain, and therefore that brain's physical and chemical condition. Awareness is regulated by cortical areas within the cerebral hemispheres, the outermost layer of the brain that separates humans from other animals by allowing for greater intellectual functioning.

Arousal is regulated solely by physiological functioning and consists of more primitive responsiveness to the world, as demonstrated by predictable reflex (involuntary) responses to stimuli. Arousal is maintained by the reticular activating system (RAS). This is not an anatomical area of the brain, but rather a network of structures (including the brainstem, the medulla, and the thalamus) and nerve pathways, which function together to produce and maintain arousal.

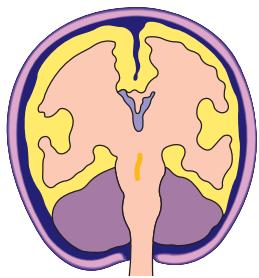
Causes and symptoms

Coma, then, is the result of something that interferes with the functioning of the cerebral cortex and/or the functioning of the structures which make up the RAS. In fact, a huge and varied number of conditions can result in coma. A good way of categorizing these conditions is to consider the anatomic and the metabolic causes of coma. Anatomic causes of coma are those conditions that disrupt the normal physical architecture of the brain structures responsible for consciousness, either at the level of



A side-view of the brain, showing movement of the reticular activating substance (RAS) essential to consciousness

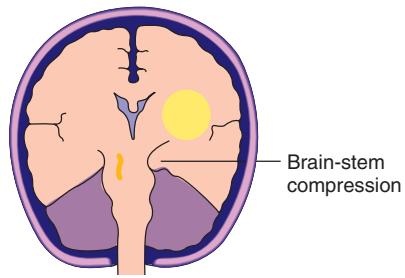
Diffuse and bilateral damage to the cerebral cortex (relative preservation of brain-stem reflexes)



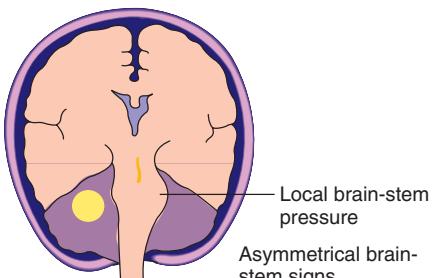
Possible causes

- Damage due to lack of oxygen or restricted blood flow, perhaps resulting from cardiac arrest, an anaesthetic accident, or shock
- Damage incurred from metabolic processes associated with kidney or liver failure, or with hypoglycemia
- Trauma damage
- Damage due to a bout with meningitis, encephalomyelitis, or a severe systemic infection

Mass lesions in this region resulting in compression of the brain-stem and damage to the reticular activating substance (RAS)

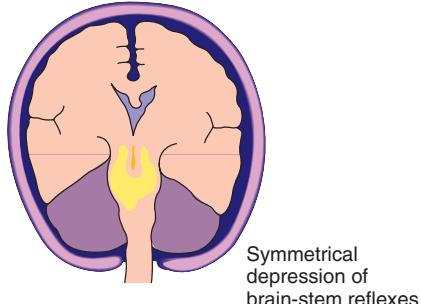


Structural lesions within this region also resulting in compression of the brain-stem and damage to the reticular activating substance (RAS)



Possible causes • Cerebellar tumors, abscesses, or hemorrhages

Lesions within the brain-stem directly suppressing the reticular activating substance (RAS)



Possible causes • Drug overdosage

The four brain conditions that result in coma. (Illustration by Hans & Cassady.)

KEY TERMS

Anatomic—Related to the physical structure of an organ or organism.

Metabolic—Refers to the chemical processes of an organ or organism.

Neuron—The cells within the body which make up the nervous system, specifically those along which information travels.

Physiological—Pertaining to the functioning of an organ, as governed by the interactions between its physical and chemical conditions.

Psychological—Pertaining to the mind, its mental processes, and its emotional makeup.

Stimulus/stimuli—Action or actions performed on an individual which predictably provoke(s) a reaction.

the cerebral cortex or the brainstem, while metabolic causes of coma consist of those conditions that change the chemical environment of the brain, thereby adversely affecting function.

There are many metabolic causes of coma, including:

- A decrease in the delivery to the brain of substances necessary for appropriate brain functioning, such as oxygen, glucose (sugar), and sodium.
- The presence of certain substances that disrupt the functioning of neurons. Drugs or alcohol in toxic quantities can result in neuronal dysfunction, as can substances normally found in the body, but that, due to some diseased state, accumulate at toxic levels. Accumulated substances that might cause coma include ammonia due to liver disease, ketones due to uncontrolled diabetes, or carbon dioxide due to a severe **asthma** attack.
- The changes in chemical levels in the brain due to the electrical derangements caused by seizures.

Diagnosis

As in any neurologic condition, history and examination form the cornerstone of diagnosis when the patient is in a coma; however, history must be obtained from family, friends, or EMS. The Glasgow Coma Scale is a system of examining a comatose patient. It is helpful for evaluating the depth of the coma, tracking the patient's progress, and predicting (somewhat) the ultimate outcome of the coma. The Glasgow Coma Scale assigns a different number of points for exam results in three different categories: opening the eyes, verbal response (using words or voice to respond), and motor response (moving a part of the body). Fifteen is the largest possible number of total points, indicating the highest level of functioning. The highest level of functioning would be demonstrated by an individual who spontaneously opens his/her eyes, gives appropriate answers to questions about his/her situation, and can carry out a command (such as "move your leg" or "nod your head"). Three is the least possible number of total points and would be given to a patient for whom not even a painful stimulus is sufficient to provoke a response. In the middle are those patients who may be able to respond, but who require an intense or painful stimulus, and whose response may demonstrate some degree of brain malfunctioning (such as a person whose only response to **pain** in a limb is to bend that limb in toward the body). When performed as part of the admission examination, a Glasgow score of three to five points often suggests that the patient has likely suffered fatal brain damage, while eight or more points indicates that the patient's chances for recovery are good. Expansion of the pupils and respiratory pattern are also important. Metabolic causes of coma are diagnosed from blood work and **urinalysis** to evaluate blood chemistry, drug screen, and blood cell abnormalities that may indicate infection. Anatomic causes of coma are diagnosed from **computed tomography scans (CT)** or **magnetic resonance imaging (MRI)** scans.

Treatment

Coma is a medical emergency, and attention must first be directed to maintaining the patient's respiration and circulation, using intubation and ventilation, administration of intravenous fluids or blood as needed, and other supportive care. If head trauma has not been excluded, the neck should be stabilized in the event of fracture. It is obviously extremely important for a physician to determine quickly the cause of a coma, so that potentially reversible conditions are treated immediately. For example, an infection may be treated with **antibiotics**; a **brain tumor** may be removed; and brain swelling from an injury can be reduced with certain medications. Various metabolic disorders can be addressed by supplying the individual with the correct amount of oxygen, glucose, or sodium; by treating the underlying disease in liver disease, asthma, or diabetes; and by halting seizures with medication. Because of their low incidence of side effects and potential for prompt reversal of coma in certain conditions, glucose, the B-vitamin thiamine, and Narcan (to counteract any narcotic-type drugs) are routinely given.

Prognosis

Some conditions that cause coma can be completely reversed, restoring the individual to his or her original level of functioning. However, if areas of the brain have been sufficiently damaged due to the severity or duration of the condition which led to the coma, the individual may recover from the coma with permanent disabilities, or may even never regain consciousness. Take, for example, the situation of someone whose coma was caused by brain injury in a car accident. Such an injury can result in one of three outcomes. In the event of a less severe brain injury, with minimal swelling, an individual may indeed recover consciousness and regain all of his or her original abilities. In the event of a more severe brain injury, with swelling that resulted in further pressure on areas of the brain, an individual may regain consciousness, but may have some degree of impairment. The impairment may be physical (such as **paralysis** of a leg) or may even result in a change in the individual's intellectual functioning and/or personality. The most severe types of brain injury, short of death, result in states in which the individual loses all ability to function and remains deeply unresponsive. An individual who has suffered such a severe brain injury may remain in a coma indefinitely. This condition is termed **persistent vegetative state**.

Outcome from a coma is therefore quite variable and depends a great deal on the cause and duration of the coma. In the case of drug poisonings, extremely high rates of recovery can be expected following prompt medical attention. Patients who have suffered head injuries tend to do better than do patients whose coma was caused by other types of medical illnesses. Leaving out those people whose coma followed drug **poisoning**, only about 15% of patients who remain in a coma for more than just a few hours make a good recovery. Those adult patients who remain in a coma for greater than four weeks have almost no chance of eventually regaining their previous level of functioning. On the other hand, children and young adults have regained functioning even after two months in a coma.

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ORGANIZATIONS

American Academy of Neurology. 1080 Montreal Ave., St. Paul, MN 55116. (612) 695-1940. <<http://www.aan.com>>. Coma Recovery Association, Inc. 570 Elmont Rd., Suite 104, Elmont, NY 11003. (516) 355-0951.

Rosalyn Carson-DeWitt, MD

Combat neurosis see **Post-traumatic stress disorder**

Common cold

Definition

The common cold is a viral infection of the upper respiratory system, including the nose, throat, sinuses, eustachian tubes, trachea, larynx, and bronchial tubes. Although over 200 different viruses can cause a cold, 30–50% are caused by a group known as rhinoviruses. Almost all colds clear up in less than two weeks without complications.

Description

Colds, sometimes called rhinovirus or coronavirus infections, are the most common illness to strike any part of the body. It is estimated that the average person has more than 50 colds during a lifetime. Anyone can get a cold, although pre-school and grade school children catch them more frequently than adolescents and adults. Repeated exposure to viruses causing colds creates partial immunity.

Although most colds resolve on their own without complications, they are a leading cause of visits to the doctor and of time lost from work and school. Treating symptoms of the common cold has given rise to a multi-million dollar industry in over-the-counter medications.

Cold season in the United States begins in early autumn and extends through early spring. Although it is not true that getting wet or being in a draft causes a cold (a person has to come in contact with the virus to catch a cold), certain conditions may lead to increased susceptibility. These include:

- fatigue and overwork
- emotional **stress**
- poor **nutrition**
- **smoking**
- living or working in crowded conditions

Colds make the upper respiratory system less resistant to bacterial infection. Secondary bacterial infection may lead to middle ear infection, **bronchitis**, **pneumonia**, sinus infection, or **strep throat**. People with chronic lung disease, **asthma**, diabetes, or a weakened immune system are more likely to develop these complications.

Causes and symptoms

Colds are caused by more than 200 different viruses. The most common groups are rhinoviruses and coronaviruses. Different groups of viruses are more infectious at different seasons of the year, but knowing the exact virus causing the cold is not important in treatment.

People with colds are contagious during the first two to four days of the infection. Colds pass from person to person in several ways. When an infected person coughs, sneezes, or speaks, tiny fluid droplets containing the virus are expelled. If these are breathed in by other people, the virus may establish itself in their noses and airways.

Colds may also be passed through direct contact. If a person with a cold touches his runny nose or watery eyes, then shakes hands with another person some of the virus is transferred to the uninfected person. If that person then touches his mouth, nose, or eyes, the virus is transferred to an environment where it can reproduce and cause a cold.

Finally, cold viruses can be spread through inanimate objects (door knobs, telephones, toys) that become contaminated with the virus. This is a common method of transmission in child care centers. If a child with a cold touches his runny nose, then plays with a toy, some of the virus may be transferred to the toy. When another child plays with the toy a short time later, he may pick up some of the virus on his hands. The second child then touches his contaminated hands to his eyes, nose, or mouth and transfers some of the cold virus to himself.

Once acquired, the cold virus attaches itself to the lining of the nasal passages and sinuses. This causes the infected cells to release a chemical called histamine. Histamine increases the blood flow to the infected cells, causing swelling, congestion, and increased mucus production. Within one to three days the infected person begins to show cold symptoms.

The first cold symptoms are a tickle in the throat, runny nose, and sneezing. The initial discharge from the nose is clear and thin. Later it changes to a thick yellow or greenish discharge. Most adults do not develop a **fever** when they catch a cold. Young children may develop a low fever of up to 102°F (38.9°C).

In addition to a runny nose and fever, signs of a cold include coughing, sneezing, nasal congestion, **headache**, muscle ache, chills, **sore throat**, hoarseness, watery

eyes, tiredness, and lack of appetite. The **cough** that accompanies a cold is usually intermittent and dry.

Most people begin to feel better four to five days after their cold symptoms become noticeable. All symptoms are generally gone within ten days, except for a dry cough that may linger for up to three weeks.

Colds make people more susceptible to bacterial infections such as strep throat, middle ear infections, and sinus infections. A person whose cold does not begin to improve within a week; or who experiences chest **pain**, fever for more than a few days, difficulty breathing, bluish lips or fingernails, a cough that brings up greenish-yellow or grayish sputum, skin rash, swollen glands, or whitish spots on the tonsils or throat should consult a doctor to see if they have acquired a secondary bacterial infection that needs to be treated with an antibiotic.

People who have **emphysema**, chronic lung disease, diabetes, or a weakened immune system—either from diseases such as **AIDS** or leukemia, or as the result of medications, (**corticosteroids**, **chemotherapy** drugs)—should consult their doctor if they get a cold. People with these health problems are more likely to get a secondary infection.

Diagnosis

Colds are diagnosed by observing a person's symptoms. There are no laboratory tests readily available to detect the cold virus. However, a doctor may do a **throat culture** or blood test to rule out a secondary infection.

Influenza is sometimes confused with a cold, but flu causes much more severe symptoms and generally a fever. **Allergies** to molds or pollens also can make the nose run. Allergies are usually more persistent than the common cold. An allergist can do tests to determine if the cold-like symptoms are being caused by an allergic reaction. Also, some people get a runny nose when they go outside in winter and breathe cold air. This type of runny nose is not a symptom of a cold.

Treatment

There are no medicines that will cure the common cold. Given time, the body's immune system will make antibodies to fight the infection, and the cold will be resolved without any intervention. **Antibiotics** are useless against a cold. However, a great deal of money is spent by pharmaceutical companies in the United States promoting products designed to relieve cold symptoms. These products usually contain **antihistamines**, **decongestants**, and/or pain relievers.

Antihistamines block the action of the chemical histamine that is produced when the cold virus invades the

cells lining the nasal passages. Histamine increases blood flow and causes the cells to swell. Antihistamines are taken to relieve the symptoms of sneezing, runny nose, itchy eyes, and congestion. Side effects are **dry mouth** and drowsiness, especially with the first few doses. Antihistamines should not be taken by people who are driving or operating dangerous equipment. Some people have allergic reactions to antihistamines. Common over-the-counter antihistamines include Chlor-Trimeton, Dimetapp, Tavist, and Actifed. The generic name for two common antihistamines are chlorpheniramine and diphenhydramine.

Decongestants work to constrict the blood flow to the vessels in the nose. This can shrink the tissue, reduce congestion, and open inflamed nasal passages, making breathing easier. Decongestants can make people feel jittery or keep them from sleeping. They should not be used by people with heart disease, high blood pressure, or **glaucoma**. Some common decongestants are Neo-Synepherine, Novafed, and Sudafed. The generic names of common decongestants include phenylephrine, phenylpropanolamine, pseudoephedrine, and in nasal sprays naphazoline, oxymetazoline and xylometazoline.

Many over the counter medications are combinations of both antihistamines and decongestants; an ache and pain reliever, such as **acetaminophen** (Datril, Tylenol, Panadol) or ibuprofen (Advil, Nuprin, Motrin, Medipren); and a cough suppressant (dextromethorphan). Common combination medications include Tylenol Cold and Flu, Triaminic, Sudafed Plus, and Tavist D. **Aspirin** should not be given to children with a cold because of its association with a risk of **Reye's syndrome**, a serious disease.

Nasal sprays and nose drops are other products promoted for reducing nasal congestion. These usually contain a decongestant, but the decongestant can act more quickly and strongly than ones found in pills or liquids because it is applied directly in the nose. Congestion returns after a few hours.

People can become dependent on nasal sprays and nose drops. If used for a long time, users may suffer withdrawal symptoms when these products are discontinued. Nasal sprays and nose drops should not be used for more than a few days. Check the label for recommendations on length and frequency of use.

People react differently to different cold medications and may find some more helpful than others. A medication may be effective initially, then lose some of its effectiveness. Children sometimes react differently than adults. Over-the-counter cold remedies should not be given to infants without consulting a doctor first.

Cold Remedies

	Symptoms	Side effects
Antihistamines	Congestion Itchy eyes Runny nose Sneezing Stuffy nose	Drowsiness Dry mouth and eyes
Decongestants	Congestion Stuffy nose	Insomnia Rapid heart beat Stimulation

Care should be taken not to exceed the recommended dosages, especially when combination medications or nasal sprays are taken. Individuals should determine whether they wish to use any of these drugs. None of them shorten or cure a cold. At best they help a person feel more comfortable. People who are confused about the drugs in any over-the-counter cold remedies should ask their pharmacist for an explanation.

In addition to the optional use of over the counter cold remedies, there are some self-care steps that people can take to ease their discomfort. These include:

- drinking plenty of fluids, but avoiding acidic juices, which may irritate the throat
- gargling with warm salt water—made by adding one teaspoon of salt to 8 oz of water—for a sore throat
- not smoking
- getting plenty of rest
- using a cool-mist room humidifier to ease congestion and sore throat
- rubbing Vaseline or other lubricant under the nose to prevent irritation from frequent nose blowing
- for babies too young to blow their noses, the mucus should be suctioned gently with an infant nasal aspirator, it may be necessary to soften the mucus first with a few drops of salt water

Alternative treatment

Alternative practitioners emphasize that people get colds because their immune systems are weak. They point out that everyone is exposed to cold viruses, but not everyone gets every cold. The difference seems to be in the ability of the immune system to fight infection. Prevention focuses on strengthening the immune system by eating a healthy diet low in sugars and high in fresh fruits and vegetables, practicing **meditation** to reduce stress, and getting regular moderate **exercise**.

Once cold symptoms appear, some naturopathic practitioners believe the symptoms should be allowed to

KEY TERMS

Bronchial tubes—The major airways to the lungs and their main branches.

Coronavirus—a genus of viruses that cause respiratory disease and gastroenteritis.

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.

Eustachian tube—A thin tube between the middle ear and the pharynx. Its purpose is to equalize pressure on either side of the ear drum.

Rhinovirus—A virus that infects the upper respiratory system and causes the common cold.

run their course without interference. Others suggest the following:

- Inhaling a steaming mixture of lemon oil, thyme oil, eucalyptus, and tea tree oil (*Melaleuca* spp.). (**Aromatherapy**)
- Gargling with a mixture of water, salt, and turmeric powder or astringents such as alum, sumac, sage, and bayberry to ease a sore throat. (**Ayurvedic medicine**)
- Taking coneflower (*Echinacea* spp.) or goldenseal (*Hydrastis canadensis*). Other useful herbs to reduce symptoms include yarrow (*Achillea millefolium*), eyebright (*Euphrasia officinalis*), garlic (*Allium sativum*), and onions (*Allium cepa*). (**Herbal**)
- Microdoses of *Viscum album*, *Natrum muriaticum*, *Allium cepa*, or *Nux vomica*. (**Homeopathy**)
- Taking yin chiao (sometimes transliterated as yinquiao) tablets that contain honeysuckle and forsythia when symptoms appear. Natural herb loquat syrup for cough and sinus congestion and Chinese ephedra (*ma-huang*) for runny nose. (**Chinese traditional medicine**)
- The use of zinc lozenges every two hours along with high doses of vitamin C is suggested. Some practitioners also suggest eliminating dairy products for the duration of the cold. (**Nutritional therapy**).

The use of zinc lozenges may be moving toward acceptance by practitioners of traditional medicine. In 1996 the Cleveland Clinic tested zinc gluconate lozenges and found using zinc in the first 24 hours after cold symptoms occurred shortened the duration of symptoms.

The mechanism by which zinc worked was not clear, but additional studies are underway.

Prognosis

Given time, the body will make antibodies to cure itself of a cold. Most colds last a week to 10 days. Most people start feeling better within four or five days. Occasionally a cold will lead to a secondary bacterial infection that causes strep throat, bronchitis, pneumonia, sinus infection, or a middle ear infection. These conditions usually clear up rapidly when treated with an antibiotic.

Prevention

It is not possible to prevent colds because the viruses that cause colds are common and highly infectious. However, there are some steps individuals can take to reduce their spread. These include:

- washing hands well and frequently, especially after touching the nose or before handling food
- covering the mouth and nose when sneezing
- disposing of used tissues properly
- avoiding close contact with someone who has a cold during the first two to four days of their infection
- not sharing food, eating utensils, or cups with anyone
- avoiding crowded places where cold germs can spread
- eating a healthy diet and getting adequate sleep

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Tish Davidson

Common variable immunodeficiency

Definition

Common variable immunodeficiency is an immunodeficiency disorder characterized by a low level of antibodies. Patients with this disease are subject to recurring infections.

Description

Immunodeficiency means that the immune system is deficient in one or more of its components and is unable to respond effectively. Common variable immunodeficiency is the most common of the immunodeficiency disorders. Patients with this disease have frequent infections, especially those caused by the same microorganism. Recurring infections are an indication that the immune system is not responding normally and developing immunity to reinfection. Patients with common variable immunodeficiency have a normal number of B cells, the lymphocytes that make antibodies. In approximately one-third of these patients, the number of B cells in the blood that have IgG antibodies on their surface is lower than normal, but there are normal numbers of B cells in their bone marrow. B cells with IgG antibodies on their surface are capable of responding to microorganisms. The lack of IgG on the surface of the B cells means that they are not prepared to fight infection. The T-cell lymphocytes, those cells responsible for cellular immunity, are usually normal, although some cell signal components may be lacking.

Causes and symptoms

The cause of common variable immunodeficiency is not known, although some forms seem to be hereditary. The main symptom is recurring infections that tend to be chronic rather than acute. Patients may also develop **diarrhea** and, as a consequence of the diarrhea, do not absorb food efficiently. This can lead to malnourishment that can aggravate the disorder. Common variable immunodeficiency normally appears in children after the age of 10. **Autoimmune disorders** such as **rheumatoid arthritis**, **thyroiditis**, and **systemic lupus erythematosus** and certain cancers such as lymphomas and leukemias may be associated with common variable immunodeficiency.

Diagnosis

As is true of most immunodeficiency disorders, one of the first signs that the patient has the condition is recurrent infections. Patients with common variable immunodeficiency are subject to recurrent infections, especially those caused by microbes that don't normally cause disease in normal persons. The main diagnostic test that distinguishes common variable immunodeficiency from other immunodeficiency diseases is the low antibody level despite the normal number of B cells. Antibody levels are tested in the serum by a procedure called electrophoresis. This procedure both quantifies the amount of antibody present and identifies the various classes of antibodies. The main class of antibody for fighting infectious diseases is IgG.

Treatment

There is no treatment that will cure the disorder. Treatment for common variable immunodeficiency aims at boosting the body's immune response and preventing or controlling infections. Immune serum, obtained from donated blood, is given as a source of antibodies to boost the immune response. Immune serum is obtained from donated blood. It contains whatever antibodies the donors had in their blood. Consequently, it may not contain all the antibodies that the patient needs and may lack antibodies specific for some of the recurring infections that these patients suffer. **Antibiotics** are used routinely at the first sign of an infection to help the patient eliminate infectious microorganisms.

Prognosis

With good medical care, people with common variable immunodeficiency usually have a normal life span.

Prevention

The disease itself cannot be prevented, but patients and their families can take precautions to prevent the recurrent infections commonly associated with it. For example, good hygiene and **nutrition** are important, as is avoiding crowds or other people who have active infections.

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John T. Lohr, PhD

Complement deficiencies

Definition

Complement deficiencies are a group of disorders in which there is a reduced level of specific proteins, complement, involved in proper immune functioning.

Description

Complement plays several functions in immunity. It can poke holes in bacteria, kill bacteria that are first targeted by antibodies, or, working with antibodies, point

KEY TERMS

Autoimmune diseases—A group of diseases, like rheumatoid arthritis and systemic lupus erythematosus, in which immune cells turn on the body, attacking various tissues and organs.

Hereditary angioedema—A complement deficiency characterized by lymphatic vessel blockages that cause temporary swelling (edema) of areas of the skin, mucous membranes, and, sometimes, internal organs.

Leucocyte adhesion deficiency syndrome—A complement deficiency syndrome characterized by recurrent infections of the skin, mucous membranes, and gastrointestinal tract and the absence of pus formation. This disorder is sometimes apparent at birth when separation of the umbilical cord takes longer than normal.

Meningitis—An inflammation of the lining surrounding the brain and spinal cord.

Paroxysmal nocturnal hemoglobinuria (PNH)—A rare complement disorder characterized by episodes of red blood cell destruction (hemolysis) and blood in the urine (hemoglobinuria) that is worse at night.

Systemic lupus erythematosus—An autoimmune disease in which the immune system attacks the body's connective tissue. A butterfly-shaped facial rash is characteristic.

White blood cells—Cells that are key in immune defense. There are various types, including those that engulf and kill invading bacteria.

out which bacteria need to be engulfed by white blood cells. Without sufficient complement, the body is prone to frequent infections, like **pneumonia** or **meningitis**, or other illnesses, including autoimmune diseases, like **systemic lupus erythematosus**. Since there are more than 20 different types of complement, the disease that results depends on the specific complement that is lacking.

Cause and symptoms

A defect in the complement system can be genetic, but a secondary complement deficiency can also result from ailments that involve a lot of protein loss, including serious **burns**, liver or kidney disease, and autoimmune diseases, like lupus. Symptoms vary depending on the specific complement deficiency and the disease that results. Some peo-

ple remain healthy with no symptoms at all. Others, who suffer from frequent infections, may develop a high **fever**, **diarrhea**, headaches with a stiff neck, or a **cough** with **chest pain**. If an autoimmune disease develops, like lupus, the person may lose weight, suffer from a rash, and have joint pain. Other symptoms of complement deficiency diseases (like hereditary angioedema, paroxysmal nocturnal hemoglobinuria, or leukocyte adhesion deficiency syndrome) include abdominal and back pain, skin infections, **edema** or swelling of the face and red bumps on the skin.

Diagnosis

There are blood tests that determine the activity of the complement system. The two most common screening tests, CH50 and APH50, tell the physician which group of complement components have a defect. More specific blood tests for the individual complement components (e.g., C3 or C4 complement) are then performed. Other specialized blood tests, including C1 esterase level, Ham test, and a white **blood count**, may also be performed.

Treatment

There is no way to treat the actual complement deficiency. However, **antibiotics** are used to treat infections and vaccinations are given to reduce the risk of disease. Often, the person is vaccinated against infections that include **influenza**, pneumonia, and meningitis. In some cases, (e.g., a specific disease called paroxysmal nocturnal hemoglobinuria [PNH]), a bone marrow transplant may be recommended.

Alternative treatment

There is no alternative treatment for complement problems.

Prognosis

Since complement deficiencies include a wide range of disorders, the prognoses can also vary widely. Some patients remain healthy their entire life. Others are hospitalized frequently because of infections which, if not properly treated, can be fatal. Those with autoimmune diseases could have a normal life expectancy. There are some complement deficiencies, that have a high mortality rate. In those cases, **death** may occur within 10 years after diagnosis.

Prevention

There is currently no way to prevent complement deficiencies.

Resources

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Jeanine Barone, Physiologist

Complete blood count see **Blood count**

Computed tomography scans

Definition

Computed tomography (CT) scans are completed with the use of a 360-degree x-ray beam and computer production of images. These scans allow for cross-sectional views of body organs and tissues.

Purpose

CT scans are used to image a wide variety of body structures and internal organs. Since the 1990s, CT equipment has become more affordable and available. In some diagnoses, CT scans have become the first imaging exam of choice. Because the computerized image is so sharp, focused, and three-dimensional, many tissues can be better differentiated than on standard x rays. Common CT indications include:

- Sinus studies. The CT scan can show details of **sinusitis** and bone **fractures**. Physicians may order a CT of the sinuses to provide an accurate map for surgery.
- Brain studies. Brain scans can detect hematomas, tumors, and strokes. The introduction of CT scanning, especially spiral CT, has helped reduce the need for more invasive procedures such as cerebral **angiography**.
- Body scans. CT scans of the body will often be used to observe abdominal organs, such as the liver, kid-

neys, adrenal glands, spleen, and lymph nodes, and extremities.

- Aorta scans. CT scans can focus on the thoracic or abdominal aorta to locate aneurysms and other possible aortic diseases.
- Chest scans. CT scans of the chest are useful in distinguishing tumors and in detailing accumulation of fluid in chest infections.

Precautions

Pregnant women or those who could possibly be pregnant should not have a CT scan unless the diagnostic benefits outweigh the risks. Pregnant patients should particularly avoid full body or abdominal scans. If the exam is necessary for obstetrics purposes, technologists are instructed not to repeat films if there are errors. Pregnant patients receiving CT or any x-ray exam away from the abdominal area may be protected by a lead apron; most radiation, known as scatter, travels through the body and is not blocked by the apron.

Contrast agents are often used in CT exams and the use of these agents should be discussed with the medical professional prior to the procedure. Patients should be asked to sign a consent form concerning the administration of contrast. One of the common contrast agents, iodine, can cause allergic reactions. Patients who are known to be allergic to iodine (or shellfish) should inform the physician prior to the CT scan.

Description

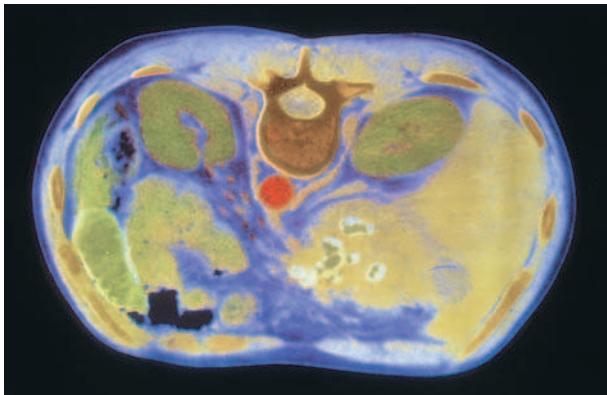
Computed tomography, also called CT scan, CAT scan, or computerized axial tomography, is a combination of focused x-ray beams and computerized production of an image. Introduced in the early 1970s, this radiologic procedure has advanced rapidly and is now widely used, sometimes in the place of standard x rays.

CT equipment

A CT scan may be performed in a hospital or outpatient imaging center. Although the equipment looks large and intimidating, it is very sophisticated and fairly comfortable. The patient is asked to lie on a gantry, or narrow table, that slides into the center of the scanner. The scanner looks like a doughnut and is round in the middle, which allows the x-ray beam to rotate around the patient. The scanner section may also be tilted slightly to allow for certain cross-sectional angles.

CT procedure

The patient will feel the gantry move very slightly as the precise adjustments for each sectional image are



Colorized CT scan of human abdomen—*aorta is dead center/red* (Photo Researchers. Reproduced by permission.)

made. A technologist watches the procedure from a window and views the images on a computer screen.

It is essential that the patient lie very still during the procedure to prevent motion blurring. In some studies, such as chest CTs, the patient will be asked to hold his or her breath during image capture.

Following the procedure, films of the images are usually printed for the radiologist and referring physician to review. A radiologist can also interpret CT exams on a special computer screen. The procedure time will vary in length depending on the area being imaged. Average study times are from 30 to 60 minutes. Some patients may be concerned about claustrophobia, but the width of the “doughnut” portion of the scanner is such that many patients can be reassured of openness.

The CT image

While traditional x rays image organs in two dimensions, with the possibility that organs in the front of the body are superimposed over those in the back, CT scans allow for a more three-dimensional effect. Some have compared CT images to slices in a loaf of bread. Precise sections of the body can be located and imaged as cross-sectional views. The screen before the technologist shows a computer’s analysis of each section detected by the x-ray beam. Thus, various densities of tissue can be easily distinguished.

Contrast agents

Contrast agents are often used in CT exams and in other radiology procedures to illuminate certain details of anatomy which may not be easily seen. Some contrasts are natural, such as air or water. Other times, a water-based contrast agent is administered for specific diagnostic purposes. Barium sulfate is commonly used in

gastroenterology procedures. The patient may drink this contrast, or receive it in an enema. Oral and rectal contrast are usually given when examining the abdomen or lungs, and not given when scanning the brain or chest. Iodine is the most widely used intravenous contrast agent and is given through an intravenous needle.

If contrast agents are used in the CT exam, these will be administered several minutes before the study begins. Abdominal CT patients may be asked to drink a contrast medium. Some patients may experience a salty taste, flushing of the face, warmth or slight nausea, or **hives** from an intravenous contrast injection. Technologists and radiologists have equipment and training to help patients through these minor reactions and to handle more severe reactions. Severe reactions to contrast are rare, but do occur.

Spiral CT

Spiral CT, also called helical CT, is a newer version of CT scanning which is continuous in motion and allows for three-dimensional recreation of images. For example, traditional CT allows the technologist to take slices at very small and precise intervals one after the other. Spiral CT allows for a continuous flow of images, without stopping the scanner to move to the next image slice. A major advantage of spiral CT is the ability to reconstruct images anywhere along the length of the study area. The procedure also speeds up the imaging process, meaning less time for the patient to lie still. The ability to image contrast more rapidly after it is injected, when it is at its highest level, is another advantage of spiral CT’s high speed.

Some facilities will have both spiral and conventional CT available. Although spiral is more advantageous for many applications, conventional CT is still a superior and precise method for imaging many tissues and structures. The physician will evaluate which type of CT works best for the specific exam purpose.

Preparation

If a contrast medium is administered, the patient may be asked to fast from about four to six hours prior to the procedure. Patients will usually be given a gown (like a typical hospital gown) to be worn during the procedure. All metal and jewelry should be removed to avoid artifacts on the film.

Aftercare

No aftercare is generally required following a CT scan. Immediately following the exam, the technologist will continue to watch the patient for possible adverse contrast reactions. Patients are instructed to advise the

technologist of any symptoms, particularly respiratory difficulty. The site of contrast injection will be bandaged and may feel tender following the exam. Hives may develop later and usually do not require treatment.

Risks

Radiation exposure from a CT scan is similar to, though higher than, that of a conventional x ray. Although this is a risk to pregnant women, the exposure to other adults is minimal and should produce no effects. Although severe contrast reactions are rare, they are a risk of many CT procedures.

Normal results

Normal findings on a CT exam show bone, the most dense tissue, as white areas. Tissues and fat will show as various shades of gray, and fluids will be gray or black. Air will also look black. Intravenous, oral, and rectal contrast appear as white areas. The radiologist can determine if tissues and organs appear normal by the sensitivity of the gray shadows. In CT, the images that can cut through a section of tissue or organ provide three-dimensional viewing for the radiologist and referring physician.

Abnormal results

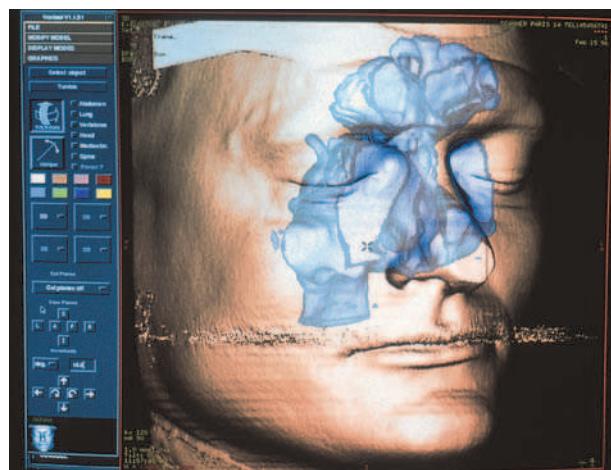
Abnormal results may show different characteristics of tissues within organs. Accumulations of blood or other fluids where they do not belong may be detected. Radiologists can differentiate among types of tumors throughout the body by viewing details of their makeup.

Sinus studies

The increasing availability and lowered cost of CT scanning has led to its increased use in sinus studies, either as a replacement for a sinus x ray or as a follow-up to an abnormal sinus radiograph. The sensitivity of CT allows for location of areas of sinus infection, particularly chronic infection. CT scans can show the extent and location of tiny fractures to the sinus and nasal bones. Foreign bodies in the sinus and nasal area are also easily detected by CT. CT imaging of the sinuses is important in evaluating trauma or disease of the sphenoid bone (the wedge shaped bone at the base of the skull). Sinus tumors will show as shades of gray indicating the difference in their density from that of normal tissues in the area.

Brain studies

The precise differences in density allowed by CT scan can clearly show tumors, strokes, or lesions in the brain area as altered densities. These lighter or darker areas on the



Computerized CT scan of facial sinuses. (Pascal Goetgheluck. Photo Researchers. Reproduced by permission.)

image may indicate a tumor or hematoma within the brain and skull area. Different types of tumors can be identified by the presence of **edema**, by the tissue's density, or by studying blood vessel location and activity. The speed and convenience of CT often allows for detection of hemorrhage before symptoms even occur. Congenital abnormalities in children, such as **hydrocephalus**, may also be confirmed with CT. Hydrocephalus is suggested by enlargement of the fluid structures called ventricles of the brain.

Body scans

The body scan can identify abnormal body structures and organs. Throughout the body, a CT may indicate tumors or cysts, enlarged lymph nodes, abnormal collections of fluids, blood or fat, and metastasis of **cancer**. Tumors resulting from metastasis are different in makeup than primary tumors, or those that originate in the location of study. Fractures or damage to soft tissues and ligaments will be more easily seen on the sensitive images produced by CT scanning, though CT is not usually done for these. Liver conditions, such as **cirrhosis** or abscessed or **fatty liver**, may be observed on the body scan.

CT of the aorta

CT provides the ability to see and measure the thickness of the aortal wall, which is very helpful in diagnosing aortic aneurysms. The use of contrast will help see details within the aorta. In addition, density can identify calcification, and this helps differentiate between acute and chronic problems. An abnormal CT scan may indicate signs of aortic clots. Aortic rupture is suggested by signs such as a hematoma around the aorta or the escape of blood from its cavity.

KEY TERMS

Aneurysm—The bulging of the blood vessel wall. Aortic aneurysms are the most dangerous. Aneurysms can break and cause bleeding.

Contrast (agent, medium)—A substance injected into the body that illuminates certain structures that would otherwise be hard to see on the radiograph (film).

Gantry—A name for the couch or table used in a CT scan. The patient lies on the gantry while it slides into the x-ray scanner portion.

Hematoma—A collection of blood that has escaped from the vessels. It may clot and harden, causing pain to the patient.

Hydrocephalus—A collection of fluid on or around the brain. The pressure from the spinal fluid causes the ventricles to widen.

Metastasis—Secondary cancer, or cancer that has spread from one body organ or tissue to another.

Radiologist—A medical doctor specially trained in radiology (x ray) interpretation and its use in the diagnosis of disease and injury.

Spiral CT—Also referred to as helical CT, this method allows for continuous 360-degree x-ray image capture.

Thoracic—Refers to the chest area. The thorax runs between the abdomen and neck and is encased in the ribs.

Chest scans

In addition to those findings that may indicate aortic aneurysms, chest CT studies can show other problems in the heart and lungs, and distinguish between an **aortic aneurysm** and a tumor adjacent to the aorta. The computer will not only show differences between air, water, tissues, and bone, but will also assign numerical values to the various densities. Coin-sized lesions in the lungs may be indicative of **tuberculosis** or tumors. CT will help distinguish among the two. Enlarged lymph nodes in the chest area may indicate **Hodgkin's disease**. Spiral CT is particularly effective at identifying pulmonary emboli (clots in the lung's blood vessels).

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American College of Radiology. 1891 Preston White Drive, Reston, VA 22091. (800) 227-5463. <<http://www.acr.org>>.

Teresa Norris, RN

Computerized axial tomography see
Computed tomography scans

Concussion

Definition

Concussion is a trauma-induced change in mental status, with confusion and **amnesia**, and with or without a brief loss of consciousness.

Description

A concussion occurs when the head hits or is hit by an object, or when the brain is jarred against the skull, with sufficient force to cause temporary loss of function in the higher centers of the brain. The injured person may remain conscious or lose consciousness briefly, and is disoriented for some minutes after the blow. According to the Centers for Disease Control and Prevention, approximately 300,000 people sustain mild to moderate sports-related brain injuries each year, most of them young men between 16 and 25.

While concussion usually resolves on its own without lasting effect, it can set the stage for a much more serious condition. "Second impact syndrome" occurs when a person with a concussion, even a very mild one, suffers a second blow before fully recovering from the first. The brain swelling and increased intracranial pressure that can result is potentially fatal. More than 20 such cases have been reported since the syndrome was first described in 1984.

Causes and symptoms

Causes

Most concussions are caused by motor vehicle accidents and **sports injuries**. In motor vehicle accidents, concussion can occur without an actual blow to the head. Instead, concussion occurs because the skull suddenly decelerates or stops, which causes the brain to be jarred against the skull. Contact sports, especially football,

hockey, and boxing, are among those most likely to lead to concussion. Other significant causes include falls, collisions, or blows due to bicycling, horseback riding, skiing, and soccer.

The risk of concussion from football is extremely high, especially at the high school level. Studies show that approximately one in five players suffer concussion or more serious brain injury during their brief high-school careers. The rate at the collegiate level is approximately 1 in 20. Rates for hockey players are not known as certainly, but are believed to be similar.

Concussion and lasting brain damage is an especially significant risk for boxers, since the goal of the sport is, in fact, to deliver a concussion to the opponent. For this reason, the American Academy of Neurology has called for a ban on boxing. Repeated concussions over months or years can cause cumulative **head injury**. The cumulative brain injuries suffered by most boxers can lead to permanent brain damage. Multiple blows to the head can cause “punch-drunk” syndrome or **dementia pugilistica**, as evidenced by Muhammed Ali, whose parkinsonism is a result of his career in the ring.

Young children are likely to suffer concussions from falls or collisions on the playground or around the home. **Child abuse** is, unfortunately, another common cause of concussion.

Symptoms

Symptoms of concussion include:

- headache
- disorientation as to time, date, or place
- confusion
- dizziness
- vacant stare or confused expression
- incoherent or incomprehensible speech
- incoordination or weakness
- amnesia for the events immediately preceding the blow
- nausea or vomiting
- double vision
- ringing in the ears

These symptoms may last from several minutes to several hours. More severe or longer-lasting symptoms may indicate more severe brain injury. The person with a concussion may or may not lose consciousness from the blow; if so, it will be for several minutes at the most. More prolonged unconsciousness indicates more severe brain injury.

The severity of concussion is graded on a three-point scale, used as a basis for treatment decisions.

KEY TERMS

Amnesia—A loss of memory that may be caused by brain injury, such as concussion.

Parkinsonism—A neurological disorder that includes a fine tremor, muscular weakness and rigidity, and an altered way of walking.

- Grade 1: no loss of consciousness, transient confusion, and other symptoms that resolve within 15 minutes.
- Grade 2: no loss of consciousness, transient confusion, and other symptoms that require more than 15 minutes to resolve.
- Grade 3: loss of consciousness for any period.

Days or weeks after the accident, the person may show signs of:

- headache
- poor attention and concentration
- memory difficulties
- anxiety
- depression
- sleep disturbances
- light and noise intolerance

The occurrence of such symptoms is called “post-concussion syndrome.”

Diagnosis

It is very important for those attending a person with concussion to pay close attention to the person’s symptoms and progression immediately after the accident. The duration of unconsciousness and degree of confusion are very important indicators of the severity of the injury and help guide the diagnostic process and treatment decisions.

A doctor, nurse, or emergency medical technician may make an immediate assessment based on the severity of the symptoms; a **neurologic exam** of the pupils, coordination, and sensation; and brief tests of orientation, memory, and concentration. Those with very mild concussions may not need to be hospitalized or have expensive diagnostic tests. Questionable or more severe cases may require **computed tomography scan** (CT) or **magnetic resonance imaging** (MRI) scans to look for brain injury.

Treatment

The symptoms of concussion usually clear quickly and without lasting effect, if no further injury is sus-

tained during the healing process. Guidelines for returning to sports activities are based on the severity of the concussion.

A grade 1 concussion can usually be treated with rest and continued observation alone. The person may return to sports activities that same day, but only after examination by a trained professional, and after all symptoms have completely resolved. If the person sustains a second concussion of any severity that same day, he or she should not be allowed to continue contact sports until he or she has been symptom-free, during both rest and activity, for one week.

A person with a grade 2 concussion must discontinue sports activity for the day, should be evaluated by a trained professional, and should be observed closely throughout the day to make sure that all symptoms have completely cleared. Worsening of symptoms, or continuation of any symptoms beyond one week, indicates the need for a CT or MRI scan. Return to contact sports should only occur after one week with no symptoms, both at rest and during activity, and following examination by a physician. Following a second grade 2 concussion, the person should remain symptom-free for two weeks before resuming contact sports.

A person with a grade 3 concussion (involving any loss of consciousness, no matter how brief) should be examined by a medical professional either on the scene or in an emergency room. More severe symptoms may warrant a CT or MRI scan, along with a thorough neurological and physical exam. The person should be hospitalized if any abnormalities are found or if confusion persists. Prolonged unconsciousness and worsening symptoms require urgent neurosurgical evaluation or transfer to a trauma center. Following discharge from professional care, the patient is closely monitored for neurological symptoms which may arise or worsen. If headaches or other symptoms worsen or last longer than one week, a CT or MRI scan should be performed. Contact sports are avoided for one week following unconsciousness of only seconds, and for two weeks for unconsciousness of a minute or more. A person receiving a second grade 3 concussion should avoid contact sports for at least a month after all symptoms have cleared, and then only with the approval of a physician. If signs of brain swelling or bleeding are seen on a CT or MRI scan, the athlete should not return to the sport for the rest of the season, or even indefinitely.

For someone who has sustained a concussion of any severity, it is critically important that he or she avoid the possibility of another blow to the head until well after all symptoms have cleared to prevent second-impact syndrome. The guidelines above are designed to minimize the risk of this syndrome.

Prognosis

Concussion usually leaves no lasting neurological problems. Nonetheless, symptoms of **post-concussion syndrome** may last for weeks or even months.

Studies of concussion in contact sports have shown that the risk of sustaining a second concussion is even greater than it was for the first if the person continues to engage in the sport.

Prevention

Many cases of concussion can be prevented by using appropriate protective equipment. This includes seat belts and air bags in automobiles, and helmets in all contact sports. Helmets should also be worn when bicycling, skiing, or horseback riding. Soccer players should avoid heading the ball when it is kicked at high velocity from close range. Playground equipment should be underlaid with soft material, either sand or special matting.

The value of high-contact sports such as boxing, football, or hockey should be weighed against the high risk of brain injury during a young person's participation in the sport. Steering a child's general enthusiasm for sports into activities less apt to produce head impacts may reduce the likelihood of brain injury.

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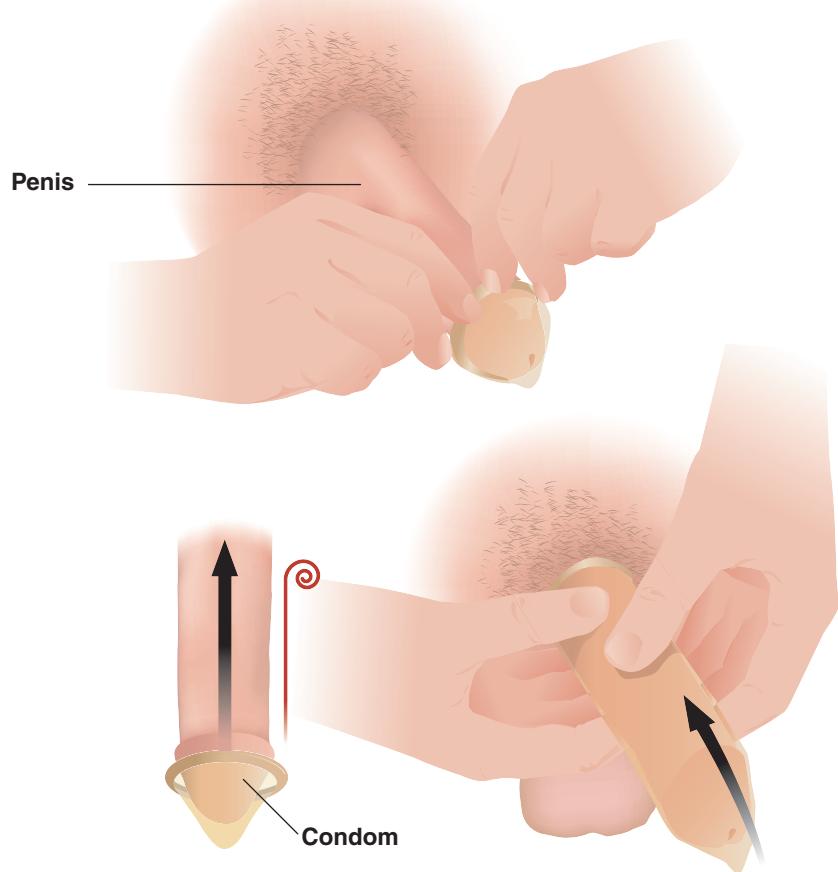
American Academy of Neurology. 1080 Montreal Ave., St. Paul, MN 55116. (612) 695-1940. <<http://www.aan.com>>.

Richard Robinson

Condom

Definition

Male condoms are thin sheaths of latex (rubber), polyurethane (plastic), or animal tissue that are rolled onto an erect penis immediately prior to intercourse. They are commonly called "safes" or "rubbers." Female condoms are made of polyurethane and are inserted into the vaginal



A condom is most effective when it is placed on the penis correctly without trapping air between the penis and the condom.
(Illustration by Argosy, Inc.)

canal before sexual relations. The open end covers the outside of the vagina, and the closed ring fits over the cervix (opening into the uterus). Both types of condoms collect the male semen at ejaculation, acting as a barrier to fertilization. Condoms also perform as barriers to the exchange of bodily fluids and are subsequently an important tool in the prevention of **sexually transmitted diseases** (STDs).

Purpose

Both male and female condoms are used to prevent **pregnancy** and to protect against STDs such as human **immunodeficiency** virus (HIV), **gonorrhea**, chlamydia, and **syphilis**. To accomplish these goals, the condom must be applied and removed correctly.

Precautions

Male and female condoms should not be used together as there is a risk that one of them may come off. The

male condom should not be snug on the tip of the penis. A space of about 0.5 in should be left at the end to avoid the possibility of it breaking during sexual intercourse. The penis must be withdrawn quickly after ejaculation to prevent the condom from falling off as the penis softens. The condom should therefore always be removed while the penis is still erect to prevent the sperm from spilling into the vagina.

Description

Male condoms made from animal tissue and linen have been in use for centuries. Latex condoms were introduced in the late 1800s and gained immediate popularity because they were inexpensive and effective. At that time, they were primarily used to protect against STDs. A common complaint made by many consumers is that condoms reduce penis sensitivity and impair orgasm. Both men and women may develop **allergies** to

KEY TERMS

Ejaculate—To expel semen.

Semen—The thick whitish liquid released from the penis during sexual intercourse. It contains sperm and other secretions.

Sperm or spermatozoa—The part of the semen that is generative—can cause fertilization of the female ovum.

Spermicide—An agent that is destructive to sperm.

Vagina—The genital canal in the female, leading from the vulva to the uterus.

the latex. Consumer interest in female condoms has been slight.

Male condoms may be purchased lubricated, ribbed, or treated with spermicide (a chemical that kills sperm). To be effective, condoms must be removed carefully so as not to “spill” the contents into the vaginal canal. Condoms that leak or break do not provide protection against pregnancy or disease.

If used correctly, male condoms have an effectiveness rate of about 90% for preventing pregnancy, but this rate can be increased to about 99% if used with a spermicide. (Several types of spermicides are available; they can be purchased in the form of contraceptive creams and jellies, foams, or films.) Benefits associated with this type of contraceptive device include easy availability (no prescription is required), convenience of use, and lack of serious side effects. The primary disadvantage is that sexual activity must be interrupted in order to put the condom on.

Female condoms, when used correctly and at every instance of intercourse, were shown to prevent pregnancy in over 95% of women surveyed over the course of six months. When used inconsistently, the female condom was shown to have a failure rate of 21% in the same study. One benefit of the female condom is that it may be inserted immediately before sexual intercourse or up to eight hours prior, so that sexual activity does not need to be interrupted for its insertion. One study performed by a manufacturer of the female condom indicated that 50–75% of couples in numerous countries found the barrier acceptable for use.

Condoms provide better protection against STDs than any other contraceptive method. One study conducted in the 1990s indicated that out of 123 couples with one HIV-positive partner, not one healthy individual con-

tracted the disease when condoms were used with every instance of sexual intercourse. A similar 1993 study showed that out of 171 couples with one HIV-positive partner, all but two individuals were protected against HIV transmission with condom use. In addition to HIV, condoms provide effective transmission against gonorrhea, chlamydia, syphilis, **chancroid**, and **trichomoniasis**. A measure of protection is also provided against **hepatitis B** virus (HBV), human papillomavirus (HPV), and herpes simplex virus (HSV).

Before purchasing a condom, check the expiration date. Prior to use, examine the condom for holes. If a lubricant is going to be used, it should be water soluble because petroleum jellies, such as Vaseline, and other oil based lubricants can weaken latex. It is also important to note that condoms made from animal tissue or plastic are not recommended as a protection against STDs.

Resources

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Stephanie Dionne

Conduct disorder

Definition

Conduct disorder (CD) is a behavioral and emotional disorder of childhood and adolescence. Children with conduct disorder act inappropriately, infringe on the rights of others, and violate the behavioral expectations of others.

Description

CD is present in approximately 9% of boys and 2–9% of girls under the age of 18. Children with conduct disorder act out aggressively and express anger inappropriately. They engage in a variety of antisocial and destructive acts, including violence towards people and animals, destruction of property, lying, stealing,

truancy, and running away from home. They often begin using and abusing drugs and alcohol, and having sex at an early age. Irritability, temper tantrums, and low self-esteem are common personality traits of children with CD.

Causes and symptoms

There are two sub-types of CD, one beginning in childhood and the other in adolescence. There is no known cause. Researchers and physicians suggest that this disease may be caused by the following:

- poor parent-child relationships
- dysfunctional families
- drug abuse
- physical abuse
- poor relationships with other children
- cognitive problems leading to school failures
- brain damage
- biological defects

Difficulty in school is an early sign of potential conduct disorder problems. While the patient's IQ tends to be in the normal range, they can have trouble with verbal and abstract reasoning skills and may lag behind their classmates, and consequently, feel as if they don't "fit in." The frustration and loss of self-esteem resulting from this academic and social inadequacy can trigger the development of CD.

A dysfunctional home environment can be another major contributor to CD. An emotionally, physically, or sexually abusive home environment, a family history of antisocial personality disorder, or parental substance abuse can damage a child's perceptions of himself and put him on a path toward negative behavior. Other less obvious environmental factors can also play a part in the development of conduct disorder. Long-term studies have shown that maternal **smoking** during **pregnancy** may be linked to the development of CD in boys. Animal and human studies point out that nicotine can have undesirable effects on babies. These include altered structure and function of their nervous systems, learning deficits, and behavioral problems. In a study of 177 boys ages seven-12 years, those with mothers who smoked over one half a package of cigarettes daily while pregnant were more apt to have a CD than those with mothers who did not smoke.

Other conditions that may cause or co-exist with CD include **head injury**, substance abuse disorder, major depressive disorder, and attention deficit hyperactivity disorder (**ADHD**). Thirty to fifty percent of children

diagnosed with ADHD, a disorder characterized by a persistent pattern of inattention and/or hyperactivity, also have CD.

CD is defined as a repetitive behavioral pattern of violating the rights of others or societal norms. Three of the following criteria, or symptoms, are required over the previous 12 months for a diagnosis of CD (one of the three must have occurred in the past six months):

- bullies, threatens, or intimidates others
- picks fights
- has used a dangerous weapon
- has been physically cruel to people
- has been physically cruel to animals
- has stolen while confronting a victim (for example, mugging or extortion)
- has forced someone into sexual activity
- has deliberately set a fire with the intention of causing damage
- has deliberately destroyed property of others
- has broken into someone else's house or car
- frequently lies to get something or to avoid obligations
- has stolen without confronting a victim or breaking and entering (e.g., shoplifting or forgery)
- stays out at night; breaks curfew (beginning before 13 years of age)
- has run away from home overnight at least twice (or once for a lengthy period)
- is often truant from school (beginning before 13 years of age).

Diagnosis

CD is diagnosed and treated by a number of social workers, school counselors, psychiatrists, and psychologists. Genuine diagnosis may require psychiatric expertise to rule out such conditions as **bipolar disorder** or ADHD. A comprehensive evaluation of the child should ideally include interviews with the child and parents, a full social and medical history, a cognitive evaluation, and a psychiatric exam. One or more clinical inventories or scales may be used to assess the child for conduct disorder—including the Youth Self-Report, the Overt Aggression Scale (OAS), Behavioral Assessment System for Children (BASC), Child Behavior Checklist (CBCL), and Diagnostic Interview Schedule for Children (DISC). The tests are verbal and/or written and are administered in both hospital and outpatient settings.

KEY TERMS

ADHD—Attention deficit hyperactivity disorder; a disorder characterized by a persistent pattern of inattention and/or hyperactivity.

Major depressive disorder—A mood disorder characterized by profound feelings of sadness or despair.

Treatment

Treating conduct disorder requires an approach that addresses both the child and his environment. Behavioral therapy and psychotherapy can help a child with CD to control his anger and develop new coping skills. Family **group therapy** may also be effective in some cases. Parents should be counseled on how to set appropriate limits with their child and be consistent and realistic when disciplining. If an abusive home life is at the root of the conduct problem, every effort should be made to move the child into a more supportive environment. Parent training programs are increasing in number.

For children with coexisting ADHD, substance abuse, depression, or **learning disorders**, treating these conditions first is preferred, and may result in a significant improvement to the CD condition. In all cases of CD, treatment should begin when symptoms first appear. Recent studies have shown Ritalin to be a useful drug for both ADHD and CD.

When aggressive behavior is severe, mood stabilizing medication, including lithium (Cibalith-S, Eskalith, Lithane, Lithobid, Lithonate, Lithotabs), carbamazepine (Tegretol, Atretol), and propranolol (Inderal), may be an appropriate option for treating the aggressive symptoms. However, placing the child into a structured setting or treatment program such as a psychiatric hospital may be just as beneficial for easing aggression as medication.

Prognosis

The prognosis for children with CD is not bright. Follow-up studies of conduct disordered children have shown a high incidence of antisocial personality disorder, affective illnesses, and chronic criminal behavior later in life. However, proper treatment of co-existing disorders, early identification and intervention, and long-term support may improve the outlook significantly.

Prevention

A supportive, nurturing, and structured home environment is believed to be the best defense against CD. Children with learning disabilities and/or difficulties in school should get immediate and appropriate academic assistance. Addressing these problems when they first appear helps to prevent the frustration and low self-esteem that may lead to CD later on.

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American Academy of Child and Adolescent Psychiatry (AACAP). 3615 Wisconsin Ave. NW, Washington, DC 20016. (202) 966-7300. <<http://www.aacap.org>>.

Children and Adults with Attention Deficit Disorder (CH.A.D.D.). 8181 Professional Place, Suite 201

Paula Anne Ford-Martin

Conductive hearing loss see **Hearing loss**

Condylomata acuminata see **Genital warts**

Cone biopsy see **Cervical conization**

Congenital adrenal hyperplasia

Definition

Congenital adrenal hyperplasia is (CAH) a genetic disorder characterized by a deficiency in the hormones cortisol and aldosterone and an over-production of the

hormone androgen, which is present at birth and affects sexual development.

Description

CAH is a form of adrenal insufficiency in which the enzyme that produces two important adrenal steroid hormones, cortisol and aldosterone, is deficient. Because cortisol production is impeded, the adrenal gland instead overproduces androgens (male steroid hormones). Females with CAH are born with an enlarged clitoris and normal internal reproductive tract structures. Males have normal genitals at birth. CAH causes abnormal growth for both sexes; patients will be tall as children and short as adults. Females develop male characteristics, and males experience premature sexual development.

In its most severe form, called salt-wasting CAH, a life-threatening adrenal crisis can occur if the disorder is untreated. Adrenal crisis can cause **dehydration**, **shock**, and **death** within 14 days of birth. There is also a mild form of CAH that occurs later in childhood or young adult life in which patients have partial enzyme deficiency.

CAH, a genetic disorder, is the most common adrenal gland disorder in infants and children, occurring in one in 10,000 total births worldwide. It affects both females and males. It is also called adrenogenital syndrome.

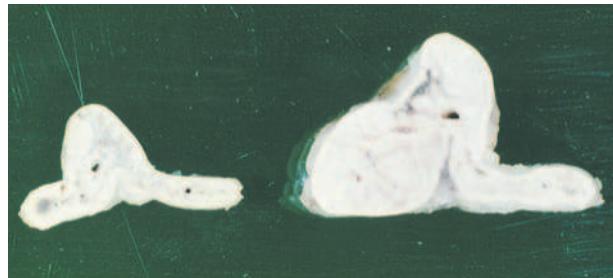
Causes and symptoms

CAH is an inherited disorder. It is a recessive disease, which means that a child must inherit one copy of the defective gene from each parent who is a carrier; when two carriers have children, each **pregnancy** carries a 25% risk of producing an affected child.

In females, CAH produces an enlarged clitoris at birth and masculinization of features as the child grows, such as deepening of the voice, facial hair, and failure to menstruate or abnormal periods at **puberty**. Females with severe CAH may be mistaken for males at birth. In males, the genitals are normal at birth, but the child becomes muscular, the penis enlarges, pubic hair appears, and the voice deepens long before normal puberty, sometimes as early as two to three years of age.

In the severe salt-wasting form of CAH, newborns may develop symptoms shortly after birth, including vomiting, dehydration, electrolyte (a compound such as sodium or calcium that separates to form ions when dissolved in water) changes, and cardiac arrhythmia.

In the mild form of CAH, which occurs in late childhood or early adulthood, symptoms include premature development of pubic hair, irregular menstrual periods,



Adrenal cortical hyperplasia. The adrenal on the right is normal, that on the left shows hyperplasia. (Photo Researchers, Inc. Reproduced by permission.)

unwanted body hair, or severe **acne**. However, sometimes there are no symptoms.

Diagnosis

CAH is diagnosed by a careful examination of the genitals and blood and urine tests that measure the hormones produced by the adrenal gland. A number of states in the United States perform a hormonal test (a heel prick blood test) for CAH and other inherited diseases within a few days of birth. In questionable cases, **genetic testing** can provide a definitive diagnosis. For some forms of CAH, prenatal diagnosis is possible through chronic villus sampling in the first trimester and by measuring certain hormones in the amniotic fluid during the second trimester.

Treatment

The goal of treatment for CAH is to return the androgen levels to normal. This is usually accomplished through drug therapy, although surgery is an alternative. Lifelong treatment is required.

Drug therapy consists of a cortisol-like steroid medication called a glucocorticoid. Oral hydrocortisone is prescribed for children, and prednisone or dexamethasone is prescribed for older patients. For patients with salt-wasting CAH, fludrocortisone, which acts like aldosterone (the missing hormone), is also prescribed. Infants and small children may also receive salt tablets, while older patients are told to eat salty foods. Medical therapy achieves hormonal balance most of the time, but CAH patients can have periods of fluctuating hormonal control that lead to increases in the dose of steroids prescribed. Side effects of steroids include stunted growth. Steroid therapy should not be suddenly stopped, since adrenal insufficiency results.

Patients with CAH should see a pediatric endocrinologist frequently. The endocrinologist will assess height,

KEY TERMS

Adrenal glands—The two endocrine glands located above the kidney that secrete hormones and epinephrine.

Aldosterone—A hormone secreted by the adrenal glands that is important for maintaining salt and water balance in the body.

Androgens—Steroid hormones that cause masculinization.

Congenital—Present at birth.

Cortisol—A steroid hormone secreted by the adrenal cortex that is important for maintenance of body fluids, electrolytes, and blood sugar levels.

Hormone—A chemical messenger produced by the endocrine glands or certain other cells. Hormones are usually carried in the blood stream and regulate some metabolic activities.

Steroids—Hormones, including aldosterone, cortisol, and androgens, derived from cholesterol that share a four-ring structure.

weight, and blood pressure, and order an annual x ray of the wrist (to assess bone age), as well as assess blood hormone levels. CAH patients with the milder form of the disorder are usually effectively treated with hydrocortisone or prednisone, if they need medical treatment at all.

Females with CAH who have masculine external genitalia require surgery to reconstruct the clitoris and/or vagina. This is usually performed between the ages of one and three.

An experimental type of drug therapy—a three-drug combination, with an androgen blocking agent (flutamide), an aromatase inhibitor (testolactone), and low dose hydrocortisone—is currently being studied by physicians at the National Institutes of Health. Preliminary results are encouraging, but it will be many years before the safety and effectiveness of this therapy is fully known.

Adrenalectomy, a surgical procedure to remove the adrenal glands, is a more radical treatment for CAH. It was widely used before the advent of steroids. Today, it is recommended for CAH patients with little or no enzyme activity and can be accomplished by **laparoscopy**. This is a minimally invasive type of surgery done through one or more small 1 in (2.5 cm) incisions and a laparoscope, an instrument with a fiber-optic light containing a tube with

openings for surgical instruments. Adrenalectomy is followed by hormone therapy, but in lower doses than CAH patients not treated surgically receive.

Prognosis

CAH can be controlled and successfully treated in most patients as long as they remain on drug therapy.

Prevention

Prenatal therapy, in which a pregnant woman at risk for a second CAH child is given dexamethasone to decrease secretion of androgens by the adrenal glands of the female fetus, has been in use for about 10 years. This therapy is started in the first trimester when fetal adrenal production of androgens begins, but before prenatal diagnosis is done that would provide definitive information about the sex of the fetus and its disease status. This means that a number of fetuses are exposed to unnecessary steroid treatment in order to prevent the development of male-like genitals in female fetuses with CAH. Several hundred children have undergone this treatment with no major adverse effects, but its long-term risks are unknown. Since there is very little data on the effectiveness and safety of prenatal therapy, it should only be offered to patients who clearly understand the risks and benefits and who are capable of complying with strict monitoring and follow-up throughout pregnancy and after the child is born.

Parents with a family history of CAH, including a child who has CAH, should seek **genetic counseling**. Genetic testing during pregnancy can provide information on the risk of having a child with CAH.

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Jennifer Sisk

Congenital amputation

Definition

Congenital amputation is the absence of a fetal limb or fetal part at birth. This condition may be the result of the constriction of fibrous bands within the membrane that surrounds the developing fetus (amniotic band syndrome) or the exposure to substances known to cause **birth defects** (teratogenic agents). Other factors, including genetics, may also play a role.

Description

An estimated one in 2,000 babies are born with all or part of a limb missing, ranging from a missing part of a finger to the absence of both arms and both legs. Congenital amputation is the least common reason for amputation. However, there are occasional periods in history where the number of congenital amputations increased. For example, the thalidomide tragedy of the early 1960s occurred after pregnant mothers in western Europe were given a tranquilizer containing the drug. The result was a drastic increase in the number of babies born with deformed limbs. In this example, the birth defect usually presented itself as very small, deformed versions of normal limbs. More recently, birth defects as a result of radiation exposure near the site of the Chernobyl disaster in Russia have left numerous children with malformed or absent limbs.

Causes and symptoms

The exact cause of congenital amputations is unknown. However, according to the March of Dimes, most birth defects have one or more genetic factors and one or more environmental factors. It is also known that most birth defects occur in the first three months of **pregnancy**, when the organs of the fetus are forming. Within these crucial first weeks, frequently prior to when a woman is aware of the pregnancy, the developing fetus is most susceptible to substances that can cause birth defects (teratogens). Exposure to teratogens can cause congenital amputation. In other cases, tight amniotic bands may constrict the developing fetus, preventing a

limb from forming properly, if at all. It is estimated that this amniotic band syndrome occurs in between one in 12,000 and one in 15,000 live births.

An infant with congenital amputation may be missing an entire limb or just a portion of a limb. Congenital amputation resulting in the complete absence of a limb beyond a certain point (and leaving a stump) is called transverse deficiency or amelia. Longitudinal deficiencies occur when a specific part of a limb is missing; for example, when the fibula bone in the lower leg is missing, but the rest of the leg is intact. Phocomelia is the condition in which only a mid-portion of a limb is missing, as when the hands or feet are attached directly to the trunk.

Diagnosis

Many cases of congenital amputation are not diagnosed until the baby is born. Ultrasound examinations may reveal the absence of a limb in some developing fetuses, but routine ultrasounds may not pick up signs of more subtle defects. However, if a doctor suspects that the fetus is at risk for developing a limb deficiency (for example, if the mother has been exposed to radiation), a more detailed ultrasound examination may be performed.

Treatment

Successful treatment of a child with congenital amputation involves an entire medical team, including a pediatrician, an orthopedist, a psychiatrist or psychologist, a prosthetist (an expert in making prosthetics, or artificial limbs), a social worker, and occupational and physical therapists. The accepted method of treatment is to fit the child early with a functional prosthesis because this leads to normal development and less wasting away (atrophy) of the muscles of the limbs present. However, some parents and physicians believe that the child should be allowed to learn to play and perform tasks without a prosthesis, if possible. When the child is older, he or she can be involved in the decision of whether or not to be fitted for a prosthesis.

Recently, there have been cases in which physicians have detected amniotic band constriction interfering with limb development fairly early in its course. In 1997, doctors at the Florida Institute for Fetal Diagnosis and Therapy reported two cases in which minimally invasive surgery freed constricting amniotic bands and preserved the affected limbs.

Alternative treatment

Prevention of birth defects begins with building the well-being of the mother before pregnancy. Prenatal care

KEY TERMS

Prosthesis—An artificial replacement for a missing part of the body.

Teratogen—Any substance, agent, or process that interferes with normal prenatal development, causing the formation of one or more developmental abnormalities of the fetus.

should be strong and educational so that the mother understands both her genetic risks and her environmental risks. Several disciplines in alternative therapy also recommend various supplements and **vitamins** that may reduce the chances of birth defects. If a surgical procedure is planned, naturopathic and homeopathic pre- and post-surgical therapies can speed recovery.

Prognosis

A congenital limb deficiency has a profound effect on the life of the child and parents. However, occupational therapy can help the child learn to accomplish many tasks. In addition, some experts believe that early fitting of a prosthesis will enhance acceptance of the prosthesis by the child and parents.

Prevention

Studies have suggested that a multivitamin including **folic acid** may reduce birth defects, including congenital abnormalities. **Smoking**, drinking alcohol, and eating a poor diet while pregnant may increase the risk of congenital abnormalities. Daily, heavy exposure to chemicals may be dangerous while pregnant.

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Jeffrey P. Larson, RPT

Congenital bladder diverticulum see
Congenital bladder anomalies

Congenital bladder anomalies

Definition

The two most common congenital bladder abnormalities are exstrophy and congenital diverticula. An exstrophic bladder is one that is open to the outside and turned inside-out, so that its inside is visible at birth, protruding from the lower abdomen. A diverticulum is an extension of a hollow organ, usually shaped like a pouch with a narrow opening.

Description

During fetal development, folds enclose tissues and organs and eventually fuse at the edges to form sealed compartments. Both in the front and the back, folds eventually become major body structures. In the back, the entire spinal column folds in like a pipe wrapped in a pillow. In the front, the entire lower urinary system is folded in.

- Exstrophy of the bladder represents a failure of this folding process to complete itself, so the organs form with more or less of their front side missing and open to the outside. At the same time, the front of the pelvic bone is widely separated. The abdominal wall is open, too. In fact, the defect often extends all the way to the penis in the male or splits the clitoris in the female.
- A congenital bladder diverticulum represent an area of weakness in the bladder wall through which extrudes some of the lining of the bladder. (A small balloon squeezed in a fist will create diverticula-like effect between the fingers.) Bladder diverticula may be multiple, and they often occur at the ureterovesical junction—the entrance of the upper urinary system into the bladder. In this location, they may cause urine to reflux into the ureter and kidney, leading to infection and possible kidney damage.

Causes and symptoms

As with many **birth defects**, the causes are not well known. Lack of prenatal care and **nutrition** has been linked to many birth defects; however, beyond the avoidance of known teratogens (anything that can cause a birth defect), there is little prevention possible. Exstrophy is rare, occurring in about one in 40,000 births. Diverticula are more common, but less serious.

If left untreated, the patient with bladder exstrophy will have no control over urination and is more likely to develop **bladder cancer**. Diverticula, particularly if it causes urine reflux, may lead to chronic infection and its subsequent consequences.

Diagnosis

A major consideration with congenital abnormalities is that they tend to be multiple. Further, each one is unique in its extent and severity. Exstrophy can involve the rectum and large bowel and coexist with hernias. The obvious bladder exstrophy seen at birth will prompt immediate action and a search for other anomalies.

Diverticula are not visible and will be detected only if they cause trouble. They are usually found in an examination for the cause of recurring urinary infections. X rays of the urinary system or a **cystoscopy** (examination with a telescope-like instrument) will identify them. Often, the two procedures are done together: a urologist will perform the cystoscopy, then a radiologist will instill a contrast agent into the bladder and take x rays.

Treatment

Surgery is necessary and can usually produce successful results. If possible, the surgery must be done within 48 hours of birth. Prior to surgery, the exposed organs must be protected and all related defects identified and managed. Delay in the surgery leads to the frequent need to divert the urine into the bowel because the partially repaired bladder cannot control the flow. After surgery, the likelihood of infection requires monitoring.

Alternative treatment

After surgery ongoing precautions to reduce frequency of infection may need to be used. Cranberry juice has the ability to keep bacteria from adhering to the membranes and can help prevent infection whenever there is increased risk. There are botanical and homeopathic treatments available, however consultation by a trained practitioner is recommended before treatment.

Prognosis

With immediate surgery, three-quarters of patients can be successfully repaired. They will have control of their urine and no long-term consequences. The rate of infection is greater for those with congenital bladder anomalies, since any abnormality in the urinary system predisposes it to invasion by bacteria.

Prevention

Birth defects often have no precisely identified cause, therefore prevention is limited to general measures such as early and continuous prenatal care, appropriate nutrition, and a healthy lifestyle.

KEY TERMS

Congenital—Present at birth.

Cystoscopy—Examination of the urinary bladder with a thin telescope-like instrument.

Exstrophy—Being turned inside out combined with being outside the body.

Diverticulum—A pouch extending from a hollow organ.

Radiologist—A physician who specializes in creating images of the internal organs of the body.

Teratogen—Any agent that can cause birth defects.

Ureter—The tube that transports urine from the kidney to the bladder.

Ureterovesical junction—The joining of the ureter to the bladder.

Urologist—A surgeon who specializes in diseases of the urinary system.

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J. Ricker Polsdorfer, MD

Congenital brain defects

Definition

Congenital brain defects are a group of disorders of brain development.

Description

Brain development begins shortly after conception and continues throughout the growth of a fetus. A com-

plex genetic program coordinates the formation, growth, and migration of billions of neurons, or nerve cells, and their development into discrete, interacting brain regions. Interruption of this program, especially early in development, can cause structural defects in the brain. In addition, normal brain formation requires proper development of the surrounding skull, and skull defects may lead to brain malformation. Congenital brain defects may be caused by inherited genetic defects, spontaneous mutations within the genes of the embryo, or effects on the embryo due to the mother's infection, trauma, or drug use.

Early on in development, a flat strip of tissue along the back of the fetus rolls up to form a tube. This so-called "neural tube" develops into the spinal cord, and at one end, the brain. Closure of the tube is required for subsequent development of the tissue within. Anencephaly (literally "without brain"), results when the topmost portion of the tube fails to close. Anencephaly is the most common severe malformation seen in stillborn births. It is about four times more common in females than males. Anencephaly is sometimes seen to run in families, and for parents who have conceived one anencephalic fetus, the risk of a second is as high as 5%. Fewer than half of babies with anencephaly are born alive, and survival beyond the first month is rare.

Encephalocele is a protrusion of part of the brain through a defect in the skull. The most common site for encephalocele is along the front-to-back midline of the skull, usually at the rear, although frontal encephaloceles are more common among Asians. Pressure within the skull pushes out cranial tissue. The protective layer over the brain, the meninges, grows to cover the protrusion, as does skin in some cases. Defects in skull closure are thought to cause some cases of encephalocele, while defects in neural tube closure may cause others. Encephaloceles may be small and contain little or no brain tissue, or may be quite large and contain a significant fraction of the brain.

Failure of neural-tube closure below the level of the brain prevents full development of the surrounding vertebral bones and leads to **spina bifida**, or a divided spinal column. Incomplete closure causes protrusion of the spinal cord and meninges, called meningomyelocele. Some cases of spina bifida are accompanied by another defect at the base of the brain, known as the Arnold-Chiari malformation or Chiari II malformation. For reasons that are unclear, part of the cerebellum is displaced downward into the spinal column. Symptoms may be present at birth or delayed until early childhood.

The Dandy-Walker malformation is marked by incomplete formation, or absence of, the central section of the cerebellum, and the growth of cysts within the lowest of the brain's ventricles. The ventricles are fluid-filled cavities within the brain, through which cerebrospinal fluid (CSF) normally circulates. The cysts may block the exit of the fluid, causing **hydrocephalus**. Symptoms may be present at birth or delayed until early childhood.

Soon after closure of the neural tube, the brain divides into two halves, or hemispheres. Failure of division is termed holoprosencephaly (literally "whole forebrain"). Holoprosencephaly is almost always accompanied by facial and cranial deformities along the midline, including cleft lip, cleft palate, fused eye sockets and a single eye (cyclopia), and deformities of the limbs, heart, gastrointestinal tract, and other internal organs. Most infants are either stillborn or die soon after birth. Survivors suffer from severe neurological impairments.

The normal ridges and valleys of the mature brain are formed after cells from the inside of the developing brain migrate to the outside and multiply. When these cells fail to migrate, the surface remains smooth, a condition called lissencephaly ("smooth brain"). Lissencephaly is often associated with facial abnormalities including a small jaw, a high forehead, a short nose, and low-set ears.

If damaged during growth, especially within the first 20 weeks, brain tissue may stop growing, while tissue around it continues to form. This causes an abnormal cleft or groove to appear on the surface of the brain, called schizencephaly (literally "split brain"). This cleft should not be confused with the normal wrinkled brain surface, nor should the name be mistaken for **schizophrenia**, a mental disorder. Generalized destruction of tissue or lack of brain development may lead to hydranencephaly, in which cerebrospinal fluid fills much of the space normally occupied by the brain. Hydranencephaly is distinct from hydrocephalus, in which CSF accumulates within a normally-formed brain, putting pressure on it and possibly causing skull expansion.

Excessive brain size is termed megalencephaly (literally "big brain"). Megalencephaly is defined as any brain size above the 98th percentile within the population. Some cases are familial, and may be entirely benign. Others are due to metabolic or neurologic disease. The opposite condition, microcephaly, may be caused by failure of the brain to develop, or by intrauterine infection, drug toxicity, or brain trauma.

Causes and symptoms

Causes

Congenital brain defects may have genetic, infectious, toxic, or traumatic causes. In most cases, no certain cause can be identified.

GENETIC CAUSES. Some brain defects are caused by trisomy, the inclusion of a third copy of a chromosome normally occurring in pairs. Most trisomies occur because of improper division of the chromosomes during formation of eggs or sperm. Trisomy of chromosome 9 can cause some cases of Dandy-Walker and Chiari II malformation. Some cases of holoprosencephaly are caused by trisomy of chromosome 13, while others are due to abnormalities in chromosomes 7 or 18. Individual gene defects, either inherited or spontaneous, are responsible for other cases of congenital brain malformations.

DRUGS. Drugs known to cause congenital brain defects when used by the mother during critical developmental periods include:

- anticonvulsant drugs
- retinoic acid and tretinoin
- warfarin
- alcohol
- cocaine

OTHER. Other causes of congenital brain defects include:

- intrauterine infections, including cytomegalovirus, **rubella**, herpes simplex, and varicella zoster
- maternal **diabetes mellitus**
- maternal **phenylketonuria**
- fetal trauma

Symptoms

Besides the features listed above, symptoms of congenital brain defects may include:

- Chiari II malformation: impaired swallowing and gag reflex, loss of the breathing reflex, facial **paralysis**, uncontrolled eye movements (**nystagmus**), impaired balance and gait.
- Dandy-Walker malformation: symptoms of hydrocephalus, lack of muscle tone or “floppiness,” seizures, spasticity, deafness, irritability, **visual impairment**, deterioration of consciousness, paralysis.
- Lissencephaly: lack of muscle tone, seizures, developmental delay, spasticity, **cerebral palsy**.
- Hydranencephaly: irritability, spasticity, seizures, temperature oscillations.

KEY TERMS

Amniocentesis—Removal of fluid from the sac surrounding a fetus for purposes of diagnosis.

Cerebrospinal fluid—Fluid produced within the brain for nutrient transport and structural purposes. CSF circulates through the ventricles, open spaces within the brain, and drains through the membranes surrounding the brain.

Congenital—Defect present at birth.

Fetus—The unborn human, developing in a woman’s uterus, from the eighth week after fertilization to birth.

- megalencephaly due to neurological or metabolic disease: **mental retardation**, seizures.

Diagnosis

Congenital brain defects are diagnosed either from direct **physical examination** or imaging studies including **computed tomography scans** (CT) and **magnetic resonance imaging** (MRI) scans. **Electroencephalography** (EEG) may be used to reveal characteristic abnormalities.

Prenatal diagnosis of neural tube defects causing anencephaly or meningocele is possible through ultrasound examination and maternal blood testing for alpha-fetoprotein, which is almost always elevated. Ultrasound can also be used to diagnose Dandy-Walker and Chiari II malformations. **Amniocentesis** may reveal trisomies or other chromosomal abnormalities.

Treatment

Meningocele may be treated with surgery to close the open portion of the spinal cord. Surgery for encephalocele is possible only if there is a minimal amount of brain tissue protruding. Malformations associated with hydrocephalus (Dandy-Walker, Chiari II, and some cases of hydranencephaly) may be treated by installation of a drainage shunt for cerebrospinal fluid. Drugs may be used to treat some symptoms of brain defects, including seizures and spasticity.

Prognosis

Most congenital brain defects carry a very poor prognosis. Surgical treatment of meningocele and encephalocele may be successful, with lasting neurologi-

cal deficiencies that vary in severity. Early treatment of hydrocephalus may prevent more severe brain damage.

Prevention

Some cases of congenital brain defects can be prevented with good maternal **nutrition**, including **folic acid** supplements. Folic acid is a vitamin that has been shown to reduce the incidence of neural tube defects. Pregnant women should avoid exposure to infection, especially during the first trimester. Abstention from drugs and alcohol during **pregnancy** may reduce risk. **Genetic counseling** is advisable for parents who have had one child with anencephaly, since the likelihood of having another is increased.

Resources

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Richard Robinson

Congenital defects see **Birth defects**

Congenital hip dysplasia

Definition

A condition of abnormal development of the hip, resulting in hip joint instability and potential dislocation of the thigh bone from the socket in the pelvis. This condition has been more recently termed developmental hip dysplasia, as it often develops over the first few weeks, months, or years of life.

Description

Congenital hip dysplasia is a disorder in children that is either present at birth or shortly thereafter. During gestation, the infant's hip should be developing with the head of the thigh bone (femur) sitting perfectly centered in its shallow socket (acetabulum). The acetabulum should cover the head of the femur as if it were a ball sitting inside of a cup. In the event of congenital hip dysplasia, the development of the acetabulum in an infant

allows the femoral head to ride upward out of the joint socket, especially when weight bearing begins.

Causes and symptoms

Clinical studies show a familial tendency toward hip dysplasia, with more females affected than males. This disorder is found in many cultures around the world. However, statistics show that the Native American population has a high incidence of hip dislocation. This has been documented to be due to the common practice of swaddling and using cradleboards for restraining the infants. This places the infant's hips into extreme adduction (brought together). The incidence of congenital hip dysplasia is also higher in infants born by caesarian and breech position births. Evidence also shows a greater chance of this hip abnormality in the first born compared to the second or third child. Hormonal changes within the mother during **pregnancy**, resulting in increased ligament laxity, is thought to possibly cross over to the placenta and cause the baby to have lax ligaments while still in the womb. Other symptoms of complete dislocation include a shortening of the leg and limited ability to abduct the leg.

Diagnosis

Because the abnormalities of this hip problem often vary, a thorough **physical examination** is necessary for an accurate diagnosis of congenital hip dysplasia. The hip disorder can be diagnosed by moving the hip to determine if the head of the femur is moving in and out of the hip joint. One specific method, called the Ortolani test, begins with each of the examiners' hands around the infant's knees, with the second and third fingers pointing down the child's thigh. With the legs abducted (moved apart), the examiner may be able to discern a distinct clicking sound with motion. If symptoms are present with a noted increase in abduction, the test is considered positive for hip joint instability. It is important to note this test is only valid a few weeks after birth.

The Barlow method is another test performed with the infant's hip brought together with knees in full bent position. The examiner's middle finger is placed over the outside of the hipbone while the thumb is placed on the inner side of the knee. The hip is abducted to where it can be felt if the hip is sliding out and then back in the joint. In older babies, if there is a lack of range of motion in one hip or even both hips, it is possible that the movement is blocked because the hip has dislocated and the muscles have contracted in that position. Also in older infants, hip dislocation is evident if one leg looks shorter than the other.

X-ray films can be helpful in detecting abnormal findings of the hip joint. X rays may also be helpful in finding the proper positioning of the hip joint for treatments of

casting. Ultrasound has been noted as a safe and effective tool for the diagnosis of congenital hip dysplasia. Ultrasound has advantages over x rays, as several positions are noted during the ultrasound procedure. This is in contrast to only one position observed during the x ray.

Treatment

The objective of treatment is to replace the head of the femur into the acetabulum and, by applying constant pressure, to enlarge and deepen the socket. In the past, stabilization was achieved by placing rolled cotton diapers or a pillow between the thighs, thereby keeping the knees in a frog like position. More recently, the Pavlik harness and von Rosen splint are commonly used in infants up to the age of six months. A stiff shell cast may be used, which achieves the same purpose, spreading the legs apart and forcing the head of the femur into the acetabulum. In some cases, in older children between six to 18 months, surgery may be necessary to reposition the joint. Also at this age, the use of closed manipulation may be applied successfully, by moving the leg around manually to replace joint. Operations are not only performed to reduce the dislocation of the hip, but also to repair a defect in the acetabulum. A cast is applied after the operation to hold the head of the femur in the correct position. The use of a home **traction** program is now more common. However, after the age of eight years, surgical procedures are primarily done for **pain** reduction measures only. Total hip surgeries may be inevitable later in adulthood.

Alternative treatment

Nonsurgical treatments include **exercise** programs, orthosis (a force system, often involving braces), and medications. A physical therapist may develop a program that includes strengthening, range-of-motion exercises, pain control, and functional activities. **Chiropractic** medicine may be helpful, especially the procedures of closed manipulations, to reduce the dislocated hip joint.

Prognosis

Unless corrected soon after birth, abnormal stresses cause malformation of the developing femur, with a characteristic limp or waddling gait. If cases of congenital hip dysplasia go untreated, the child will have difficulty walking, which could result in life-long pain. In addition, if this condition goes untreated, the abnormal hip positioning will force the acetabulum to locate to another position to accommodate the displaced femur.

Prevention

Prevention includes proper prenatal care to determine the position of the baby in the womb. This may be

KEY TERMS

Acetabulum—The large cup-shaped cavity at the junction of pelvis and femur or thigh bone.

Orthosis—A force system designed to control or correct or compensate for a bone deformity, deforming forces, or forces absent from the body.

helpful in preparing for possible breech births associated with hip problems. Avoiding excessive and prolonged infant hip adduction may help prevent strain on the hip joints. Early diagnosis remains an important part of prevention of congenital hip dysplasia.

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ORGANIZATIONS

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (914) 428-7100. <<http://www.modimes.org>>.

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Congenital lobar emphysema

Definition

Congenital lobar emphysema is a chronic disease that causes respiratory distress in infants.

Description

Congenital lobar emphysema, also called infantile lobar emphysema, is a respiratory disease that occurs in infants when air enters the lungs but cannot leave easily. The lungs become over-inflated, causing respiratory function to decrease and air to leak out into the space around the lungs.

KEY TERMS

Congenital—A disease or condition that is present at birth.

Emphysema—A condition in which the air sacs in the lungs become overinflated, causing a decrease in respiratory function.

Lobar—Relating to a lobe, a rounded projecting part of the lungs.

Half of the cases of congenital lobar emphysema occur in the first four weeks of life, and three-quarters occur in infants less than six months old. Congenital lobar emphysema is more common in boys than in girls.

Each person has two lungs, right and left. The right lung is divided into three sections, called lobes, and the left lung into two lobes. Congenital lobar emphysema usually affects only one lobe, and this is usually an upper lobe. It occurs most frequently in the left upper lobe, followed by the right middle lobe.

Causes and symptoms

The cause of congenital lobar emphysema often cannot be identified. The airway may be obstructed or the infant's lungs may not have developed properly. Congenital lobar emphysema is almost never of genetic origin.

Symptoms of congenital lobar emphysema include:

- shortness of breath
- wheezing
- lips and fingernail beds that have a bluish tinge

Diagnosis

Congenital lobar emphysema is usually identified within the first two weeks of the infant's life. It is diagnosed by respiratory symptoms and a **chest x ray**, which shows the over-inflation of the affected lobe and may show a blocked air passage.

Treatment

For infants with no, mild, or intermittent symptoms, no treatment is necessary. For more serious cases of congenital lobar emphysema, surgery is necessary, usually a lobectomy to remove the affected lung lobe.

Alternative treatment

Alternative treatments that may be helpful for congenital lobar emphysema are aimed at supporting and

strengthening the patient's respiratory function. Vitamin and mineral supplementation may be recommended as may herbal remedies such as lobelia (*Lobelia inflata*) that strengthen the lungs and enhance their elasticity. Homeopathic constitutional care may also be beneficial for this condition.

Prognosis

Surgery for congenital lobar emphysema has excellent results.

Prevention

Congenital lobar emphysema cannot be prevented.

Resources

BOOKS

Baum, Gerald L., et al., eds. *Textbook of Pulmonary Diseases*. Philadelphia: Lippincott-Raven, 1998.

ORGANIZATIONS

American Lung Association. 1740 Broadway, New York, NY 10019. (800) 586-4872. <<http://www.lungusa.org>>.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

National Jewish Center for Immunology and Respiratory Medicine. 1400 Jackson St., Denver, CO 80206. (800) 222-5864. <<http://www.nationaljewish.org/main.html>>.

Lori De Milto

Congenital megacolon see **Hirschsprung's disease**

Congenital thymic hypoplasia see **DiGeorge syndrome**

Congenital ureter anomalies

Definition

The ureter drains urine from the kidney into the bladder. It is not simply a tube but an active organ that propels urine forward by muscular action. It has a valve at its bottom end that prevents urine from flowing backward into the kidney. Normally, there is one ureter on each side of the body for each kidney. However, among the many abnormalities of ureteral development, duplication is quite common. Ureters may also be malformed in a variety of ways—some harmful, others not.

Description

The urogenital system, for some reason, is more likely than any other to have **birth defects**, and they can occur in endless variety. Ureters can be duplicated completely or partially, they can be in the wrong place, they can be deformed, and they can end in the wrong place. The trouble these abnormalities bring is directly related to their effect on the flow of urine. As long as urine flows normally through them, and only in one direction, no harm is done.

- Duplication of ureters is quite common, either in part or completely. Kidneys are sometimes duplicated as well. Someone may have four kidneys and four ureters or two kidneys, half of each drained by a separate ureter, or a single kidney with two, three, or four ureters attached. As long as urine can flow easily in the correct direction, such malformations may never be detected. If, however, one of the ureters has a dead end, a stricture or stenosis (narrowing), or a leaky ureterovesical valve (valve between the ureter and bladder), infection is the likely result.
- Stricture or stenosis of a ureter prevents urine from flowing freely. Whenever flow is obstructed in the body—urine, bile, mucus, or any other liquid—infestation follows. Ureters can be obstructed anywhere along their course, though the ureterovesical valve is the most common place.
- A ureter may have an ectopic (out of place) orifice (opening)—it may enter the bladder, or even another structure, where it does not belong and therefore without an adequate valve to control reflux.
- The primary ureter, or a duplicate, may not even reach the bladder, but rather terminate in a dead end. Urine will stagnate there and eventually cause infection.
- A ureter can be perfectly normal but in the wrong place, such as behind the vena cava (the large vein in the middle of the abdomen). A so-called retrocaval ureter may be pinched by the vena cava so that flow is hindered. Other aberrant locations may also lead to compression and impaired flow.

Besides infection, urine that backs up will cause the ureter and the kidney to dilate. Eventually, the kidney will stop functioning because of the back pressure. This condition is called hydronephrosis—a kidney swollen with urine.

Causes and symptoms

The causes of birth defects are multiple and often unknown. Furthermore, the precise cause of specific birth defects has only rarely been identified. Such is the case with congenital ureteral anomalies.

KEY TERMS

Congenital—Present at birth.

Contrast agent—A chemical or other substance placed in the body to show structures that would not otherwise be visible on x ray or other imaging studies.

Cystoscopy—Looking into the urinary bladder with a thin telescope-like instrument.

Ectopic—Out of place.

Septicemia—A serious whole body infection spreading through the blood stream.

Ureterovesical valve—A sphincter (an opening controlled by a circular muscle), located where the ureter enters the bladder, that keeps urine from flowing backward toward the kidney.

Urogenital—Both the urinary system and the sexual organs, which form together in the developing embryo.

Practically the only symptom generated by ureteral abnormalities is urinary tract infection. A lower tract infection—in the bladder—is called **cystitis**. In children, it may cause **fever** and systemic symptoms, but in adults it causes only cloudy, burning, and frequent urine. Upper tract infections, on the other hand, can be serious for both adults and children, causing high fevers, back **pain**, severe generalized discomfort, and even leading to kidney failure or septicemia (infection spreading throughout the body by way of the blood stream).

In rare cases, urine from an ectopic ureter will bypass the bladder and dribble out of the bottom somewhere, through a natural orifice like the vagina or a completely separate unnatural opening.

Diagnosis

Serious or recurrent urinary infections will prompt a search for underlying abnormalities. **Cystoscopy** (looking into the bladder with a thin telescope-like instrument) and x rays with a contrast agent to illuminate the urinary system will usually identify the defect. **Computed tomography scans** (CT) and **magnetic resonance imaging** (MRI) scans may provide additional information. Urine cultures to identify the infecting germs will be repeated frequently until the problem is corrected.

Treatment

Sometimes the recurring infections caused by flow abnormalities can be treated with repeated and changing

courses of **antibiotics**. Over time, the infecting germs develop resistance to most treatments, especially the safer ones. If it can be done with acceptable risk, it is better to repair the defect surgically. Urologists have an arsenal of approaches to urine drainage that range from simply reimplanting a ureter into the bladder, in such a way that an effective valve is created, to building a new bladder out of a piece of bowel.

Alternative treatment

There are botanical and homeopathic treatments available for urinary tract infection. None can take the place of correcting a problem that is occurring because of a malformed or dysfunctional organ system. Once correction of the cause is addressed and there is unimpeded flow of urine, adequate fluid intake can contribute to prevention of future infections.

Prognosis

As long as damage to the kidneys from infection or back pressure has not become significant, the surgical repair of troublesome ureteral defects produces excellent long-term results in the great majority of cases. Monitoring for recurrent infections is always a good idea, and occasional checking of kidney function will detect hidden ongoing damage.

Resources

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J. Ricker Polsdorfer, MD

Congestive cardiomyopathy

Definition

Cardiomyopathy is an ongoing disease process that damages the muscle wall of the lower chambers of the heart. Congestive cardiomyopathy is the most common

form of cardiomyopathy. In congestive cardiomyopathy, also called dilated cardiomyopathy, the walls of the heart chambers stretch (dilate) to hold a greater volume of blood than normal. Congestive cardiomyopathy is the final stage of many heart diseases and the most common condition resulting in congestive **heart failure**.

Description

About 50,000 Americans develop cardiomyopathy each year. Of those, 87% have congestive cardiomyopathy. Primary cardiomyopathy accounts for only 1% of all deaths from heart disease.

When the heart muscle is damaged by a disease process, it cannot pump enough blood to meet the body's needs. Uninjured areas of the walls of the two lower heart chambers (called ventricles) stretch to make up for the lost pumping action. At first, the enlarged chambers allow more blood to be pumped with less force. The stretched muscle can also contract more forcefully. Over time, the heart muscle continues to stretch, ultimately becoming weaker. The heart is forced to work harder to pump blood by beating faster. Eventually it cannot keep up, and blood backs up into the veins, legs, and lungs. When this happens, the condition is called congestive heart failure.

Congestive cardiomyopathy usually affects both ventricles. Blood backed up into the lungs from the left ventricle causes fluid to congest the lung tissue. This is called **pulmonary edema**. When the right ventricle fails to pump enough blood, blood backs up into the veins causing **edema** in the legs, feet, ankles, and abdomen.

Causes and symptoms

Congestive cardiomyopathy may be caused by a number of conditions. Cardiomyopathy with a known cause is called secondary cardiomyopathy. When no cause can be identified, it is called primary cardiomyopathy or idiopathic cardiomyopathy. About 80% of all cases of cardiomyopathy do not have a known cause. Many heart specialists think that many cases of idiopathic congestive cardiomyopathy may be caused by a viral infection. Because cardiomyopathy may occur many years after a viral infection and viruses sometimes go undetected in laboratory tests, it is difficult to know if a virus is the cause. Some people have a weak heart from advanced **coronary artery disease** that causes heart muscle damage. This is sometimes called ischemic cardiomyopathy.

Conditions that can cause congestive cardiomyopathy are:

- Coronary artery disease
- Infections

KEY TERMS

Angiotensin-converting enzyme (ACE) inhibitor—A drug that relaxes blood vessel walls and lowers blood pressure.

Atherosclerosis—Buildup of a fatty substance called a plaque inside blood vessels.

Cardiac catheterization—A diagnostic test for evaluating heart disease; a catheter is inserted into an artery and passed into the heart.

Cardiomyopathy—Disease of the heart muscle.

Congestive cardiomyopathy—Also called dilated cardiomyopathy; cardiomyopathy in which the walls of the heart chambers stretch, enlarging the heart ventricles so they can hold a greater volume of blood than normal.

Coxsackievirus B—A type of virus in the group Enterovirus that causes an infection similar to polio, but without paralysis.

Digitalis—A drug that helps the heart muscle to have stronger pumping action.

Dilated cardiomyopathy—Also called congestive cardiomyopathy; cardiomyopathy in which the walls of the heart chambers stretch, enlarging the heart ventricles so they can hold a greater volume of blood than normal.

Diuretic—A type of drug that helps the kidneys eliminate excess salt and water.

Edema—Swelling caused by fluid buildup in tissues.

Granulomatous myocarditis—Also called giant cell myocarditis, this noninfectious inflammation of the heart causes large areas of tissue death in the heart muscle, ventricular enlargement, and clots inside the heart chambers.

Idiopathic cardiomyopathy—Cardiomyopathy without a known cause.

Sarcoidosis—A chronic disease that causes formation of abnormal areas containing inflammatory cells, called granulomas, in any organ or tissue; in the heart, large areas of the heart muscle can be involved, causing cardiomyopathy.

Vasodilator—Any drug that relaxes blood vessel walls.

Ventricle—One of the two lower chambers of the heart.

Wegener's granulomatosis—A disease usually affecting males that causes the infiltration of inflammatory cells and tissue death in the lungs, kidneys, blood vessels, heart, and other tissues.

- noninfectious inflammatory conditions
- alcohol and other drugs or toxins
- **hypertension**
- nutritional and metabolic disorders
- **pregnancy**.

Coronary artery disease is one of the most common causes of congestive cardiomyopathy. In coronary artery disease, the arteries supplying blood to the heart become narrowed or blocked. When blood flow to an area of the heart is completely blocked, the person has a **heart attack**. The heart muscle suffers damage when its blood supply is reduced or blocked. Significant recurrent muscle damage can occur silently. This damage can lead to congestive cardiomyopathy.

Infections caused by bacteria, viruses, and other microorganisms can involve the heart, causing inflammation of the heart muscle (**myocarditis**). The inflammation may damage the heart muscle and cause congestive cardiomyopathy. In the United States, the coxsackievirus

B is the most common cause of viral congestive cardiomyopathy.

Myocarditis can also be caused by noninfectious disorders. For example, the conditions **sarcoidosis**, granulomatous myocarditis, and **Wegener's granulomatosis** cause inflammation and tissue **death** in the heart muscle.

Years of drinking excessive amounts of alcohol can weaken the heart muscle, leading to congestive cardiomyopathy. Other drugs and toxins, such as **cocaine**, pesticides, and other chemicals, may have the same effect.

High blood pressure (hypertension) puts extra pressure on blood vessels and the heart. This increased pressure makes the heart work harder to pump blood, which may thicken and damage the chamber walls.

Severe nutritional deficiencies can weaken the heart muscle and affect its pumping ability. Certain disorders of metabolism, including **diabetes mellitus** and thyroid disorders, can also lead to congestive cardiomyopathy.

Occasionally, inflammation of the heart muscle and congestive cardiomyopathy may develop late in pregnan-

cy or shortly after a woman gives birth. This type of congestive cardiomyopathy is called peripartum cardiomyopathy. The cause of congestive cardiomyopathy in pregnancy is not known.

Congestive cardiomyopathy usually is a chronic condition, developing gradually over time. Patients with early congestive cardiomyopathy may not have symptoms. The most common symptoms are **fatigue** and **shortness of breath** on exertion. Unfortunately, **sudden cardiac death** is not uncommon with this condition. It stems from irregular heart rhythms in the ventricles (ventricular **arrhythmias**).

Patients with more advanced congestive cardiomyopathy may also have chest or abdominal pains, extreme tiredness, **dizziness**, and swelling of the legs and ankles.

Diagnosis

Diagnosis of congestive cardiomyopathy is based on:

- symptoms
- medical history
- **physical examination**
- **chest x ray**
- electrocardiogram (ECG; also called EKG)
- echocardiogram
- **cardiac catheterization**

The diagnosis is based on the patient's symptoms, a complete physical examination, and tests that detect abnormalities of the heart chambers. The physician listens to the heart with a stethoscope to detect abnormal heart rhythms and heart sounds. A heart murmur might mean that the heart valves are not closing properly due to the ventricles being enlarged.

A chest x ray can show if the heart is enlarged and if there is fluid in the lungs. Abnormalities of heart valves and other structures may also be seen on a chest x ray.

An electrocardiogram provides a record of electrical changes in the heart muscle during the heartbeat. It gives information on the heart rhythm and can show if the heart chamber is enlarged. An ECG can detect damage to the heart muscle and the amount of damage.

Echocardiography uses sound waves to make images of the heart. These images can show if the heart wall or chambers are enlarged and if there are any abnormalities of the heart valves. Echocardiography can also evaluate the pumping efficiency of the ventricles.

Cardiac catheterization usually is only used if a diagnosis cannot be made with other methods. 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 of the heart muscle can be removed through the catheter for examination under a microscope (biopsy). This biopsy can show the type and amount of damage to the heart muscle.

Treatment

When a patient is diagnosed with congestive cardiomyopathy, physicians try to find out the cause. If coronary artery disease is not the culprit, in most other cases a cause is not identified. When a condition responsible for the congestive cardiomyopathy is diagnosed, treatment is aimed at correcting the underlying condition. Congestive cardiomyopathy caused by drinking excess alcohol or by drugs or toxins can be treated by eliminating the alcohol or toxin completely. In some cases, the heart may recover after the toxic substance is removed from the body. Bacterial myocarditis is treated with an antibiotic to eliminate the bacteria.

There is no cure for idiopathic congestive cardiomyopathy. Medicines are given to reduce the workload of the heart and to relieve the symptoms.

One or more of the following types of medicines may be prescribed for congestive cardiomyopathy:

- **digitalis**
- **diuretics**
- **vasodilators**
- **beta blockers**
- angiotensin converting enzyme inhibitors (ACE inhibitors)
- angiotensin receptor blockers

Digitalis helps the heart muscle to have stronger pumping action. Diuretics help eliminate excess salt and water from the kidneys by making patients urinate more often. This helps reduce the swelling caused by fluid buildup in the tissues. Vasodilators, beta blockers, and ACE inhibitors lower blood pressure and expand the blood vessels so blood can move more easily through them. This action makes it easier for the heart to pump blood through the vessels.

Patients may also be given anticoagulant medications to prevent clots from forming due to pooling of blood in the heart chambers. Medicines to prevent abnormal heart rhythms (arrhythmias) may be given, but some of these drugs can also reduce the force of heart contractions. Automatic implantable cardioverter defibrillators (AICDs) can treat life-threatening arrhythmias, which are relatively common in severe cardiomyopathy.

Certain lifestyle changes may help reduce the workload on the heart and relieve symptoms. Some patients may need to change their diet, stop drinking alcohol, begin a physician-supervised **exercise** program, and/or stop **smoking**.

Severe congestive cardiomyopathy usually causes heart failure. When the heart muscle is damaged so severely that medicines cannot help, a heart transplant may be the only remaining treatment to be considered.

Prognosis

The outlook for a patient with congestive cardiomyopathy depends on the severity of the disease and the person's health. Generally, congestive cardiomyopathy worsens over time and the prognosis is not good. About 50% of patients with congestive cardiomyopathy live for five years after the diagnosis. Twenty five percent of patients are alive 10 years after diagnosis. Women with congestive cardiomyopathy live twice as long as men with the disease. Many of the deaths are caused by sudden abnormal heart rhythms.

Prevention

Because idiopathic congestive cardiomyopathy does not have a known cause, there is no sure way to prevent it. The best way to prevent congestive cardiomyopathy is to avoid known causes such as drinking excess alcohol or taking toxic drugs. Eating a nutritious diet and getting regular exercise to improve overall fitness also can help the heart to stay healthy.

Congestive cardiomyopathy may also be prevented by identifying and treating any conditions that might damage the heart muscle. These include high blood pressure and coronary artery disease. Regular blood pressure checks and obtaining immediate medical care for hypertension and symptoms of coronary artery disease, such as chest **pain**, are important to keep the heart functioning properly.

Finally, diagnosing and treating congestive cardiomyopathy before the heart becomes severely damaged may improve the outlook.

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American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

Texas Heart Institute. Heart Information Service. P.O. Box 20345, Houston, TX 77225-0345. <<http://www.tmc.edu/thi>>.

Toni Rizzo

Congestive heart failure see **Heart failure**

Congenital heart disease

Definition

Congenital heart disease, also called congenital heart defect, includes a variety of malformations of the heart or its major blood vessels that are present at birth.

Description

Congenital heart disease occurs when the heart or blood vessels near the heart do not develop properly before birth. Some infants are born with mild types of congenital heart disease, but most need surgery in order to survive. Patients who have had surgery are likely to experience other cardiac problems later in life.

Most types of congenital heart disease obstruct the flow of blood in the heart or the nearby vessels, or cause an abnormal flow of blood through the heart. Rarer types of congenital heart disease occur when the newborn has only one ventricle, or when the pulmonary artery and the aorta come out of the same ventricle, or when one side of the heart is not completely formed.

Patent ductus arteriosus

Patent ductus arteriosus refers to the opening of a passageway—or temporary blood vessel (ductus)—to carry the blood from the heart to the aorta before birth, allowing blood to bypass the lungs, which are not yet functional. The ductus should close spontaneously in the first few hours or days after birth. When it does not close in the newborn, some of the blood that should flow through the aorta then returns to the lungs. Patent ductus arteriosus is common in premature babies, but rare in full-term babies. It has also been associated with mothers who had German measles (**rubella**) while pregnant.

Hypoplastic left heart syndrome

Hypoplastic left heart syndrome, a condition in which the left side of the heart is underdeveloped, is rare, but it is the most serious type of congenital heart disease.

With this syndrome, blood reaches the aorta, which pumps blood to the entire body, only from the ductus, which then normally closes within a few days of birth. In hypoplastic left heart syndrome, the baby seems normal at birth, but as the ductus closes, blood cannot reach the aorta and circulation fails.

Obstruction defects

When heart valves, arteries, or veins are narrowed, they partly or completely block the flow of blood. The most common obstruction defects are **pulmonary valve stenosis**, **aortic valve stenosis**, and **coarctation of the aorta**. Bicuspid aortic valve and subaortic stenosis are less common.

Stenosis is a narrowing of the valves or arteries. In pulmonary stenosis, the pulmonary valve does not open properly, forcing the right ventricle to work harder. In aortic stenosis, the improperly formed aortic valve is narrowed. As the left ventricle works harder to pump blood through the body, it becomes enlarged. In coarctation of the aorta, the aorta is constricted, reducing the flow of blood to the lower part of the body and increasing blood pressure in the upper body.

A bicuspid aortic valve has only two flaps instead of three, which can lead to stenosis in adulthood. Subaortic stenosis is a narrowing of the left ventricle below the aortic valve, that limits the flow of blood from the left ventricle.

Septal defects

When a baby is born with a hole in the septum (the wall separating the right and left sides of the heart), blood leaks from the left side of the heart to the right, or from a higher pressure zone to a lower pressure zone. A major leakage can lead to enlargement of the heart and failing circulation. The most common types of septal defects are **atrial septal defect**, an opening between the two upper heart chambers, and **ventricular septal defect**, an opening between the two lower heart chambers. Ventricular septal defect accounts for about 15% of all cases of congenital heart disease in the United States.

Cyanotic defects

Heart disorders that cause a decreased, inadequate amount of oxygen in blood pumped to the body are called cyanotic defects. Cyanotic defects, including tricuspid arteriosus, total anomalous pulmonary venous return, **tetralogy of Fallot**, **transposition of the great arteries**, and tricuspid atresia, result in a blue discoloration of the skin due to low oxygen levels. About 10% of cases of

congenital heart disease in the United States are tetralogy of Fallot, which includes four defects. The major defects are a large hole between the ventricles, which allows oxygen-poor blood to mix with oxygen-rich blood, and narrowing at or beneath the pulmonary valve. The other defects are an overly muscular right ventricle and an aorta that lies over the ventricular hole.

In transposition (reversal of position) of the great arteries, the pulmonary artery and the aorta are reversed, causing oxygen-rich blood to re-circulate to the lungs while oxygen-poor blood goes to the rest of the body. In tricuspid atresia, the baby lacks a tricuspid valve and blood cannot flow properly from the right atrium to the right ventricle.

Other defects

Ebstein's anomaly is a rare congenital syndrome that causes malformed tricuspid valve leaflets, which allow blood to leak between the right ventricle and the right atrium. It also may cause a hole in the wall between the left and right atrium. Treatment often involves repairing the tricuspid valve. Ebstein's anomaly may be associated with maternal use of the psychiatric drug lithium during pregnancy.

Brugada syndrome is another rare congenital heart defect that appears in adulthood and may cause sudden death if untreated. Symptoms, which include rapid, uneven heart beat, often appear at night. Scientists believe that Brugada syndrome is caused by mutations in the gene SCN5A, which involves cardiac sodium channels.

Infants born with DiGeorge sequence can have heart defects such as a malformed aortic arch and tetralogy of Fallot. Researchers believe DiGeorge sequence is most often caused by mutations in genes in the region 22q11.

Marfan syndrome is a connective tissue disorder that causes tears in the aorta. Since the disease also causes excessive bone growth, most Marfan syndrome patients are over six feet tall. In athletes, and others, it can lead to sudden death. Researchers believe the defect responsible for Marfan's syndrome is found in gene FBN1, on chromosome 15.

About 32,000 infants are born every year with congenital heart disease, which is the most common birth defect. About half of these cases require medical treatment. More than one million people with heart defects are currently living in the United States.

Causes and symptoms

In most cases, the causes of congenital heart disease are unknown. Genetic and environmental factors and lifestyle habits can all be involved. The likelihood of hav-

ing a child with a congenital heart disease increases if the mother or father, another child, or another relative had congenital heart disease or a family history of sudden death. Viral infections, such as German measles, can produce congenital heart disease. Women with diabetes and **phenylketonuria** also are at higher risk of having children with congenital heart defects. Many cases of congenital heart disease result from the mother's excessive use of alcohol or taking illegal drugs, such as **cocaine**, while pregnant. The mother's exposure to certain anticonvulsant and dermatologic drugs during pregnancy can also cause congenital heart disease. There are many genetic conditions, such as **Down syndrome**, which affect multiple organs and can cause congenital heart disease.

Symptoms of congenital heart disease in general include: **shortness of breath**, difficulty feeding in infancy, sweating, **cyanosis** (bluish discoloration of the skin), heart murmur, respiratory infections that recur excessively, stunted growth, and limbs and muscles that are underdeveloped.

Symptoms of specific types of congenital heart disease are as follows:

- Patent ductus arteriosus: quick tiring, slow growth, susceptibility to **pneumonia**, rapid breathing. If the ductus is small, there are no symptoms.
- Hypoplastic left heart syndrome: ashen color, rapid and difficult breathing, inability to eat.
- Obstruction defects: cyanosis (skin that is discolored blue), chest **pain**, tiring easily, **dizziness or fainting**, congestive **heart failure**, and high blood pressure.
- Septal defects: difficulty breathing, stunted growth. Sometimes there are no symptoms.
- Cyanotic defects: cyanosis, sudden rapid breathing or unconsciousness, and shortness of breath and fainting during **exercise**.

Diagnosis

Echocardiography and cardiac **magnetic resonance imaging** (MRI) are used to confirm congenital heart disease when it is suggested by the symptoms and **physical examination**. An echocardiograph will display an image of the heart that is formed by sound waves. It detects valve and other heart problems. Fetal echocardiography is used to diagnose congenital heart disease in utero, usually after 20 weeks of pregnancy. Between 10 and 14 weeks of pregnancy, physicians also may use an ultrasound to look for a thickness at the nuchal translucency, a pocket of fluid in back of the embryo's neck, which may indicate a cardiac defect in 55% of cases. Cardiac MRI, a scanning method that uses magnetic

KEY TERMS

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. Many congenital heart defects affect the aorta.

Congenital—Refers to a disorder that is present at birth.

Cyanotic—Marked by bluish discoloration of the skin due to a lack of oxygen in the blood. It is one of the types of congenital heart disease.

Ductus—The blood vessel that joins the pulmonary artery and the aorta. When the ductus does not close at birth, it causes a type of congenital heart disease called patent ductus arteriosus.

Electrocardiograph (ECG, EKG)—A test used to measure electrical impulses coming from the heart in order to gain information about its structure or function.

Hypoplastic—Incomplete or underdevelopment of a tissue or organ. Hypoplastic left heart syndrome is the most serious type of congenital heart disease.

Neuchal translucency—A pocket of fluid at the back of an embryo's neck visible via ultrasound that, when thickened, may indicate the infant will be born with a congenital heart defect.

Septal—Relating to the septum, the thin muscle wall dividing the right and left sides of the heart. Holes in the septum are called septal defects.

Stenosis—The constricting or narrowing of an opening or passageway.

fields and radio waves, can help physicians evaluate congenital heart disease, but is not always necessary. Physicians also may use a chest x ray to look at the size and location of the heart and lungs, or an electrocardiograph (ECG), which measures electrical impulses to create a graph of the heart beat.

Treatment

Congenital heart disease is treated with drugs and/or surgery. Drugs used include **diuretics**, which aid the baby in excreting water and salts, and digoxin, which strengthens the contraction of the heart, slows the heart-beat, and removes fluid from tissues.

Surgical procedures seek to repair the defect as much as possible and restore circulation to as close to

normal as possible. Sometimes, multiple surgical procedures are necessary. Surgical procedures include: arterial switch, balloon atrial septostomy, **balloon valvoplasty**, Damus-Kaye-Stansel procedure, Fontan procedure, pulmonary artery banding, Ross procedure, shunt procedure, and venous switch or intra-atrial baffle.

Arterial switch, to correct transposition of the great arteries, involves connecting the aorta to the left ventricle and connecting the pulmonary artery to the right ventricle. Balloon atrial septostomy, also done to correct transposition of the great arteries, enlarges the atrial opening during heart catheterization. Balloon valvoplasty uses a balloon-tipped catheter to open a narrowed heart valve, improving the flow of blood in pulmonary stenosis. It is sometimes used in aortic stenosis. Transposition of the great arteries can also be corrected by the Damus-Kaye-Stansel procedure, in which the pulmonary artery is cut in two and connected to the ascending aorta and the farthest section of the right ventricle.

For tricuspid atresia and pulmonary atresia, the Fontan procedure connects the right atrium to the pulmonary artery directly or with a conduit, and the atrial defect is closed. Pulmonary artery banding, narrowing the pulmonary artery with a band to reduce blood flow and pressure in the lungs, is used for ventricular septal defect, atrioventricular canal defect, and tricuspid atresia. Later, the band can be removed and the defect corrected with open-heart surgery.

To correct aortic stenosis, the Ross procedure grafts the pulmonary artery to the aorta. For tetralogy of Fallot, tricuspid atresia, or pulmonary atresia, the shunt procedure creates a passage between blood vessels, sending blood into parts of the body that need it. For transposition of the great arteries, venous switch creates a tunnel inside the atria to re-direct oxygen-rich blood to the right ventricle and aorta and venous blood to the left ventricle and pulmonary artery.

When all other options fail, some patients may need a heart transplant. Children with congenital heart disease require lifelong monitoring, even after successful surgery. The American Heart Association recommends regular dental check-ups and the preventive use of **antibiotics** to protect patients from heart infections, or **endocarditis**. Since children with congenital heart disease have slower growth, **nutrition** is important. Physicians may also limit their athletic activity.

Prognosis

The outlook for children with congenital heart disease has improved markedly in the past two decades. Many types of congenital heart disease that would have

been fatal can now be treated successfully. Research on diagnosing heart defects when the fetus is in the womb may lead to future treatment to correct defects before birth. Promising new prevention methods and treatments include genetic screening and the cultivation of cardiac tissue in the laboratory that could be used to repair congenital heart defects.

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ORGANIZATIONS

American Heart Association. 7272 Greenville Ave., Dallas, TX 75231-4596. (214) 373-6300 or (800) 242-8721. <inquire@heart.org>. <http://www.americanheart.org>.

Congenital Heart Disease Information and Resources. 1561 Clark Dr., Yardley, PA 19067. <http://www.tchin.org>.

Texas Heart Institute Heart Information Service. P.O. Box 20345, Houston, TX 77225-0345. (800) 292-2221. <http://www.tmc.edu/thi/his.html>.

Melissa Knopper

Conjunctivitis

Definition

Conjunctivitis is an inflammation or redness of the lining of the white part of the eye and the underside of the eyelid (conjunctiva) that can be caused by infection, allergic reaction, or physical agents like infrared or ultraviolet light.

Description

Conjunctivitis is the inflammation of the conjunctiva, a thin, delicate membrane that covers the eyeball and lines the eyelid. Conjunctivitis is an extremely common eye problem because the conjunctiva is continually

exposed to microorganisms and environmental agents that can cause infections or allergic reactions. Conjunctivitis can be acute or chronic depending upon how long the condition lasts, the severity of symptoms, and the type of organism or agent involved. It can also affect one or both eyes and, if caused by infection, can be very easily transmitted to others during close physical contact, particularly among children in a daycare center. Other names for conjunctivitis include pink eye and red eye.

Causes and symptoms

Conjunctivitis may be caused by a viral infection, such as a cold, acute respiratory infection, or disease such as **measles**, herpes simplex, or herpes zoster. Symptoms include mild to severe discomfort in one or both eyes, redness, swelling of the eyelids, and watery, yellow, or green discharge. Symptoms may last anywhere from several days to two weeks. Infection with an adenovirus, however, may also cause a significant amount of pus-like discharge and a scratchy, foreign body-type of sensation in the eye. This may also be accompanied by swelling and tenderness of the lymph nodes near the ear.

Bacterial conjunctivitis can occur in adults and children and is caused by organisms such as *Staphylococcus*, *Streptococcus*, and *Hemophilus*. Symptoms of bacterial conjunctivitis include a pus-like discharge and crusty eyelids after awakening. Redness of the conjunctiva can be mild to severe and may be accompanied by swelling. Persons with symptoms of conjunctivitis who are sexually active may possibly be infected with the bacteria that cause either **gonorrhea** or chlamydia. There may be large amounts of pus-like discharge, and symptoms may include intolerance to light (photophobia), watery mucous discharge, and tenderness in the lymph nodes near the ear that may persist for up to three months.

Conjunctivitis may also be caused by environmental hazards, such as wind, smoke, dust, and allergic reactions caused by pollen, dust, or grass. Symptoms range from **itching** and redness to a mucous discharge. Persons who wear contact lenses may develop allergic conjunctivitis caused by the various eye solutions and foreign proteins contained in them.

Other less common causes of conjunctivitis include exposure to sun lamps or the electrical arcs used during welding, and problems with inadequate drainage of the tear ducts.

Diagnosis

An accurate diagnosis of conjunctivitis centers on taking a patient history to learn when symptoms began, how long the condition has been going on, the symptoms



This person has severe conjunctivitis, most likely caused by an allergic reaction. (Custom Medical Stock Photo. Reproduced by permission.)

experienced, and other predisposing factors, such as upper respiratory complaints, **allergies**, **sexually transmitted diseases**, herpes simplex infections, and exposure to persons with pink eye. It may be helpful to learn whether an aspect of an individual's occupation may be the cause, for example, welding. Diagnostic tests are usually not indicated unless initial treatment fails or an infection with gonorrhea or chlamydia is suspected. In such cases, the discharge may be cultured and Gram stained to determine the organism responsible for causing the condition. Cultures and smears are relatively painless.

Treatment

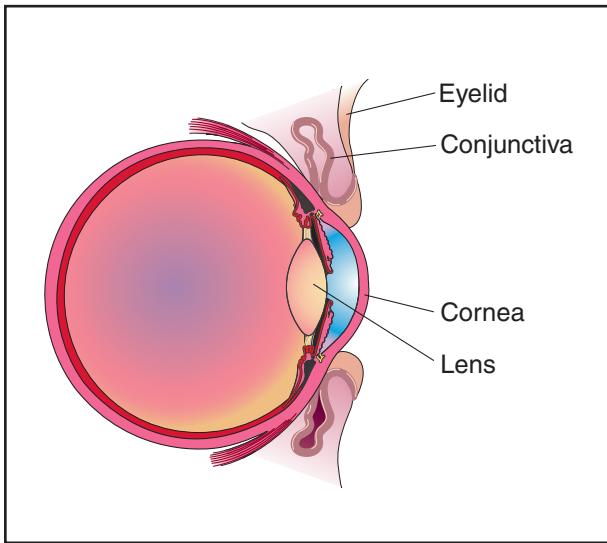
The treatment of conjunctivitis depends on what caused the condition. In all cases, warm compresses applied to the affected eye several times a day may help to reduce discomfort.

Conjunctivitis due to a viral infection, particularly those due to adenoviruses, are usually treated by applying warm compresses to the eye(s) and applying topical antibiotic ointments to prevent secondary bacterial infections.

Viral conjunctivitis caused by herpes simplex should be referred to an ophthalmologist. Topical steroids are commonly prescribed in combination with antiviral therapy.

In cases of bacterial conjunctivitis, a physician may prescribe an antibiotic eye ointment or eye drops containing sodium sulfacetamide (Sulamyd) to be applied daily for seven to 14 days. If, after 72 hours, the condition does not improve, a physician or primary care provider should be notified because the bacteria involved may be resistant to the antibiotic used or the cause may not be bacterial.

For cases of conjunctivitis caused by a gonococcal organism, a physician may prescribe an intramuscular injection of ceftriaxone (Rocephin) and a topical antibi-



Conjunctivitis is the inflammation of the conjunctiva, a thin, delicate membrane that covers the eyeball and lines the eyelid. It may be caused by a viral infection, such as a cold or acute respiratory infection, or by such diseases as measles, herpes simplex, or herpes zoster. (Illustration by Electronic Illustrators Group.)

otic ointment containing erythromycin or bactracin to be applied four times daily for two to three weeks. Sexual partners should also be treated.

With accompanying chlamydia infection, a topical antibiotic ointment containing erythromycin (Ilotycin) may be prescribed to be applied 1-2 times daily. In addition, oral erythromycin or tetracycline therapy may be indicated for three to four weeks. Here again, sexual partners should also be treated.

Allergic conjunctivitis can be treated by removing the allergic substance from a person's environment, if possible; by applying cool compresses to the eye; and by administering eye drops four to six times daily for four days. Also, the antihistamine diphenhydramine hydrochloride (Benadryl) may help to relieve itchy eyes.

Alternative treatment

Conjunctivitis caused by gonococcal and chlamydial infection usually requires conventional medical treatment. With bacterial, viral, and allergic conjunctivitis, however, alternative options can be helpful. Internal immune enhancement with supplementation can aid in the resolution of bacterial and viral conjunctivitis. Removal of the allergic agent is an essential step in treating allergic conjunctivitis. As with any of the recommended treatments, however, if no improvement is seen within 48–72 hours, a physician should be consulted.

Homeopathically, there are a number of acute remedies designed to treat conjunctivitis. These include *Pulsatilla* (windflower, *Pulsatilla nigricans*), *Belladonna*, and eyebright (*Euphrasia officinalis*). Eye drops, prepared with homeopathic remedies and/or herbs, can be a good substitute for pharmaceutical eye drops. Eye washes can also be made. Herbal eyewashes made with eyebright (1 tsp. dried herb steeped in 1 pint of boiling water) or chamomile (*Matricaria recutita*; 2–3 tsp in 1 pt of boiling water) may be helpful. Eyewashes should be strained and cooled before use, and close attention should be paid to make sure that any solution put into the eye is sterile.

Other simple home remedies may help relieve the discomfort associated with conjunctivitis. A boric acid eyewash can be used to clean and soothe the eyes. A warm compress applied to the eyes for five to 10 minutes three times a day can help relieve the discomfort of bacterial and viral conjunctivitis. A cool compress or cool, damp tea bags placed on the eyes can ease the discomfort of allergic conjunctivitis.

Prognosis

If treated properly, the prognosis for conjunctivitis is good. Conjunctivitis caused by an allergic reaction should clear up once the allergen is removed. However, allergic conjunctivitis will likely recur if the individual again comes into contact with the particular allergen. Conjunctivitis caused by bacteria or a virus, if treated properly, is usually resolved in 10–14 days. If there is no relief of symptoms in 48–72 hours, or there is moderate to severe eye pain, changes in vision, or the conjunctivitis is suspected to be caused by herpes simplex, a physician should be notified immediately. If untreated or if treatment fails and is not corrected, conjunctivitis may cause visual impairment by spreading to other parts of the eye, such as the cornea.

Prevention

Conjunctivitis can, in many cases, be prevented, or at least the course of the disease can be shortened by following some simple practices.

- Frequently wash hands using antiseptic soap, and use single-use towels during the disease to prevent spreading the infection.
- Avoid chemical irritants and known allergens.
- If in an area where welding occurs, use the proper protective eye wear and screens to prevent damaging the eyes.
- Use a clean tissue to remove discharge from eyes, and wash hands to prevent the spread of infection.

KEY TERMS

Adenovirus—A virus that affects the upper respiratory tract.

Chlamydia—The most common bacterial sexually transmitted disease in the United States that often accompanies gonorrhea and is known for its lack of evident symptoms in the majority of women.

Gonococcal—The bacteria *Neisseria gonorrhoeae* that causes gonorrhea, a sexually transmitted infection of the genitals and urinary tract. The gonococcal organism may occasionally affect the eye, causing blindness if not treated.

Herpes simplex virus—A virus that can cause fever and blistering on the skin, mucous membranes, or genitalia.

Herpes zoster virus—Acute inflammatory virus that attacks the nerve cells on the root of each spinal nerve with skin eruptions along a sensory nerve ending.

Staphylococcus—A bacterial organism, looking much like a cluster of grapes, that can infect various body systems.

Streptococcus—An organism that causes infections of either the upper respiratory or gastrointestinal tract.

- If medication is prescribed, finish the course of **antibiotics**, as directed, to make sure that the infection is cleared up and does not recur.
- Avoid contact, such as vigorous physical activities, with other persons until symptoms resolve.

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Lisa Papp, RN

Consciousness disorders see **Coma**

Constipation

Definition

Constipation is an acute or chronic condition in which bowel movements occur less often than usual or consist of hard, dry stools that are painful or difficult to pass. Bowel habits vary, but an adult who has not had a bowel movement in three days or a child who has not had a bowel movement in four days is considered constipated.

Description

Constipation is one of the most common medical complaints in the United States. Constipation can occur at any age, and is more common among individuals who resist the urge to move their bowels at their body's signal. This often happens when children start school or enter daycare and feel shy about asking permission to use the bathroom.

Constipation is more common in women than in men and is especially apt to occur during **pregnancy**. Age alone does not increase the frequency of constipation, but elderly people (especially women) are more likely to suffer from constipation.

Although this condition is rarely serious, it can lead to:

- bowel obstruction
- chronic constipation
- hemorrhoids (a mass of dilated veins in swollen tissue around the anus)
- hernia (a protrusion of an organ through a tear in the muscle wall)
- spastic colitis (**irritable bowel syndrome**, a condition characterized by alternating periods of **diarrhea** and constipation)
- laxative dependency

Chronic constipation may be a symptom of colorectal **cancer**, depression, diabetes, diverticulosis (small pouches in the muscles of the large intestine), **lead poisoning**, or **Parkinson's disease**.

In someone who is elderly or disabled, constipation may be a symptom of bowel impaction, a more serious condition in which feces are trapped in the lower part of the large intestine. A doctor should be called if an elderly or disabled person is constipated for a week or more or if a child seems to be constipated.

A doctor should be notified whenever constipation occurs after starting a new prescription, vitamin, or mineral supplement or is accompanied by blood in the stools, changes in bowel patterns, or **fever** and abdominal **pain**.

Causes and symptoms

Constipation usually results from not getting enough **exercise**, not drinking enough water, or from a diet that does not include an adequate amount of fiber-rich foods like beans, bran cereals, fruits, raw vegetables, rice, and whole-grain breads.

Other causes of constipation include anal fissure (a tear or crack in the lining of the anus); **chronic kidney failure**; **colon or rectal cancer**; depression; **hypercalcemia** (abnormally high levels of calcium in the blood); **hypothyroidism** (underactive thyroid gland); illness requiring complete bed rest; irritable bowel syndrome; and **stress**.

Constipation can also be a side effect of:

- aluminum salts in **antacids**
- antihistamines
- antipsychotic drugs
- aspirin
- belladonna (*Atropa belladonna*, source of atropine, a medication used to relieve spasms and dilate the pupils of the eye)
- beta blockers (medications used to stabilize irregular heartbeat, lower high blood pressure, reduce chest pain)
- blood pressure medications
- calcium channel blockers (medication prescribed to treat high blood pressure, chest pain, some types of irregular heartbeat and **stroke**, and some non-cardiac diseases)
- diuretics (drugs that promote the formation and secretion of urine)
- iron or calcium supplements
- narcotics (potentially addictive drugs that relieve pain and cause mood changes)

- tricyclic antidepressants (medications prescribed to treat chronic pain, depression, headaches, and other illnesses)

An adult who is constipated may feel bloated, have a **headache**, swollen abdomen, or pass rock-like feces; or strain, bleed, or feel pain during bowel movements. A constipated baby may strain, cry, draw the legs toward the abdomen, or arch the back when having a bowel movement.

Diagnosis

Everyone becomes constipated once in a while, but a doctor should be notified if significant changes in bowel patterns last for more than a week or if symptoms continue more than three weeks after increasing activity and fiber and fluid intake.

The patient's observations and medical history help a primary care physician diagnose constipation. The doctor uses his fingers to see if there is a hardened mass in the abdomen, and may perform a **rectal examination**. Other diagnostic procedures include a **barium enema**, which reveals blockage inside the intestine; laboratory analysis of blood and stool samples for internal bleeding or other symptoms of systemic disease; and a **sigmoidoscopy** (examination of the sigmoid area of the colon with a flexible tube equipped with a magnifying lens).

Physical and psychological assessments and a detailed history of bowel habits are especially important when an elderly person complains of constipation.

Treatment

If changes in diet and activity fail to relieve occasional constipation, an over-the-counter laxative may be used for a few days. Preparations that soften stools or add bulk (bran, psyllium) work more slowly but are safer than Epsom salts and other harsh **laxatives** or herbal laxatives containing senna (*Cassia senna*) or buckthorn (*Rhamnus purshiana*), which can harm the nerves and lining of the colon.

A woman who is pregnant should never use a laxative. Neither should anyone who is experiencing abdominal pain, nausea, or vomiting.

A warm-water or mineral oil enema can relieve constipation, and a non-digestible sugar (lactulose) or special electrolyte solution is recommended for adults and older children with stubborn symptoms.

If a patient has an impacted bowel, the doctor inserts a gloved finger into the rectum and gently dislodges the hardened feces.

Alternative treatment

Initially, alternative practitioners will suggest that the patient drink an adequate amount of water each day (six to eight glasses), exercise on a regular basis, and eat a diet high in soluble and insoluble fibers. Soluble fibers include pectin, flax, and gums; insoluble fibers include psyllium and brans from grains like wheat and oats. Fresh fruits and vegetables contain both soluble and insoluble fibers. Castor oil, applied topically to the abdomen and covered by a heat source (a heating pad or hot water bottle), can help relieve constipation when used nightly for 20–30 minutes.

Acupressure

This needless form of **acupuncture** is said to relax the abdomen, ease discomfort, and stimulate regular bowel movements when diet and exercise fail to do so. After lying down, the patient closes his eyes and takes a deep breath. For two minutes, he applies gentle fingertip pressure to a point about two and one-half inches below the navel.

Accupressure can also be applied to the outer edges of one elbow crease and maintained for 30 seconds before pressing the crease of the other elbow. This should be done three times a day to relieve constipation.

Aromatherapy

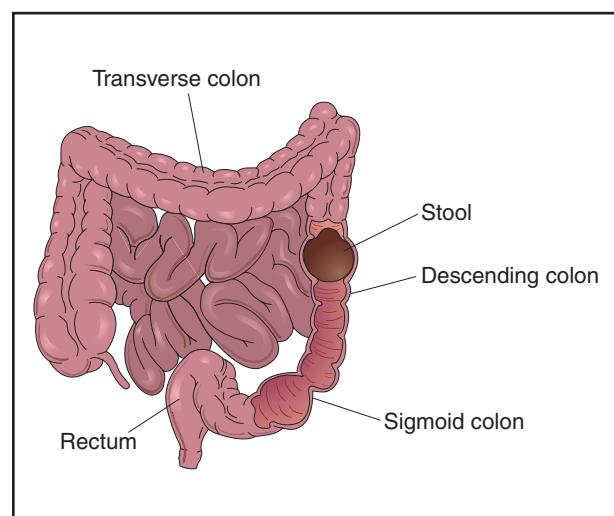
Six drops of rosemary (*Rosmarinus officinalis*) and six drops of thyme (*Thymus* spp.) diluted by 1 oz of almond oil, olive oil, or another carrier oil can relieve constipation when used to massage the abdomen.

Herbal therapy

A variety of herbal therapies can be useful in the treatment of constipation. Several herbs, including chamomile (*Matricaria recutita*), dandelion (*Taraxacum mongolicum*), and burdock (*Arctium lappa*), act as bitters, stimulating the movement of the digestive and excretory systems. There are also “laxative” herbs that assist with bowel movement. Two of these are senna (*Cassia senna*) and buckthorn (*Rhamnus purshiana*). These “laxative” herbs are stronger acting on elimination than bitters and can sometimes cause cramping (mixing them with a calming herb like fennel or caraway can help reduce cramping). Both senna and buckthorn are powerful herbs that are best used with direction from an experienced practitioner, since they can have adverse side effects and the patient may become dependent on them.

Homeopathy

Homeopathy also can offer assistance with constipation. There are acute remedies for constipation that can



Constipation is an acute or chronic condition in which bowel movements occur less often than usual or consist of hard, dry stools that are painful or difficult to pass. (Illustration by Electronic Illustrators Group).

be found in one of the many home remedy books on homeopathic medicine. A constitutional prescription also can help rebalance someone who is struggling with constipation.

Massage

Massaging the leg from knee to hip in the morning, at night, and before trying to move the bowels is said to relieve constipation. There is also a specific Swedish massage technique that can help relieve constipation.

Yoga

The knee-chest position, said to relieve gas and stimulate abdominal organs, involves:

- standing straight with arms at the sides
- lifting the right knee toward the chest
- grasping the right ankle with the left hand
- pulling the leg as close to the chest as possible
- holding the position for about eight seconds
- repeating these steps with the left leg

The cobra position, which can be repeated as many as four time a day, involves:

- lying on the stomach with legs together
- placing the palms just below the shoulders, holding elbows close to the body
- inhaling, then lifting the head (face forward) and chest off the floor

- keeping the navel in contact with the floor
- looking as far upward as possible
- holding this position for three to six seconds
- exhaling and lowering the chest

Prognosis

Changes in diet and exercise usually eliminate the problem.

Prevention

Most Americans consume between 11–18 g of fiber a day. Consumption of 30 grams of fiber and between six and eight glasses of water each day can generally prevent constipation.

Thirty-five grams of fiber a day (an amount equal to five servings of fruits and vegetables, and a large bowl of high-fiber cereal) can relieve constipation.

Daily use of 500 mg vitamin C and 400 mg magnesium can prevent constipation. If symptoms do occur, each dosage can be increased by 100 mg a day, up to a maximum of 5,000 mg vitamin C and 1,000 mg magnesium. Use of preventive doses should be resumed after relief occurs, and vitamin C should be decreased to the pre-diarrhea dosage if the patient develops diarrhea.

Sitting on the toilet for 10 minutes at the same time every day, preferably after a meal, can induce regular bowel movements. This may not become effective for a few months, and it is important to defecate whenever necessary.

Fiber supplements containing psyllium (*Plantago psyllium*) usually become effective within about 48 hours and can be used every day without causing dependency. Powdered flaxseed (*Linum usitatissimum*) works the same way. Insoluble fiber, like wheat or oat bran, is as effective as psyllium but may give the patient gas at first.

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Maureen Haggerty

Constitutional homeopathic remedies see
Homeopathic remedies, constitutional prescribing

Consumption see **Tuberculosis**

Contact dermatitis

Definition

Contact **dermatitis** is the name for any skin inflammation that occurs when the skin's surface comes in contact with a substance originating outside the body. There are two kinds of contact dermatitis, irritant and allergic.

Description

Thousands of natural and man-made substances can cause contact dermatitis, which is the most common skin condition requiring medical attention and the foremost source of work-related disease. Florists, domestic workers, hairdressers, food preparers, and employees in industry, construction, and health care are the people most at risk of contracting work-related contact dermatitis. Americans spend roughly \$300 million a year in their quest for relief from contact dermatitis, not counting the considerable sums devoted by governments and businesses to regulating and policing the use of skin-threatening chemicals in the workplace. But exactly how many people suffer from contact dermatitis remains unclear; a 1997 article in the *Journal of the American Medical Association* notes that figures ranging from 1% to 15% have been put forward for Western industrial nations.

Causes and symptoms

Irritant contact dermatitis (ICD) is the more commonly reported of the two kinds of contact dermatitis, and is seen in about 80% of cases. It can be caused by soaps, detergents, solvents, adhesives, fiberglass, and other substances that are able to directly injure the skin. Most attacks are slight and confined to the hands and forearms, but can affect any part of the body that comes

in contact with an irritating substance. The symptoms can take many forms: redness, **itching**, crusting, swelling, blistering, oozing, dryness, scaliness, thickening of the skin, and a feeling of warmth at the site of contact. In extreme cases, severe blistering can occur and open sores can form. Jobs that require frequent skin exposure to water, such as hairdressing and food preparation, can make the skin more susceptible to ICD.

Allergic contact dermatitis (ACD) results when repeated exposure to an allergen (an allergy-causing substance) triggers an immune response that inflames the skin. Tens of thousands of drugs, pesticides, cosmetics, food additives, commercial chemicals, and other substances have been identified as potential allergens. Fewer than 30, however, are responsible the majority of ACD cases. Common culprits include poison ivy, poison oak, and poison sumac; fragrances and preservatives in cosmetics and personal care products; latex items such as gloves and condoms; and formaldehyde. Many people find that they are allergic to the nickel in inexpensive jewelry. ACD is usually confined to the area of skin that comes in contact with the allergen, typically the hands or face. Symptoms range from mild to severe and resemble those of ICD; a patch test may be needed to determine which kind of contact dermatitis a person is suffering from.

Diagnosis

Diagnosis begins with a **physical examination** and asking the patient questions about his or her health and daily activities. When contact dermatitis is suspected, the doctor attempts to learn as much as possible about the patient's hobbies, workplace duties, use of medications and cosmetics, etc.—anything that might shed light on the source of the disease. In some cases, an examination of the home or workplace is undertaken. If the dermatitis is mild, responds well to treatment, and does not recur, ordinarily the investigation is at an end. More difficult cases require patch testing to identify the allergen.

Two methods of patch testing are currently used. The most widely used method, the Finn chamber method, employs a multiwell, aluminum patch. Each well is filled with a small amount of the allergen being tested and the patch is taped to normal skin on the patient's upper back. After 48 hours, the patch is removed and an initial reading is taken. A second reading is made a few days later. The second method of patch testing involves applying a small amount of the test substance to directly to normal skin and covering it with a dressing that keeps air out and keeps the test substance in (occlusive dressing). After 48 hours, the dressing is taken off to see if a reaction has occurred. Identifying the allergen may require repeated testing, can take weeks or months, and is not always suc-



The abdomen of a male patient afflicted with contact dermatitis, triggered by an allergic reaction to a nickel belt buckle. (Photograph by Dr. P. Marazzi, Custom Medical Stock Photo. Reproduced by permission.)

cessful. Moreover, patch testing works only with ACD, though it is considered an essential step in ruling out ICD.

Treatment

The best treatment for contact dermatitis is to identify the allergen or irritating substance and avoid further contact with it. If the culprit is, for instance, a cosmetic, avoidance is a simple matter, but in some situations, such as an allergy to an essential workplace chemical for which no substitute can be found, avoidance may be impossible or force the sufferer to find new work or make other drastic changes in his or her life. Barrier creams and protective clothing such as gloves, masks, and long-sleeved shirts are ways of coping with contact dermatitis when avoidance is impossible, though they are not always effective.

For the symptoms themselves, treatments in mild cases include cool compresses and nonprescription lotions and ointments. When the symptoms are severe, **corticosteroids** applied to the skin or taken orally are used. Contact dermatitis that leads to a bacterial skin infection is treated with **antibiotics**.

Alternative treatment

Herbal remedies have been used for centuries to treat skin disorders including contact dermatitis. An experienced herbalist can recommend the remedies that

KEY TERMS

Antibiotics—Substances used against microorganisms that cause infection.

Corticosteroids—A group of anti-inflammatory substances often used to treat skin conditions.

Immune response—The protective reaction by the immune system against foreign antigens (substances that the body perceives as potentially dangerous). The immune system combats disease by neutralizing or destroying antigens.

will be most effective for an individual's condition. Among the herbs often recommended are:

- Burdock (*Arctium lappa*) minimizes inflammation and boosts the immune system. It is taken internally as a tea or tincture (a concentrated herbal extract prepared with alcohol).
- Calendula (*Calendula officinalis*) is a natural antiseptic and anti-inflammatory agent. It is applied topically in a lotion, ointment, or oil to the affected area.
- Aloe (*Aloe barbadensis*) soothes skin irritations. The gel is applied topically to the affected area.

A homeopath treating a patient with contact dermatitis will do a thorough investigation of the individual's history and exposures before prescribing a remedy. One homeopathic remedy commonly prescribed to relieve the itching associated with contact dermatitis is *Rhus toxicodendron* taken internally three to four times daily.

Poison ivy, poison oak, and poison sumac are common culprits in cases of allergic contact dermatitis. Following exposure to these plants, rash development may be prevented by washing the area with soap and water within 15 minutes of exposure. The leaves of jewelweed (*Impatiens spp.*), which often grows near poison ivy, may neutralize the poison-ivy allergen if rubbed on the skin right after contact. Several topical remedies may help relieve the itching associated with allergic contact dermatitis, including the juice of plantain leaves (*Plantago major*); a paste made of equal parts of green clay and goldenseal root (*Hydrastis canadensis*); a paste made of salt, water, clay, and peppermint (*Mentha piperita*) oil; and calamine lotion.

Prognosis

If the offending substance is promptly identified and avoided, the chances of a quick and complete recovery are excellent. Otherwise, symptom management—not cure—is the best doctors can offer. For some people,

contact dermatitis becomes a chronic and disabling condition that can have a profound effect on employability and quality of life.

Prevention

Avoidance of known or suspected allergens or irritating substances is the best prevention. If avoidance is difficult, barrier creams and protective clothing can be tried. Skin that comes in contact with an offending substance should be thoroughly washed as soon as possible.

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Howard Baker

Contact lenses see **Eye glasses and contact lenses**

Continent urinary diversion see **Urinary diversion surgery**

Continuous ambulatory electrocardiography see **Holter monitoring**

Continuous positive airway see **Inhalation therapies**

Contraception

Definition

Contraception (birth control) prevents **pregnancy** by interfering with the normal process of ovulation, fer-



Various types of contraception. (Photo Researchers, Inc. Reproduced by permission.)

tilization, and implantation. There are different kinds of birth control that act at different points in the process.

Purpose

Every month, a woman's body begins the process that can potentially lead to pregnancy. An egg (ovum) matures, the mucus that is secreted by the cervix (a cylindrical-shaped organ at the lower end of the uterus) changes to be more inviting to sperm, and the lining of the uterus grows in preparation for receiving a fertilized egg. Any woman who wants to prevent pregnancy must use a reliable form of birth control.

Birth control (contraception) is designed to interfere with the normal process and prevent the pregnancy that could result. There are different kinds of birth control that act at different points in the process, from ovulation, through fertilization, to implantation. Each method has its own side effects and risks. Some methods are more reliable than others.

Although there are many different types of birth control, they can be divided into a few groups based on how they work. These groups include:

- Hormonal methods—These use medications (hormones) to prevent ovulation. Hormonal methods include birth control pills (**oral contraceptives**), Depo Provera injections and Norplant.
- Barrier methods—These methods work by preventing the sperm from getting to and fertilizing the egg. Barri-

Types Of Contraceptives

Effectiveness	Predicted (%)	Actual (%)
Birth control pills	99.9	97
Condoms	98	88
Depo Provera	99.7	99.7
Diaphragm	94	82
IUDs	99.2	97
Norplant	99.7	99.7
Tubal sterilization	99.8	99.6
Spermicides	97	79
Vasectomy	99.9	99.9

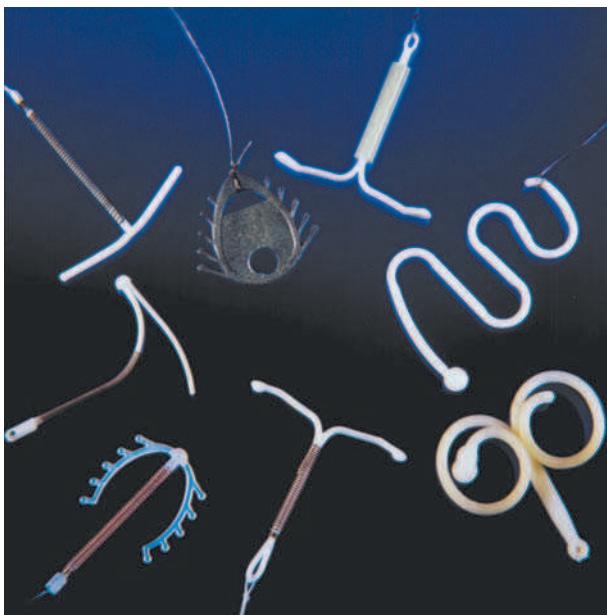
er methods include the **condom**, diaphragm, and cervical cap. The condom is the only form of birth control that also protects against **sexually transmitted diseases**, including HIV (the virus that causes **AIDS**).

- Spermicides—These medications kill sperm on contact. Most spermicides contain nonoxynol-9. Spermicides come in many different forms such as jelly, foam, tablets, and even a transparent film. All are placed in the vagina. Spermicides work best when they are used at the same time as a barrier method.
- Intrauterine devices—Intrauterine contraceptive devices (IUDs) are inserted into the uterus, where they stay from one to 10 years. An **IUD** prevents the fertilized egg from implanting in the lining of the uterus, and may have other effects as well.
- Tubal sterilization—Tubal sterilization is a permanent form of contraception for women. Each fallopian tube is either tied or burned closed. The sperm cannot reach the egg, and the egg cannot travel to the uterus.
- Vasectomy—is the male form of sterilization, and should also be considered permanent. In **vasectomy**, the vas defrens, the tiny tubes that carry the sperm into the semen, are cut and tied off. Thus, no sperm can get into the semen.

Unfortunately, there is no perfect form of birth control. Only abstinence (not having sexual intercourse) can protect against unwanted pregnancy with 100% reliability. The failure rates, which means the rates of pregnancy, for most forms of birth control are quite low. However, some forms of birth control are more difficult or inconvenient to use than others. In actual practice, the birth control methods that are more difficult or inconvenient have much higher failure rates because they are not used faithfully.

Description

All the different forms of birth control have one thing in common. They are only effective if used faithfully. Birth control pills will work only if taken every day; the



A variety of intrauterine contraceptive devices. The probability of a pregnancy for year of use is about 2 to 3%. IUDs made with copper coils should be replaced every 3 to 5 years. (Photo Researchers, Inc. Reproduced by permission.)

diaphragm is effective only if used during every episode of sexual intercourse. The same is true for condoms and the cervical cap. Some methods are automatically working every day, no matter what. These methods include Depo Provera, Norplant, the IUD, and tubal sterilization.

There are many different ways to use birth control. They can be divided into several groups:

- By mouth (oral)—Birth control pills must be taken by mouth every day.
- Injected—Depo Provera is a hormonal medication that is given by injection every three months.
- Implanted—Norplant is a long-acting hormonal form of birth control that is implanted under the skin of the upper arm.
- Vaginal—Spermicides and barrier methods work in the vagina.
- Intra-uterine—The IUD is inserted into the uterus.
- Surgical—Tubal sterilization is a form of surgery. A doctor must perform the procedure in a hospital or surgical clinic. Many women need general anesthesia.

The methods of birth control differ from each other in the timing of when they are used. Some methods of birth control must be used specifically at the time of sexual intercourse (condoms, diaphragm, cervical cap, spermicides). All other methods of birth control must be working all the time to provide protection (hormonal

methods, IUDs, tubal sterilization).

Precautions

There are risks associated with some forms of birth control. Some of the risks of each method are listed below:

- Birth control pills—The hormone (estrogen) in birth control pills can increase the risk of **heart attack** in women over 40 who smoke.
- IUD—The IUD can increase the risk of serious pelvic infection. The IUD can also injure the uterus by poking into or through the uterine wall. Surgery might be needed to fix this.
- Tubal sterilization—“Tying the tubes” is a surgical procedure and has all the risks of any other surgery, including the risks of anesthesia, infection, and bleeding.

Preparation

No specific preparation is needed before using contraception. However, a woman must be sure that she is not already pregnant before using a hormonal method or having an IUD placed.

Aftercare

No aftercare is needed.

Risks

Many methods of birth control have side effects. Knowing the side effects can help a woman to determine which method of birth control is right for her.

- Hormonal methods—The hormones in birth control pills, Depo Provera, and Norplant can cause changes in menstrual periods, changes in mood, weight gain, **acne**, and headaches. In addition, it may take many months to begin ovulating again once a woman stops using Depo Provera or Norplant.
- Barrier methods—A woman must insert the diaphragm in just the right way to be sure that it works properly. Some women get more urinary tract infections if they use a diaphragm. This is because the diaphragm can press against the urethra, the tube that connects the bladder to the outside.
- Spermicides—Some women and men are allergic to spermicides or find them irritating to the skin.
- IUD—The IUD is a foreign body that stays inside the uterus, and the uterus tries to get it out. A woman may have heavier menstrual periods and more menstrual cramping with an IUD in place.

KEY TERMS

Fallopian tubes—The thin tubes that connect the ovary to the uterus. Ova (eggs) travel from the ovary to the uterus. If the egg has been fertilized, it can implant in the uterus.

Fertilization—The joining of the sperm and the egg; conception.

Implantation—The process in which the fertilized egg embeds itself in the wall of the uterus.

Ovulation—The release of an egg (ovum) from the ovary.

- **Tubal sterilization**—Some women report increased menstrual discomfort after **tubal ligation**. It is not known if this is related to the tubal ligation itself.

There is no perfect form of birth control. Every method has a small failure rate and side effects. Some methods carry additional risks. However, every method of birth control can be effective if used properly.

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Contractures

Definition

Contractures are the chronic loss of joint motion due to structural changes in non-bony tissue. These non-bony tissues include muscles, ligaments, and tendons.

Description

Contractures can occur at any joint of the body. This joint dysfunction may be a result of **immobilization** from injury or disease; nerve injury, such as spinal cord damage and **stroke**; or muscle, tendon, or ligament disease.

Causes and symptoms

There are a number of pathologies and diseases that can lead to joint contractures. The primary causes resulting in a joint contraction are muscle imbalance, **pain**, prolonged bed rest, and immobilization. Because of the frequency of **fractures** and surgery, immobilization is the most frequent cause of joint contractures. Symptoms include a significant loss of motion to any specific joint that results in immobility. If the contracture is of a significant degree, pain can result even without any voluntary joint movement.

Diagnosis

Manual testing of joint mobility by a healthcare professional skilled in joint mobilization techniques (e.g. a physical therapist) will identify indications of restricted structures within the joint. Measuring the motion of the joint with a device termed a "goniometer" can be useful if the decrease of motion can be shown to be a proven result of a joint contracture. X rays can be of some benefit in the diagnosis of contractures, because a visible decrease in joint space may indicate a tight, contracted joint. Most physicians will make the diagnosis after a thorough **physical examination** involving physical and manual testing of the joint motion.

Treatment

Manual techniques

Joint mobilization and stretching of soft tissues is a common technique used to increase joint elasticity. Structures are stretched in similar directions to those which take place upon normal joint motion. Some healthcare professionals may use some form of heat prior to the stretching and mobilization. If appropriate, **exercise** may follow manual techniques to help maintain the additional motion achieved.

Mechanical techniques

Devices known as continuous passive motion machines are very popular, especially following surgery of joints. Continuous passive motion machines (CPM) are specifically adjusted to each individual's need. This method is administered within the first 24–72 hours after the injury or surgery. The joint is mechanically moved through the patient's tolerable motion. CPM machines have been proved to accelerate the return motion process, allowing patients more function in less time.

KEY TERMS

Mobilization—Making movable, restoring the power of motion in a joint. Movement which increases joint mobility.

Muscle tone—Also termed tonus; the normal state of balanced tension in the tissues of the body, especially the muscles.

Casting or splinting

Casting or splinting techniques are used to provide a constant stretch to the soft tissues surrounding a joint. It is most effective when used to increase motion of a joint from prolonged immobilization. It is also popular for treating contractures resulting from an increase in muscle tone from nerve injury. After an initial holding cast is applied for seven to 10 days, a series of positional casts are applied at weekly intervals. Before the application of each new cast, the joint is moved as much as can be tolerated by the patient, and measured by a goniometer. When as much motion as possible is obtained after stretching, another final cast is applied to maintain the newly acquired motion.

Surgery

In some cases the contracture may be severe and not respond to conservative treatment. In this event, manipulation of the joint under a general anesthesia may be necessary.

Alternative treatment

In some areas of the body, **chiropractic** techniques have been found to be useful to improve motion. **Massage therapy** can be beneficial by promoting additional circulation to joint structures, causing better elasticity. **Yoga** can help prevent as well as rehabilitate a contracture and can facilitate the return of joint mobility.

Prognosis

Prognosis of contractures will depend upon the cause of the contracture. In general, the earlier the treatment for the contracture begins, the better the prognosis.

Prevention

Prevention of contractures and deformities from **spinal cord injury**, fracture, and immobilization is achieved through a program of positioning, splinting if

appropriate, and range-of-motion exercises either manually or mechanically aided. These activities should be started as early as possible for optimal results.

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ORGANIZATIONS

The American College of Rheumatology. 1800 Century Place, Suite 250, Atlanta, GA 30345. (404) 633-3777. <<http://www.rheumatology.org>>.

American Physical Therapy Association. 1111 North Fairfax St., Alexandria, Virginia 22314. (800) 999-2782. <<https://www.apta.org>>.

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Conversion disorder see **Somatoform disorders**

Cooley's anemia see **Thalassemia**

Cooling treatments

Definition

Cooling treatments lower body temperature in order to relieve **pain**, swelling, constriction of blood vessels, and to decrease the likelihood of cellular damage by slowing the metabolism. Sponge baths, cold compresses, and cold packs are all wet cooling treatments. Dry treatments, such as ice bags and chemical cold packs, are also used to lower body temperature.

Purpose

The most common reason for cooling a body is **fever** or hyperthermia (extremely high fever). The body can sustain temperatures up to 104°F (40°C) with relative safety; however, when temperatures rise above 104°F (40°C), damage to the brain, muscles, blood, and kidneys is increasingly likely. Cooling treatments are also applied immediately following sprains, **bruises**, **burns**, eye injuries, and muscle spasms to help alleviate the resulting swelling, pain, and discoloration of the skin.

Cooling treatments slow chemical reactions within the body. For this reason, cooling tissues below normal temperature (98.6°F/37°C) can prevent injury from inadequate oxygen or **nutrition**. Cold water drowning victims suffering from **hypothermia** (cooling of the body below its normal temperature) have been successfully resuscitated after long periods underwater without medical complications because of this effect. For the past 40 years, heart surgeons have been experimenting with hypothermia to protect tissues from lack of blood circulation during an operation. Neurosurgeons are also working with hypothermia to protect the very sensitive brain tissues during periods of absent or reduced blood flow.

Description

Depending on the medical need, various cooling methods are used.

- Cold packs and ice bags are placed on a localized site and provide topical relief. These compresses should be covered with a waterproof material to protect the skin. Repeated treatments produce the desired pain and swelling relief.
- Cold treatments are placed on the groin and under the arms to treat hyperthermia. Treatments are refreshed periodically until the appropriate temperature is attained.
- A tepid sponge bath relieves fever without cooling the body too fast. Eighty degrees Fahrenheit is still 20°F below body temperature and yet warm enough not to drive blood from the skin, thereby preventing the cooling from getting to the body's core. Limbs are bathed first and then the chest, abdomen, back, and buttocks.
- Perfusion of isolated regions like the brain by using cooled blood is an experimental treatment, offering promising results for the treatment of stroke.

Preparation

Topical treatments are prepared with ice, cold water (59°F/15°C), and chemical cold packs. Tepid baths should be 80–93°F (26.7–34°C).

Risks

Small children, adults with circulation problems, and the elderly are all at risk of tissue damage. Rapid cooling causes chills, which in effect raise the body's temperature by raising its metabolism. Blood clots may form from thickened blood caused by the temperature change.

Resources

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Coombs' tests

Definition

Coombs' tests are blood tests that identify the causes of anemia.

Purpose

Anemia, which literally means no blood, refers to blood with abnormally low oxygen-carrying capacity. The hemoglobin in red blood cells carries oxygen. One of the many causes of anemia is destruction of red blood cells, a process called hemolysis (*hemo* means blood and *lysis* means disintegration). A simple **blood count** detects anemia. Even the test done before a blood donation can identify anemia. To detect hemolysis requires other tests. The Coombs' tests are conducted in order to determine the cause of anemia.

One characteristic of hemolysis is the autoimmune response against the body's red blood cells. Instead of protecting the body from outside agents, the immune system attacks parts of its own body with a deluge of antibodies. Autoimmunity is thought to be the cause of many collagen-vascular diseases, including **rheumatoid arthritis** and **systemic lupus erythematosus**. It is also the cause of the autoimmune hemolytic **anemias**. The Coombs' tests detect the antibodies responsible for the destruction of the red blood cells.

Causes of autoimmune **hemolytic anemia** include:

- drugs such as penicillin, methyldopa (lowers blood pressure), and quinidine (treats heart rhythm disturbances)
- cancers of the lymph system—Hodgkin's disease and lymphomas
- virus infections
- collagen-vascular diseases
- mismatched blood transfusions
- Rh incompatibility between a mother and fetus. This disease is called erythroblastosis fetalis

KEY TERMS

Antibody—A protein made by the immune system and used as a weapon against foreign invaders in the body.

Antigen—The chemical that stimulates an immune response.

Anemia—Reduced oxygen-carrying capacity of the blood, due to too little hemoglobin or too few red blood cells.

Collagen-vascular disease—Various diseases inflaming and destroying connective tissue.

Hematologist—Physician who specializes in diseases of the blood.

Hemoglobin—The red pigment in blood that carries oxygen.

Hemolysis—Breaking apart red blood cells.

Rh—A blood typing group, like the ABO system. When a mother is Rh negative and her baby is Rh positive, she may develop antibodies to the baby's blood that will cause it to hemolyze.

Many times the cause cannot be identified.

Description

There are two Coombs' tests. A direct Coombs' test detects the two different antigens that might induce hemolysis in the patient's red blood cells. An indirect Coombs' test looks for antibodies to someone else's red blood cells in the patient's serum (the blood without the cells). Combining the two tests gives clues to the origin of the hemolysis.

Preparation

No preparation is needed for this test. It will probably be among the second or third set of blood tests done after anemia is diagnosed and there is a suspicion that its cause is hemolysis.

Aftercare

Coombs' tests are done on blood that is drawn from the arm.

Risks

Taking blood for testing is the most common medical procedure performed. The worst complication is a

bruise at the site of the puncture or punctures. It is extremely rare for the needle to injure an important structure such as an artery or a nerve.

Normal results

If the Coombs' tests are negative, the anemia is unlikely to be autoimmune, and the hematologist will have to search elsewhere for a cause.

Abnormal results

If the test is positive, the antigens that react will narrow the search for a cause. Coombs' tests are also done for blood **transfusion** reactions to determine why the transfused blood did not match, and when there is a chance a newborn may have an Rh problem.

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Teresa Norris, RN

Coordination tests see **Balance and coordination tests**

COPD see **Emphysema; Chronic obstructive lung disease**

Copper deficiency see **Mineral deficiency**

Copper excess see **Wilson's disease**

Cor pulmonale

Definition

Cor pulmonale is an increase in bulk of the right ventricle of the heart, generally caused by chronic diseases or malfunction of the lungs. This condition can lead to **heart failure**.

Description

Cor pulmonale, or pulmonary heart disease, occurs in 25% of patients with chronic obstructive pulmonary dis-

ease (COPD). In fact, about 85% of patients diagnosed with cor pulmonale have COPD. Chronic **bronchitis** and **emphysema** are types of COPD. High blood pressure in the blood vessels of the lungs (**pulmonary hypertension**) causes the enlargement of the right ventricle. In addition to COPD, cor pulmonale may also be caused by lung diseases, such as **cystic fibrosis**, **pulmonary embolism**, and pneumoconiosis. Loss of lung tissue after **lung surgery** or certain chest-wall disturbances can produce cor pulmonale, as can neuromuscular diseases, such as **muscular dystrophy**. A large pulmonary thromboembolism (blood clot) may lead to acute cor pulmonale.

Causes and symptoms

Any respiratory disease or malfunction that affects the circulatory system of the lungs may lead to cor pulmonale. These circulatory changes cause the right ventricle to compensate for the extra work required to pump blood through the lungs. The right ventricle has thin walls and is crescent-shaped. The resulting pressure causes the right ventricle to dilate and bulge, eventually leading to its failure.

Cor pulmonale should be expected in any patient with COPD and other respiratory or neuromuscular diseases. Initial symptoms of cor pulmonale may actually reflect those of the underlying disease. These may include chronic coughing, **wheezing**, weakness, **fatigue**, and **shortness of breath**. **Edema** (abnormal buildup of fluid), weakness, and discomfort in the upper chest may be evident in cor pulmonale.

Diagnosis

An electrocardiograph (EKG) will show signs such as frequent premature contractions in the atria or ventricles. Chest x rays may show enlargement of the right descending pulmonary artery. This sign, along with an enlarged main pulmonary artery, indicates pulmonary artery **hypertension** in patients with COPD. **Magnetic resonance imaging** (MRI) is often the preferred method of diagnosis for cor pulmonale because it can clearly show and measure volume of the pulmonary arteries. Other tests used to support a diagnosis of cor pulmonale may include arterial **blood gas analysis**, pulmonary function tests, and **hematocrit**.

Treatment

Treatment of cor pulmonale is aimed at increasing a patient's **exercise tolerance** and improving oxygen levels of the arterial blood. Treatment is also aimed at the underlying condition that is producing cor pulmonale. Common treatments include **antibiotics** for respiratory

KEY TERMS

Ventricle—A cavity, as in the brain or heart. The right ventricle of the heart drives blood from the heart into the pulmonary artery, which supplies blood to the lungs.

infection; anticoagulants to reduce the risk of thromboembolism; and digitalis, oxygen, and **phlebotomy** to reduce red blood cell count. A low-salt diet and restricted fluids are often prescribed.

Alternative treatment

Co-management of the patient with cor pulmonale should be coordinated between the medical doctor and the alternative practitioner. The first step in treatment is to determine the cause of the condition and to evaluate all organ systems of the body. Dietary considerations, for example, a low-salt diet and reduced fluid intake aimed at reducing the edema associated with cor pulmonale, can be supportive aspects of treatment.

Prognosis

The prognosis for cor pulmonale is poor, particularly because it occurs late in the process of serious disease.

Prevention

Cor pulmonale is best prevented by prevention of COPD and other irreversible diseases that lead to heart failure. **Smoking** cessation is critically important. Carefully following the recommended course of treatment for the underlying disease may help prevent cor pulmonale.

Resources

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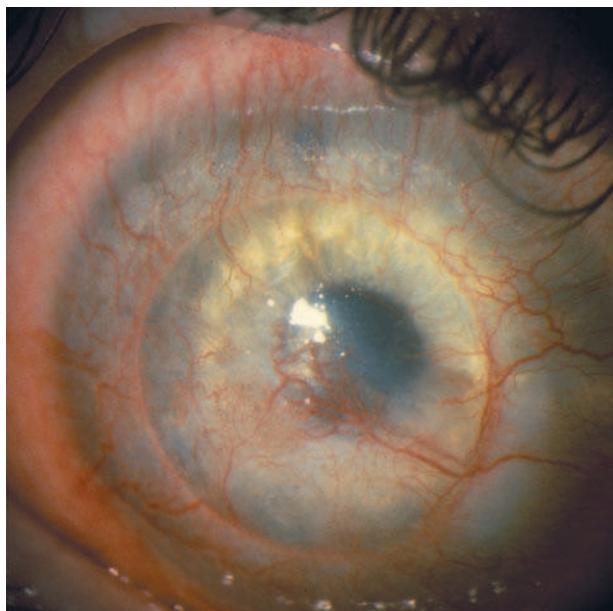
American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

J. Ricker Polsdorfer, MD

Cori's disease see **Glycogen storage diseases**

Corkscrew esophagus see **Diffuse esophageal spasm**



A close-up view of an abrasion on patient's cornea. (Photograph by Dennis R. Cain, CRA, Custom Medical Stock Photo. Reproduced by permission.)

Corneal abrasion

Definition

A corneal abrasion is a worn or scraped-off area of the outer, clear layer of the eye (cornea).

Description

The cornea is the clear, dome-shaped outer area of the eye. It lies in front of the colored part of the eye (iris) and the black hole in the iris (pupil). The outermost layer of the eyeball consists of the cornea and the white part of the eye (sclera). A corneal abrasion is basically a superficial cut or scrape on the cornea. A corneal abrasion is not as serious as a corneal ulcer, which is generally deeper and more severe than an abrasion.

Causes and symptoms

A corneal abrasion is usually the result of direct injury to the eye, often from a fingernail scratch, makeup brushes, contact lenses, foreign body, or even twigs. Patients often complain of feeling a foreign body in their eye, and they may have pain, sensitivity to light, or tearing.

Diagnosis

Ophthalmologists and optometrists, who treat eye disorders, are well qualified to diagnose corneal abrasions. The doctor will check the patient's vision (visual acuity) in both eyes with an eye chart. A patient history will also be taken, which may help to determine the cause of the abrasion. A slit lamp, which is basically a microscope and light source, will allow the doctor to see the abrasion. Fluorescein, a yellow dye, may be placed into the eye to determine the extent of the abrasion. The fluorescein will temporarily stain the affected area.

Treatment

The cornea has a remarkable ability to heal itself, so treatment is designed to minimize complications. If the abrasion is very small, the doctor might just suggest an eye lubricant and a follow-up visit the next day. A very small abrasion should heal in one to two days; others

usually in one week. However, to avoid a possible infection, an antibiotic eye drop may be prescribed. Sometimes additional eye drops may make the eye feel more comfortable. Depending upon the extent of the abrasion, some doctors may patch the affected eye. It is very important to go for the follow-up checkup to make sure an infection does not occur. Use of contact lenses should not be resumed without the doctor's approval.

Prognosis

In typical cases, the prognosis is good. The cornea will heal itself, usually within several days. A very deep abrasion may lead to scarring. If the abrasion does not heal properly, a recurrent corneal erosion (RCE) may result months or even years later. The symptoms are the same as for an abrasion (e.g., tearing, foreign body sensation, and blurred vision), but it will keep occurring. Similar or additional treatment for the RCE may be necessary.

Prevention

Everyone should wear eye protection whenever this is recommended. This should be standard practice when using power tools and playing certain sports. Goggles should even be worn when mowing the lawn, because a twig can be thrown upward toward the face. Contact lens wearers should be careful to follow their doctors' instructions on caring for and wearing their lenses. Ill-fitting or dirty lenses could lead to an abrasion, so patients should go for their prescribed checkups.

Resources

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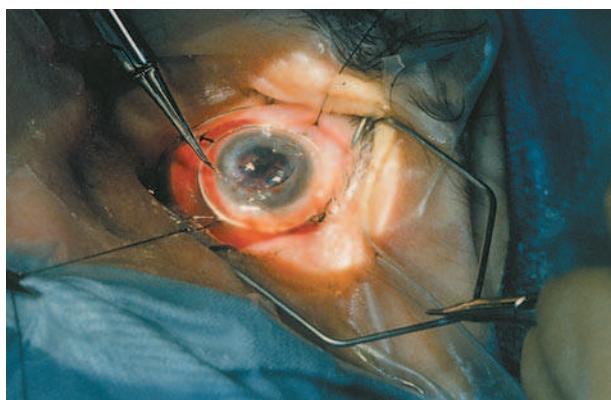
ORGANIZATIONS

- American Academy of Family Physicians. 8880 Ward Parkway, Kansas City, MO 64114. (816) 333-9700. <<http://www.aafp.org>>.

Richard H. Lampert

Corneal infection see **Keratitis**

Corneal keratoplasty see **Corneal transplantation**



A corneal transplant in progress. (Photograph by Chet Szymecki, Phototake NYC. Reproduced by permission.)

Corneal transplantation

Definition

In corneal transplant, also known as keratoplasty, a patient's damaged cornea is replaced by the cornea from the eye of a human cadaver. This is the single most common type of human transplant surgery and has the highest success rate. Eye banks acquire and store eyes from donor individuals largely to supply the need for transplant corneas.

Purpose

Corneal transplant is used when vision is lost in an eye because the cornea has been damaged by disease or traumatic injury. Some of the disease conditions that might require corneal transplant include the bulging outward of the cornea (keratoconus), a malfunction of the inner layer of the cornea (Fuchs' dystrophy), and painful swelling of the cornea (pseudophakic bullous keratopathy). Some of these conditions cause cloudiness of the cornea; others alter its natural curvature, which can also reduce the quality of vision.

Injury to the cornea can occur because of chemical burns, mechanical trauma, or infection by viruses, bacteria, fungi, or protozoa. The herpes virus produces one of the more common infections leading to corneal transplant.

Surgery would only be used when damage to the cornea is too severe to be treated with corrective lenses. Occasionally, corneal transplant is combined with other types of eye surgery (such as **cataract surgery**) to solve multiple eye problems in one procedure.

Precautions

Corneal transplant is a very safe procedure that can be performed on almost any patient who would benefit from it. Any active infection or inflammation of the eye usually needs to be brought under control before surgery can be performed.

Description

The cornea is the transparent layer of tissue at the very front of the eye. It is composed almost entirely of a special type of collagen. It normally contains no blood vessels, but because it contains nerve endings, damage to the cornea can be very painful.

In a corneal transplant, a disc of tissue is removed from the center of the eye and replaced by a corresponding disc from a donor eye. The circular incision is made using an instrument called a trephine. In one form of corneal transplant (penetrating keratoplasty), the disc removed is the entire thickness of the cornea and so is the replacement disc. Over 90% of all corneal transplants in the United States are of this type. In lamellar keratoplasty, on the other hand, only the outer layer of the cornea is removed and replaced.

The donor cornea is attached with extremely fine sutures. Surgery can be performed under anesthesia that is confined to one area of the body while the patient is awake (local anesthesia) or under anesthesia that places the entire body of the patient in a state of unconsciousness (general anesthesia.) Surgery requires 30–90 minutes.

Over 40,000 corneal transplants are performed in the United States each year. Medicare reimbursement for a corneal transplant in one eye was about \$1,200 in 1997.

A less common but related procedure called epikeratophakia involves suturing the donor cornea directly

KEY TERMS

Cadaver—The human body after death.

Cataract—A condition of cloudiness of the lens of the eye.

Cornea—The transparent layer of tissue at the very front of the eye.

Corticosteroids—Synthetic hormones widely used to fight inflammation.

Epikeratophakia—A procedure in which the donor cornea is attached directly onto the host cornea.

Epithelial cells—Cells that form a thin surface coating on the outside of a body structure.

Fibrous connective tissue—Dense tissue found in various parts of the body containing very few living cells.

Fuchs' dystrophy—A hereditary disease of the inner layer of the cornea. Treatment requires penetrating keratoplasty. The lens of the eye may also be affected and require surgical replacement at the same time as the cornea.

Glaucoma—A vision defect caused when excessive fluid pressure within the eye damages the optic nerve.

Histocompatibility antigens—Proteins scattered throughout body tissues that are unique for almost every individual.

Keratoconus—An eye condition in which the cornea bulges outward, interfering with normal vision. Usually both eyes are affected.

Pseudophakic bullous keratopathy—Painful swelling of the cornea occasionally occurring after surgery to implant an artificial lens in place of a lens affected by cataract.

Retinal detachment—A serious vision disorder in which the light-detecting layer of cells inside the eye (retina) is separated from its normal support tissue and no longer functions properly.

Trephine—A small surgical instrument that is rotated to cut a circular incision.

onto the surface of the existing host cornea. The only tissue removed from the host is the extremely thin epithelial cell layer on the outside of the host cornea. There is no permanent damage to the host cornea, and this procedure can be reversed. It is usually employed in children. In adults, the use of contact lenses can usually achieve the same goals.

Preparation

No special preparation for corneal transplant is needed. Some eye surgeons may request the patient have a complete **physical examination** before surgery. The patient may also be asked to skip breakfast on the day of surgery.

Aftercare

Corneal transplant is often performed on an outpatient basis, although some patients need brief hospitalization after surgery. The patient will wear an eye patch at least overnight. An eye shield or glasses must be worn to protect the eye until the surgical wound has healed. Eye drops will be prescribed for the patient to use for several weeks after surgery. These drops include **antibiotics** to prevent infection as well as **corticosteroids** to reduce inflammation and prevent graft rejection.

For the first few days after surgery, the eye may feel scratchy and irritated. Vision will be somewhat blurry for as long as several months.

Sutures are often left in place for six months, and occasionally for as long as two years.

Risks

Corneal transplants are highly successful, with over 90% of operations in United States achieving restoration of sight. However, there is always some risk associated with any surgery. Complications that can occur include infection, **glaucoma**, **retinal detachment**, cataract formation, and rejection of the donor cornea.

Graft rejection occurs in 5–30% of patients, a complication possible with any procedure involving tissue transplantation from another person (allograft). Allograft rejection results from a reaction of the patient's immune system to the donor tissue. Cell surface proteins called histocompatibility antigens trigger this reaction. These antigens are often associated with vascular tissue (blood vessels) within the graft tissue. Since the cornea normally contains no blood vessels, it experiences a very low rate of rejection. Generally, blood typing and **tissue typing** are not needed in corneal transplants, and no close match between donor and recipient is required. Symp-

toms of rejection include persistent discomfort, sensitivity to light, redness, or a change in vision.

If a rejection reaction does occur, it can usually be blocked by steroid treatment. Rejection reactions may become noticeable within weeks after surgery, but may not occur until 10 or even 20 years after the transplant. When full rejection does occur, the surgery will usually need to be repeated.

Although the cornea is not normally vascular, some corneal diseases cause vascularization (the growth of blood vessels) into the cornea. In patients with these conditions, careful testing of both donor and recipient is performed just as in transplantation of other organs and tissues such as hearts, kidneys, and bone marrow. In such patients, repeated surgery is sometimes necessary in order to achieve a successful transplant.

Cornea donors are carefully screened. Individuals with infectious diseases are not accepted as donors.

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- American Academy of Ophthalmology. 655 Beach Street, P.O. Box 7424, San Francisco, CA 94120-7424. <<http://www.eyenet.org>>.

Victor Leipzig, PhD



A close-up view of an ulcer on cornea. (Custom Medical Stock Photo. Reproduced by permission.)

injury from exposure and from **foreign objects**. Infection and injury cause inflammation of the cornea—a condition called **keratitis**. Tissue loss because of inflammation produces an ulcer. The ulcer can either be centrally located, thus greatly affecting vision, or peripherally located. There are about 30,000 cases of bacterial corneal ulcers in the United States each year.

Description

The most common cause of corneal ulcers is germs, but most of them cannot invade a healthy cornea with adequate tears and a functioning eyelid. They gain access because injury has impaired these defense mechanisms. A direct injury from a foreign object inoculates germs directly through the outer layer of the cornea, just as it does to the skin. A caustic chemical can inflame the cornea by itself or so damage it that germs can invade. Improper use of contact lenses has become a common cause of corneal injury. Eyelid or tear function failure is the other way to make the eye vulnerable to infection. Tears and the eyelid together wash the eye and prevent foreign material from settling in. Tears contain enzymes and other substances to help protect against infection. Certain diseases dry up tear production, leaving the cornea dry and defenseless. Other diseases paralyze or weaken the eyelids so that they cannot effectively protect and cleanse the eyes.

Causes and symptoms

Viruses, bacteria, fungi, and a protozoan called *Acanthamoeba* can all invade the cornea and damage it under suitable conditions.

- Bacteria from a common **conjunctivitis** (pink eye) rarely spread to the cornea, but can if untreated.

Corneal ulcers

Definition

The cornea, the clear front part of the eye through which light passes, is subject to many infections and to

KEY TERMS

Fluorescein—A fluorescent chemical used to examine the cornea.

Germ—A disease-causing microorganism.

Inflammation—The body's reaction to irritation.

Topical corticosteroids—Cortisone and related drugs used on the skin and in the eye, usually for allergic conditions.

- Fecal bacteria are more likely to be able to infect the cornea.
- A bacterium called *Pseudomonas aeruginosa*, which can contaminate eyedrops, is particularly able to cause corneal infection.
- A group of incomplete bacteria known as *Chlamydia* can be transmitted to the eye directly by flies or dirty hands. One form of chlamydial infection is the leading cause of blindness in developing countries and is known as Egyptian ophthalmia or **trachoma**. Another type of *Chlamydia* causes a sexually transmitted disease.
- Other sexually transmitted diseases—for example, syphilis—can affect the cornea.

The most common viruses to damage the cornea are adenoviruses and herpes viruses. Viral and fungal infections are often caused by improper use of topical **corticosteroids**. If topical corticosteroids are used in a patient with herpes simplex keratitis, the ulcer can get much worse and blindness could result.

Symptoms are obvious. The cornea is intensely sensitive, so corneal ulcers normally produce severe **pain**. If the corneal ulcer is centrally located, vision is impaired or completely absent. Tearing is present and the eye is red. It hurts to look at bright lights.

Diagnosis

The doctor will take a case history to try to determine the cause of the ulcer. This can include improper use of contact lenses; injury, such as a scratch from a twig; or severe dry eye. An instrument called a slit lamp will be used to examine the cornea. The slit lamp is a microscope with a light source that magnifies the cornea, allowing the extent of the ulcer to be seen. Fluorescein, a yellow dye, may be used to illuminate further detail. If a germ is responsible for the ulcer, identification may require scraping samples directly from the cornea, conjunctiva, and lids, and sending them to the laboratory.

Treatment

A corneal ulcer needs to be treated aggressively, as it can result in loss of vision. The first step is to eliminate infection. Broad spectrum **antibiotics** will be used before the lab results come back. Medications may then be changed to more specifically target the cause of the infection. A combination of medications may be necessary. Patients should return for their follow-up visits so that the doctor can monitor the healing process. The cornea can heal from many insults, but if it remains scarred, **corneal transplantation** may be necessary to restore vision. If the corneal ulcer is large, hospitalization may be necessary.

Prognosis

Treated early enough, corneal infections will usually resolve, perhaps even without the formation of an ulcer. However, left untreated, infections can lead to ulcers and the corneal ulcer can result in scarring or perforation of the cornea. Other problems may occur as well, including **glaucoma**. Patients with certain systemic diseases that impede healing (such as **diabetes mellitus** or **rheumatoid arthritis**) may need more aggressive treatment. The later the treatment, the more damage will be done and the more scarring will result. Corneal transplant is standard treatment with a high probability of success.

Prevention

Attentive care of contact lenses will greatly reduce the incidence of corneal damage and ulceration. Germs that cause no problems in the mouth or on the hands can damage the eye, so contact lens wearers must wash their hands before touching their lenses and must not use saliva to moisten them. Tap water should not be used to rinse the lenses. Contacts should be removed whenever there is irritation and left out until the eyes are back to normal. It is not advisable to wear contact lenses while swimming or in hot tubs. Daily wear contact lenses have been found to be less of a risk than contacts for overnight wear (extended wear). Organisms have been cultured from contact lens cases, so the cases should be rinsed in hot water and allowed to air dry. Cases should be replaced every three months. Patients should follow their doctors' schedules for replacement of the contacts.

Eye protection in the workplace, or wherever tiny particles are flying around, is essential. Ultraviolet (UV) coatings on glasses or sunglasses can help protect the eyes from the sun's rays. Goggles with UV protection should be worn when skiing or in suntanning salons to protect against UV rays. Prompt attention to any red eye should prevent progressive damage.

For people with inadequate tears, use of artificial tears eyedrops will prevent damage from drying. Eyelids that do not close adequately may temporarily have to be sewn shut to protect the eye until more lasting treatment can be instituted.

Resources

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American Academy of Ophthalmology. 655 Beach Street, P.O. Box 7424, San Francisco, CA 94120-7424. <<http://www.eyenet.org>>.

American Optometric Association. 243 North Lindbergh Blvd., St. Louis, MO 63141. (314) 991-4100. <<http://www.aoanet.org>>.

Prevent Blindness America. 500 East Remington Road, Schaumburg, IL 60173. (800) 331-2020. <<http://www.preventblindness.org>>.

J. Ricker Polsdorfer, MD



Corns on toes. (Custom Medical Stock Photo. Reproduced by permission.)

Types of corns

A hard corn is a compact lump with a thick core. Hard corns usually form on the tops of the toes, on the outside of the little toe, or on the sole of the foot.

A soft corn is a small, inflamed patch of skin with a smooth center. Soft corns usually appear between the toes.

A seed corn is the least common type of corn. Occurring only on the heel or ball of the foot, a seed corn consists of a circle of stiff skin surrounding a plug of cholesterol.

Types of calluses

A plantar callus, a callus that occurs on the sole of the foot, has a white center. Hereditary calluses develop where there is no apparent friction, run in families, and occur most often in children.

Causes and symptoms

Corns and calluses form to prevent injury to skin that is repeatedly pinched, rubbed, or irritated. The most common causes are:

- shoes that are too tight or too loose, or have very high heels
- tight socks or stockings
- deformed toes

KEY TERMS

Ayurveda—Ayurveda is a system of wholistic medicine from India that aims to bring the individual into harmony with nature. It provides guidance regarding food and lifestyle, so that healthy people can stay healthy and people with health challenges can improve their health.

Bursitis—Inflammation of a bursa, a fluid-filled cavity or sac. In the body, bursae are located at places where friction might otherwise develop.

- walking down a long hill, or standing or walking on a hard surface for a long time

Jobs or hobbies that cause steady or recurring pressure on the same spot can also cause calluses.

Symptoms include hard growths on the skin in response to direct pressure. Corns may be extremely sore and surrounded by inflamed, swollen skin.

Diagnosis

Corns can be recognized on sight. A family physician or podiatrist may scrape skin off what seems to be a callus, but may actually be a wart. If the lesion is a wart, it will bleed. A callus will not bleed, but will reveal another layer of dead skin.

Treatment

Corns and calluses do not usually require medical attention unless the person who has them has **diabetes mellitus**, poor circulation, or other problems that make self-care difficult.

Treatment should begin as soon as an abnormality appears. The first step is to identify and eliminate the source of pressure. Placing moleskin pads over corns can relieve pressure, and large wads of cotton, lamb's wool, or moleskin can cushion calluses.

Using hydrocortisone creams or soaking feet in a solution of Epsom salts and very warm water for at least five minutes a day before rubbing the area with a pumice stone will remove part or all of some calluses. Rubbing corns just makes them hurt more.

Applying petroleum jelly or lanolin-enriched hand lotion helps keep skin soft, but corn-removing ointments that contain acid can damage healthy skin. They should never be used by pregnant women or by people who are diabetic or who have poor circulation.

It is important to see a doctor if the skin of a corn or callus is cut, because it may become infected. If a corn discharges pus or clear fluid, it is infected. A family physician, podiatrist, or orthopedist may:

- remove (debride) affected layers of skin
- prescribe oral **antibiotics** to eliminate infection
- drain pus from infected corns
- inject cortisone into the affected area to decrease pain or inflammation
- perform surgery to correct toe deformities or remove bits of bone

Alternative treatment

Standing and walking correctly can sometimes eliminate excess foot pressure. Several types of bodywork can help correct body imbalances. Bodywork is a term used for any of a number of systems, including **Aston-Patterning**, the **Feldenkrais method**, and **rolfing**, that manipulate the body through massage, movement education, or meditational techniques.

Aloe (*Aloe barbadensis*) cream is an effective skin softener, and two or three daily applications of calendula (*Calendula officinalis*) salve can soften skin and prevent inflammation. One teaspoon of lemon juice mixed with one teaspoon of dried chamomile (*Matricaria recutita*) tea and one crushed garlic clove dissolves thickened skin.

An ayurvedic practitioner may recommend the following treatment:

- apply each day a paste made by combining one teaspoon of aloe vera gel with half that amount of turmeric (*Circuma longa*)
- bandage overnight
- soak in warm water for 10 minutes every morning
- massage gently with mustard (*Brassica cruciferae*) oil

Prognosis

Most corns and calluses disappear about three weeks after the pressure that caused them is eliminated. They are apt to recur if the pressure returns.

Extreme pain can change the way a person stands or walks. Such changes can, in turn, cause pain in the ankle, back, hip, or knee.

Bursitis, a painful, inflamed fluid-filled sac, can develop beneath a corn. An ulcer or broken area within a corn can reach to the bone. Infection can have serious consequences for people who have diabetes or poor circulation.

Prevention

Corns and calluses can usually be prevented by avoiding friction-causing activities and wearing shoes that fit properly, are activity-appropriate, and are kept in good repair. Soles and heels that wear unevenly may indicate a need for corrective footwear or special insoles. Socks and stockings should not cramp the toes. Gloves, kneepads, and other protective gear should also be worn as needed.

Feet should be measured, while standing, whenever buying new shoes. It is best to shop for shoes late in the day, when feet are likely to be swollen. It is also important to buy shoes with toe-wiggling room and to try new shoes on both feet.

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American Podiatric Medical Association. 9312 Old Georgetown Road, Bethesda, MD 20814-1698. (301) 571-9200. <<http://www.apma.org>>.

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Maureen Haggerty

Coronary artery bypass graft surgery

Definition

Coronary artery bypass graft surgery is a surgical procedure in which one or more blocked coronary arteries are bypassed by a blood vessel graft to restore normal blood flow to the heart. These grafts usually come from the patient's own arteries and veins located in the leg, arm, or chest.

Purpose

Coronary artery bypass graft surgery (also called coronary artery bypass surgery, CABG, and bypass oper-

ation) is performed to restore blood flow to the heart. This relieves chest pain and ischemia, improves the patient's quality of life, and in some cases, prolongs the patient's life. The goals of the procedure are to enable the patient to resume a normal lifestyle and to lower the risk of a heart attack.

The decision to perform coronary artery bypass graft surgery is a complex one, and there is some disagreement among experts as to when it is indicated. Many experts feel that it has been performed too frequently in the United States. According to the American Heart Association, appropriate candidates for coronary artery bypass graft surgery include patients with blockages in at least three major coronary arteries, especially if the blockages are in arteries that feed the heart's left ventricle; patients with angina so severe that even mild exertion causes chest pain; and patients who cannot tolerate percutaneous transluminal coronary angioplasty and do not respond well to drug therapy. It is well accepted that coronary artery bypass graft surgery is the treatment of choice for patients with severe coronary artery disease (three or more diseased arteries with impaired function in the left ventricle).

Precautions

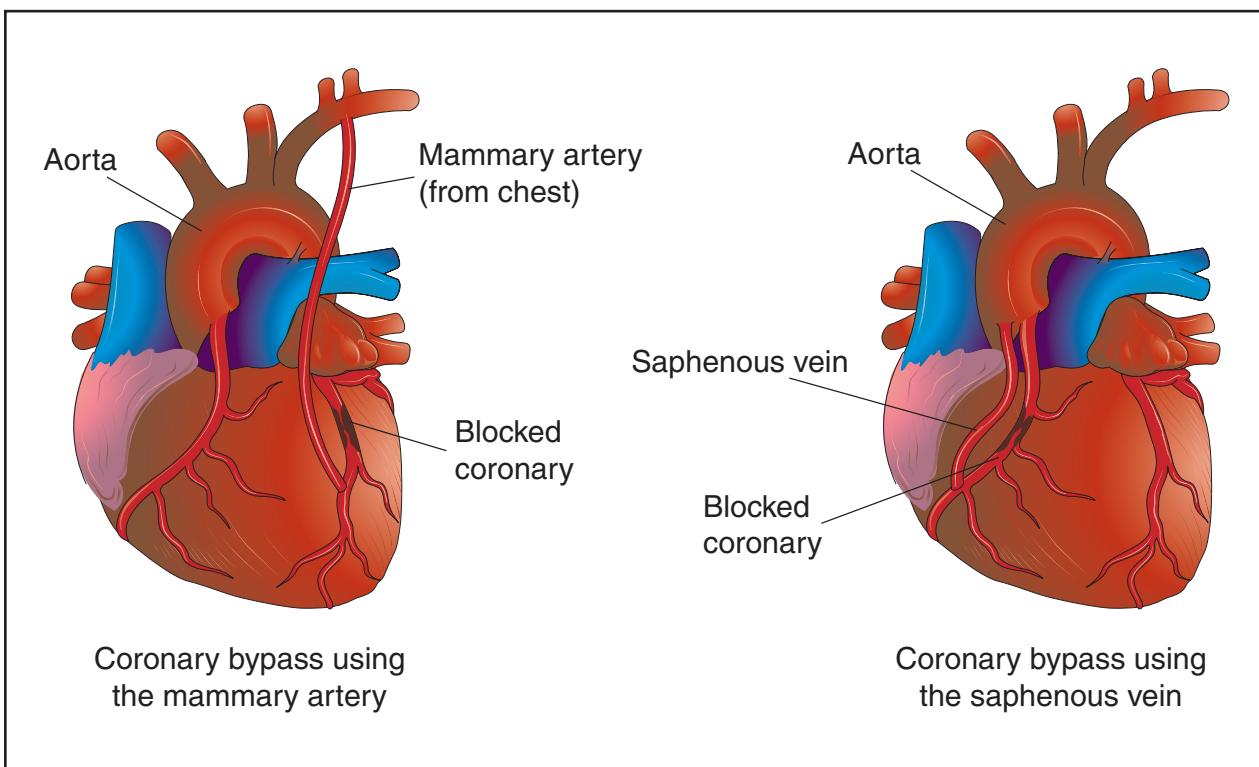
Coronary artery bypass graft surgery should ideally be postponed for three months after a heart attack. Patients should be medically stable before the surgery, if possible.

Description

Coronary artery bypass graft surgery builds a detour around one or more blocked coronary arteries with a graft from a healthy vein or artery. The graft goes around the clogged artery (or arteries) to create new pathways for oxygen-rich blood to flow to the heart.

Coronary artery bypass graft surgery is major surgery performed in a hospital. The length of the procedure depends upon the number of arteries being bypassed, but it generally takes from four to six hours—sometimes longer. The average hospital stay is four to seven days. Full recovery from coronary artery bypass graft surgery takes three to four months. Within four to six weeks, people with sedentary office jobs can return to work; people with physical jobs must wait longer and sometimes change careers.

Coronary artery bypass graft surgery is widely performed in the United States. The American Heart Association estimates that 573,000 coronary artery bypass graft surgeries were performed on 363,000 patients in 1995. Seventy-four percent of these procedures were performed on men and 44% on men and women under the age of 65 (1995 data). The estimated average cost of this procedure in 1995 was \$44,820.



Coronary artery bypass graft surgery builds a detour around one or more blocked coronary arteries with a graft from a healthy vein or artery. The graft goes around the clogged artery (or arteries) to create new pathways for oxygen-rich blood to flow to the heart. (Illustration by Electronic Illustrators Group.)

Procedure

The surgery team for coronary artery bypass graft surgery includes the cardiovascular surgeon, assisting surgeons, a cardiovascular anesthesiologist, a perfusion technologist (who operates the heart-lung machine), and specially trained nurses. After general anesthesia is administered, the surgeon removes the veins or prepares the arteries for grafting. If the saphenous vein is to be used, a series of incisions are made in the patient's thigh or calf. More commonly, a segment of the internal mammary artery will be used and the incisions are made in the chest wall. The surgeon then makes an incision from the patient's neck to navel, saws through the breastbone, and retracts the rib cage open to expose the heart. The patient is connected to a heart-lung machine, also called a cardiopulmonary bypass pump, that cools the body to reduce the need for oxygen and takes over for the heart and lungs during the procedure. The heart is then stopped and a cold solution of potassium-enriched normal saline is injected into the aortic root and the coronary arteries to lower the temperature of the heart, which prevents damage to the tissue.

Next, a small opening is made just below the blockage in the diseased coronary artery. Blood will be redirect-

ed through this opening once the graft is sewn in place. If a leg vein is used, one end is connected to the coronary artery and the other to the aorta. If a mammary artery is used, one end is connected to the coronary artery while the other remains attached to the aorta. The procedure is repeated on as many coronary arteries as necessary. Most patients who have coronary artery bypass graft surgery have at least three grafts done during the procedure.

Electric shocks start the heart pumping again after the grafts have been completed. The heart-lung machine is turned off and the blood slowly returns to normal body temperature. After implanting pacing electrodes (if needed) and inserting a chest tube, the surgeon closes the chest cavity.

Success rate of coronary artery bypass graft surgery

About 90% of patients experience significant improvements after coronary artery bypass graft surgery. Patients experience full relief from chest pain and resume their normal activities in about 70% of the cases; the remaining 20% experience partial relief. In 5–10% of coronary artery bypass graft surgeries, the bypass graft stops supplying blood to the bypassed artery within one year. Younger peo-

ple who are healthy except for the heart disease do well with bypass surgery. Patients who have poorer results from coronary artery bypass graft surgery include those over the age of 70, those who have poor left ventricular function, or are undergoing a repeat surgery or other procedures concurrently, and those who continue **smoking**, do not treat **high cholesterol** or other coronary risk factors, or have another debilitating disease.

Long term, symptoms recur in only about 3–4% of patients per year. Five years after coronary artery bypass graft surgery, survival expectancy is 90%, at 10 years it is about 80%, at 15 years it is about 55%, and at 20 years it is about 40%.

Angina recurs in about 40% of patients after about 10 years. In most cases, it is less severe than before the surgery and can be controlled by drug therapy. In patients who have had vein grafts, 40% of the grafts are severely obstructed 10 years after the procedure. Repeat coronary artery bypass graft surgery may be necessary, and is usually less successful than the first surgery.

Minimally invasive coronary artery bypass graft surgery

There are two new types of minimally invasive coronary artery bypass graft surgery: port-access coronary artery bypass (also called PACAB or PortCAB) and minimally invasive coronary artery bypass (also called MID-CAB). These procedures are minimally invasive because they do not require the neck-to-navel incision, sawing through the breastbone, or opening the rib cage to expose the heart. Both procedures enable surgeons to work on the coronary arteries through small chest holes called ports and other small incisions. Port-access coronary artery bypass requires the use of a heart-lung machine but minimally invasive coronary artery bypass does not. Advantages of these procedures over standard coronary artery bypass graft surgery include a shorter hospital stay, a shorter recovery period, and lower costs.

Port-access coronary artery bypass enables surgeons to perform bypasses through smaller incisions. Using a video monitor to view the procedure, the surgeon passes instruments through ports in the patient's chest to perform the bypass. Mammary arteries or leg veins are used for the grafts. Minimally invasive coronary artery bypass is performed on a beating heart and is appropriate only for bypasses of one or two arteries. Small ports are made in the patient's chest, along with a small incision directly over the coronary artery to be bypassed. Generally, the surgeon uses a mammary artery for the bypass.

Early data on outcomes for port-access coronary artery bypass and minimally invasive coronary artery bypass are favorable. Mortality rates with port-access coronary artery

bypass and minimally invasive coronary artery bypass are both less than 3%—about the same as in standard coronary artery bypass graft surgery. One clinical trial indicated that survival at seven years was the same in minimally invasive coronary artery bypass and standard coronary artery bypass graft surgery, but that another intervention was necessary five times more often with minimally invasive coronary artery bypass than with standard coronary artery bypass graft surgery. The American Heart Association Council on Cardio-Thoracic and Vascular Surgery feels that both procedures appear promising but that further study is needed. More data covering longer term outcomes are necessary in order to fully assess these procedures.

Preparation

The patient is usually admitted to the hospital the day before the coronary artery bypass graft surgery is scheduled. Coronary **angiography** has been previously performed to show the surgeon where the arteries are blocked and where the grafts might best be positioned. The patient is given a blood-thinning drug—usually heparin—that helps to prevent blood clots. The evening before the surgery, the patient showers with antiseptic soap and is shaved from chin to toes. After midnight, food and fluids are restricted. A sedative is prescribed on the morning of surgery and sometimes the night before. Heart monitoring begins.

Aftercare

The patient recovers in a surgical intensive care unit for at least the first two days after the surgery. He or she is connected to chest and breathing tubes, a mechanical ventilator, a heart monitor and other monitoring equipment, and a urinary catheter. The breathing tube and ventilator are usually removed within six hours of surgery, but the other tubes remain in place as long as the patient is in the intensive care unit. Drugs are prescribed to control pain and to prevent unwanted blood clotting. The patient is closely monitored. Vital signs and other parameters, such as heart sounds and oxygen and carbon dioxide levels in arterial blood, are checked frequently. The chest tube is checked to ensure that it is draining properly. The patient is fed intravenously for the first day or two. Daily doses of **aspirin** are started within six to 24 hours after the procedure. Chest physiotherapy is started after the ventilator and breathing tube are removed. The therapy includes coughing, turning frequently, and taking deep breaths. Other exercises will be encouraged to improve the patient's circulation and prevent complications due to prolonged bed rest.

If there are no complications, the patient begins to resume a normal routine around the second day. This includes eating regular food, sitting up, and walking

KEY TERMS

Aorta—The main artery which carries blood from the heart to the rest of the body. The aorta is the largest artery in the body.

Graft—To implant living tissue surgically. In coronary artery bypass graft surgery, healthy veins or arteries are grafted to coronary arteries.

Mammary artery—A chest wall artery that descends from the aorta and is commonly used for bypass grafts.

Saphenous vein—A long vein in the thigh or calf commonly used for bypass grafts.

Ventricles—The left and right ventricles are the large chambers of the heart. The ventricles propel blood to the lungs and the rest of the body.

around a little bit. Before being released from the hospital, the patient usually spends a few days under observation in a non-surgical unit. During this time, counseling is usually provided on eating right and starting a light **exercise** program to keep the heart healthy. Patients should eat a lot of fruits, vegetables, grains, and non-fat or low-fat dairy products, and reduce fats to less than 30% of all calories. An exercise program will usually be tailored for the patient, who will be encouraged to participate in a **cardiac rehabilitation** program where exercise will be supervised by professionals. Cardiac **rehabilitation** programs, offered by hospitals and other organizations, may also include classes on heart-healthy living.

Full recovery from coronary artery bypass graft surgery takes three to four months and is a gradual process. Upon release from the hospital, the patient will feel weak because of the extended bed rest in the hospital. Within a few weeks, the patient should begin to feel stronger.

While the incision scar from coronary artery bypass graft surgery heals, which takes one to two months, it may be sore. The scar should not be bumped, scratched, or otherwise disturbed. An exercise test is often conducted after the patient leaves the hospital to determine how effective the surgery was and to confirm that progressive exercise is safe.

Risks

Coronary artery bypass graft surgery is major surgery and patients may experience any of the complications associated with major surgery. The risk of **death** during

coronary artery bypass graft surgery is two to three percent. Possible complications include graft closure and development of blockages in other arteries, long-term development of atherosclerotic disease of saphenous vein grafts, abnormal heart rhythms, high or low blood pressure, blood clots that can lead to a **stroke** or heart attack, infections, and depression. There is a higher risk for complications in patients who are heavy smokers, patients who have serious lung, kidney, or metabolic problems, or patients who have a reduced supply of blood to the brain.

Resources

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ORGANIZATIONS

- American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>. Texas Heart Institute. Heart Information Service. P.O. Box 20345, Houston, TX 77225-0345. <<http://www.tmc.edu/thi>>.

Lori De Milto

Coronary artery disease

Definition

Coronary artery disease is a narrowing or blockage of the arteries and vessels that provide oxygen and nutri-

ents to the heart. It is caused by **atherosclerosis**, an accumulation of fatty materials on the inner linings of arteries. The resulting blockage restricts blood flow to the heart. When the blood flow is completely cut off, the result is a **heart attack**.

Description

Coronary artery disease, also called coronary heart disease or heart disease, is the leading cause of **death** for both men and women in the United States. According to the American Heart Association, in 1995 one in every 4.8 deaths in the United States was caused by coronary artery disease. About every 29 seconds, one American will have a heart attack; about every minute, one American will die from a heart attack. Fourteen million Americans have active symptoms of coronary artery disease (heart attack or chest pains). Many millions more have silent coronary disease, the first indication of which can be sudden death.

Coronary artery disease occurs when the coronary arteries become partially blocked or clogged. This blockage limits the flow of blood from the coronary arteries, which are the major arteries supplying oxygen-rich blood to the heart. The coronary arteries expand when the heart is working harder and needs more oxygen. Arteries would expand, for example, when a person is climbing stairs, exercising, or having sex. If the arteries are unable to expand, the heart is deprived of oxygen (**myocardial ischemia**). When the blockage is limited, chest **pain** or pressure, called **angina**, may occur. When the blockage cuts off the flow of blood, the result is heart attack (**myocardial infarction** or heart muscle death).

Healthy coronary arteries are clean, smooth, and slick. The artery walls are flexible and can expand to let more blood through when the heart needs to work harder. The disease process in arteries is thought to begin with an injury to the linings and walls of the arteries. This injury makes them susceptible to atherosclerosis and blood clots (**thrombosis**).

Causes and symptoms

Coronary artery disease is usually caused by atherosclerosis. Cholesterol and other fatty substances accumulate on the inner wall of the arteries. They attract fibrous tissue, blood components, and calcium and harden into artery-clogging plaques. Atherosclerotic plaques often form blood clots that can also block the coronary arteries (**coronary thrombosis**). Congenital defects and muscle spasms can also block blood flow. Recent research indicates that infection from organisms such as chlamydia bacteria may be responsible for some cases of coronary artery disease.

A number of major contributing factors increase the risk of developing coronary artery disease. Some of these can be changed and some cannot. People with more risk factors are more likely to develop coronary artery disease.

Major risk factors

Major risk factors significantly increase the chance of developing coronary artery disease. Those that cannot be changed are:

- Heredity—People whose parents have coronary artery disease are more likely to develop it. African-Americans are also at increased risk because they experience a higher rate of severe **hypertension** than whites do.
- Sex—Men are more likely to have heart attacks than women are and to have them at a younger age. Over age 60, however, women have coronary artery disease at a rate equal to that of men.
- Age—Men who are 45 years of age and older and women who are 55 years of age and older are more likely to have coronary artery disease. Occasionally, coronary disease may strike a person in the 30s. Older people (those over 65) are more likely to die of a heart attack. Older women are twice as likely as older men to die within a few weeks of a heart attack.

Major risk factors that can be changed are:

- Smoking—Smoking increases both the chance of developing coronary artery disease and the chance of dying from it. Smokers are two to four times more likely than are non-smokers to die of sudden heart attack. They are more than twice as likely as non-smokers to have a heart attack. They are also more likely to die within an hour of a heart attack. Second hand smoke may also increase risk.
- High cholesterol—Dietary sources of cholesterol are meat, eggs, and other animal products. The body also produces it. Age, sex, heredity, and diet affect one's blood cholesterol. Total blood cholesterol is considered high at levels above 240 mg/dL and borderline at 200–239 mg/dL. High-risk levels of low-density lipoprotein (LDL cholesterol) begin at 130–159 mg/dL, depending on other risk factors. Risk of developing coronary artery disease increases steadily as blood cholesterol levels increase above 160 mg/dL. When a person has other risk factors, the risk multiplies.
- High blood pressure—High blood pressure makes the heart work harder and weakens it over time. It increases the risk of heart attack, **stroke**, kidney failure, and congestive **heart failure**. A blood pressure of 140 over 90 or above is considered high. As the numbers rise, high blood pressure goes from Stage 1 (mild) to Stage 4 (very severe). In combination with **obesity**, **smoking**,

high cholesterol, or diabetes, high blood pressure raises the risk of heart attack or stroke several times.

- Lack of physical activity—Lack of **exercise** increases the risk of coronary artery disease. Even modest physical activity, like walking, is beneficial if done regularly.
- Diabetes mellitus—The risk of developing coronary artery disease is seriously increased for diabetics. More than 80% of diabetics die of some type of heart or blood vessel disease.

Contributing risk factors

Contributing risk factors have been linked to coronary artery disease, but their significance is not known yet. Contributing risk factors are:

- **Obesity**—Excess weight increases the strain on the heart and increases the risk of developing coronary artery disease even if no other risk factors are present. Obesity increases blood pressure and blood cholesterol and can lead to diabetes.
- **Stress** and anger—Some scientists believe that stress and anger can contribute to the development of coronary artery disease and increase the blood's tendency to form clots (thrombosis). Stress, the mental and physical reaction to life's irritations and challenges, increases the heart rate and blood pressure and can injure the lining of the arteries. Evidence shows that anger increases the risk of dying from heart disease. The risk of heart attack is more than double after an episode of anger.

Chest pain (angina) is the main symptom of coronary heart disease but it is not always present. Other symptoms include **shortness of breath**, chest heaviness, tightness, pain, a burning sensation, squeezing, or pressure either behind the breastbone or in the arms, neck, or jaws. Many people have no symptoms of coronary artery disease before having a heart attack; 63% of women and 48% of men who died suddenly of coronary artery disease had no previous symptoms of the disease, according to the American Heart Association.

Diagnosis

Diagnosis begins with a visit to the physician, who will take a medical history, discuss symptoms, listen to the heart, and perform basic screening tests. These tests will measure weight, blood pressure, blood lipid levels, and **fasting** blood glucose levels. Other diagnostic tests include resting and exercise electrocardiogram, **echocardiography**, radionuclide scans, and coronary **angiography**. The treadmill exercise (stress) test is an appropriate screening test for those with high risk factors even when they feel well.

An electrocardiogram (ECG) shows the heart's activity and may reveal a lack of oxygen (ischemia). Electrodes covered with conducting jelly are placed on the patient's chest, arms, and legs. They send impulses of the heart's activity through an oscilloscope (a monitor) to a recorder that traces them on paper. The test takes about 10 minutes and is performed in a physician's office. A definite diagnosis cannot be made from **electrocardiography**. About 50% of patients with significant coronary artery disease have normal resting electrocardiograms. Another type of electrocardiogram, known as the exercise **stress test**, measures how the heart and blood vessels respond to exertion when the patient is exercising on a treadmill or a stationary bike. This test is performed in a physician's office or an exercise laboratory. It takes 15–30 minutes. It is not perfectly accurate. It sometimes gives a normal reading when the patient has a heart problem or an abnormal reading when the patient does not.

If the electrocardiogram reveals a problem or is inconclusive, the next step is exercise echocardiography or nuclear scanning (angiography). Echocardiography, cardiac ultrasound, uses sound waves to create an image of the heart's chambers and valves. A technician applies gel to a hand-held transducer, then presses it against the patient's chest. The heart's sound waves are converted into an image that can be displayed on a monitor. It does not reveal the coronary arteries themselves, but can detect abnormalities in heart wall motion caused by coronary disease. Performed in a cardiology outpatient diagnostic laboratory, the test takes 30–60 minutes.

Radionuclide angiography enables physicians to see the blood flow of the coronary arteries. Nuclear scans are performed by injecting a small amount of radiopharmaceutical such as thallium into the bloodstream. A device that uses gamma rays to produce an image of the radioactive material (gamma camera) records pictures of the heart. Radionuclide scans are not dangerous. The radiation exposure is about the same as that in a **chest x ray**. The tiny amount of radioactive material used disappears from the body in a few days. Radionuclide scans cost about four times as much as exercise stress tests but provide more information.

In radionuclide angiography, a scanning camera passes back and forth over the patient who lies on a table. Radionuclide angiography is usually performed in a hospital's nuclear medicine department and takes 30–60 minutes. Thallium scanning is usually done in conjunction with an exercise stress test. When the stress test is finished, thallium or sestamibi is injected. The patient resumes exercise for one minute to absorb the thallium. For patients who cannot exercise, cardiac blood flow and heart rate may be increased by intravenous dipyridamole (Persantine) or adenosine. Thallium scanning is done

twice, immediately after injecting the radiopharmaceutical and again four hours (and maybe 24 hours) later. It is usually performed in a hospital's nuclear medicine department. Each scan takes 30–60 minutes.

Coronary angiography is the most accurate method for making a diagnosis of coronary artery disease, but it is also the most invasive. It is a form of **cardiac catheterization** that shows the heart's chambers, great vessels, and coronary arteries using x-ray technology. During coronary angiography the patient is awake but sedated. ECG electrodes are placed on the patient's chest and an intravenous line is inserted. A local anesthetic is injected into the site where the catheter will be inserted. The cardiologist inserts a catheter into a blood vessel and guides it into the heart. A contrast dye is injected to make the heart visible on x-ray cinematography. Coronary angiography is performed in a cardiac catheterization laboratory either in an outpatient or inpatient surgery unit. It takes from 30 minutes to two hours.

Treatment

Coronary artery disease can be treated many ways. The choice of treatment depends on the severity of the disease. Treatments include lifestyle changes and drug therapy, percutaneous transluminal coronary **angioplasty**, and coronary artery bypass surgery. Coronary artery disease is a chronic disease requiring lifelong care. Angioplasty or bypass surgery is not a "cure."

People with less severe coronary artery disease may gain adequate control through lifestyle changes and drug therapy. Many of the lifestyle changes that prevent disease progression—a low-fat, low-cholesterol diet, weight loss if needed, exercise, and not smoking—also help prevent the disease from developing. These lifestyle changes are discussed in more detail under prevention.

Drugs such as nitrates, beta-blockers, and calcium-channel blockers relieve chest pain and complications of coronary artery disease, but they cannot clear blocked arteries. Nitrates (nitroglycerin) improve blood flow to the heart. Beta-blockers (acebutolol, propranolol) reduce the amount of oxygen required by the heart during stress. One type of calcium-channel blocker (verapamil, diltiazem hydrochloride) helps keep the arteries open and reduces blood pressure. **Aspirin** helps prevent blood clots from forming on plaques, reducing the likelihood of a heart attack. Cholesterol-lowering medications are also indicated in most cases.

Percutaneous transluminal coronary angioplasty and bypass surgery are procedures that enter the body (invasive procedures) to improve blood flow in the coronary arteries. Percutaneous transluminal coronary angioplasty,

KEY TERMS

Atherosclerosis—A process in which the walls of the coronary arteries thicken due to the accumulation of plaque in the blood vessels. Atherosclerosis is the cause of coronary artery disease.

Angina—Chest pain that happens when diseased blood vessels restrict the flow of blood to the heart. Angina is often the first symptom of coronary artery disease.

Beta-blocker—A drug that blocks some of the effects of fight-or-flight hormone adrenaline (epinephrine and norepinephrine), slowing the heart rate and lowering the blood pressure.

Calcium-channel blocker—A drug that blocks the entry of calcium into the muscle cells of small blood vessels (arterioles) and keeps them from narrowing.

Coronary arteries—The main arteries that provide blood to the heart. The coronary arteries surround the heart like a crown, coming out of the aorta, arching down over the top of the heart, and dividing into two branches. These are the arteries in which coronary artery disease occurs.

HDL cholesterol—High-density lipoprotein cholesterol is a component of cholesterol that helps protect against heart disease. HDL is nicknamed "good" cholesterol

LDL Cholesterol—Low-density lipoprotein cholesterol is the primary cholesterol molecule. High levels of LDL increase the risk of coronary heart disease. LDL is nicknamed "bad" cholesterol.

Plaque—A deposit of fatty and other substances that accumulate in the lining of the artery wall.

Triglyceride—A fat that comes from food or is made from other energy sources in the body. Elevated triglyceride levels contribute to the development of atherosclerosis.

usually called coronary angioplasty, is a non-surgical procedure. A catheter tipped with a balloon is threaded from a blood vessel in the thigh into the blocked artery. The balloon is inflated, compressing the plaque to enlarge the blood vessel and open the blocked artery. The balloon is deflated, and the catheter is removed. Coronary angioplasty is performed by a cardiologist in a hospital and generally requires a stay of one or two days. Coronary angioplasty is successful about 90% of the

time, but for one-third of patients the artery narrows again within six months. The procedure can be repeated. It is less invasive and less expensive than coronary artery bypass surgery.

In coronary artery bypass surgery, a healthy artery or vein from an arm, leg, or chest wall is used to build a detour around the coronary artery blockage. The healthy vessel then supplies oxygen-rich blood to the heart. Bypass surgery is major surgery. It is appropriate for those patients with blockages in two or three major coronary arteries, those with severely narrowed left main coronary arteries, and those who have not responded to other treatments. It is performed in a hospital under general anesthesia. A heart-lung machine is used to support the patient while the healthy vein or artery is attached past the blockage to the coronary artery. About 70% of patients who have bypass surgery experience full relief from angina; about 20% experience partial relief. Only about 3–4% of patients per year experience a return of symptoms. Survival rates after bypass surgery decrease over time. At five years after surgery, survival expectancy is 90%; at 10 years about 80%, at 15 years about 55%, and at 20 years about 40%.

Three semi-experimental surgical procedures for unblocking coronary arteries are currently being studied. **Atherectomy** is a procedure in which the cardiologist shaves off and removes strips of plaque from the blocked artery. In laser angioplasty, a catheter with a laser tip is inserted into the affected artery to burn or break down the plaque. A metal coil called a stent can be implanted permanently to keep a blocked artery open. Stenting is becoming more common.

Alternative treatment

Natural therapies may reduce the risk of certain types of heart disease, but once symptoms appear, conventional medical attention is necessary. A healthy diet (including cold-water fish as a source of essential fatty acids) and exercise, important components of conventional prevention and treatment strategies, also are emphasized in alternative approaches to coronary artery disease. Herbal medicine has a variety of remedies that may have a beneficial effect on coronary artery disease. For example, ginger (*Zingiber officinale*) may help reduce cholesterol. Garlic (*Allium sativum*), ginger, and hot red or chili peppers are all circulatory enhancers that can help prevent blood clots. **Yoga** and other bodywork, massage, relaxation therapies, and talking therapies may also help prevent coronary artery disease and stop, or even reverse, the progression of atherosclerosis. Vitamin and mineral therapy to reduce, reverse, or protect against coronary artery disease includes chromium; calcium and

magnesium; B-complex **vitamins**; the anti-oxidant vitamins C and E; selenium; and zinc. **Traditional Chinese medicine** may recommend herbal remedies, massage, **acupuncture**, and dietary modification.

Prognosis

In many cases, coronary artery disease can be successfully treated. Advances in medicine and healthier lifestyles have caused a substantial decline in death rates from coronary artery disease since the mid-1980s. New diagnostic techniques enable doctors to identify and treat coronary artery disease in its earliest stages. New technologies and surgical procedures have extended the lives of many patients who would otherwise have died. Research on coronary artery disease continues.

Prevention

A healthy lifestyle can help prevent coronary artery disease and help keep it from progressing. A heart-healthy lifestyle includes eating right, regular exercise, maintaining a healthy weight, no smoking, moderate drinking, no recreational drugs, controlling hypertension, and managing stress. **Cardiac rehabilitation** programs are excellent to help prevent recurring coronary problems for people who are at risk and who have had coronary events and procedures.

Eat right

A healthy diet includes a variety of foods that are low in fat, especially saturated fat, low in cholesterol, and high in fiber. It includes plenty of fruits and vegetables and limited sodium. Some foods are low in fat but high in cholesterol and some are low in cholesterol but high in fat. Saturated fat raises cholesterol and, in excessive amounts, increases the amount of the clot-forming proteins in blood. Polyunsaturated and monounsaturated fats are good for the heart. Fat should comprise no more than 30% of total daily calories.

Cholesterol, a waxy substance containing fats, is found in foods such as meat, eggs, and other animal products. It is also produced in the liver. Soluble fiber can help lower cholesterol. Dietary cholesterol should be limited to about 300 milligrams per day. Many popular lipid-lowering drugs can reduce LDL cholesterol by an average of 25–30% when used with a low-fat, low-cholesterol diet.

Fruits and vegetables are rich in fiber, vitamins, and **minerals**. They are low-calorie and nearly fat free. Vitamin C and beta-carotene, found in many fruits and vegetables, keep LDL-cholesterol from turning into a form that damages coronary arteries.

Excess sodium can increase the risk of high blood pressure. Many processed foods contain large amounts of sodium. Limit daily intake to about 2,400 milligrams, about the amount in a teaspoon of salt.

The “Food Guide” Pyramid developed by the U.S. Departments of Agriculture and Health and Human Services provides easy-to-follow guidelines for daily heart-healthy eating. It recommends six to 11 servings of bread, cereal, rice, and pasta; three to five servings of vegetables; two to four servings of fruit; two to three servings of milk, yogurt, and cheese; and two to three servings of meat, poultry, fish, dry beans, eggs, and nuts. Fats, oils, and sweets should be used sparingly. Canola and olive oil are better for the heart than other cooking oils. Coronary patients should be on a strict diet.

Exercise regularly

Aerobic exercise can lower blood pressure, help control weight, and increase HDL (“good”) cholesterol. It may keep the blood vessels more flexible. The Centers for Disease Control and Prevention and the American College of Sports Medicine recommend moderate to intense aerobic exercise lasting about 30 minutes four or more times per week for maximum heart health. Three 10-minute exercise periods are also beneficial. Aerobic exercise—activities such as walking, jogging, and cycling—uses the large muscle groups and forces the body to use oxygen more efficiently. It can also include everyday activities such as active gardening, climbing stairs, or brisk housework. People with coronary artery disease or risk factors should consult a doctor before beginning an exercise program.

Maintain a desirable body weight

About one quarter of all Americans are overweight and nearly one-tenth are obese, according to the Surgeon General’s Report on **Nutrition** and Health. People who are 20% or more over their ideal body weight have an increased risk of developing coronary artery disease. Losing weight can help reduce total and LDL cholesterol, reduce triglycerides, and boost HDL cholesterol. It may also reduce blood pressure. Eating right and exercising are two key components of losing weight.

Avoid recreational drugs

Do not smoke or use tobacco. Smoking has many adverse effects on the heart. It increases the heart rate, constricts major arteries, and can create irregular heartbeats. It raises blood pressure, contributes to the development of plaque, increases the formation of blood clots, and causes blood platelets to cluster and impede blood flow. Heart damage caused by smoking can be repaired by quitting. Even heavy smokers can return to heart

health. Several studies have shown that ex-smokers face the same risk of heart disease as non-smokers within five to 10 years after they quit.

Drink in moderation. Modest consumption of alcohol may actually protect against coronary artery disease because alcohol appears to raise levels of HDL (“good”) cholesterol. The American Heart Association defines moderate consumption as one ounce of alcohol per day, roughly one cocktail, one 8-ounce glass of wine, or two 12-ounce glasses of beer. However, even moderate drinking can increase risk factors for heart disease for some people (by raising blood pressure, for example). Excessive drinking is always bad for the heart. It usually raises blood pressure and can poison the heart and cause abnormal heart rhythms or even heart failure.

Do not use other recreational drugs. Commonly used recreational drugs, particularly **cocaine** and “crack,” can seriously harm the heart and should never be used.

Seek treatment for hypertension

High blood pressure, one of the most common and serious risk factors for coronary artery disease, can be completely controlled through lifestyle changes and medication. Moderate hypertension can be controlled by reducing dietary intake of sodium and fat, exercising regularly, managing stress, abstaining from smoking, and drinking alcohol in moderation. People for whom these changes do not work or people with severe hypertension may be helped by many categories of medication.

Manage stress

Everyone experiences stress, the mental and physical reaction to life’s irritations and challenges. Stress can sometimes be avoided and when it is inevitable, it can be controlled. Techniques for controlling stress include: taking life more slowly, spending more time with family and friends, thinking positively, getting enough sleep, exercising, and practicing relaxation techniques.

Resources

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- American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.
- National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.
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Lori De Milton

Coronary disease see **Coronary artery disease**

Coronary heart disease see **Coronary artery disease**

Coronary stenting

Definition

A coronary stent is an artificial support device used in the coronary artery to keep the vessel open.

Purpose

The coronary stent is a relatively new tool used to keep coronary arteries expanded, usually following a balloon **angioplasty**. Balloon angioplasty is used in patients

with **coronary artery disease**. In this disease, the blood vessels on the heart become narrow. When this happens, the oxygen supply is reduced to the heart muscle. The primary cause of coronary artery disease is fat deposits blocking the arteries (**atherosclerosis**). In many cases, balloon angioplasty is unsuccessful and the vessel closes after the procedure (restenosis). By forming a rigid support, the stent can prevent restenosis and reduce the need for coronary bypass surgery. The stent is usually a stainless steel mesh tube. Since the stent will be placed inside an artery, the device comes in various sizes to match the size of the artery.

Precautions

Any foreign object in the body, like a stent, will increase the risk of thrombosis. Anticlotting medication is given to prevent this complication.

Description

Coronary stenting usually follows balloon angioplasty, which requires inserting a balloon catheter into the femoral artery in the upper thigh. When this catheter is positioned at the location of the blockage in the coronary artery, it is slowly inflated to widen that artery, and is then removed. The stent catheter is then threaded into the artery and the stent is placed around a deflated balloon. When this is correctly positioned in the coronary artery, the balloon is inflated, expanding the stent against the walls of the coronary artery. The balloon catheter is removed, leaving the stent in place to hold the coronary artery open. A cardiac **angiography** will follow to insure that the stent is keeping the artery open.

Alternative procedures

Balloon angioplasty and coronary stenting are performed to relieve the symptoms of coronary artery disease. By the time coronary artery disease progresses and requires balloon angioplasty, there is no alternative to balloon angioplasty other than coronary bypass surgery. Coronary bypass surgery carries greater risks. However, since coronary artery disease can be related to high fat diets, smoking, and lack of exercise, changes in lifestyle may reduce the risk of developing the disease. Various medications for cholesterol, high blood pressure, and diabetes also can help treat or prevent coronary artery disease.

Preparation

Before the stent is inserted, the patient will probably be instructed to take **aspirin** for several days. Aspirin can help decrease the possibility of blood clots forming at the

KEY TERMS

Balloon angioplasty—The use of a balloon attached to a catheter to widen an artery that has become narrowed. As the balloon is inflated, it opens the artery.

Cardiac angiography—A procedure used to visualize blood vessels of the heart. A catheter is used to inject a dye into the vessels; the vessels can then be seen by x ray.

Catheter—A long thin flexible tube that can be inserted into the body; in this case, it is threaded to the heart.

Restenosis—The narrowing of a blood vessel after it has been opened, usually by balloon angioplasty.

Thrombosis—The development of a blood clot in the vessels. This thrombosis may clog a blood vessel and stop the flow of blood.

stent. Because anesthesia will be used during the procedure, the patient should not eat or drink after midnight of the previous day.

Aftercare

Following the procedure, blood thinners (anticoagulants) will be given through a needle in a vein for about 24 hours. The patient should remain flat and still for awhile to allow the femoral artery to heal from the insertion of the catheter. Medication to control blood clotting should be taken after the patient is discharged from the hospital. A special diet may also be recommended that is low in vitamin K and cholesterol. With time, the patient should begin light exercise, like walking. It is important that no **magnetic resonance imaging** (MRI) tests are given for six months because the magnetic field may move the stent.

Risks

Although coronary stents greatly reduce the risk of restenosis following balloon angioplasty, there is still some risk that the stented artery may close. Thrombosis, bleeding, and artery damage are also risks.

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American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

OTHER

AdvocateHealthCare. <<http://www.advocatehealth.com>>.

Cindy L. A. Jones, PhD

Coronary thrombosis see **Heart attack**

Coronavirus infection see **Common cold**

Corticosteroids

Definition

Corticosteroids are a group of natural and synthetic analogues of the hormones secreted by the hypothalamic-anterior pituitary-adrenocortical (HPA) axis, more commonly referred to as the pituitary gland. These include glucocorticoids, which are anti-inflammatory agents with a large number of other functions; mineralocorticoids, which control salt and water balance primarily through action on the kidneys; and corticotropins, which control secretion of hormones by the pituitary gland.

Purpose

Glucocorticoids have multiple effects, and are used for a large number of conditions. They affect glucose utilization, fat metabolism, and bone development, and are potent anti-inflammatory agents. They may be used for replacement of natural hormones in patients with pituitary deficiency (**Addison's disease**), as well as for a wide number of other conditions including, but not limited to, arthritis, **asthma**, anemia, various cancers, and skin inflammations. Additional uses include inhibition of **nausea and vomiting** after **chemotherapy**, treatment of **septic shock**, treatment of spinal cord injuries, and treatment of hirsutism (excessive hair growth). The choice of drug will vary with the condition. Cortisone and hydrocortisone, which have both glucocorticoid and mineralocorticoid effects, are the drugs of choice for replacement therapy of natural hormone deficiency. Synthetic compounds, which have greater anti-inflammatory effects and less effect on salt and water balance, are usually preferred for other purposes. These compounds include dexamethasone, which is almost exclusively glucocorticoid in its actions, as well as prednisone, prednisolone,

KEY TERMS

Hallucination—A false or distorted perception of objects, sounds, or events that seems real. Hallucinations usually result from drugs or mental disorders.

Hormone—A substance that is produced in one part of the body, then travels through the bloodstream to another part of the body where it has its effect.

Inflammation—Pain, redness, swelling, and heat that usually develop in response to injury or illness.

Ointment—A thick, spreadable substance that contains medicine and is meant to be used on the outside of the body.

Pregnancy category—A system of classifying drugs according to their established risks for use during pregnancy. Category A: controlled human studies have demonstrated no fetal risk. Category B: animal studies indicate no fetal risk, but no human studies; or adverse effects in animals, but not in well-controlled human studies. Category C: no adequate human or animal studies; or adverse fetal effects in animal studies, but no available human data. Category D: evidence of fetal risk, but benefits outweigh risks. Category X: evidence of fetal risk. Risks outweigh any benefits.

betamethasone, trimacinalone, and others. Glucocorticoids are formulated in oral dosage forms, topical creams and ointments, oral and nasal inhalations, rectal foams, and ear and eye drops.

Mineralocorticoids control the retention of sodium in the kidneys. In mineralocorticoid deficiency, there is excessive loss of sodium through the kidneys, with resulting water loss. Fludrocortisone (Florinef) is the only drug available for treatment of mineralocorticoid deficiency, and is available only in an oral dosage form.

Corticotropin (ACTH, adrenocorticotrophic hormone) stimulates the pituitary gland to release cortisone. A deficiency of corticotrophic hormone will have the same effects as a deficiency of cortisone. The hormone, which is available under the brand names Acthar and Actrel, is used for diagnostic testing, to determine the cause of a glucocorticoid deficiency, but is rarely used for replacement therapy since direct administration of glucocorticoids may be easier and offers better control over dosages.

Recommended dosage

The dosage of glucocorticoids varies with the drug, route of administration, condition being treated, and patient. Consult specific references.

Fludrocortisone, for use in replacement therapy, is normally dosed at 0.1 mg/day. Some patients require higher doses. It should normally be administered in conjunction with cortisone or hydrocortisone.

ACTH, when used for diagnostic purposes, is given as 10–25 units dissolved in 500 ml of 5% dextrose injection-infused IV over eight hours. A long-acting form, which may be used for replacement therapy, is given by subcutaneous (SC) or intramuscular (IM) injection at a dose of 40 to 80 units every 24–72 hours.

Precautions

Glucocorticoids

The most significant risk associated with administration of glucocorticoids is suppression of natural corticosteroid secretion. When the hormones are administered, they suppress the secretion of ACTH, which in turn reduces the secretion of the natural hormones. The extent of suppression varies with dose, drug potency, duration of treatment, and individual patient response. While suppression is seen primarily with drugs administered systemically, it can also occur with topical drugs such as creams and ointments, or drugs administered by inhalation. Abrupt cessation of corticosteroids may result in acute adrenal crisis (Addisonian crisis) that is marked by **dehydration** with severe vomiting and **diarrhea, hypotension**, and loss of consciousness. Acute adrenal crisis is potentially fatal.

Chronic overdose of glucocorticoids leads to Cushingoid syndrome, which is clinically identical to **Cushing's syndrome** and differs only in that in Cushingoid, the excessive steroids are from drug therapy rather than excessive glandular secretion. Symptoms vary, but most people have upper body **obesity**, rounded face, increased fat around the neck, and thinning arms and legs. In its later stages, this condition leads to weakening of bones and muscles with rib and spinal column **fractures**.

The short term adverse effects of corticosteroids are generally mild, and include **indigestion**, increased appetite, **insomnia**, and nervousness. There are also a very large number of infrequent adverse reactions, the most significant of which is drug induced-paranoia. Delirium, depression, menstrual irregularity, and increased hair growth are also possible. Consult detailed reviews for further information.

Long-term use of topical glucocorticoids can result in thinning of the skin. Oral steroid inhalations may cause

fungal overgrowth in the oral cavity. Patients must be instructed to rinse their mouths carefully after each dose. Corticosteroids are **pregnancy** category C. The drugs have caused congenital malformations in animal studies, including cleft palate. Breastfeeding should be avoided.

Mineralocorticoids

Because fludrocortisone has glucocorticoid activity as well as mineralocorticoid action, the same hazards and precautions apply to fludrocortisone as to the glucocorticoids. Overdose of fludrocortisone may also cause **edema, hypertension, and congestive heart failure.**

Corticotropins

Corticotropin has all the same risks as the glucocorticoids. Prolonged use may cause reduced response to the stimulatory effects of corticotropin.

Warnings and contraindications

Use corticosteroids with caution in patients with the following conditions:

- osteoporosis or any other bone disease
- current or past tuberculosis
- glaucoma or cataracts
- infections of any type (virus, bacteria, fungus, amoeba)
- sores in the nose or recent nose surgery (if using nasal spray forms of corticosteroids)
- underactive or overactive thyroid
- liver disease
- stomach or intestine problems
- diabetes
- heart disease
- high blood pressure
- high cholesterol
- kidney disease or kidney stones
- myasthenia gravis
- systemic lupus erythematosus (SLE)
- emotional problems
- skin conditions that cause the skin to be thinner to bruise more easily

Interactions

Corticosteroids have many drug interactions. Consult specific references.

Resources

ORGANIZATIONS

American Academy of Allergy, Asthma and Immunology. 611 East Wells Street, Milwaukee, WI 53202. (414) 272-6071. <<http://www.aaaai.org>>.

Asthma and Allergy Foundation of America. 1125 15th Street NW, Suite 502, Washington, DC 20005. (800) 727-8462. <<http://www.aafa.org>>.

National Heart, Lung and Blood Institute. National Institutes of Health. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov/nhlbi/nhlbi.htm>>.

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Corticotropin test see **Adrenocortotropic hormone test**

Cortisol tests

Definition

This test is a measure of serum cortisol (also known as hydrocortisone), or urine cortisol (also known as urinary free cortisol), an important hormone produced by a pair of endocrine glands called the adrenal glands.

Purpose

This test is performed on patients who may have malfunctioning adrenal glands. Blood and urine cortisol, together with the determination of adrenocortotropic hormone (ACTH), are the three most important tests in the investigation of **Cushing's syndrome** (caused by an overproduction of cortisol) and **Addison's disease** (caused by the underproduction of cortisol).

Precautions

Increased levels of cortisol are associated with **pregnancy**. Physical and emotional **stress** can also elevate cortisol levels. Drugs that may cause increased levels of cortisol include estrogen, **oral contraceptives**, amphetamines, cortisone, and spironolactone (Aldactone). Drugs that may cause decreased levels include androgens, aminoglutethimide, betamethasone, and other steroid medications, danazol, lithium, levodopa, metyrapone and phenytoin (Dilantin).

Description

Cortisol is a potent hormone known as a glucocorticoid that affects the metabolism of carbohydrates, proteins, and fats, but especially glucose. Cortisol increases blood sugar levels by stimulating the release of glucose from glucose stores in cells. It also acts to inhibit insulin, thus affecting glucose transport into cells.

The hypothalamus (an area of the brain), the pituitary gland (sometimes called the “master gland”), and

KEY TERMS

Addison's disease—A rare disorder in which symptoms are caused by a deficiency of hydrocortisone (cortisol) and aldosterone, two corticosteroid hormones normally produced by a part of the adrenal glands called the adrenal cortex. Symptoms include weakness, tiredness, vague abdominal pain, weight loss, skin pigmentation and low blood pressure.

Adrenal glands—A pair of endocrine glands (glands that secrete hormones directly into the bloodstream) that are located on top of the kidneys.

Adrenocorticotropic hormone (ACTH)—Also called corticotropin, this hormone is produced by the pituitary gland to stimulate the adrenal cortex to release various corticosteroid hormones.

Cushing's syndrome—A hormonal disorder caused by an abnormally high level of corticosteroid hormones that are produced by the adrenal glands. Corticosteroid hormones control the body's use of nutrients and the excretion of salts and water in the urine. Symptoms include high blood sugar levels, a moon face, weight gain, and increased blood pressure

the adrenal glands coordinate the production of cortisol. After corticotropin-releasing hormone (CRH) is made in the hypothalamus, CRH stimulates the pituitary to produce adrenocorticotropic hormone (ACTH). The production of ACTH in turn stimulates a part of the adrenal glands known as the adrenal cortex to produce cortisol. Rising levels of cortisol act as a negative feedback to curtail further production of CRH and ACTH, thus completing an elaborate feedback mechanism.

There are two methods for evaluating cortisol: blood and urine. The most reliable index of cortisol secretion is the 24-hour urine sample collection, but when blood levels are required or requested by the physician, plasma cortisol should be measured in the morning and again in the afternoon. Cortisol levels normally rise and fall during the day in what is called a diurnal variation, so that cortisol is at its highest level between 6–8 A.M. and gradually falls, reaching its lowest point around midnight. One reason for ordering blood cortisol levels versus a 24-hour urine collection is that sometimes the earliest sign of adrenal malfunction is the loss of this diurnal variation, even though the cortisol levels are not yet elevated. For example, individuals with Cushing's syndrome often have upper normal plasma cortisol levels in the morning and exhibit no decline as the day progresses.

Preparation

When testing for cortisol levels through the blood, a blood specimen is usually collected at 8 A.M. and again at 4 P.M. It should be noted that normal values may be transposed in individuals who have worked during the night and slept during the day for long periods of time.

When testing for cortisol level through the urine, a 24-hour urine sample is collected, refrigerated, and sent to the reference laboratory for examination.

Risks

Risks for the blood test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or feeling lightheaded after venipuncture, or hematoma (blood accumulating under the puncture site).

Normal results

Reference ranges for cortisol vary from laboratory to laboratory but are usually within the following ranges for blood:

- adults (8 A.M.): 6–28 mg/dL; adults (4 P.M.): 2–12 mg/dL
- child one to six years (8 A.M.): 3–21 mg/dL; child one to six years (4 P.M.): 3–10 mg/dL
- newborn: 1/24 mg/dL

Reference ranges for cortisol vary from laboratory to laboratory, but are usually within the following ranges for 24-hour urine collection:

- adult: 10–100 mg/24 hours
- adolescent: 5–55 mg/24 hours
- child: 2–27 mg/24 hours

Abnormal results

Increased levels of cortisol are found in Cushing's syndrome, excess thyroid (**hyperthyroidism**), **obesity**, ACTH-producing tumors, and high levels of stress.

Decreased levels of cortisol are found in Addison's disease, conditions of low thyroid, and **hypopituitarism**, in which pituitary activity is diminished.

Resources

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Cosmetic dentistry

Definition

Cosmetic dentistry includes a variety of dental treatments aimed at improving the appearance of the teeth.

Purpose

The purpose of cosmetic dentistry is to improve the appearance of the teeth using bleaching, bonding, veneers, reshaping, orthodontics, or implants.

Description

Bleaching is done to lighten teeth that are stained or discolored. It entails the use of a bleaching solution applied by a dentist or a gel in a tray that fits over the teeth used at home under a dentist's supervision. Bonding involves applying tooth-colored plastic putty, called composite resin, to the surface of chipped or broken teeth. This resin is also used to fill cavities in front teeth (giving a more natural-looking result) and to fill gaps between teeth. Veneers are thin, porcelain shells that cover the front of the teeth. They can improve the appearance of damaged, discolored, misshapen, or misaligned teeth. Reshaping involves the removal of enamel from a misshapen tooth so that it matches other teeth. Orthodontics uses braces to correct the position of crowded or misaligned teeth. Implants are artificial teeth which are attached directly to the jaw to replace missing teeth.

Preparation

Bleaching involves having a custom-made bleaching tray made by the dentist. This tray is worn at home for several hours each day or night. Teeth slowly become white over a period of one to six weeks. Bleaching can also be done in a dentist's office. A heat- or light-activated bleaching solution is applied to six to eight teeth per visit.

Bonding involves etching the surface of the tooth so composite resin can adhere. The dentist then contours the resin to the right shape, and smooths and polishes the resin after it is hard and dry.

To prepare for the application of a veneer, a thin layer of enamel is removed from the tooth (so that the finished tooth will be flush with surrounding teeth) and an impression of the tooth is taken from which the veneer will be created. Before a veneer is applied, the tooth is etched with an acid solution and an adhesive resin is painted on the tooth. The veneer is then applied, the resin is hardened with a bonding light, and the dentist polishes the veneer.

KEY TERMS

Bleaching—Technique used to brighten stained teeth.

Bonding—Rebuilding, reshaping, and covering tooth defects using tooth-colored materials.

Composite resin—Plastic material matching natural tooth color used to replace missing parts of a tooth.

During cosmetic reshaping, some enamel is removed from the uneven tooth so it more closely matches other teeth.

Orthodontics involves applying braces to the teeth, and wires are threaded through the braces. These wires are adjusted to gradually move the teeth to the desired new positions. Over time, crowded or misaligned teeth are straightened.

Implants are more secure and natural looking than dentures or bridgework, but are much more expensive. First an anchor for the implant is attached to the jaw bone. This surgery can take several hours. About six months later, after the bone around the anchor has healed, a post is attached to the anchor, and an artificial tooth is attached to the post. The whole process may take about nine months to complete.

Aftercare

Periodic touch-up may be needed to keep the teeth white if the teeth have been bleached or bonded. Also, the resin used in bonded teeth can be chipped by ice, popcorn kernels, or hard candy, requiring repair. Veneered teeth may need to be reveneered after five to 12 years. Once orthodontic braces are removed, regular visits to the orthodontist are advised because teeth can shift position. Implanted teeth require regular dental checkups to ensure that the anchor and post are stable.

Risks

After teeth are bleached, they may darken faster if exposed to staining products such as coffee or tobacco. Some patients experience increased sensitivity to cold while teeth are being bleached, but the sensitivity usually disappears shortly after completion of the treatment.

Bonded teeth, like bleached teeth, may also stain more easily than natural teeth. Bonding materials also chip easily.

Because cosmetic reshaping involves the removal of enamel, the process is irreversible because enamel cannot be replaced once it is removed.

The anchors of implanted teeth can loosen and cause pain; regular dental checkups are recommended.

Normal results

Cosmetic dentistry can improve the appearance of stained, chipped, misshapen, or crowded teeth.

Resources

ORGANIZATIONS

American Dental Association. 211 E. Chicago Ave., Chicago, IL 60611. (312) 440-2500. <<http://www.ada.org>>.

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Cosmetic surgery see **Plastic, cosmetic, and reconstructive surgery**

Costochondritis

Definition

Costochondritis is an inflammation and associated tenderness of the cartilage (i.e., the costochondral joints) that attaches the front of the ribs to the breastbone.

Description

Costochondritis causes **pain** in the lower rib area or upper breastbone. Some patients fear they are having a **heart attack**. The most severe pain is usually between the breast and the upper abdomen. The pain may be greater when in sitting or reclining positions. **Stress** may aggravate this condition. Generally the third or fourth ribs are affected. However, any of the seven costochondral junctions may be affected, and more often than not more than one site is involved. The inflammation can involve cartilage areas on both sides of the sternum, but usually is on one side only. Costochondritis should be distinguished from Tietze Syndrome, which is an inflammation involving the same area of the chest, but also includes swelling.

Causes and symptoms

The causes of costochondritis are not well-understood and may be difficult to establish. The most likely causes include injury, repetitive minor trauma, and unusual excessive physical activity.

KEY TERMS

Inflammation—Process whereby the immune system reacts to infection or other stimulus, characterized by pain, swelling, redness, and warmth of the affected part

The primary symptom of costochondritis is severe chest wall pain, which may vary in intensity. The pain becomes worse with trunk movement, deep breathing, and/or exertion, and better with decreased movement, quiet breathing, or changing of position. It is usually localized but may radiate extensively from the chest area. The pain has been described as sharp, nagging, aching, or pressure-like.

Diagnosis

Diagnosis is based on pain upon palpation (gentle pressing) of the affected joints. Swelling is not associated with costochondritis. Diagnosis is also dependent on the exclusion of other causes, including heart attack or bacterial or fungal infections found in IV drug users or post-operative **thoracic surgery** patients.

Treatment

The goals of treatment are to reduce inflammation and to control pain. To accomplish these goals, nonsteroidal anti-inflammatory agents (NSAIDs) are used, with ibuprofen usually selected as the drug of choice. Other NSAIDS options are flurbiprofen, mefenamic acid, ketoprofen, and naproxen. Additional treatment recommendations include the use of local heat, **biofeedback**, and gentle stretching of the pectoralis muscles two to three times a day.

For more difficult cases, where the patient continues to exhibit pain and discomfort, cortisone injections are used as therapy.

Alternative treatment

Supplements that are used to reduce inflammation have been used to treat costochondritis. Examples of such supplements include ginger root, evening primrose oil, bromelain, vitamin E, omega-3 oils, and white willow bark. Glucosamine/chondroitin sulfate, which may aid in the healing of cartilage, has also been used. Other alternative therapies include **acupuncture** and massages.

Prognosis

The prognosis for recovery from costochondritis is good. For most patients, the condition lessens in six

months to a year. However, after one year, about one-half of patients continue with some discomfort, while about one-third still report tenderness with palpation.

Prevention

Though the causes of costochondritis are not well known, avoidance of activities that may strain (e.g., the repetitive misuse of muscles) or cause trauma to the rib cage is recommended to prevent the occurrence of costochondritis. Modification of improper posture or ergonomics of the home or work place may also deter the development of this condition.

Resources

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Judith Sims

Cotrel-Dubousset spinal instrumentation see
Spinal instrumentation

Cough

Definition

A cough is a forceful release of air from the lungs that can be heard. Coughing protects the respiratory system by clearing it of irritants and secretions.

Description

While people can generally cough voluntarily, a cough is usually a reflex triggered when an irritant stimulates one or more of the cough receptors found at different points in the respiratory system. These receptors then send a message to the cough center in the brain, which in turn tells the body to cough. A cough begins with a deep breath in, at which point the opening between the vocal cords at the upper part of the larynx (glottis) shuts, trapping the air in the lungs. As the diaphragm and other muscles involved in breathing press against the lungs, the glottis suddenly opens, producing an explosive outflow of air at speeds greater than 100 mi (160 km) per hour.

In normal situations, most people cough once or twice an hour during the day to clear the airway of irritants. However, when the level of irritants in the air is high or when the respiratory system becomes infected, coughing may become frequent and prolonged. It may interfere with **exercise** or sleep, and it may also cause distress if accompanied by **dizziness**, **chest pain**, or breathlessness. In the majority cases, frequent coughing lasts one to two weeks and tapers off as the irritant or infection subsides. If a cough lasts more than three weeks it is considered a chronic cough, and physicians will try to determine a cause beyond an acute infection or irritant.

Coughs are generally described as either dry or productive. A dry cough does not bring up a mixture of mucus, irritants, and other substances from the lungs (sputum), while a productive cough does. In the case of a bacterial infection, the sputum brought up in a productive cough may be greenish, gray, or brown. In the case of an allergy or viral infection it may be clear or white. In the most serious conditions, the sputum may contain blood.

Causes and symptoms

In the majority of cases, coughs are caused by respiratory infections, including:

- colds or **influenza**, the most common causes of coughs
- **bronchitis**, an inflammation of the mucous membranes of the bronchial tubes
- croup, a viral inflammation of the larynx, windpipe, and bronchial passages that produces a bark-like cough in children
- whooping cough, a bacterial infection accompanied by the high-pitched cough for which it is named
- **pneumonia**, a potentially serious bacterial infection that produces discolored or bloody mucus
- **tuberculosis**, another serious bacterial infection that produces bloody sputum
- fungal infections, such as **aspergillosis**, **histoplasmosis**, and **cryptococcoses**

Environmental pollutants, such as cigarette smoke, dust, or smog, can also cause a cough. In the case of cigarette smokers, the nicotine present in the smoke paralyzes the hairs (cilia) that regularly flush mucus from the respiratory system. The mucus then builds up, forcing the body to remove it by coughing. Post-nasal drip, the irritating trickle of mucus from the nasal passages into the throat caused by **allergies** or **sinusitis**, can also result in a cough. Some chronic conditions, such as **asthma**, chronic bronchitis, **emphysema**, and **cystic fibrosis**, are characterized in part by a cough. A condition in which stomach acid backs up into the esophagus (gastro-

KEY TERMS

Antitussives—Drugs used to suppress coughing.

Expectorant—Drug used to thin mucus.

Gastroesophageal reflux—Condition in which stomach acid backs up into the esophagus.

Glottis—The opening between the vocal cords at the upper part of the larynx.

Larynx—A part of the respiratory tract between the pharynx and the trachea, having walls of cartilage and muscle and containing the vocal cords.

Sputum—The mixture of mucus, irritants, and other substances expelled from the lungs by coughing.

sophageal reflux) can cause coughing, especially when a person is lying down. A cough can also be a side-effect of medications that are administered via an inhaler. It can also be a side-effect of beta-blockers and ACE inhibitors, which are drugs used for treating high blood pressure.

Diagnosis

To determine the cause of a cough, a physician should take an exact medical history and perform an exam. Information regarding the duration of the cough, what other symptoms may accompany it, and what environmental factors may influence it aid the doctor in his or her diagnosis. The appearance of the sputum will also help determine what type of infection, if any, may be involved. The doctor may even observe the sputum microscopically for the presence of bacteria and white blood cells. Chest x rays may help indicate the presence and extent of such infections as pneumonia or tuberculosis. If these actions are not enough to determine the cause of the cough, a **bronchoscopy** or **laryngoscopy** may be ordered. These tests use slender tubular instruments to inspect the interior of the bronchi and larynx.

Treatment

Treatment of a cough generally involves addressing the condition causing it. An acute infection such as pneumonia may require **antibiotics**, an asthma-induced cough may be treated with the use of bronchodilators, or an antihistamine may be administered in the case of an allergy. Physicians prefer not to suppress a productive cough, since it aids the body in clearing respiratory system of infective agents and irritants. However, cough

medicines may be given if the patient cannot rest because of the cough or if the cough is not productive, as is the case with most coughs associated with colds or flu. The two types of drugs used to treat coughs are antitussives and **expectorants**.

Antitussives

Antitussives are drugs that suppress a cough. Narcotics—primarily codeine—are used as antitussives and work by depressing the cough center in the brain. However, they can cause such side effects as drowsiness, nausea, and **constipation**. Dextromethorphan, the primary ingredient in many over-the-counter cough remedies, also depresses the brain's cough center, but without the side effects associated with narcotics. Demulcents relieve coughing by coating irritated passageways.

Expectorants

Expectorants are drugs that make mucus easier to cough up by thinning it. Guaifenesin and terpin hydrate are the primary ingredients in most over-the-counter expectorants. However, some studies have shown that in acute infections, simply increasing fluid intake has the same thinning effect as taking expectorants.

Alternative treatment

Coughs due to bacterial or viral upper respiratory infections may be effectively treated with botanical and homeopathic therapies. The choice of remedy will vary and be specific to the type of cough the patient has. Some combination over-the-counter herbal and homeopathic cough formulas can be very effective for cough relief. Lingering coughs or coughing up blood should be treated by a trained practitioner.

Many health practitioners advise increasing fluids and breathing in warm, humidified air as ways of loosening chest congestion. Others recommend hot tea flavored with honey as a temporary home remedy for coughs caused by colds or flu. Various **vitamins**, such as vitamin C, may be helpful in preventing or treating conditions (including colds and flu) that lead to coughs. Avoiding of mucous-producing foods can be effective in healing a cough condition. These mucous-producing foods can vary, based on individual intolerance, but dairy products are a major mucous-producing food for most people.

Prognosis

Because the majority of coughs are related to the **common cold** or influenza, most will end in seven to 21 days. The outcome of coughs due to a more serious underlying disease depends on the pathology of that disease.

Prevention

It is important to identify and treat the underlying disease and origin of the cough. Avoid **smoking** and coming in direct contact with people experiencing cold or flu symptoms. Wash hands frequently during episodes of upper-respiratory illnesses.

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ORGANIZATIONS

National Heart, Lung and Blood Institute. PO Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

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KEY TERMS

Asthma—A disease in which the air passages of the lungs become inflamed and narrowed.

Bronchitis—Inflammation of the air passages of the lungs.

Chronic—A word used to describe a long-lasting condition. Chronic conditions often develop gradually and involve slow changes.

Emphysema—An irreversible lung disease in which breathing becomes increasingly difficult.

Mucus—Thick fluid produced by the moist membranes that line many body cavities and structures.

Phenylketonuria (PKU)—A genetic disorder in which the body lacks an important enzyme. If untreated, the disorder can lead to brain damage and mental retardation.

such as Vicks Formula 44, Drixoral Cough Liquid Caps, Sucrets Cough Control, Benylin DM and some Robitussin products. These medicines come in capsule, tablet, lozenge, and liquid forms and are available without a physician's prescription.

Recommended dosage

Regular (short-acting) capsules, lozenges, syrups, or tablets:

ADULTS AND CHILDREN OVER 12. 10-30 mg every 4-8 hours, as needed.

CHILDREN 6-12. 5-15 mg every 4-8 hours, as needed.

CHILDREN 2-6. 2.5-7.5 mg every 4-8 hours, as needed.

Children under 6 should not be given lozenges containing dextromethorphan because of the high dose of dextromethorphan in each lozenge.

CHILDREN UNDER 2. Check with child's physician.

Children under 6 should not be given lozenges containing dextromethorphan.

For extended-release oral suspension

ADULTS AND CHILDREN OVER 12. 60 mg every 12 hours, as needed.

CHILDREN 6-12. 30 mg every 12 hours, as needed.

CHILDREN 2-6. 15 mg every 12 hours, as needed.

CHILDREN UNDER 2. Check with child's physician.

Cough suppressants

Definition

Cough suppressants are medicines that prevent or stop coughing.

Purpose

Cough suppressants act on the center in the brain that controls the cough reflex. They are meant to be used only to relieve dry, hacking coughs associated with colds and flu. They should not be used to treat coughs that bring up mucus or the chronic coughs associated with **smoking, asthma, emphysema** or other lung problems.

Many cough medicines contain cough suppressants along with other ingredients. Some combinations of ingredients may cancel each other's effects. One example is the combination of cough suppressant with an expectorant—a medicine that loosens and clears mucus from the airways. The cough suppressant interferes with the ability to cough up the mucus that the expectorant loosens.

Description

The cough suppressant described here, dextromethorphan, is an ingredient in many cough medicines,

Precautions

Do not take more than the recommended daily dosage of dextromethorphan.

Dextromethorphan is not meant to be used for coughs associated with smoking, asthma, emphysema, chronic **bronchitis**, or other lung conditions. It also should not be used for coughs that produce mucus.

A lingering cough could be a sign of a serious medical condition. Coughs that last more than seven days or are associated with **fever**, **rash**, **sore throat**, or lasting **headache** should have medical attention. Call a physician as soon as possible.

People with **phenylketonuria** should be aware that some products with dextromethorphan also contain the artificial sweetener aspartame, which breaks down in the body to phenylalanine.

Anyone who has asthma or liver disease should check with a physician before taking dextromethorphan.

Women who are pregnant or breastfeeding or who plan to become pregnant should check with their physicians before taking dextromethorphan.

The dye tartrazine is an ingredient in some cough suppressant products. This dye causes allergic reactions in some people, especially those who are allergic to **aspirin**.

Side effects

Side effects are rare, but may include nausea, vomiting, stomach upset, slight drowsiness, and **dizziness**.

Interactions

Patients who take **monoamine oxidase inhibitors** (MAO inhibitors) should be aware that the co-administration of products containing dextromethorphan can cause dizziness, **fainting**, fever, nausea, and possibly **coma**. Do not take dextromethorphan unless a physician permits the use of the two drugs together.

When dextromethorphan is taken with medicines that cause drowsiness, this effect may be enhanced.

Nancy Ross-Flanigan

Coughing and deep-breathing exercises see
Chest physical therapy

Coxsackievirus infections see **Enterovirus infections**

CPK test see **Creatine kinase test**

CPR see **Cardiopulmonary resuscitation**

Crab lice see **Lice infestation**

Cradle cap see **Seborrheic dermatitis**

Cramps see **Dysmenorrhea**

Cranial arteritis see **Temporal arteritis**

Cranial manipulation see **Craniosacral therapy**

Craniopharyngioma see **Pituitary tumors**

Craniosacral therapy

Definition

Craniosacral therapy is a holistic healing practice that uses very light touching to balance the craniosacral system in the body, which includes the bones, nerves, fluids, and connective tissues of the cranium and spinal area.

Purpose

According to Upledger, craniosacral therapy is ideally suited for **attention-deficit hyperactivity disorder** (ADHD), headaches, chronic middle ear infection, **pain**, and general health maintenance. It is recommended for **autism**, **fibromyalgia**, heart disease, **osteoarthritis**, **pneumonia**, **rheumatoid arthritis**, chronic sinus infections, and **gastroenteritis** (inflammation of the lining of the stomach or small intestine). It is also used with other therapies to treat **chronic fatigue syndrome**, back pain, and menstrual irregularity. In addition, other craniosacral practitioners have reported benefits for eye dysfunction, **dyslexia**, depression, motor coordination difficulties, temporomandibular joint dysfunction (TMD), hyperactivity, **colic**, **asthma** in babies, floppy baby syndrome, **whiplash**, **cerebral palsy**, certain **birth defects**, and other central nervous system disorders.

Description

Origins

The first written reference to the movement of the spinal nerves and its importance in life, clarity, and “bringing quiet to the heart” is found in a 4,000-year-old text from China. Craniosacral work was referred to as “the art of listening.” Bone setters in the Middle Ages also sensed the subtle movements of the body. They used these movements to help reset **fractures** and dislocations and to treat headaches.

In the early 1900s, the research of Dr. William Sutherland, an American osteopathic physician, detailed the movement of the cranium and pelvis. Before his research it was believed that the cranium was a solid immovable mass. Sutherland reported that the skull is actually made up of 22 separate and movable bones that are connected by layers of tissue. He called his work cranial **osteopathy**. Nephi Cotton, an American chiropractor and contemporary of Sutherland, called this approach craniology. The graduates of these two disciplines have refined and enhanced these original approaches and renamed their work as sacro-occipital technique, cranial **movement therapy**, or craniosacral therapy.

Dr. John Upledger, an osteopathic physician, and others at the Department of Biomechanics at Michigan State University, College of Osteopathic Medicine learned of Sutherland's research and developed it further. He researched the clinical observations of various osteopathic physicians. This research provided the basis for Upledger's work that he named craniosacral therapy.

Craniosacral therapy addresses the craniosacral system. This system includes the cranium, spine, and sacrum that are connected by a continuous membrane of connective tissue deep inside the body, called the dura mater. The dura mater also encloses the brain and the central nervous system. Sutherland noticed that cerebral spinal fluid rises and falls within the compartment of the dura manta. He called this movement the primary respiratory impulse; today it is known as the craniosacral rhythm (CSR) or the cranial wave.

Craniosacral therapists can most easily feel the CSR in the body by lightly touching the base of the skull or the sacrum. During a session, they feel for disturbances in the rate, amplitude, symmetry, and quality of flow of the CSR. A therapist uses very gentle touch to balance the flow of the CSR. Once the cerebrospinal fluid moves freely, the body's natural healing responses can function.

A craniosacral session generally lasts 30–90 minutes. The client remains fully clothed and lays down on a massage table while the therapist gently assesses the flow of the CSR. Upledger describes several techniques which may be used in a craniosacral therapy session. The first is energy cyst release. "This technique is a hands-on method of releasing foreign or disruptive energies from the patient's body. Energy cysts may cause the disruption of the tissues and organs where they are located." The therapist feels these cysts in the client's body and gently releases the blockage of energy.

Sutherland first wrote about a second practice called direction of energy. In this technique the therapist intends energy to pass from one of his hands, through the patient, into the other hand.

WILLIAM SUTHERLAND (1873–1954)

William Garner Sutherland studied osteopathy under its founder, Andrew Taylor Still. Dr. Sutherland made his own important discovery while examining the sutures of cranial bones the skull bones that protect the brain. What he noticed is that the sutures were designed for motion. Sutherland termed this motion the *Breath of Life*. Through his experiments and research he determined that primary respiration was essential to all other physiological functions.

When Sutherland developed his techniques for craniosacral therapy, he wanted it to serve as a vehicle for listening to the body's rhythmic motions and treat the patterns of inertia when those motions become congested. He believed that the stresses—any physical or emotional trauma—created an imbalance in the body that needed correction to restore it to full health. The therapy is a hands-on method so that the therapist can feel the subtleties of the patterns of movement and inertia. Sutherland felt that this was the way to encourage self-healing and restoration of the body's own mechanisms, taking a holistic approach to creating optimal health.

The Craniosacral Therapy Educational Trust, based on Sutherland's pioneering work, is located at 10 Normington Close, Leigham Court Road, London SW16 2QS, United Kingdom. The phone number is 07000 785778.

The third technique is called myofascial release. This is a manipulative form of bodywork that releases tension in the fascia or connective tissue of the body. This form of bodywork uses stronger touch.

Upledger's fourth technique is position of release. This involves following the client's body into the positions in which an injury occurred and holding it there. When the rhythm of the CSR suddenly stops the therapist knows that the trauma has been released.

The last technique is somatoemotional release. This technique was developed by Upledger and is an offshoot of craniosacral therapy. It is used to release the mind and body of the residual effects of trauma and injury that are "locked in the tissues."

The cost of a session varies due to the length of time needed and the qualifications of the therapist. The cost may be covered by insurance when the therapy is performed or prescribed by a licensed health care provider.

Precautions

This gentle approach is extremely safe in most cases. However, craniosacral therapy is not recommended in

cases of acute systemic infections, recent skull fracture, intracranial hemorrhage or aneurysm, or herniation of the medulla oblongata (brain stem). Craniosacral therapy does not preclude the use of other medical approaches.

Side effects

Some people may experience mild discomfort after a treatment. This may be due to re-experiencing a trauma or injury or a previously numb area may come back to life and be more sensitive. These side effects are temporary.

Research and general acceptance

More than 40 scientific papers have been published that document the various effects of craniosacral therapy. There are also 10 authoritative textbooks on this therapy. The most notable scientific papers include Viola M. Fryman's work documenting the successful treatment of 1,250 newborn children with birth defects. Edna Lay and Stephen Blood showed the effects on TMD, and John Wood documented results with psychiatric disorders. The American Dental Association has found craniosacral therapy to be an effective adjunct to orthodontic work. However, the conventional medical community has not endorsed these techniques.

Resources

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ORGANIZATIONS

Milne Institute Inc. P.O. Box 2716, Monterey, CA 93942-2716. (831) 649-1825. Fax: (831) 649-1826. <milneinst@aol.com>. <<http://www.milne institute.com>>.

Upledger Institute. 11211 Prosperity Farms Road, Palm Beach Gardens, FL 33410. (800) 233-5880. Fax: (561) 622-4771. <<http://www.upledger.com>>.

OTHER

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Linda Chrisman

Craniotomy

Definition

Surgical removal of part of the skull to expose the brain.

Purpose

A craniotomy is the most commonly performed surgery for brain **tumor removal**. It may also be done to remove a blood clot and control hemorrhage, inspect the brain, perform a biopsy, or relieve pressure inside the skull.

Precautions

Before the operation, the patient will have undergone diagnostic procedures such as **computed tomography scans** (CT) or **magnetic resonance imaging** (MRI) scans to determine the underlying problem that required the craniotomy and to get a better look at the brain's structure. Cerebral **angiography** may be used to study the blood supply to the tumor, aneurysm, or other brain lesion.

Description

There are two basic ways to open the skull:

- a curving incision from behind the hairline, in front of the ear, arching above the eye
- at the nape of the neck around the occipital lobe

The surgeon marks with a felt tip pen a large square flap on the scalp that covers the surgical area. Following this mark, the surgeon makes an incision into the skin as far as the thin membrane covering the skull bone. Because the scalp is well supplied with blood, the surgeon will have to seal many small arteries. The surgeon then folds back a skin flap to expose the bone.

Using a high speed hand drill or an automatic craniotome, the surgeon makes a circle of holes in the skull, and pushes a soft metal guide under the bone from one hole to the next. A fine wire saw is then moved along the guide channel under the bone between adjacent holes. The surgeon saws through the bone until the bone flap can be removed to expose the brain.

After the surgery for the underlying cause is completed, the piece of skull is replaced and secured with pieces of fine, soft wire. Finally, the surgeon sutures the membrane, muscle, and skin of the scalp.

Preparation

Before the surgery, patients are usually given drugs to ease **anxiety**, and other medications to reduce the risk

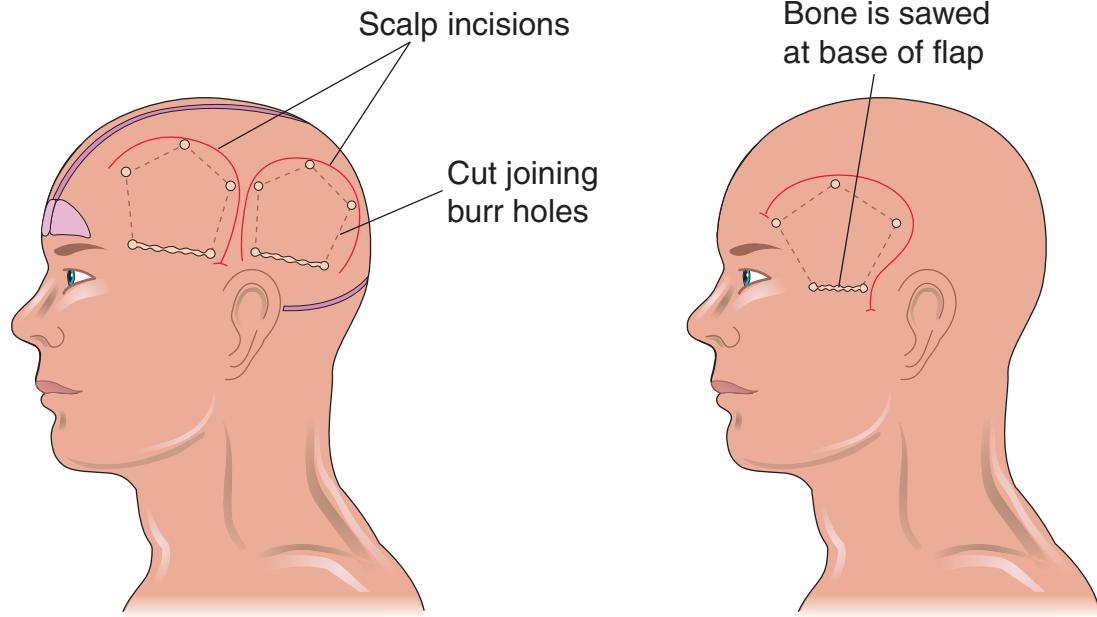


Figure A

Figure B

A craniotomy is the most commonly performed surgery for brain tumor removal. There are two basic ways to open the skull: a curving incision from behind the hairline in front of the ear and at the nape of the neck (figure A). To reach the brain, the surgeon uses a hand drill to make holes in the skull, pushing a soft metal guide under the bone. The bone is sawed through until the bone flap can be removed to expose the brain (figure B). (Illustration by Electronic Illustrators Group.)

of swelling, seizures, and infection after the operation. Fluids may be restricted, and a diuretic may be given before and during surgery if the patient has a tendency to retain water. A catheter is inserted before the patient goes to the operating room.

The scalp is shaved in the operating room right before surgery; this is done so that any small nicks in the skin won't have a chance to become infected before the operation.

Aftercare

Oxygen, painkillers, and drugs to control swelling and seizures are given after the operation. Codeine may be given to relieve the **headache** that may occur as a result of stretching or irritation of the nerves of the scalp that happens during the craniotomy. Some type of drainage from the head may be in place, depending on the reason for the surgery.

Patients are usually out of bed within a day and out of the hospital within a week. Headache and **pain** from the scalp wound can be controlled with medications.

The bandage on the skull should be changed regularly. Sutures closing the scalp will be removed, but soft

wires used to reattach the skull are permanent and require no further attention. The patient should avoid getting the scalp wet until all the sutures have been removed. A clean cap or scarf can be worn until the hair grows back.

Risks

Accessing the area of the brain that needs repair may damage other brain tissue. Therefore, the procedure carries with it some risk of brain damage that could leave the patient with some loss of brain function. The surgeon performing the operation can give the patient an assessment of the risk of his or her particular procedure.

Normal results

While every patient's experience is different depending on the reason for the surgery, age, and overall health, if the surgery has been successful, recovery is usually rapid because of the good supply of blood to the area.

Abnormal results

Possible complications after craniotomy include:

KEY TERMS

Craniotome—A type of surgical drill used to operate on the skull. It has a self-controlled system that stops the drill when the bone is penetrated.

- swelling of the brain
- excessive intracranial pressure
- infection
- seizures

Resources

BOOKS

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- The Surgery Book: An Illustrated Guide to 73 of the Most Common Operations*. Ed. Robert M. Younson, et al. New York: St. Martin's Press, 1993.

Carol A. Turkington

Creatine kinase test

Definition

The creatine kinase test measures the blood levels of certain muscle and brain enzyme proteins.

Purpose

Creatine kinase (CK or CPK) is an enzyme (a type of protein) found in muscle and brain. Normally, very little CK is found circulating in the blood. Elevated levels indicate damage to either muscle or brain; possibly from a myocardial infarction (**heart attack**), muscle disease, or **stroke**.

There are three types, or isoforms, of CK:

- CK-I, or BB, is produced primarily by brain and smooth muscle.
- CK-II, or MB, is produced primarily by heart muscle.
- CK-III, or MM, is produced primarily by skeletal muscle.

Precautions

No special precautions are necessary, except in patients with a bleeding disorder.

Description

A small amount of blood is drawn and used for laboratory analysis.

Preparation

Physical activity may cause a rise in CK levels, especially the CK-III fraction. Therefore, patients should not engage in strenuous physical activity the day of the test. The patient should report any recent injections, falls, or **bruises** that have occurred, as these may elevate CK levels as well.

Aftercare

No aftercare is required, except to keep the puncture site clean while it heals.

Risks

There are no risks to this test beyond the very slight risk of infection at the puncture site.

Normal results

In females, total CK should be 10–79 units per liter (U/L). In males, total CK should be 17–148 U/L. CK levels are reduced in the first half of **pregnancy**, and increased in the second half. CK levels are elevated in newborns.

The distribution of isoenzymes should be:

- CK-I: 0%
- CK-II: 0–5%
- CK-III: 95–100%

Abnormal results

Elevation of CK-I may be seen in stroke, extreme shock, or **brain tumor**.

Elevation of CK-II is seen after a myocardial infarction. It begins to rise three to six hours after the heart attack, and may peak within 24 hours. It should then return to normal. For this reason, it is a useful marker for recent myocardial infarction, but not for one which occurred more than a day before the test.

Elevation of CK-III indicates skeletal muscle damage. This may occur from normal **exercise**, trauma, or muscle disease. CK levels may be very high early on in **muscular dystrophy**, but may fall to normal later as muscle tissue is lost. Elevated CK is also seen in myositis, myoglobinuria, **toxoplasmosis**, and **trichinosis**. **Hypothyroidism** may also cause elevated CK.

KEY TERMS

Skeletal muscles—Muscles which move the skeleton. All of the muscles under voluntary control are skeletal muscles.

Smooth muscles—Muscles that surround the linings of the digestive system, airways, and circulatory system.

Resources

BOOKS

Corbett, Jane Vincent. *Laboratory Tests and Diagnostic Procedures with Nursing Diagnoses*. 2nd ed. Los Altos, CA: Appleton & Lange, 1987.

Richard Robinson

Creatine phosphokinase test see **Creatine kinase test**

Creatinine test

Definition

Creatine is an important compound produced by the body. It combines with phosphorus to make a high-energy phosphate compound in the body. Creatine phosphate is used in skeletal muscle contraction.

Purpose

The creatinine test is used to diagnose impaired kidney function and to determine renal (kidney) damage.

Precautions

A diet high in meat content can cause transient elevations of serum creatinine. Some drugs that may increase creatinine values include gentamicin, cimetidine, heavy-metal chemotherapeutic agents (e.g., cisplatin), and other drugs toxic to the kidneys, such as the cephalosporins.

Description

The creatinine test is used to measure the amount of creatinine in the blood. Because creatinine is a nonprotein end-product of creatine phosphate, which is used in

skeletal muscle contraction, the daily production of creatine, and the following product, creatinine, depends on muscle mass, which fluctuates very little.

Creatinine is excreted entirely by the kidneys, and therefore is directly related to renal function. When the kidneys are functioning normally, the serum creatinine level should remain constant and normal. Slight increases in creatine levels can appear after meals, especially after ingestion of large quantities of meat, and some diurnal variation may occur, with a low point at 7 A.M. and a peak at 7 P.M. Serious renal disorders, such as **glomerulonephritis**, **pyelonephritis**, and urinary obstruction, will cause abnormal elevations.

The creatinine level is interpreted in conjunction with another kidney function test called the Blood Urea Nitrogen (BUN). The serum creatinine level has much the same significance as the BUN but tends to rise later. Because of this, determinations of creatinine help to chronicle a disease process. Generally, a doubling of creatinine suggests a 50% reduction in kidney filtration rate.

Preparation

The creatinine test requires a blood sample. It is recommended that the patient be **fasting** (nothing to eat or drink) for at least eight hours before the test. The physician may also require that ascorbic acid (vitamin C), **barbiturates**, and **diuretics** be withheld for 24 hours.

Risks

Risks for this test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or feeling lightheaded after venipuncture, or hematoma (blood accumulating under the puncture site).

Normal results

Normal values can vary from laboratory to laboratory, but are generally in the following ranges:

- Adult female: 0.5–1.1 mg/dL
- Adult male: 0.6–1.2 mg/dL
- Adolescent: 0.5–1.0 mg/dL
- Child: 0.3–0.7 mg/dL
- Infant: 0.2–0.4 mg/dL
- Newborn: 0.3–1.2 mg/dL

Note that variations between sources for serum creatinine normal ranges are greater than for other important tests. For example, due to the greater amount of muscle mass generally present, males normally demonstrate higher creatinine levels than females. Also, because the kidney filtration rate normally increases in **pregnancy**, serum creatinine should be slightly less during such peri-

KEY TERMS

Glomerulonephritis—Glomerulonephritis is an inflammation of the filtering units of the kidney (glomeruli). The condition hinders removal of waste products, salt, and water from the bloodstream, leading to serious complications. It is the most common cause of renal failure.

Pyelonephritis—Pyelonephritis is an inflammation of the kidney itself, usually caused by a bacterial infection. In its most serious form, complications can include high blood pressure (hypertension) and renal failure.

ods. In older patients, creatinine is reduced because of decreased muscle mass. Similarly, other patients may have creatinine levels in which muscle abnormalities must be taken into consideration, such as long-term corticosteroid therapy, high thyroid (**hyperthyroidism**), **muscular dystrophy**, or **paralysis**.

Abnormal results

Two to 4 mg/dL indicate the presence of impairment of renal function. Greater than 4 mg/dL indicates serious impairment in renal function.

Resources

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Janis O. Flores

Creeping eruption see **Cutaneous larva migrans**

CREST syndrome see **Scleroderma**

Cretinism see **Hypothyroidism**

Creutzfeldt-Jakob disease

Definition

Creutzfeldt-Jakob disease (CJD) is a transmissible, rapidly progressing, fatal neurodegenerative disorder

called a spongiform degeneration that seems to be related to “mad cow disease.”

Description

Before 1995, Creutzfeldt-Jakob disease was little-known outside of the medical profession; even within it, many practitioners did not know much about it. Most doctors had never seen a case. With the recognition of a so-called “new variant” or simply variant form of CJD with the strong possibility that those with it became infected simply by eating contaminated beef, CJD has become one of the most talked-about diseases in the world. Additionally, the radical theory that the infectious agent is a normal protein that has been changed in its form has also sparked much interest.

First described in the first part of the twentieth century independently by Creutzfeldt and Jakob, CJD is a neurodegenerative disease causing a rapidly progressing **dementia** ending in **death**, usually within eight months of the onset of symptoms. It is also a very rare disease, affecting only about one in every million in the population throughout the world. In the United States, CJD is thought to affect about 250 people each year. CJD affects adults primarily between ages 50 and 75.

Spongiform encephalopathies

The most obvious pathologic feature of CJD is the formation of numerous fluid-filled spaces in the brain (vacuoles) resulting in a sponge-like appearance. CJD is one of several human “spongiform encephalopathies,” diseases that produce this characteristic change in brain tissue. Others are kuru; Gerstmann-Straussler-Scheinker disease, a genetic predominantly characterized by cerebellar ataxia (a kind of movement disorder); and fatal familial **insomnia**, associated with progressive insomnia, autonomic system dysfunction, and weakness caused by motor system dysfunction.

Kuru was prevalent among the Fore people in Papua, New Guinea, and spread from infected individuals after their deaths through the practice of ritual cannibalism, in which the relatives of the dead person honored him by consuming his organs, including the brain. Discovery of the infectious nature of kuru won the Nobel Prize for Carleton Gadjusek in 1976. The incubation period for kuru was between four to 30 years or more. While kuru has virtually disappeared following the cessation of these cannibalistic practices, several new cases continue to arise each year.

Cases of CJD have been grouped into three types: familial, iatrogenic, and sporadic.

- Familial CJD, representing 5–15% of cases, is inherited in an autosomal dominant manner, meaning that either parent may pass along the disease to a child, who may then develop CJD later in life.
- Iatrogenic CJD occurs when a person is infected during a medical procedure, such as organ donation, blood **transfusion**, or brain surgery. The rise in organ donation has increased this route of transmission; grafts of infected corneas and dura mater (the tissue covering the brain) have been shown to transmit CJD. Another source is hormones concentrated from the pituitary glands of cadavers, some of whom carried CJD, for use in people with growth hormone deficiencies. Iatrogenic infection from exposure to nerve-containing tissue represents a small fraction of all cases. The incubation period between exposure to the infectious agent is very long and is estimated to be from less than 10 to more than 30 years. It remains unlikely, but not impossible, that blood from patients with CJD is infectious to others by transfusion.
- Sporadic CJD represents at least 85% of all cases. Sporadic cases have no identifiable source of infection. Death usually follows first symptoms within eight months.

Animal forms and “mad cow disease”

Six forms of spongiform encephalopathies are known to occur in other mammals: scrapie in sheep, recognized for more than 200 years; chronic wasting disease in elk and mule deer in Wyoming and Colorado; transmissible mink encephalopathy; exotic ungulate encephalopathy in some types of zoo animals; feline spongiform encephalopathy in domestic cats; and bovine spongiform encephalopathy (BSE) in cows.

BSE was first recognized in Britain in 1986. Besides the spongiform changes in the brain, BSE causes dementia-like behavioral changes—hence the name “mad cow disease.” BSE was thought to be an altered form of scrapie, transmitted to cows when they were fed sheep offal (slaughterhouse waste) as part of their feed, but it is now thought to be more likely to be a primary cattle disease spread by contaminated feed.

The use of slaughterhouse offal in animal feed has been common in many countries and has been practiced for at least 50 years. The trigger for the BSE epidemic in Great Britain seems to have come in the early 1980s, when the use of organic solvents for preparation of offal was altered there. It is possible that these solvents had been destroying the agent called a prion, thereby preventing infection, and that the change in preparation procedure opened the way for the agent to “jump species” and cause BSE in cows that consumed scrapie-infected meal. The slaughter of infected (but not yet visibly sick) cows

at the end of their useful farm lives, and the use of their carcasses for feed, spread the infection rapidly and widely. For at least a year after BSE was first recognized in British herds, infected bovine remains continued to be incorporated into feed, spreading the disease still further. Although milk from infected cows has never been shown to pass the infectious agent, passage from infected mother to calf may have occurred through unknown means.

Beginning in 1988, the British government took steps to stop the spread of BSE, banning the use of bovine offal in feed and other products and ordering the slaughter of infected cows. By then, the slow-acting agent had become epidemic in British herds. In 1992, it was diagnosed in over 25,000 animals (1% of the British herd). By mid-1997, the cumulative number of BSE cases in the United Kingdom had risen to more than 170,000. The feeding ban did stem the tide of the epidemic; however, the number of new cases each week fell from a peak of 1,000 in 1993 to less than 300 two years later.

The export of British feed and beef to member countries was banned by the European Union, but cases of BSE had developed in Europe by then as well; however, by mid-1997, only about 1,000 cases had been identified. In 1989, the United States banned import of British beef and began monitoring United States herds in 1990. To date, no BSE has been detected in the United States, and only one case has been reported in North America in a cow imported to Canada from Great Britain.

Variant CJD: The human equivalent of mad cow disease?

From the beginning of the BSE epidemic, scientists and others in Britain feared that BSE might jump species again to infect humans who had consumed infected beef. This, however, had never occurred in scrapie from sheep, a disease known from hundreds of years. In 1996, the first report of this possibility occurred and this fear seemed to be realized with the first cases of a new variant of Creutzfeldt-Jacob disease, termed nvCJD, now just vCJD. Its victims are much younger than the 60–65 year old average for CJD, and the time from symptom onset to death has averaged 12 months or more instead of eight. The disease appears to cause more psychiatric symptoms early on. EEG abnormalities characteristic of CJD are not typically seen in vCJD.

As of July 2001, the total number of human cases of vCJD is 102. It is of major concern that the number of cases per year seems to be increasing by a factor of 1.35 each year. Almost all the cases have been found in Great Britain with three in France, one in Ireland, and one suspected in Hong Kong (who spent time in Great Britain).

Evidence is growing stronger that vCJD is in fact caused by BSE:

- almost all of the cases so far have occurred in Great Britain, the location of the BSE epidemic.
- BSE injected into monkeys produces a disease very similar to vCJD
- BSE and vCJD produce the same brain lesions after the same incubation period when injected into laboratory mice
- brain proteins isolated from vCJD victims, but not from the other forms of CJD, share similar molecular characteristics with brain proteins of animals that died from BSE

Many researchers now treat the BSE-vCJD connection as solidly established.

Assuming that BSE is the source, the question that has loomed from the beginning has been is how many people will eventually be affected. Epidemiological models of infectious disease produce estimates ranging from less than one hundred (a level already broken) to tens of thousands or more, depending on the assumptions used by the modelers. The incubation period of vCJD in humans is not known, nor are the genetic and environmental risk factors that influence susceptibility, nor the quantity of infectious agent needed to cause the disease. It is estimated that between one and two million infected cattle have been eaten by humans, most in the earliest stages of the epidemic. Estimates cannot be based on the very few cases that have developed so far. These cases could represent the very few people with the right combination of exposure and susceptibility to a relatively fast-developing infection, or they could be the first few victims of a slower-acting, more highly infectious agent.

Causes and symptoms

Causes

It is clear that Creutzfeldt-Jakob disease is caused by an infectious agent, but it is not yet clear what type of agent that is. Originally assumed to be a virus, evidence is accumulating that, instead, CJD is caused by a protein called a prion (PREE-on, for “proteinaceous infectious particle”) transmitted from victim to victim. The other spongiform encephalopathies are also hypothesized to be due to prion infection.

If this hypothesis is proved true, it would represent one of the most radical new ideas in biology since the discovery of deoxyribonucleic acid (DNA). All infectious diseases, in fact all life, uses nucleic acids—DNA or ribonucleic acid (RNA)—to code the instructions needed for reproduction. Inactivation of the nucleic acids

destroys the capacity to reproduce. However, when these same measures are applied to infected tissue from spongiform encephalopathy victims, infectivity is not destroyed. Furthermore, purification of infected tissue to concentrate the infectious fraction yields protein, not nucleic acid. While it remains possible that some highly stable nucleic acid remains hidden within the purified protein, this is seemingly less and less likely as further experiments are done. The “prion hypothesis,” as it is called, is now widely accepted, at least provisionally, by most researchers in the field. The most vocal proponent of the hypothesis, Stanley Prusiner, was awarded the Nobel Prize in 1997 for his work in the prion diseases.

A prion is an altered form of a normal brain protein. The normal protein has a helical shape along part of its length. In the prion form, a sheet structure replaces the helix. According to the hypothesis, when the normal form interacts with the prion form, some of its helical part is converted to a sheet, thus creating a new prion capable of transforming other normal forms. In this way, the disease process resembles crystallization more than typical viral infection, in which the virus commands the host’s cellular machinery to reproduce more of the virus. Build-up of the sheet form causes accumulation of abnormal protein clumps and degeneration of brain cells, which is thought to cause the disease.

The brain protein affected by the prion, called PrP, is part of the membrane of brain cells, but its exact function is unknown. It is composed of about 250 subunits, called amino acids, coded for by a gene on chromosome 20. Slight genetic differences, called polymorphisms, give rise to two slightly different normal protein forms: sub-unit 129 is a “methionine” in one form, but is “valine” in the other. A person may have all of one, all of the other, or a mixture of the two, depending on their genetic inheritance. Both forms have the normal helical structure, and function normally. However, susceptibility to prion conversion is influenced by subunit 129: a person with a mixture of forms is more resistant to conversion. All the cases of vCJD tested have had just methionine at 129. Exposure to the infectious agent is, of course, still required for disease development. Prion diseases are not contagious in the usual sense, and transmission from an infected person to another person requires direct inoculation of infectious material.

Familial CJD, on the other hand, does not require exposure, but develops through the inheritance of other, more disruptive mutations in the gene for the normal PrP protein. Researchers believe these mutations increase the likelihood that the protein may more spontaneously “flip” to the sheet form; once created, these can then convert other normal-form molecules. The other two inherited human prion diseases, Gerstmann-Straussler-

Scheinker disease and fatal familial insomnia, involve different mutations in the same gene.

The large majority of CJD cases are sporadic, meaning they have no known route of infection or genetic link. Causes of sporadic CJD are likely to be diverse and may include spontaneous genetic mutation, spontaneous protein changes, or unrecognized exposure to infectious agents. It is highly likely that future research will identify more risk factors associated with sporadic CJD.

Symptoms

About one in four people with CJD begin their illness with weakness, changes in sleep patterns, weight loss, or loss of appetite or sexual drive. A person with CJD may first complain of visual disturbances, including double vision, blurry vision, or partial loss of vision. Some visual symptoms are secondary to cortical blindness related to death of nerve cells in the occipital lobe of the brain responsible for vision. This form of visual loss is unusual in that patients may be unaware that they are unable to see. These symptoms may appear weeks to months before the onset of dementia.

The most characteristic symptom of CJD is rapidly progressing dementia, or loss of mental function. Dementia is marked by:

- memory losses
- impaired abstraction and planning
- language and comprehension disturbances
- poor judgment
- disorientation
- decreased attention and increased restlessness
- personality changes and psychosis
- hallucinations

Muscle spasms and jerking movements, called myoclonus, are also a prominent symptom of CJD. Balance and coordination disturbance (ataxia), is common in CJD, and is more pronounced in nvCJD. Stiffness, difficulty moving, and other features representing **Parkinson's disease** are seen and can progress to akinetic mutism, which is a state of being unable to speak or move.

Diagnosis

CJD is diagnosed by a clinical neurological exam and **electroencephalography** (EEG), which shows characteristic spikes called triphasic sharp waves. **Magnetic resonance imaging** (MRI) or **computed tomography scans** (CT) should be done to exclude other forms of dementia, and in CJD typically shows atrophy or loss of brain tissue. Lumbar puncture, or spinal tap, may be

done to rule out other causes of dementia (as cell count, chemical analysis, and other routine tests are normal in CJD) and to identify elevated levels of marker proteins known as 14-3-3. Another marker, neuron-specific enolase, may also be increased in CJD. CJD is conclusively diagnosed after death by brain **autopsy**. Scientists are investigating whether testing lymphatic tissue such as the tonsil may be an early tool in vCJD diagnosis. Additionally, recent studies have suggested that other blood tests may be useful as well.

Treatment

There is no cure for CJD, and no treatment that slows the progression of the disease. Drug therapy and nursing care are aimed at minimizing psychiatric symptoms and increasing patient comfort. However, the rapid progression of CJD frustrates most attempts at treatment, since decreasing cognitive function and more prominent behavioral symptoms develop so quickly. Despite the generally grim prognosis, a few CJD patients progress more slowly and live longer than the average; for these patients, treatment will be more satisfactory. Scientists are investigating whether some medicines that can "break" the abnormal protein form may be useful and whether a vaccine could help.

Prognosis

Creutzfeldt-Jakob disease is invariably fatal, with death following symptom onset by an average of eight months. About 5% of patients live longer than two years. Death from vCJD has averaged approximately 12 months after onset.

Prevention

There is no known way to prevent sporadic CJD, by far the most common type. Not everyone who inherits the gene mutation for familial CJD will develop the disease, but at present, there is no known way to predict who will and who won't succumb. The incidence of iatrogenic CJD has fallen with recognition of its sources, the development of better screening techniques for infected tissue, and the use of sterilization techniques for surgical instruments that inactivate prion proteins.

Strategies for prevention of vCJD are a controversial matter, as they involve a significant sector of the agricultural industry and a central feature of the diet in many countries. The infectious potential of contaminated meat is unknown, because the ability to detect prions within meat is limited. Surveillance of North American herds strongly suggests there is no BSE here, and strict regulations on imports of European livestock make future outbreaks highly unlikely. Therefore, avoidance of all meat

KEY TERMS

Autosomal dominant inheritance—A pattern of inheritance in which a trait will be expressed if the gene is inherited from either parent.

Encephalopathy—Brain disorder characterized by memory impairment and other symptoms.

Iatrogenic—Caused by a medical procedure.

Nucleic acids—The cellular molecules DNA and RNA that act as coded instructions for the production of proteins and are copied for transmission of inherited traits.

originating in North America, simply on grounds of BSE risk, is a personal choice unsupported by current data. The ban on the export of British beef continues in countries of the European Union, although some herds in these countries have developed low levels of infection as well.

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- Creutzfeldt-Jakob Disease Foundation. P.O. Box 611625, North Miami, FL 33261-1625. <<http://members.aol.com/crjakob/contact.html>>.
 The UK Creutzfeldt-Jakob Disease Surveillance Unit. <<http://www.cjd.ed.ac.uk/index.htm>>.

Larry I. Lutwick, MD

Cri du chat syndrome

Definition

Cri du chat syndrome occurs when a piece of chromosomal material is missing from a particular region on

chromosome 5. Individuals with this syndrome have unusual facial features, poor muscle tone(hypotonia), small head size (microcephaly), and **mental retardation**. A classic feature of the syndrome is the cat-like cry made by infants with this disorder.

Description

Dr. Jerome Lejeune first described cri du chat syndrome in 1963. The syndrome is named for the cat-like cry made by infants with this genetic disorder. *Cri du chat* means "cry of the cat" in French. This unusual cry is caused by abnormal development of the larynx (organ in the throat responsible for voice production). Cri du chat syndrome is also called "5p minus syndrome" because it is caused by a deletion, or removal, of genetic material from chromosome 5. The deletion that causes cri du chat syndrome occurs on the short or "p" arm of chromosome 5. This deleted genetic material is vital for normal development. Absence of this material results in the features associated with cri du chat syndrome.

A high-pitched mewing cry during infancy is a classic feature of cri du chat. Infants with cri du chat also typically have low birth weight, slow growth, a small head (microcephaly) and poor muscle tone (hypotonia). Infants with cri du chat may have congenital heart defects. Individuals with cri du chat syndrome have language difficulties, delayed motor skill development, and mental retardation. Behavioral problems may also develop as the child matures.

It has been estimated that cri du chat syndrome occurs in one of every 50,000 live births. According to the 5p minus Society, approximately 50–60 children are born with cri du chat syndrome in the United States each year. It can occur in all races and in both sexes.

Causes and symptoms

Cri du chat is the result of a chromosome abnormality—a deleted piece of chromosomal material on chromosome 5. In 90% of patients with cri du chat syndrome, the deletion is sporadic. This means that it happens randomly and is not hereditary. If a child has cri du chat due to a sporadic deletion, the chance the parents could have another child with cri du chat is 1%. In approximately 10% of patients with cri du chat, there is a hereditary chromosomal rearrangement that causes the deletion. If a parent has this rearrangement, the risk for them to have a child with cri du chat is greater than 1%.

An abnormal larynx causes the unusual cat-like cry made by infants that is a hallmark feature of the syndrome. As children with cri du chat get older, the cat-like cry becomes less noticeable. This can make the diagnosis more difficult in older patients. In addition to the cat-like

KEY TERMS

Aminocentesis—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.

Centromere—The centromere is the constricted region of a chromosome. It performs certain functions during cell division.

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 early embryo. 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 and consists 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.

Congenital—Refers to a disorder that is present at birth.

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.

Hypotonia—Reduced or diminished muscle tone.

Karyotyping—A laboratory procedure in which chromosomes are separated from cells, stained and arranged so that their structure can be studied under the microscope.

Microcephaly—An abnormally small head.

cry, individuals with cri du chat also have unusual facial features. These facial differences can be very subtle or more obvious. Microcephaly (small head size) is common. During infancy many patients with cri du chat do not gain weight or grow normally. Approximately 30% of infants with cri du chat have a congenital heart defect. Hypotonia (poor muscle tone) is also common, leading to problems with eating and slow, but normal, development. Mental retardation is present in all patients with cri du chat, but the degree of mental retardation varies between patients.

Diagnosis

During infancy, the diagnosis of cri du chat syndrome is strongly suspected if the characteristic cat-like cry is heard. If a child has this unusual cry or other features seen in cri du chat syndrome, chromosome testing should be performed. Chromosome analysis provides the definitive diagnosis of cri du chat syndrome and can be performed from a blood test. Chromosome analysis, also called "karyotyping," involves staining the chromosomes and examining them under a microscope. In some cases the deletion of material from chromosome 5 can be easily seen. In other cases, further testing must be performed. FISH (fluorescence in-situ hybridization) is a special technique that detects very small deletions. The majority of the deletions that cause cri du chat syndrome can be identified using the FISH technique.

Cri du chat syndrome can be detected before birth if the mother undergoes **amniocentesis** testing or chorionic villus sampling(CVS). This testing would only be recommended if the mother or father is known to have a chromosome rearrangement, or if they already have a child with cri du chat syndrome.

Treatment

Currently, there is no cure for cri du chat syndrome. Treatment consists of supportive care and developmental therapy.

Prognosis

Individuals with cri du chat have a 10% mortality during infancy due to complications associated with congenital heart defects, hypotonia, and feeding difficulties. Once these problems are controlled, most individuals with cri du chat syndrome have a normal lifespan. The degree of mental retardation can be severe. However, a recent study suggested that the severity is somewhat affected by the amount of therapy received.

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- 5p- Society. 7108 Katella Ave. #502, Stanton, CA 90680. (888) 970-0777. <<http://www.fivepminus.org>>.
- Alliance of Genetic Support Groups. 4301 Connecticut Ave. NW, Suite 404, Washington, DC 20008. (202) 966-5557. Fax: (202) 966-8553. <<http://www.geneticalliance.org>>.
- Cri du Chat Society. Dept. of Human Genetics, Box 33, MCV Station, Richmond VA 23298. (804) 786-9632.
- Cri du Chat Syndrome Support Group. <<http://www.cridchat.u-net.com>>.
- National Organization for Rare Disorders (NORD). P.O. Box 8923, New Fairfield, CT 06812-8923. (203) 746-6518 or (800) 999-6673. Fax: (203) 746-6481. <<http://www.rarediseases.org>>.

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- OMIM—Online Mendelian Inheritance in Man*. <<http://www.ncbi.nlm.nih.gov/Omim>>.

Holly Ann Ishmael, MS

Crib death see Sudden infant death syndrome

Crohn's disease

Definition

Crohn's disease is a type of inflammatory bowel disease (IBD), resulting in swelling and dysfunction of the intestinal tract.

Description

Crohn's disease involves inflammation of the intestine, especially the small intestine. Inflammation refers to swelling, redness, and loss of normal function. There is evidence that the inflammation is caused by various products of the immune system that attack the body itself instead of helpfully attacking a foreign invader (a virus or bacteria, for example). The inflammation of Crohn's disease most commonly affects the last part of the ileum (a section of the small intestine), and often includes the

large intestine (the colon). However, inflammation may also occur in other areas of the gastrointestinal tract, affecting the mouth, esophagus, or stomach. Crohn's disease differs from **ulcerative colitis**, the other major type of IBD, in two important ways:

- The inflammation of Crohn's disease may be discontinuous, meaning that areas of involvement in the intestine may be separated by normal, unaffected segments of intestine. The affected areas are called "regional enteritis," while the normal areas are called "skip areas."
- The inflammation of Crohn's disease affects all the layers of the intestinal wall, while ulcerative colitis affects only the lining of the intestine.

Also, ulcerative colitis does not usually involve the small intestine; in rare cases it involves the terminal ileum (so-called "backwash" ileitis).

In addition to inflammation, Crohn's disease causes ulcerations, or irritated pits in the intestinal wall. These pits occur because the inflammation has made areas of tissue shed.

Crohn's disease may be diagnosed at any age, although most diagnoses are made between the ages of 15–35. About 0.02–0.04% of the population suffers from this disorder, with men and women having an equal chance of being stricken. Whites are more frequently affected than other racial groups, and people of Jewish origin are between three and six times more likely to suffer from IBD. IBD runs in families; an IBD patient has a 20% chance of having other relatives who are fellow sufferers.

Crohn's disease is a chronic disorder. While the symptoms can be improved, a patient will not be completely cured of the underlying disease.

Causes and symptoms

The cause of Crohn's disease is unknown. No infectious agent (virus, bacteria, or fungi) has been identified as the cause of Crohn's disease. Still, some researchers have theorized that some type of infection may have originally been responsible for triggering the immune system, resulting in the continuing and out-of-control cycle of inflammation that occurs in Crohn's disease. Other evidence for a disorder of the immune system includes the high incidence of other immune disorders that may occur along with Crohn's disease.

The first symptoms of Crohn's disease include **diarrhea**, **fever**, abdominal **pain**, inability to eat, weight loss, and **fatigue**. Some patients have severe pain that mimics **appendicitis**. It is rare, however, for patients to notice blood in their bowel movements. Because Crohn's disease severely limits the ability of the affected intestine to absorb

the nutrients from food, a patient with Crohn's disease can have signs of **malnutrition**, depending on the amount of intestine affected and the duration of the disease.

The combination of severe inflammation, ulceration, and scarring that occurs in Crohn's disease can result in serious complications, including obstruction, **abscess** formation, and fistula formation.

An obstruction is a blockage in the intestine. This obstruction prevents the intestinal contents from passing beyond the point of the blockage. The intestinal contents "back up," resulting in **constipation**, vomiting, and intense pain. Although rare in Crohn's disease (because of the increased thickness of the intestinal wall due to swelling and scarring), a severe bowel obstruction can result in an intestinal wall perforation (a hole in the intestine). Such a hole in the intestinal wall would allow the intestinal contents, usually containing bacteria, to enter the abdomen. This complication could result in a severe, life-threatening infection.

Abcess formation is the development of a walled-off pocket of infection. A patient with an abscess will have bouts of fever, increased abdominal pain, and may have a lump or mass that can be felt through the wall of the abdomen.

Fistula formation is the formation of abnormal channels. These channels may connect one area of the intestine to another neighboring section of intestine. Fistulas may join an area of the intestine to the vagina or bladder, or they may drain an area of the intestine through the skin. Abscesses and fistulas commonly affect the area around the anus and rectum (the very last portions of the colon allowing waste to leave the body). These abnormal connections allow the bacteria that normally live in the intestine to enter other areas of the body, causing potentially serious infections.

Patients suffering from Crohn's disease also have a significant chance of experiencing other disorders. Some of these may relate specifically to the intestinal disease, and others appear to have some relationship to the imbalanced immune system. The faulty absorption state of the bowel can result in **gallstones** and **kidney stones**. Inflamed areas in the abdomen may press on the tube that drains urine from the kidney to the bladder (the ureter). Ureter compression can make urine back up into the kidney, enlarge the ureter and kidney, and can potentially lead to kidney damage. Patients with Crohn's disease also frequently suffer from:

- arthritis (inflammation of the joints)
- spondylitis (inflammation of the vertebrae, the bones of the spine)
- ulcers of the mouth and skin



A barium x-ray showing the colon of a patient with Crohn's disease where the large and small intestines join (bottom left). (Custom Medical Stock Photo. Reproduced by permission.)

- painful, red bumps on the skin
- inflammation of several eye areas
- inflammation of the liver, gallbladder, and/or the channels (ducts) that carry bile between and within the liver, gallbladder, and intestine

The chance of developing **cancer** of the intestine is greater than normal among patients with Crohn's disease, although this chance is not as high as among those patients with ulcerative colitis.

Diagnosis

Diagnosis is first suspected based on a patient's symptoms. Blood tests may reveal an increase in certain types of white blood cells, an indication that some type of inflammation is occurring in the body. The blood tests may also reveal anemia and other signs of malnutrition due to malabsorption (low blood protein; variations in

KEY TERMS

Abscess—A walled-off pocket of pus caused by infection.

Endoscope—A medical instrument that can be passed into an area of the body (the bladder or intestine, for example) to allow examination of that area. The endoscope usually has a fiber-optic camera that allows a greatly magnified image to be shown on a television screen viewed by the operator. Many endoscopes also allow the operator to retrieve a small sample (biopsy) of the area being examined to more closely view the tissue under a microscope.

Fistule—An abnormal channel that creates an open passageway between two structures that do not normally connect.

Gastrointestinal tract—The entire length of the digestive system, running from the stomach, through the small intestine, large intestine, and out the rectum and anus.

Immune system—The body system responsible for producing various cells and chemicals that fight infection by viruses, bacteria, fungi, and other foreign invaders. In autoimmune disease, these cells and chemicals turn against the body itself.

Inflammation—The result of the body's attempts to fight off and wall off an area that is infected. Inflammation results in the classic signs of redness, heat, swelling, and loss of function.

Obstruction—A blockage.

Ulceration—A pitted area or break in the continuity of a surface such as skin or mucous membrane.

the amount of calcium, potassium, and magnesium present in the blood; changes in certain markers of liver function). Stool samples may be examined to make sure that no infectious agent is causing the diarrhea, and to see if the waste contains blood.

During an endoscopic exam, a doctor passes a flexible tube with a tiny, fiber-optic camera device through the rectum and into the colon. The doctor can then carefully examine the lining of the intestine for signs of inflammation and ulceration that might suggest Crohn's disease. A tiny sample (a biopsy) of the intestine can also be taken through the endoscope, and the tissue will be examined under a microscope for evidence of Crohn's disease.

X rays can be helpful for diagnosis, and also for determining how much of the intestine is involved in the disease. For these x rays, the patient must either drink a chalky solution containing barium, or receive a **barium enema** (a solution that is administered through the rectum). Barium helps to "light up" the intestine, allowing more detail to be seen on the resulting x rays.

While Crohn's disease and ulcerative colitis are similar, they are also very different. Although it can be difficult to determine whether a patient has Crohn's disease or ulcerative colitis, it is important to make every effort to distinguish between these two diseases. Because the long-term complications of the diseases are different, treatment will depend on careful diagnosis of the specific IBD present.

Treatment

Treatments for Crohn's disease try to reduce the underlying inflammation, the resulting malabsorption/malnutrition, the uncomfortable symptoms of crampy abdominal pain and diarrhea, and the possible complications (obstructions, abscesses, and fistulas).

Inflammation can be treated with a drug called sulfasalazine. Sulfasalazine is made up of two parts. One part is related to the sulfa **antibiotics**; the other part is a form of the anti-inflammatory chemical, salicylic acid (related to **aspirin**). Sulfasalazine is not well absorbed from the intestine, so it stays mostly within the intestine, where it is broken down into its components. It is believed that the salicylic acid component actively treats Crohn's disease by fighting inflammation. Some patients do not respond to sulfasalazine, and require steroid medications (such as prednisone). Steroids, however, must be used carefully to avoid the complications of these drugs, including increased risk of infection and weakening of bones (**osteoporosis**). Some very potent immunosuppressive drugs, which interfere with the products of the immune system and can hopefully decrease inflammation, may be used for those patients who do not improve on steroids.

A new drug called infliximab (Remicade) appears to be a powerful treatment for Crohn's disease, particularly for patients who have not responded well to other forms of treatment. Infliximab is administered through infusion, and consists of a monoclonal antibody that interferes with the inflammatory process mediated by tumor necrosis factor-alpha (TNF- α). Patients taking infliximab seem to be able to decrease their use of steroid medications, and require fewer surgical interventions. Furthermore, infliximab is the first medication approved for treating fistulas. Unfortunately, infliximab can only be used on a short-term basis, because its interference with TNF- α activity can also predispose patients to serious infection. More

research is needed to try to harness the benefits of infliximab, while avoiding the potential complications.

Serious cases of malabsorption/malnutrition may need to be treated by providing nutritional supplements. These supplements must be in a form that can be absorbed from the damaged, inflamed intestine. Some patients find that certain foods are hard to digest, including milk, large quantities of fiber, and spicy foods. When patients are suffering from an obstruction, or during periods of time when symptoms of the disease are at their worst, they may need to drink specially formulated, high-calorie liquid supplements. Those patients who are severely ill may need to receive their **nutrition** through a needle inserted in a vein (intravenously), or even by a tiny tube (a catheter) inserted directly into a major vein in the chest.

A number of medications are available to help decrease the cramping and pain associated with Crohn's disease. These include loperamide, tincture of opium, and codeine. Some fiber preparations (methylcellulose or psyllium) may be helpful, although some patients do not tolerate them well.

The first step in treating an obstruction involves general attempts to decrease inflammation with sulfasalazine, steroids, or immunosuppressive drugs. A patient with a severe obstruction will have to stop taking all food and drink by mouth, allowing the bowel to "rest." Abscesses and other infections will require antibiotics. Surgery may be required to repair an obstruction that does not resolve on its own, to remove an abscess, or to repair a fistula. Such surgery may involve the removal of a section of the intestine. In extremely severe cases of Crohn's disease that do not respond to treatment, a patient may need to have the entire large intestine removed (an operation called a colectomy). In this case, a piece of the remaining small intestine is pulled through an opening in the abdomen. This bit of intestine is fashioned surgically to allow a special bag to be placed over it. This bag catches the body's waste, which no longer can be passed through the large intestine and out of the anus. This opening, which will remain in place for life, is called an ileostomy.

Prognosis

Crohn's disease is a life-long illness. The severity of the disease can vary, and a patient can experience periods of time when the disease is not active and he or she is symptom-free. However, the complications and risks of Crohn's disease tend to increase over time. Well over 60% of all patients with Crohn's disease will require surgery, and about half of these patients will require more than one operation over time. About 5–10% of all

Crohn's patients will die of their disease, primarily due to massive infection.

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ORGANIZATION

Crohn's & Colitis Foundation of America, Inc. 386 Park Avenue South, 17th Floor, New York, NY 10016-8804. (800) 932-2423.

Rosalyn S. Carson-DeWitt

Cromolyn see **Antiasthmatic drugs**

Cross-eye see **Strabismus**

Cross-gender identification see **Gender identity disorder**

Croup

Definition

Croup is a common childhood ailment. Typically, it arises from a viral infection of the larynx (voice box) and is associated with mild upper respiratory symptoms such as a runny nose and **cough**. The key symptom is a harsh barking cough. Croup is usually not serious, and most children recover within a few days. In a small percentage of cases, a child develops breathing difficulties and may need medical attention.

Description

At one time, the term croup was primarily associated with **diphtheria**, a life-threatening respiratory infection. Owing to widespread vaccinations, diphtheria has become rare in the United States, and croup currently

refers to a mild viral infection of the larynx. Croup is also known as laryngotracheitis, a medical term that describes the inflammation of the trachea (windpipe) and larynx.

Parainfluenza viruses are the typical root cause of the infection, but **influenza** (flu) and cold viruses may sometimes be responsible. All of these viruses are highly contagious and easily transmitted between individuals via sneezing and coughing. Children between the ages of three months and six years are usually affected, with the greatest incidence at one to two years of age. Croup can occur at any time of the year, but it is most typical during early autumn and winter. The characteristic harsh barking of a croupy cough can be very distressing, but it rarely indicates a serious problem. Most children with croup can be treated very effectively at home; however, 1–5% may require medical treatment.

Croup may sometimes be confused with more serious conditions, such as **epiglottitis** or bacterial tracheitis. These ailments arise from bacterial infection and must receive medical treatment.

Causes and symptoms

Owing to an upper respiratory viral infection, the larynx and trachea may become inflamed or swollen. The hallmark sign of croup is a harsh, barking cough. This cough may be preceded by one to three days of symptoms that resemble a slight cold. A croupy cough is often accompanied by a runny nose, hoarseness, and a low **fever**. When the child inhales, there may be a raspy or high-pitched noise, called **stridor**, owing to the narrowed airway and accumulated mucus. In the presence of stridor, medical attention is required.

However, the airway rarely narrows so much that breathing is impeded. Symptoms usually abate completely within a few days. Medical treatment may be sought if the child's symptoms do not respond to home treatment.

Emergency medical treatment is required immediately if the child has difficulty breathing, swallowing, or talking; develops a high fever (103°F/39.4°C or more); seems unalert or confused; or has pale or blue-tinged skin.

Diagnosis

Croup is diagnosed based on the symptoms. If symptoms are particularly severe, or do not respond to treatment, an x ray of the throat area is done to assess the possibility of epiglottitis or other blockage of the airway.

Treatment

Home treatment is the usual method of managing croup symptoms. It is important that the child is kept

comfortable and calm to the best degree possible, because crying can make symptoms seem worse. Humid air can help a child with croup feel more comfortable. Recommended methods include sitting in a steamy bathroom with the hot water running or using a cool-water vaporizer or humidifier. Breathing may also be eased by going outside into cooler air. The child should drink frequently in order to stay well hydrated. To treat any fever, the child may be given an appropriate dose of **acetaminophen** (like Tylenol). **Antihistamines** and **decongestants** are ineffective in treating croup. Children under the age of 18 should not be given aspirin, as it may cause **Reye's syndrome**, a life-threatening disease of the brain.

If the child does not respond to home treatment, medical treatment at a doctor's office or an emergency room could be necessary. Based on the severity of symptoms and the response to treatment, the child may need to be admitted to a hospital.

For immediate symptom relief, epinephrine may be administered as an inhaled aerosol. Effects last for up to two hours, but there is a possibility that symptoms may return. For that reason, the child is kept under supervision for three or more hours. Another effective drug is a glucocorticoid, dexamethasone. This drug requires more time to take effect, but is longer lasting. It can be administered orally or as an injection. Another glucocorticoid, budesonide, has been used outside the United States for treating croup. It is administered as an inhaled aerosol and has been shown to be effective; however, it is not available as a treatment option in the United States.

Of the 1–5% of children requiring medical treatment, approximately 1% need respiratory support. Such support involves intubation (inserting a tube into the trachea) and oxygen administration.

Alternative treatment

Botanical/herbal medicines can be helpful in healing the cough that is commonly associated with croup. Several herbs to consider for cough treatment include aniseed (*Pimpinella anisum*), sundew (*Drosera rotundifolia*), thyme (*Thymus vulgaris*), and wild cherry bark (*Prunus serotina*). Homeopathic medicine can be very effective in treating cases of croup. Choosing the correct remedy (a common choice is aconite or monkshood, *Aconitum napellus*) is always the key to the success of this type of treatment.

Prognosis

Croup is a temporary condition and children typically recover completely within three to six days. Children can experience one or more episodes of croup during early childhood; however, croup is rarely a dangerous condition.

KEY TERMS

Diphtheria—A serious, frequently fatal, bacterial infection that affects the respiratory tract. Vaccinations given in childhood have made diphtheria very rare in the United States.

Epiglottitis—A bacterial infection that affects the epiglottis. The epiglottis is a flap of tissue that prevents food and fluid from entering the trachea. The infection causes it to become swollen, potentially blocking the airway. Other symptoms include a high fever, nonbarking cough, muffled voice, and an inability to swallow properly (possibly indicated by drooling).

Glucocorticoid—A hormone that helps in digestion of carbohydrates and reduces inflammation.

Larynx—Commonly called the voice box, it is the area of the trachea that contains the vocal cords.

Stridor—The medical term used to describe the high-pitched or rasping noise made when air is inhaled.

Trachea—Commonly called the windpipe, it is the air pathway that connects the nose and mouth to the lungs.

Prevention

Croup is caused by highly transmissible viruses. Similar to other common childhood ailments, prevention is not applicable.

Resources

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Julia Barrett

Cryoglobulin test

Definition

Cryoglobulin is an abnormal blood protein associated with several diseases. Testing for cryoglobulin is done

when a person has symptoms of this protein or is being evaluated for one of the associated diseases.

Purpose

Cryoglobulin clumps in cold temperatures. This physical characteristic causes people with cryoglobulin to have symptoms during cold weather: blanching, numbness, and pain in their fingers or toes (Raynaud's phenomenon); bleeding into the skin (purpura); and pain in joints (arthralgia). People with these symptoms or any other symptoms that appear in cold weather should be tested for cryoglobulin.

Diseases that cause the body to make extra or abnormal proteins are often associated with cryoglobulin. These diseases include cancers involving white blood cells, infections, **autoimmune disorders**, and rheumatoid diseases.

This test provides information about the cause of symptoms in a person who already has a disease process. It doesn't diagnose a specific disease or monitor the course of a disease.

Precautions

This test is not a screening test for disease in a person without symptoms.

Description

Laboratory testing for cryoglobulin is based on the fact that cryoglobulin clumps when cooled and dissolves when warmed. The test is done on a person's serum (the yellow liquid part of blood that separates from the cells after the blood clots). The serum is kept warm from the time drawn until the cells and the serum are separated in the laboratory. The serum is placed at 33.8°F (1°C) for one to seven days. If there is clumping, cryoglobulins are present. The amount of cryoglobulins is determined by measuring the amount of clumping. Negative tests are checked through seven days.

Additional testing is done to find out what kind of cryoglobulin protein is present. There are three kinds of cryoglobulin, each associated with different diseases.

The test, also called the cold sensitivity antibodies test, is covered by insurance when medically necessary. Results are usually available the following day.

Preparation

This test requires 15–20 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

KEY TERMS

Cryoglobulin—An abnormal blood protein associated with several diseases. It is characterized by its tendency to clump in cold temperatures.

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. The blood must be kept warm, at body temperature, until the laboratory can separate the cells from the serum.

Aftercare

Discomfort or bruising may occur at the puncture site or the person may feel dizzy or faint. Pressure to the puncture site until the bleeding stops reduces bruising. Warm packs to the puncture site relieve discomfort.

Normal results

Negative or absent.

Abnormal results

If the person has cryoglobulin, the amount is reported. Larger amounts of cryoglobulin are associated with cancers or abnormalities involving white blood cells, moderate amounts are associated with autoimmune disorders and rheumatoid diseases, and smaller amounts are associated with infections.

The type of cryoglobulin is also reported. Type I cryoglobulin, also called monoclonal cryoglobulinemia, is found in cancers or abnormalities of white blood cells. Type II, also called mixed cryoglobulinemia, is associated with autoimmune disorders, rheumatoid diseases, and infections, particularly chronic **hepatitis B**.

The physician must interpret the cryoglobulin result along with other test results and the patient's clinical condition and medical history.

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Nancy J. Nordenson

Cryosurgery see **Cryotherapy**

Cryotherapy

Definition

Cryotherapy is a technique that uses an extremely cold liquid or instrument to freeze and destroy abnormal skin cells that require removal. The technique has been in use since the turn of the century, but modern techniques have made it widely available to dermatologists and primary care doctors. The technique is also called cryosurgery.

Purpose

Cryotherapy can be employed to destroy a variety of benign skin growths, such as **warts**, pre-cancerous lesions (such as actinic keratoses), and malignant lesions (such as basal cell and squamous cell cancers). The goal of cryotherapy is to freeze and destroy targeted skin growths while preserving the surrounding skin from injury.

Precautions

Cryotherapy is not recommended for certain areas of the body because of the danger of destruction of tissue or unacceptable scarring. These areas include: skin that overlies nerves, the corners of the eyes, the fold of skin between the nose and lip, the skin surrounding the nostrils, and the border between the lips and the rest of the face. Lesions that are suspected or known to be **malignant melanoma** should not be treated with cryotherapy, but should instead be removed surgically. Similarly, basal cell or squamous cell carcinomas that have reappeared at the site of a previously treated tumor should also be removed surgically. If it remains unclear whether a growth is benign or malignant, a sample of tissue should be removed for analysis (biopsy) by a pathologist before any attempts to destroy the lesion with cryotherapy. Care should be taken in people with diabetes or certain circulation problems when cryotherapy is considered for growths located on their lower legs, ankles, and feet. In these patients, healing can be poor and the risk of infection can be higher than for other patients.

Description

There are three main techniques to performing cryotherapy. In the simplest technique, usually reserved for warts and other benign skin growths, the physician

will dip a cotton swab or other applicator into a cup containing a “cryogen,” such as liquid nitrogen, and apply it directly to the skin growth to freeze it. At a temperature of -320°F (-196°C), liquid nitrogen is the coldest cryogen available. The goal is to freeze the skin growth as quickly as possible, and then let it thaw slowly to cause maximum destruction of the skin cells. A second application may be necessary depending on the size of the growth. In another cryotherapy technique, a device is used to direct a small spray of liquid nitrogen or other cryogen directly onto the skin growth. Freezing may last from five to 20 seconds, depending on the size of the lesion. A second freeze-thaw cycle may be required. Sometimes, the physician will insert a small needle connected to a thermometer into the lesion to make certain the lesion is cooled to a low enough temperature to guarantee maximum destruction. In a third option, liquid nitrogen or another cryogen is circulated through a probe to cool it to low temperatures. The probe is then brought into direct contact with the skin lesion to freeze it. The freeze time can take two to three times longer than with the spray technique.

Preparation

Extensive preparation prior to cryotherapy is not required. The area to be treated should be clean and dry, but sterile preparation is not necessary. Patients should know that they will experience some **pain** at the time of the freezing, but local anesthesia is usually not required. The physician may want to reduce the size of certain growths, such as warts, prior to the cryotherapy procedure, and may have patients apply salicylic acid preparations to the growth over several weeks. Sometimes, the physician will pare away some of the tissue using a device called a curette or a scalpel.

Aftercare

Redness, swelling, and the formation of a blister at the site of cryotherapy are all expected results of the treatment. A gauze dressing is applied and patients should wash the site three or four times daily while fluid continues to ooze from the wound, usually for five to 14 days. A dry crust then forms that falls off by itself. **Wounds** on the head and neck may take four to six weeks to heal, but those on the body, arms, and legs can take longer. Some patients experience pain at the site following the treatment. This can usually be eased with **acetaminophen** (Tylenol), though in some cases a stronger pain reliever may be required.

Risks

Cryotherapy poses little risk and can be well-tolerated by elderly and other patients who are not good candidates

KEY TERMS

Actinic keratosis—A crusty, scaly pre-cancerous skin lesion caused by damage from the sun. Frequently treated with cryotherapy.

Basal cell cancer—The most common form of skin cancer; it usually appears as one or several nodules having a central depression. It rarely spreads (metastasizes), but is locally invasive.

Cryogen—A substance with a very low boiling point, such as liquid nitrogen, used in cryotherapy treatment.

Melanoma—The most dangerous form of skin cancer. It should not be treated with cryotherapy, but should be removed surgically instead.

Squamous cell cancer—A form of skin cancer that usually originates in sun-damaged areas or pre-existing lesions; at first local and superficial, it may later spread to other areas of the body.

dates for other surgical procedures. As with other surgical procedures, there is some risk of scarring, infection, and damage to underlying skin and tissue. These risks are generally minimal in the hands of experienced users of cryotherapy.

Normal results

Some redness, swelling, blistering and oozing of fluid are all common results of cryotherapy. Healing time can vary by the site treated and the cryotherapy technique used. When cryogen is applied directly to the growth, healing may occur in three weeks. Growths treated on the head and neck with the spray technique may take four to six weeks to heal; growths treated on other areas of the body may take considerably longer. Cryotherapy boasts high success rates in permanently removing skin growths; even for malignant lesions such as squamous cell and basal cell cancers, studies have shown a cure rate of up to 98%. For certain types of growths, such as some forms of warts, repeat treatments over several weeks are necessary to prevent the growth’s return.

Abnormal results

Although cryotherapy is a relatively low risk procedure, some side effects may occur as a result of the treatment. They include:

- Infection. Though uncommon, infection is more likely on the lower legs where healing can take several months.

- Pigmentary changes. Both hypopigmentation (lightening of the skin) and **hyperpigmentation** (darkening of the skin) are possible after cryotherapy. Both generally last a few months, but can be longer lasting.
- Nerve damage. Though rare, damage to nerves is possible, particularly in areas where they lie closer to the surface of the skin, such as the fingers, the wrist, and the area behind the ear. Reports suggest this will disappear within several months.

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- Young, R., and R. Sinclair. "Practical Cryosurgery." *Australian Family Physician* 26 (Sept. 1997): 1045-1047.

ORGANIZATIONS

- American Academy of Dermatology. 930 N. Meacham Road, P.O. Box 4014, Schaumburg, IL 60168-4014. (847) 330-0230. <<http://www.aad.org>>.
- American Society for Dermatologic Surgery. 930 N. Meacham Road, P.O. Box 4014, Schaumburg, IL 60168-4014. (847) 330-9830. <<http://www.asds-net.org>>.

Richard H. Camer

Cryptococcosis

Definition

Cryptococcosis is an infection caused by inhaling the fungus *Cryptococcus neoformans*. It is one of the diseases most often affecting AIDS patients. Cryptococcosis may be limited to the lungs, but frequently spreads throughout the body. Although almost any organ can be infected, the fungus is often fatal if it infects the nervous system where it causes an inflammation of the membranes covering the brain and spinal cord (**meningitis**).

Description

The fungus causing cryptococcus, *C. neoformans*, is found worldwide in soil contaminated with pigeon or other bird droppings. It has also been found on unwashed

raw fruit. Cryptococcosis is a rare disease in healthy individuals, but is the most common fungal infection affecting people with AIDS.

People with **Hodgkin's disease** or who are taking large doses of drugs that suppress the functioning of the immune system (**corticosteroids, chemotherapy** drugs) are also more susceptible to cryptococcal infection. Cryptococcosis is also called cryptococcal meningitis (when the brain is infected), Busse-Buschke disease, European **blastomycosis**, torular meningitis, or torulosis.

Causes and symptoms

Once the cryptococcal fungus reaches the lungs, three things can happen. The immune system can heal the body without medical intervention, the disease can stay localized in the lungs, or it can spread throughout the body. In healthy people with normally functioning immune systems, the body usually heals itself, and the infected person notices no symptoms and has no complications (asymptomatic). The disease does not spread from one person to another.

Cryptococcosis is an opportunistic infection that puts people with immune system diseases at higher risk of developing more serious forms of the disease. In the United States, 6–10% of all patients with AIDS get cryptococcosis.

If the body does not heal itself, the fungus begins to grow in the lungs and form nodules that can be seen on chest x rays. In the early stages of infection, an individual usually only exhibits symptoms of a respiratory infection, such as a dry **cough**, so the disease is rarely diagnosed.

The fungus can remain dormant in the lungs and produce an active infection later if the immune system is weakened. If the disease becomes active, it can cause cryptococcal **pneumonia** in the lungs. Unfortunately, however, cryptococcal pneumonia has symptoms similar to other pneumonias (cough, chest **pain**, difficulty breathing), making it difficult to accurately diagnose. The infection can spread to other parts of the body, particularly the brain and central nervous system.

Most patients are not diagnosed as having cryptococcosis until they show signs of cryptococcal meningitis, or infection of the membranes surrounding the brain and spinal cord. Symptoms appear gradually over a period of two to four weeks. **Fever** and **headache** are the most common symptoms, occurring in about 85% of patients. Nausea, vomiting, unwanted weight loss, and **fatigue** are also common. Other symptoms seen in 25–30% of patients are blurred vision, stiff neck, aversion to light, and seizures. Since the symptoms of classic

meningitis, such as stiff neck and aversion to light, do not occur in many patients, diagnosis is often delayed. In addition to meningitis, inflammation of the brain (**encephalitis**) and brain lesions called cryptococcomas or tortulomas can also develop.

In addition to the brain, the cryptococcal infection can spread to the kidneys, bone marrow, heart, adrenal glands, lymph nodes, urinary tract, blood, and skin. Often times preceding the development of cryptococcal meningitis, painless **rashes** and lesions that mimic other skin diseases, such as *molluscum contagiosum*, may develop. A small percentage of patients with brain infections show infections in other organs as well.

Diagnosis

Physicians who regularly work with AIDS patients have the most experience in diagnosing cryptococcosis. The preferred methods of diagnosis use simple and very accurate blood and cerebrospinal fluid (CSF) tests that detect the presence of an antigen produced by the fungus. The cerebrospinal fluid test is generally more sensitive to detecting the meningitis form of the infection. CSF is collected during a procedure called a lumbar puncture, during which an anesthetic is applied to a small area of the back near the spine and a needle is used to withdraw a sample of cerebrospinal fluid from the space between the vertebrae and the spinal cord. Once obtained, a small amount of ink (called India ink) is added to a sample of CSF or a sample prepared from **skin lesions**. If the fungus is present, it will become visible when the ink binds to the capsule or covering that surrounds the fungus. Faster results are obtained with the India ink test, but it is less accurate than the blood test (75–85% accuracy compared to 99% accuracy with the blood test) because some strains are not visible using this method. Antigen tests are routinely recommended for non-symptomatic patients with advanced AIDS.

Another way to diagnose cryptococcosis is to culture a sample of sputum, tissue from a **lung biopsy**, or CSF in the laboratory to isolate the fungus. Cultures are also done to assess the effectiveness of treatment.

Chest x rays are useful in assessing lung damage and may reveal a single mass or multiple distinct nodules, but the x ray alone does not lead to a definitive diagnosis of cryptococcosis.

Treatment

Once cryptococcosis is diagnosed, treatment begins with amphotericin B (Fungizone), sometimes in combination with 5-flucytosine (Ancobon). Amphotericin B is a powerful fungistatic drug with potentially toxic side



This lesion appearing on this person's body is due to exposure of the *C. neoformans* fungus. (Photo Researchers, Inc. Reproduced by permission.)

effects, such as kidney toxicity and lower concentrations of an important blood component called hemoglobin. This medication can also cause fever, chills, **nausea and vomiting**, **diarrhea**, headache, and muscle aches. Treatment is generally given intravenously during a hospital stay and continues until the patient is stable or improving (no more than two to three weeks). 5-flucytosine is given orally. Patients may also receive other medication to minimize the side effects from these drugs.

Amphotericin B, with or without 5-flucytosine, is given for several weeks until the patient is stable, after which the patient receives oral fluconazole (Diflucan). Fluconazole is a broad-spectrum antifungal drug with few serious side effects. Patient with AIDS must continue taking fluconazole for the rest of their lives to prevent a relapse of cryptococcosis. Sometimes fluconazole is given to patients with advanced AIDS as a preventative (prophylactic) measure.

Because of the high cost of fluconazole, the manufacturer of the drug, Pfizer, has established a financial assistance plan to make the drug available at lower cost to those who meet certain criteria. Patients needing this drug should ask their doctors about this program.

Prognosis

Untreated cryptococcosis is always fatal. The acute mortality rate for patients with AIDS is 10–25%. Most deaths are attributable to cryptococcal meningitis and occur within two weeks after diagnosis. For AIDS patients who do not receive continued suppressive therapy (fluconazole), the relapse rate is 50–60% within six months and a shortened life expectancy. Once the cryptococcosis infection has been successfully treated, individuals may be left with a variety of neurologic symptoms,

KEY TERMS

Adrenal gland—A pair of organs located above the kidneys. The outer tissue of the gland produces the hormones epinephrine (adrenaline) and norepinephrine, while the inner tissue produces several steroid hormones.

Amphotericin B (Fengizone)—An antifungal medication, prescribed for topical or systemic use in treating fungal infections.

Antibody—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

Antigen—A foreign protein or particle capable of eliciting an immune response.

Asymptomatic—Persons who carry a disease but who do not exhibit symptoms of the disease are said to be asymptomatic.

Biopsy—The removal of a tissue sample for diagnostic purposes.

Cerebrospinal fluid (CSF)—The clear fluid that surrounds the spinal cord and brain and acts as a shock absorber.

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.

Encephalitis—Inflammation of the brain.

Hodgkin's disease—A disease that causes chronic inflammation of the lymph nodes, spleen, liver and kidneys. It is also called malignant lymphoma.

Hydrocephalus—Build-up of fluid around the brain.

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

India ink test—A diagnostic test used to detect the cryptococcal organism *C. neoformans*. A dye, called India ink, is added to a sample of CSF fluid, and if the fungi is present, they will become visible as the dye binds to the capsule surrounding the fungus.

Lumbar puncture—Also called a spinal tap, a procedure in which a thin needle is used to withdraw a sample of cerebrospinal fluid for diagnostic purposes from the area surrounding the spine.

Meningitis—Inflammation of the membranes covering the brain and spinal cord called the meninges.

Molluscum contagiosum—A disease of the skin and mucous membranes, caused by a poxvirus and found all over the world.

Opportunistic infection—An infection that is normally mild in a healthy individual, but which takes advantage of an ill person's weakened immune system to move into the body, grow, spread, and cause serious illness.

Pneumonia—Inflammation of the lungs, typically caused by a virus, bacteria, or other organism.

such as weakness, headache, and hearing or visual loss. In addition, fluid may accumulate around the brain (**hydrocephalus**).

Prevention

The best way to prevent cryptococcosis is to stay free of HIV infection. People with suppressed immune systems should try to stay away from areas contaminated with pigeon or other bird droppings, such as the attics of old buildings, barns, and areas under bridges where pigeons roost.

Resources

ORGANIZATIONS

Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. (800) 311-3435, (404) 639-3311. <<http://www.cdc.gov>>.

National AIDS Clearinghouse. 800-458-5231.

National AIDS Hotline. 800-342-AIDS.

Project Inform. 205 13th Street, #2001, San Francisco, CA 94103. (800) 822-7422. <<http://www.projinf.org>>.

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Tish Davidson

***Cryptococcus neoformans* infection** see
Cryptococcosis

Cryptorchidism see **Undescended testes**

Cryptosporidiosis

Definition

Cryptosporidiosis refers to infection by the spine-forming protozoan known as *Cryptosporidium*. Protozoa are a group of parasites that infect the human intestine, and include the better known *Giardia*. *Cryptosporidium* was first identified in 1976 as a cause of disease in humans.

Description

Cryptosporidium are normally passed in the feces of infected persons and animals in the form of cysts. The cysts can remain in the ground and water for months, and when ingested produce symptoms after maturing in the intestine and the bile ducts. When viewed under the microscope, they appear as small bluish-staining round bodies. Most common sources of infection are other humans, water supplies, or reservoirs. These are contaminated by animals that defecate in these areas. An outbreak in Milwaukee in 1993 in which over 400,000 persons were affected was traced to the city's water supply. Cysts of *Cryptosporidium* are extremely resistant to the disinfectants that are commonly used in most water treatment plants and are incompletely removed by filtration.

Most persons who experience significant symptoms have an altered immune system, and suffer from diseases such as AIDS and cancer. However, as shown in the Milwaukee outbreak, even those with normal immunity can experience symptoms.

Causes and symptoms

Cysts of *Cryptosporidium* mature in the intestine and bile ducts within three to five days of ingestion. As noted, large-scale infections from contaminated water supplies has been documented. However, human to human transmission (such as occurs in day care centers or through sexual behavior) is also an important cause.

Many individuals can be infected without any illness, but the major symptom is diarrhea, which is often watery and incapacitating. Dehydration, low-grade fever, nausea, and abdominal cramps are frequent.

In those with a normal immune system, the disease usually lasts about 10 days. For patients with altered immunity (immunocompromised), the story is quite different, with diarrhea becoming chronic, debilitating, and even fatal.

Complications

Dehydration and malnutrition are the most common effects of infection. In about 20% of AIDS patients,

bile duct infection also occurs and causes symptoms similar to gallbladder attacks. Eighty percent or more of those with infection of the bile ducts die from the disease. The lungs and pancreas are also sometimes involved. *Cryptosporidium* are just one cause of the diarrhea wasting syndrome in AIDS, which results in severe weight loss and malnutrition.

Diagnosis

This is based on either finding the characteristic cysts in stool specimens, or on biopsy of an infected organ, such as the intestine.

Treatment

The first aim of treatment is to avoid dehydration. Oral Rehydration Solution (ORS) or intravenous fluids may be needed. Medications used to treat diarrhea by decreasing intestinal motility (Anti-Motility Agents), such as loperamide or diphenoxylate, are also useful, but should only be used with the advice of a physician.

Treatment aimed directly at *Cryptosporidium* is only partially effective, and rarely eliminates the organism. The medication most commonly used is paromomycin (Humatin), but others are presently under evaluation.

Prognosis

Cryptosporidium rarely cause a serious disease in persons with normal immune systems. Replacement of fluids is all that is usually needed. On the other hand, those with altered immune systems often suffer for months to years. Paromomycin and other drugs have been able to improve symptoms in over half of those treated. Unfortunately, many organisms are resistant, and recurrence is frequent.

Prevention

The best way to prevent cryptosporidiosis is to minimize exposure to cysts from infected humans and animals. Proper hand washing technique, especially in day care centers, is recommended.

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KEY TERMS

Anti-motility medications—Medications such as loperamide (sold as Imodium), diphenoxydate (sold as Lomotil), or medications containing codeine or narcotics that decrease the ability of the intestine to contract. This can worsen the condition of a patient with dysentery or colitis.

Cyst—A protective sac that includes either fluid or the cell of an organism. The cyst enables many organisms to survive in the environment for long periods of time without need for food or water.

Immunocompromised—A change or alteration of the immune system that normally serves to fight off infections and other illnesses. This can involve changes in antibodies that the body produces (hygogammaglobulinemia), or defect in the cells that partake in the immune response. Diseases such as AIDS and cancer exhibit changes in the body's natural immunity.

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.

Parasite—An organism that lives on or in another and takes nourishment (food and fluids) from that organism.

Protozoa—Group of extremely small single cell (unicellular) or acellular organisms that are found in moist soil or water. They tend to exist as parasites, living off other life forms.

Spore—A resistant form of certain species of bacteria, protozoa, and other organisms.

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David Kaminstein, MD

C-section see **Cesarean section**

CSF analysis see **Cerebrospinal fluid (CSF) analysis**

CT-guided biopsy

Definition

Computed tomography (CT) is a process that images anatomic information from a cross-sectional plane of the body. Biopsy is the process of taking a sample of tissue from the body for analysis. CT is commonly used in biopsies to provide images that help guide the tools or equipment necessary to perform the biopsy to the appropriate area of the body.

Purpose

CT is used in the process of performing a biopsy, such as a needle biopsy, in order to guide the needle to the site of the biopsy and to provide rapid and precise localization of the needle. CT enables imaging of areas that are normally beyond visible boundaries. This enables the physician to see the target area clearly and help to ensure that the tissue being removed is from the target lesion.

Precautions

The patient that suffers from claustrophobia will want to discuss this with their physician. This procedure

KEY TERMS

Lesion—A pathologic change in tissues.

Malignancy—A locally invasive and destructive growth.

involves the patient being placed into the CT scanner, typically a small, enclosed area. Depending on the specific type of biopsies being performed, certain anesthetics will be used, so discuss drug **allergies** with your physician.

Description

CT can assist in providing more enhanced images of a suspicious lesion. It helps to determine whether a tumor is truly solitary or not. CT can characterize the tumor and aid in the estimation of malignancy.

Preparation

Since there are many different types of biopsies, you should follow the instructions from your physician to prepare for your CT-guided biopsy. Patients who suffer from claustrophobia should discuss their concerns with the physician. In some cases, medicine can be given that will relax the patient during the procedure.

Risks

CT-guided biopsy does not increase the risk of the biopsy any more than any other radiologic imaging such as x ray.

Normal results

Because the area being biopsied, as well as the specific type of biopsy procedure can vary, results will vary. Before undergoing the procedure, notification procedure should be clearly defined.

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Kim A. Sharp

CT-myelogram see **Myelography**

CT scan see **Computed tomography scans**

Culture-fair test

Definition

A culture-fair test is a test designed to be free of cultural bias, as far as possible, so that no one culture has an advantage over another. The test is designed to not be influenced by verbal ability, cultural climate, or educational level.

Purpose

The purpose of a culture-fair test is to eliminate any social or cultural advantages, or disadvantages, that a person may have due to their upbringing. The test can be administered to anyone, from any nation, speaking any language. A culture-fair test may help identify learning or emotional problems. The duration of the test varies for the individual types of tests available, but the time is approximately between 12–18 minutes per section (a test usually has two to four sections).

A culture-fair test is often administered by employers in order to determine the best location for new employees in a large company. The wide variety of culture-fair tests available allows the administrator to select which area is most vital, whether it be general intelligence, knowledge of a specific area, or emotional stability.

Precautions

There is doubt as to whether any test can truly be culturally unbiased or can ever be made completely fair to all persons independent of culture. There are no other precautions.

Description

A culture-fair test is a non-verbal paper-pencil test that can be administered to patients as young as four

years old. The patient only needs the ability to recognize shapes and figures and perceive their respective relationships. Some examples of tasks in the test may include:

- completing series
- classifying
- solving matrices
- evaluating conditions

The culture-fair test is also often referred to as a culture-free test or unbiased test. There are many variations of the test including class, economic, and intelligence tests. The threading theme among the various tests is their design to be culturally unbiased.

Preparation

The only preparation necessary to administer the test is pre-ordered materials and a quiet and secluded location for the duration of the test.

Aftercare

Post-test treatment depends on the results of the test and the specifics of the individual patient. Any further treatment is best prescribed by the doctor.

Risks

There are no risks associated with the culture-fair test.

Normal results

The results can be compared to the key that comes with the purchase of a culture-fair test. All results should be compared to the included key.

Abnormal results

The results can be compared to the key that comes with the purchase of a culture-fair test. All results should be compared to the included key.

Resources

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Ronald Watson, PhD

Cultures for sexually transmitted diseases
see **Sexually transmitted diseases cultures**

Cushing's syndrome

Definition

Cushing's syndrome is a relatively rare endocrine (hormonal) disorder resulting from excessive exposure to the hormone cortisol. The disorder, which leads to a variety of symptoms and physical abnormalities, is most commonly caused by taking medications containing the hormone over a long period of time. A more rare form of the disorder occurs when the body itself produces an excessive amount of cortisol.

Description

The adrenals are two glands, each of which is perched on the upper part of the two kidneys. The outer part of the gland is known as the cortex; the inner part is known as the medulla. Each of these parts of the adrenal gland is responsible for producing different types of hormones. Regulation of hormone production and release from the adrenal cortex involves the pituitary gland, a small gland located at the base of the brain. After the hypothalamus (the part of the brain containing secretions important to metabolic activities) sends "releasing hormones" to the pituitary gland, the pituitary secretes a hormone called adrenocorticotrophic hormone (ACTH). The ACTH then travels through the bloodstream to the adrenal cortex, where it encourages the production and release of cortisol (sometimes called the "stress" hormone) and other adrenocortical hormones.

Cortisol, a very potent glucocorticoid—a group of adrenocortical hormones that protects the body from stress and affect protein and carbohydrate metabolism—is involved in regulating the functioning of nearly every type of organ and tissue in the body, and is considered to be one of the few hormones absolutely necessary for life. Cortisol is involved in:

- complex processing and utilization of many nutrients, including sugars (carbohydrates), fats, and proteins
- normal functioning of the circulatory system and the heart
- functioning of muscles
- normal kidney function
- production of blood cells
- normal processes involved in maintaining the skeletal system
- proper functioning of the brain and nerves
- normal responses of the immune system

Cushing's syndrome, also called hypercortisolism, has an adverse effect on all of the processes described

above. The syndrome occurs in approximately 10 to 15 out of every one million people per year, usually striking adults between the ages of 20 and 50.

Causes and symptoms

The most common cause of Cushing's syndrome is the long-term use of glucocorticoid hormones in medications. Medications such as prednisone are used in a number of inflammatory conditions. Such conditions include **rheumatoid arthritis, asthma, vasculitis, lupus**, and a variety of other **autoimmune disorders** in which the body's immune cells accidentally attack some part of the body itself. In these disorders, the glucocorticoids are used to dampen the immune response, thereby decreasing damage to the body.

Cushing's syndrome can also be caused by three different categories of disease:

- a pituitary tumor producing abnormally large quantities of ACTH
- the abnormal production of ACTH by some source other than the pituitary
- a tumor within the adrenal gland overproducing cortisol

Although it is rare, about two-thirds of endogenous (occurring within the body rather than from a source outside the body, like a medication) Cushing's syndrome is a result of Cushing's disease. The term "Cushing's disease" refers to Cushing's syndrome, which is caused by excessive secretion of ACTH by a pituitary tumor, usually an adenoma (noncancerous tumor). The pituitary tumor causes increased growth of the adrenal cortex (hyperplasia) and increased cortisol production. Cushing's disease affects women more often than men.

Tumors in locations other than the pituitary can also produce ACTH. This is called ectopic ACTH syndrome ("ectopic" refers to something existing out of its normal place). Tumors in the lung account for more than half of all cases of ectopic ACTH syndrome. Other types of tumors that may produce ACTH include tumors of the thymus, the pancreas, the thyroid, and the adrenal gland. Nearly all adrenal gland tumors are benign (noncancerous), although in rare instances a tumor may actually be cancerous.

Symptoms of cortisol excess (resulting from medication or from the body's excess production of the hormone) include:

- weight gain
- an abnormal accumulation of fatty pads in the face (creating the distinctive "moon face" of Cushing's syndrome); in the trunk (termed "truncal obesity"); and



Woman with Cushing's syndrome. (Photo Researchers, Inc. Reproduced by permission.)

over the upper back and the back of the neck (giving the individual what has been called a "buffalo hump")

- purple and pink stretch marks across the abdomen and flanks
- high blood pressure
- weak, thinning bones (osteoporosis)
- weak muscles
- low energy
- thin, fragile skin, with a tendency toward both bruising and slow healing
- abnormalities in the processing of sugars (glucose), with occasional development of actual diabetes
- kidney stones
- increased risk of infections
- emotional disturbances, including mood swings, depression, irritability, confusion, or even a complete break with reality (psychosis)

KEY TERMS

Adenoma—A type of noncancerous (benign) tumor that often involves the overgrowth of certain cells of the type normally found within glands.

Adrenocorticotrophic hormone (ACTH)—A pituitary hormone that stimulates the cortex of the adrenal glands to produce adrenal cortical hormones.

Cortisol—A hormone secreted by the cortex of the adrenal gland. Cortisol regulates the function of nearly every organ and tissue in the body.

Ectopic—In an abnormal position.

Endocrine—Pertaining to a gland that secretes directly into the bloodstream.

Gland—A collection of cells whose function is to release certain chemicals (hormones) that are important to the functioning of other, sometimes distantly located, organs or body systems.

Glucocorticoids—General class of adrenal cortical hormones that are mainly active in protecting against stress and in protein and carbohydrate metabolism.

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

Hypothalamus—the part of the brain containing secretions important to metabolic activities.

Pituitary—A gland located at the base of the brain, the pituitary produces a number of hormones, including hormones that regulate growth and reproductive function.

- irregular menstrual periods in women
- decreased sex drive in men and difficulty maintaining an erection
- abnormal hair growth in women (in a male pattern, such as in the beard and mustache area), as well as loss of hair from the head (receding hair line)

Diagnosis

Diagnosing Cushing's syndrome can be complex. Diagnosis must not only identify the cortisol excess, but also locate its source. Many of the symptoms listed above can be attributed to numerous other diseases. Although a number of these symptoms seen together would certainly suggest Cushing's syndrome, the symp-

toms are still not specific to Cushing's syndrome. Following a review of the patient's medical history, **physical examination**, and routine blood tests, a series of more sophisticated tests is available to achieve a diagnosis.

24-hour free cortisol test

This is the most specific diagnostic test for identifying Cushing's syndrome. It involves measuring the amount of cortisol present in the urine over a 24-hour period. When excess cortisol is present in the bloodstream, it is processed by the kidneys and removed as waste in the urine. This 24-hour free cortisol test requires that an individual collect exactly 24-hours' worth of urine in a single container. The urine is then analyzed in a laboratory to determine the quantity of cortisol present. This technique can also be paired with the administration of dexamethasone, which in a normal individual would cause urine cortisol to be very low. Once a diagnosis has been made using the 24-hour free cortisol test, other tests are used to find the exact location of the abnormality causing excess cortisol production.

Dexamethasone suppression test

This test is useful in distinguishing individuals with excess ACTH production due to a pituitary adenoma from those with ectopic ACTH-producing tumors. Patients are given dexamethasone (a synthetic glucocorticoid) orally every six hours for four days. Low doses of dexamethasone are given during the first two days; for the last two days, higher doses are administered. Before dexamethasone is administered, as well as on each day of the test, 24-hour urine collections are obtained.

Because cortisol and other glucocorticoids signal the pituitary to decrease ACTH, the normal response after taking dexamethasone is a drop in blood and urine cortisol levels. Thus, the cortisol response to dexamethasone differs depending on whether the cause of Cushing's syndrome is a pituitary adenoma or an ectopic ACTH-producing tumor.

However, the dexamethasone suppression test may produce false-positive results in patients with conditions such as depression, alcohol abuse, high estrogen levels, acute illness, and stress. On the other hand, drugs such as phenytoin and phenobarbital may produce false-negative results. Thus, patients are usually advised to stop taking these drugs at least one week prior to the test.

Corticotropin-releasing hormone (CRH) stimulation test

The CRH stimulation test is given to help distinguish between patients with pituitary adenomas and

those with either ectopic ACTH syndrome or cortisol-secreting adrenal tumors. In this test, patients are given an injection of CRH, the corticotropin-releasing hormone that causes the pituitary to secrete ACTH. In patients with pituitary adenomas, blood levels of ACTH and cortisol usually rise. However, in patients with ectopic ACTH syndrome, this rise is rarely seen. In patients with cortisol-secreting adrenal tumors, this rise almost never occurs.

Petrosal sinus sampling

Although this test is not always necessary, it may be used to distinguish between a pituitary adenoma and an ectopic source of ACTH. Petrosal sinus sampling involves drawing blood directly from veins that drain the pituitary. This test, which is usually performed with local anesthesia and mild **sedation**, requires inserting tiny, flexible tubes (catheters) through a vein in the upper thigh or groin area. The catheters are then threaded up slowly until they reach veins in an area of the skull known as the petrosal sinuses. X rays are typically used to confirm the correct position of the catheters. Often CRH is also given during the test to increase the accuracy of results.

When blood tested from the petrosal sinuses reveals a higher ACTH level than blood drawn from a vein in the forearm, the likely diagnosis is a pituitary adenoma. When the two samples show similar levels of ACTH, the diagnosis indicates ectopic ACTH syndrome.

Radiologic imaging tests

Imaging tests such as **computed tomography scans** (CT) and **magnetic resonance imaging** (MRI) are only used to look at the pituitary and adrenal glands after a firm diagnosis has already been made. The presence of a pituitary or adrenal tumor does not necessarily guarantee that it is the source of increased ACTH production. Many healthy people with no symptoms or disease whatsoever have noncancerous tumors in the pituitary and adrenal glands. Thus, CT and MRI is often used to image the pituitary and adrenal glands in preparation for surgery.

Treatment

The choice of a specific treatment depends on the type of problem causing the cortisol excess. Pituitary and adrenal adenomas are usually removed surgically. Malignant adrenal tumors always require surgical removal.

Treatment of ectopic ACTH syndrome also involves removing all of the cancerous cells that are producing ACTH. This may be done through surgery, **chemotherapy** (using combinations of cancer-killing drugs), or **radiation therapy** (using x rays to kill **cancer** cells), depend-

ing on the type of cancer and how far it has spread. Radiation therapy may also be used on the pituitary (with or without surgery) for patients who cannot undergo surgery, or for patients whose surgery did not successfully decrease pituitary release of ACTH.

There are a number of drugs that are effective in decreasing adrenal production of cortisol. These medications include mitotane, ketoconazole, metyrapone, trilostane, aminoglutethimide, and **mifepristone**. These drugs are sometimes given prior to surgery in an effort to reverse the problems brought on by cortisol excess. However, the drugs may also need to be administered after surgery (sometimes along with radiation treatments) in patients who continue to have excess pituitary production of ACTH.

Because pituitary surgery can cause ACTH levels to drop too low, some patients require short-term treatment with a cortisol-like medication after surgery. Patients who need adrenal surgery may also require glucocorticoid replacement. If the entire adrenal gland has been removed, the patient must take oral glucocorticoids for the rest of his or her life.

Prognosis

Prognosis depends on the source of the problem. When pituitary adenomas are identified as the source of increased ACTH leading to cortisol excess, about 80% of patients are cured by surgery. When cortisol excess is due to some other form of cancer, the prognosis depends on the type of cancer and the extent of its spread.

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ORGANIZATION

- Cushing's Support and Research Foundation, Inc. 65 East India Row, Suite 22B, Boston, MA 02110. (617) 723-3674. <<http://www.world.std.com>>.
- National Adrenal Disease Foundation. 505 Northern Boulevard, Suite 200, Great Neck, NY 11021. (516) 487-4992. <<http://www.medhelp.org>>.
- National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health, Bethesda, MD 20892-2560. <<http://www.ninds.nih.gov>>.
- Pituitary Network Association. 16350 Ventura Boulevard, #231, Encino, CA 91436. (805)499-9973. <<http://www.pituitary.org>>.

Rosalyn Carson-DeWitt



Linear red rashes around a patient's knee caused by burrowing larvae of the dog hookworm *Ancylostoma brasiliense*. (Photograph by Dr. P. Marazzi, Custom Medical Stock Photo. Reproduced by permission.)

are able to penetrate human skin (even through solid material, such as a beach towel). The larvae are commonly found in shaded, moist, or sandy areas (such as beaches, a child's sandbox, or areas underneath a house), where they are easily picked up by bare feet or buttocks.

In minor infestations, there may be no symptoms at all. In more severe cases, a red elevation of the skin (papule) appears within a few hours after the larvae have penetrated the skin. This usually arises first in areas that are in contact with the soil, such as the feet, hands, and buttocks.

Between a few days and a few months after infection, the larvae begin to migrate beneath the skin, leaving extremely itchy red lines that may be accompanied by blisters. These red lines usually appear at the top of the sole of the foot or on the buttocks.

Typically, the larvae travel through the bloodstream, to the lungs, and then migrate into the mouth where they are swallowed and attach to the small intestine lining. There they mature into adult worms. In cases where the larvae migrate through the lungs, they can produce anemia, cough, and pneumonia, in addition to the itchy rash.

Diagnosis

The condition can be diagnosed by microscopic inspection of feces which can reveal hookworm eggs. In addition visual inspection of the skin would reveal tell-tale itchy red lines and blisters.

Treatment

People without intestinal symptoms do not need treatment, since the worms will eventually die or be

Cutaneous larva migrans

Definition

Cutaneous larva migrans is a parasitic skin disease caused by a hookworm larvae that usually infests dogs, cats, and other animals. Humans can pick up the infection by walking barefoot on soil or beaches contaminated with animal feces.

Description

Cutaneous larva migrans (also called "creeping eruption" or "ground itch") is found in southeastern and Gulf states, and in tropical developing countries.

The hookworms that cause the condition are small, round blood-sucking worms that infest about 700 million people around the world. Cutaneous larva migrans occurs most often among children, those who crawl beneath raised buildings, and sunbathers who lie down on wet sand contaminated with hookworm larvae.

Causes and symptoms

After an animal passes feces that are infested with hookworm eggs, the eggs hatch into infective larvae that

KEY TERMS

Larvae—Immature forms of certain worms.

excreted. Thiabendazole or albendazole are used to treat the infestation. Mild infections can be treated by applying one of the drugs to the skin along the tracks and the normal skin surrounding the area. Thiabendazole also can be given internally, but taken this way it can cause side effects including **dizziness**, nausea, and vomiting.

Prognosis

No matter how severe an infestation, with adequate treatment patients recover completely. However, if the patient scratches the lesions open, the areas can become vulnerable to bacterial infection.

Prevention

In the United States, the prevalence of dogs and cats with hookworms is the reason why the infective larvae are found so commonly in soil and sand. The play habits of children, together with their attraction to pets, puts them at high risk for hookworm infection and cutaneous larvae migrans.

Human hookworm infestation can be prevented by practicing good personal hygiene, deworming pets, and not allowing children to play in potentially contaminated environments.

Resources

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Carol A. Turkington

Description

CTCL, also known as mycosis fungoides, is a **cancer** of the white blood cells that primarily affects the skin and only secondarily affects other sites. This disease involves the uncontrollable proliferation of T-lymphocytes known as T-helper cells, so named because of their role in the immune response. T-helper cells are characterized by the presence of a protein receptor on its surface called CD4. Accordingly, T-helper cells are said to be CD4+.

The proliferation of T-helper cells results in the penetration, or infiltration, of these abnormal cells into the epidermal layer of the skin. The skin reacts with slightly scaling lesions that itch, although the sites of greatest infiltration do not necessarily correspond to the sites of the lesions. The lesions are most often located on the trunk, but can be present on any part of the body. In the most common course of the disease, the patchy lesions progress to palpable plaques that are deeper red and have more defined edges. As the disease worsens, skin tumors develop that are often mushroom-shaped, hence the name mycosis fungoides. Finally, the cancer progresses to extracutaneous involvement, often in the lymph nodes or the viscera.

CTCL is a rare disease, with an annual incidence of about 0.29 cases per 100,000 persons in the United States. It is about half as common in Eastern Europe. However, this discrepancy may be attributed to a differing physician awareness of the disease rather than a true difference in occurrence. In the United States, there are about 500 to 600 new cases a year and about 100 to 200 deaths. Usually seen in older adults, the median age at diagnosis is 55 to 60 years, and it strikes twice as many men as women.

Causes and symptoms

The cause of CTCL is unknown. Exposure to chemicals or pesticides has been suggested but the most recent study on the subject failed to show a connection between exposure and development of the disease. The ability to isolate various viruses from cell lines grown from cells of CTCL patients raises the question of a viral cause, but studies have been unable to confirm these suspicions.

The symptoms of CTCL are seen primarily in the skin, with itchy red patches or plaques and, usually over time, mushroom-shaped skin tumors. Any part of the skin can be involved and the extent and distribution of the rash or tumors vary greatly from patient to patient. The only really universal symptom of the disease is the itch and this symptom is usually what brings the patient to the doctor for treatment. If the disease spreads outside of the skin, the symptoms include swelling of the lymph

Cutaneous T-cell lymphoma

Definition

Cutaneous T-cell lymphoma (CTCL) is a malignancy of the T-helper (CD4+) cells of the immune system.

nodes, usually most severe in those draining the areas with skin involvement. Spread to the viscera is most often manifested as disorders of the lungs, upper digestive tract, central nervous system, or liver but virtually any organ can be shown to be involved at **autopsy**.

Diagnosis

Diagnosis of CTCL is often difficult in the early stages because of its slow progression and ability to mimic many other benign skin conditions. The early patches of CTCL resemble eczema, **psoriasis**, and **contact dermatitis**. In a further complication, the early manifestations of the disease can respond favorably to the topical corticosteroid treatments prescribed for these skin disorders. This has the unfortunate result of the disease being missed and the patient remaining untreated for years. CTCL is most likely discovered when a physician maintains a suspicion about the disease, performs multiple skin biopsies, and provides close follow-up after the initial presentation.

Skin biopsies showing penetration of abnormal cells into the epidermal tissue are necessary to make a firm diagnosis of CTCL. Several molecular studies can also help support the diagnosis. The first looks at the cellular proteins seen on the surface of the abnormal cells. Many cases of CTCL show the retention of the CD4+ protein, but the loss of other proteins usually seen on the surface of mature CD4+ cells, such as Leu-8 or Leu-9. The abnormal cells also show unusual rearrangements at the genetic level for the gene that encodes the T-cell receptors. These rearrangements can be identified using Southern blot analysis. The information from the molecular tests, combined with the presence of abnormal cells in the epidermis, strongly supports the CTCL diagnosis.

Treatment

Treatment of CTCL depends on the stage of the disease. The current staging of this disease was first presented at the International Consensus Conference on CTCL in 1997. The staging attempts to show the complex interaction between the various outward symptoms of the disease and prognosis. The system has seven clinical stages based on skin involvement (tumor = T), lymph node involvement (LN), and presence of visceral metastases (M).

The first stage, IA, is characterized by plaques covering less than 10% of the body (T1) and no visceral involvement (M0). Lymph node condition at this stage can be uninolved, reactive to the skin disease, or dermatopathic (biopsies showing CTCL involvement) but not enlarged (LN0-2). The shorthand expression of this stage is therefore T1, LN0-2, M0. The next stage, IB, dif-

fers from IA in that greater than 10% of the body is covered by plaques (T2, LN0-2, M0). Stage IIA occurs with any amount of plaques in addition to the ability to palpate the lymph node and the lymph uninvolved, reactive, or dermatopathic (T1-2, LN0-2, M0).

Treatments applied to the skin are preferred for patients having these preliminary stages of the disease, commonly topical **chemotherapy** with mechlorethamine hydrochloride (nitrogen mustard) or **phototherapy** of psoralen plus ultraviolet A (PUVA). Topical chemotherapy involves application to the skin of nitrogen mustard, an alkylating agent, in a concentration of 10–20 mg/dL in an aqueous or ointment base. Treatment of affected skin is suggested at a minimum and application over the entire skin surface is often recommended. Care needs to be taken that coverage of involved skin is adequate, as patients who self-apply the drug often cannot reach all affected areas. The most common side effect is skin hypersensitivity to the drug. Nearly all patients respond favorably to this treatment, with a 32–61% complete response rate, based on amount of skin involvement. Unfortunately, only 10–15% of patients maintain a complete response rate after discontinuing the treatment.

Phototherapy involves treatment with an orally administered drug, 8-methoxysoralen, that renders the skin sensitive to long-wave ultraviolet light (UVA), followed by controlled exposure to the radiation. During the initial treatment period, which may last as long as six months, patients are treated two to three times weekly. This is reduced to about once monthly after initial clearing of the lesions. Redness of the skin and blistering are the most common side effects of the treatment and are much more common in patients presenting with overall skin redness, or erythroderma, so lower intensities of light are usually used in this case. About 50% of all patients experience complete clearance with this treatment. Some patients with very fair skin and limited skin involvement can successfully treat themselves at home with special lamps and no psoralen.

The next stage, IIB, involves one or more cutaneous tumors, in combination with absent or present palpable lymph nodes, lymph uninvolved, reactive, or dermatopathic, and no visceral involvement (T3, LN0-2, M0). Stage III is characterized by erythroderma, an abnormal redness over widespread areas of the skin (T4, LN0-2, M0).

For more extensive disease, **radiation therapy** is an effective treatment option. It is generally used after the topical treatments have proven ineffective. Individual plaques or tumors can be treated using electrons, orthovoltage x rays, or megavoltage photons with exposure in the range of 15 to 25 Gy. Photon therapy has proven particularly useful once the lymph nodes are involved. Anoth-

er possibility is total-skin electron beam therapy (TSEB), although the availability of this treatment method is limited. It involves irradiation of the entire body with energized electrons. Side effects of this treatment include loss of finger and toe nails, acute redness of the skin, and inability to sweat for about six to 12 months after therapy. Almost all patients respond favorably to radiation treatment and any reoccurrence is usually much less severe.

Combinations of different types of treatments is a very common approach to the management of CTCL. Topical nitrogen mustard or PUVA is often used after completion of radiation treatment to prolong the effects. The addition of genetically engineered interferon to PUVA therapy significantly increases the percentage of patients showing a complete response. Furthermore, although treatments using chemotherapy drugs alone, such as deoxycytidine or etretinate, have been disappointing for CTCL, combining these drugs with interferon has shown promising results. Interferon has also been combined with retinoid treatments, although the mechanism of action of retinoids (Vitamin A analogues) against CTCL is unknown.

The final two stages of the disease are IVA and IVB. IVA presents as any amount of skin involvement, absent or present palpable lymph nodes, no visceral involvement, and lymph that contains large clusters of convoluted cells or obliterated nodes (T1-4, LN3-4). IVB differs in the addition of palpable lymph nodes and visceral involvement (T1-4, LN3-4, M1). All of the treatment methods described above are appropriate for the final two stages of the disease.

Alternative treatment

Itching of the skin is one of the most troublesome symptoms of CTCL. One alternative treatment for itchiness is the application of a brewed solution of chickweed that is applied to the skin using cloth compresses. Another suggested topical application is a mixture of vitamin E, vitamin A, unflavored yogurt, honey, and zinc oxide. Evening primrose oil applied topically is also claimed to reduce itch and promote healing.

Prognosis

The prognosis for CTCL is dependent on the stage of the disease. Prognosis is very good if the disease has only progressed to Stage IA, with a mean survival of 20 or more years. At this point, the disease is a very low mortality risk to the patient, with most deaths occurring to persons in this group unrelated to CTCL. For patients diagnosed at stages IB and IIA, the median survival is about 12 years. The disease in both of these stages involves intermediate risk to the patient. Patients in stage III and IVA

KEY TERMS

Alkylating agent—A chemical that alters the composition of the genetic material of rapidly dividing cells, such as cancer cells, causing selective cell death; used as a topical chemotherapeutic agent to treat CTCL.

Erythroderma—An abnormal reddening of the entire skin surface.

T-helper cells—A cellular component of the immune system that plays a major role in ridding the body of bacteria and viruses, characterized by the presence of the CD4 protein on its surface; the type of cell that divides uncontrollable with CTCL.

Total-skin electron beam therapy—A method of radiation therapy used to treat CTCL that involves bombarding the entire body surface with high-energy electrons.

have a mean life expectancy of about five years. At these later stages, the disease is high risk, with most deaths occurring by infection due to the depleted immune system of the later-stage patient. Once a patient has reached stage IVB, the mean life expectancy is one year.

Prevention

Studies have been unable to link CTCL to any environmental or genetic factors, so prevention at this time is not possible.

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

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Cutis laxa

Definition

Cutis laxa (Latin for loose or lax skin) is a connective tissue disorder in which the skin lacks elasticity and hangs in loose folds.

Description

Cutis laxa is extremely rare; less than a few hundred cases worldwide have been described.

The several forms of cutis laxa are divided into primary cutis laxa, which is present from birth and is hereditary, secondary cutis laxa, which arises later in life and may be either hereditary, and acquired cutis laxa, which arises later in life and is not hereditary. Loose skin, the primary and most obvious symptom of these diseases, is caused by underlying defects in connective tissue structure, which also cause more serious internal problems in vocal cords, bones, cartilage, blood vessels, bladder, kidney, digestive system, and lungs. The loose skin is particularly obvious on the face, and children with the disorder look sad or mournful.

There are four genetic forms of the disease: sex-linked, autosomal dominant, and two types of autosomal recessive inheritance. The recessive forms are the most common and are usually more severe than the other forms.

Causes and Symptoms

Sex-linked cutis laxa is caused by a defective gene on the X chromosome. In addition to loose skin, its symptoms are mild **mental retardation**, loose joints, bone abnormalities (like hooked nose, pigeon breast, and funnel breast), frequent loose stools, urinary tract blockages, and deficiencies in lysyl oxidase, an enzyme required for the formation of properly functioning connective tissue. (But the defective gene does not code for lysyl oxidase.)

Autosomal dominant cutis laxa is caused by a defective gene carried on an autosomal (not sex-linked) chromosome. Its symptoms are loose, hanging skin, missing elastic fibers, premature **aging**, and pulmonary **emphysema**. Only a few families are known with cutis laxa inherited as a dominant trait.

Autosomal recessive cutis laxa type 1 is caused by a defective gene on chromosome 5. Symptoms include emphysema; diverticula in the esophagus, duodenum, and bladder; lax and dislocated joints; tortuous arteries; hernias; lysyl oxidase deficiencies; and retarded growth.

Autosomal recessive cutis laxa type 2 is also inherited as a recessive trait. In addition to the loose skin, this form of the disease is characterized by bone abnormali-

ties, the delayed joining of the cranial (skull) bones, hip dislocation, curvature of the spine, flat feet, and excessive **tooth decay**.

Acquired cutis laxa tends to follow (and may be caused by) severe illness characterized by **fever**, inflammation, and a severe skin rash (**erythema multiforme**); an injury to the nerves that control blood vessel dilation and contraction; or an autoimmune condition.

Diagnosis

The signs of cutis laxa are very obvious, and it is usually easy to diagnose by examining the skin. The determination of which form of cutis laxa is present is aided by information about the associated symptoms and by family histories.

Treatment

There is no effective cure for any of these disorders. Complications are treated by appropriate specialists, for example, cardiologists, gastroenterologists, rheumatologists, and dermatologists. Plastic surgery can be helpful for cosmetic purposes, but the skin may become loose again.

Prognosis

The prognosis for cutis laxa varies with the form of the disorder. The effects may be relatively mild with individuals living a fairly normal, full life, or the disease may be fatal.

Prevention

The inherited forms of cutis laxa are genetically determined and are not currently preventable. **Genetic counseling** can be helpful for anyone with a family history of cutis laxa. The cause of acquired cutis laxa is not known, so no preventive measures can be taken.

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ORGANIZATIONS

British Coalition of Heritable Disorders of Connective Tissue.
Rochester House, 5 Aldershot Road, Fleet, Hampshire GU13 9NG, United Kingdom. (012) 52-810472.
National Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse, National Institutes of Health. 1 AMS Circle, Bethesda, Maryland 20892-3675. (877) 226-4267. <<http://www.nih.gov/niams/healthinfo/info.htm>>.

KEY TERMS

Autosomal—Refers to the 22 pairs (in humans) of chromosomes not involved with sex determination.

Connective tissue—Tissue that supports and binds other tissue; much of it occurs outside of cells (extra-cellular) and consists of fibrous webs of the polymers, elastin and collagen. Cutis laxa is associated with defects in these fibers.

Diverticula—Pouches in the walls of organs.

Dominant trait—A genetic trait where one copy of the gene is sufficient to yield an outward display of the trait; dominant genes mask the presence of recessive genes; dominant traits can be inherited from only one parent.

Duodenum—The uppermost part of the small intestine, about 10 in (25 cm) long.

Esophagus—The tube connecting the throat to the stomach, about 10 in (25 cm) long.

Funnel breast (also known as pectus excavatum)—A condition where there is a hollow depression in the lower part of the chest.

Gene—A portion of a DNA molecule that either

codes for a protein or RNA molecule or has a regulatory function.

Lysyl oxidase—An enzyme required for the crosslinking of elastin and collagen molecules to form properly functioning connective tissue; present in relatively low levels in at least some forms of cutis laxa.

Pigeon breast (also known as pectus carinatum)—A chest shape with a central projection resembling the keel of a boat.

Recessive trait—An inherited trait that is outwardly obvious only when two copies of the gene for that trait are present; an individual displaying a recessive trait must have inherited one copy of the defective gene from each parent.

Sex-linked—Refers to genes or traits carried on one of the sex chromosomes, usually the X.

Tortuous arteries—Arteries with many bends and twists.

X chromosome—One of the two types of sex chromosomes; females have two X chromosomes, while males have one X chromosome and one Y chromosome.

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OMIM Homepage, Online Mendelian Inheritance in Man.
<http://www.ncbi.nlm.nih.gov/Omim>.

Lorraine Lica, PhD

Cuts see **Wounds**

CVA see **Stroke**

CVS see **Chorionic villus sampling**



This elderly woman's lips turned purple due to central cyanosis, a condition most commonly due to slow blood circulation, leading to a bluish skin coloration. (Photo Researchers, Inc. Reproduced by permission.)

Cyanosis

Definition

Cyanosis is a physical sign causing bluish discoloration of the skin and mucous membranes. Cyanosis is caused by a lack of oxygen in the blood. Cyanosis is associated with cold temperatures, **heart failure**, lung diseases, and smothering. It is seen in infants at birth as a result of heart defects, **respiratory distress syndrome**, or lung and breathing problems.

Description

Blood contains a red pigment (hemoglobin) in its red blood cells. Hemoglobin picks up oxygen from the lungs, then circulates it through arteries and releases it to cells through tiny capillaries. After giving up its oxygen, blood cir-

KEY TERMS

Hemoglobin—A colored substance (pigment) in the blood that carries oxygen to tissues and gives blood its red color.

Respiratory distress syndrome—Also known as hyaline membrane disease, this is a condition of premature infants in which the lungs are imperfectly expanded due to a lack of a substance on the lungs that reduces tension.

culates back to the lungs through capillaries and veins. Hemoglobin, as well as blood, is bright red when it contains oxygen, but appears dark or “bluish” after it gives up oxygen.

The blue discoloration of cyanosis is seen most readily in the beds of the fingernails and toenails, and on the lips and tongue. It often appears transiently as a result of slowed blood flow through the skin due to the cold. As such, it is not a serious symptom. However, in other cases, cyanosis is a serious symptom of underlying disease.

Causes and symptoms

The blue color of the skin and mucous membranes is caused by a lack of oxygen in the blood. Low blood oxygen may be caused by poor blood circulation, or heart or breathing problems. It can also be caused by being in a low-oxygen environment or by **carbon monoxide poisoning**. More rarely, cyanosis can be present at birth as a sign of **congenital heart disease**, in which some of the blood is not pumped to the lungs where oxygen would make the blood a bright red color. Instead, the blood goes to the rest of the body and remains unoxygenated. Cyanosis also may be caused by **poisoning** from chemicals, drugs, or contaminated food and water.

Other signs of low blood oxygen may accompany cyanosis, including feeling lightheaded or **fainting**.

Treatment

Treatment of the underlying disease can restore proper color to the skin.

Prognosis

If the underlying condition (such as heart or lung disease) can be properly treated, the skin will return to its normal shade.

Resources

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Carol A. Turkington

Cyclic vomiting syndrome

Definition

Cyclic vomiting syndrome (CVS) is a rare disorder characterized by recurring periods of vomiting in an otherwise normal child.

Description

Children in the pre-school or early school years are most susceptible to CVS, although it can appear anywhere from infancy to adulthood. This disorder was identified a century ago, but its cause is still unknown. Episodes can be triggered by emotional **stress** or infections, can last hours or days, and can return at any time. Abdominal **pain** is a frequent feature.

Causes and symptoms

The cause of CVS is still a mystery. Similarities to migraine suggest a common cause, but as yet no firm evidence has surfaced. Patients can usually identify some factor that precedes an attack. Vomiting can be protracted and lead to complications such as **dehydration**, chemical imbalances, tearing and burning, and bleeding of the esophagus (swallowing tube). Between attacks, there is no sign of any illness.

Diagnosis

The most important and difficult aspect of CVS is to be sure there is not an acute and life-threatening event in progress. So many diseases can cause vomiting—from bowel obstruction to epilepsy—that an accurate and timely diagnosis is critical. Because there is no way to prove the diagnosis of CVS, the physician must instead disprove every other diagnosis. This can be tedious, expensive, exhausting, and involve almost every system in the body. The first episode may be diagnosed as a stomach flu when nothing more serious turns up. Only after several episodes and several fruitless searches for a cause will a physician normally consider the diagnosis of CVS.

Treatment

Several different medications have given good results in small trials. The **antimigraine drugs** amitriptyline and cyproheptadine performed well for one study group. Propanolol is sometimes effective, and erythromycin helped several patients in one study, not because it is an antibiotic but because it irritates the stomach and encourages it to move its contents forward instead of in reverse.

Alternative treatment

Constitutional homeopathic medicine can work well in treating CVS because it addresses rebalancing the whole person, not just the symptoms.

Prognosis

The disease may go on for many years without a change in pattern. If the acute complications of prolonged vomiting can be successfully prevented or managed, most patients can lead normal lives between episodes. Medications may ease the symptoms during attacks.

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J. Ricker Polsdorfer, MD

Cyclobenzaprine see **Muscle relaxants**

Cyclophosphapha see **Anticancer drugs**

Cyclospora infection see **Cyclosporiasis**

Cyclosporiasis

Definition

Cyclosporiasis refers to infection by the spore-forming protozoan known as *Cyclospora*. Protozoa are a group of parasites that infect the human intestine. Parasites are organisms that live in another body, called the host, and get food and liquids from that host. This parasite is a member of the group of protozoa known as coccidia, to which *Cryptosporidium* also belongs. This group of parasites infects the human intestine, and causes chronic recurrent infections in those with altered immunity or AIDS. Even in people with normal immune function, *Cyclospora* can cause prolonged bouts of **diarrhea** and other gastrointestinal symptoms.

Description

Until recently, *Cyclospora* was considered to be a form of algae. The parasite causes a common form of waterborne infectious diarrhea throughout the world. Just how the parasite gets into water sources is not yet clear. It is known that ingestion of small cysts in contaminated water leads to disease.

Causes and symptoms

Symptoms begin after an incubation period of about a day or so following ingestion of cysts. A brief period of flu-like illness characterized by weakness and low-grade **fever** is followed by watery diarrhea, nausea, loss of appetite, and muscle aches. In some patients, symptoms may wax and wane for weeks, and there are those in whom nausea and burping may predominate. It is also believed that infection can occur without any symptoms at all.

In patients with abnormal immunity (immunocompromised patients), such as those with AIDS and **cancer**, prolonged diarrhea and severe weight loss often become a major problem. The bile ducts are also susceptible to infection in AIDS patients.

Diagnosis

The disease should be suspected in anyone with a history of prolonged or recurrent diarrhea. The parasite is identified either by staining stool specimens or by apply-

KEY TERMS

Anti-motility medications—Medications such as loperamide (sold as Imodium), diphenoxydate (sold as Lomotil), or medications containing codeine or narcotics that decrease the ability of the intestine to contract. This can worsen the condition of a patient with dysentery or colitis.

Cyst—A protective sac that includes either fluid or the cell of an organism. The cyst enables many organisms to survive in the environment for long periods of time without need for food or water.

Immunocompromised—A change or alteration of the immune system that normally serves to fight off infections other illnesses. This can involve changes in antibodies that the body produces (hygogammaglobulinemia), or a defect in the cells that partake in the immune response. Diseases such as AIDS and cancer exhibit changes in the body's natural immunity.

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.

Parasite—An organism that lives on or in another and takes nourishment (food and fluids) from that organism.

Protozoa—Group of extremely small single cell (unicellular) or acellular organisms that are found in moist soil or water. They tend to exist as parasites, living off other life forms.

Spore—A resistant form of certain species of bacteria, protozoa, and other organisms.

ing certain fluorescent ultraviolet techniques to find the characteristic cysts. Biopsy of an infected organ such as the intestine through an endoscope is another way to make the diagnosis.

Treatment

The first aim of treatment as with any severe diarrheal illness is to avoid **dehydration** and **malnutrition**. Oral Rehydration Solution (ORS) or intravenous fluids are sometimes needed. Medications used to treat diarrhea by decreasing intestinal motility, such as loperamide or diphenoxydate are also useful, but should only be used with the advice of a physician.

The use of the medication, trimethoprim-sulfamethoxazole (Bactrim) for one week can be successful in treating intestinal infections and prevents relapse in those with a normal immune system. The same medicine can be prescribed to treat infections of both the intestine or bile ducts in immunocompromised individuals, but maintenance or continuous treatment is often needed.

Prognosis

The outlook is quite good for individuals in whom a diagnosis is made. Even without treatment, symptoms usually do not last much more than a month or so except in cases with altered immunity. Fortunately, treatment is usually successful even in those patients.

Prevention

Aside from a waterborne source as the origin of infection, little else is known about how the parasite is transmitted. Therefore, little can be done regarding prevention, except to maintain proper hand washing techniques and hygiene.

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David Kaminstein, MD

Cyclosporine see **Immunosuppressant drugs**

Cystectomy

Definition

Cystectomy is a surgical procedure to remove the bladder.

Purpose

Cystectomy is performed to treat **cancer** of the bladder. Radiation and **chemotherapy** are also used to treat **bladder cancer**. Surgery is used to remove cancer when it is in the muscle of the bladder.

Precautions

Cystectomy is an aggressive treatment that may not be appropriate for patients with superficial tumors that respond to more conservative treatment.

Description

Cystectomy is a major surgical operation. The patient is placed under general anesthesia. An incision is made across the lower abdomen. The ureters are located, tied and cut. The ureters connect the kidneys to the bladder. Cutting them frees the bladder for removal. The bladder and associated organs are removed. In men, the prostate is removed with the bladder. In women, the uterus, fallopian tubes, ovaries, and part of the vagina are removed with the bladder. The bladder collects urine from the kidneys for excretion at a later time. Since the bladder is removed, a new method must be created to remove the urine. A small piece of the small intestine is removed, cleaned, and tied at one end to form a tube. The other end is used to form a stoma, an opening through the abdominal wall to the outside. The ureters are then connected to the tube. Urine produced by the kidneys now flows down the ureters, into the tube, and through the stoma. The patient wears a bag to collect the urine.

Preparation

The medical team will discuss the procedure and tell the patient where the stoma will appear and what it will look like. The patient receives instruction on caring for a stoma and bag. Counseling may be initiated. A period of **fasting** and an enema may be required.

KEY TERMS

Ureters—Tubes that connect the kidneys to the bladder. Urine produced by the kidneys passes through the ureters to the bladder.

Aftercare

After the operation, the patient is given fluid-based **nutrition** until the intestines begin to function normally again. **Antibiotics** are given to prevent infection of the incision sites. The nature of the organs removed mean that there will be major lifestyle changes for the person undergoing the operation. Men will become impotent because nerves controlling penile erection are cut during removal of the bladder. In women, **infertility** is a consequence because the ovaries and uterus are removed. However, most women who undergo cystectomy are postmenopausal and past their childbearing years.

Both men and women are fitted with an external bag that connects to the stoma and collects the urine. The bag is generally worn around the waist under the clothing. It takes a period of adjustment to get used to wearing the bag. Because there is no bladder, urine is excreted as it is produced, essentially continuously. The stoma must be treated properly to ensure that it does not become infected or blocked. Patients must be trained to care for their stoma. Often there is a period of psychological adjustment to the major change in life style created by the stoma and bag. Patients should be prepared for this by discussion with their physician.

Risks

As with any major surgery, there is a risk of infection; in this case, infection of the intestine is especially dangerous as it can lead to **peritonitis** (inflammation of the membrane lining the abdomen).

Normal results

The bladder is successfully removed and a stoma created. Intestinal function returns to normal and the patient learns proper care of the stoma and bag. He or she adjusts to lifestyle changes and returns to a normal routine of work and recreation, some sports excluded.

Abnormal results

The patient develops an infection at the incision site. The patient does not make a successful psychological adjustment to the long term consequences of **impotence**.

and urinary diversion. In some women, the vagina is constricted, which may require a secondary procedure.

Resources

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John T. Lohr, PhD

Cystic fibrosis

Definition

Cystic fibrosis (CF) is an inherited disease that affects the lungs, digestive system, sweat glands, and male fertility. Its name derives from the fibrous scar tissue that develops in the pancreas, one of the principal organs affected by the disease.

Description

Cystic fibrosis affects the body's ability to move salt and water in and out of cells. This defect causes the lungs and pancreas to secrete thick mucus, blocking passageways and preventing proper function.

CF affects approximately 30,000 children and young adults in the United States, and about 3,000 babies are born with CF every year. CF primarily affects people of white northern-European descent; rates are much lower in non-white populations.

Many of the symptoms of CF can be treated with drugs or nutritional supplements. Close attention to and prompt treatment of respiratory and digestive complications have dramatically increased the expected life span of a person with CF. While several decades ago most children with CF died by age two, today about half of all people with CF live past age 31. That median age is expected to grow as new treatments are developed, and it is estimated that a person born in 1998 with CF has a median expected life span of 40 years.

Causes and symptoms

Causes

Cystic fibrosis is a genetic disease, meaning it is caused by a defect in the person's genes. Genes, found in

the nucleus of all the body's cells, control cell function by serving as the blueprint for the production of proteins. Proteins carry out a wide variety of functions within cells. The gene that, when defective, causes CF, is called the CFTR gene, which stands for cystic fibrosis transmembrane conductance regulator. A simple defect in this gene leads to all the consequences of CF. There are over 500 known defects in the CFTR gene that can cause CF. However, 70% of all people with a defective CFTR gene have the same defect, known as delta-F508.

Much as sentences are composed of long strings of words, each made of letters; genes can be thought of as long strings of chemical words, each made of chemical letters, called nucleotides. Just as a sentence can be changed by rearranging its letters, genes can be mutated, or changed, by changes in the sequence of their nucleotide letters. The gene defects in CF are called point mutations, meaning that the gene is mutated only at one small spot along its length. In other words, the delta-F508 mutation is a loss of one "letter" out of thousands within the CFTR gene. As a result, the CFTR protein made from its blueprint is made incorrectly, and cannot perform its function properly.

The CFTR protein helps to produce mucus. Mucus is a complex mixture of salts, water, sugars, and proteins that cleanses, lubricates, and protects many passageways in the body, including those in the lungs and pancreas. The role of the CFTR protein is to allow chloride ions to exit the mucus-producing cells. When the chloride ions leave these cells, water follows, thinning the mucus. In this way, the CFTR protein helps to keep mucus from becoming thick and sluggish, thus allowing the mucus to be moved steadily along the passageways to aid in cleansing.

In CF, the CFTR protein cannot allow chloride ions out of the mucus-producing cells. With less chloride leaving, less water leaves, and the mucus becomes thick and sticky. It can no longer move freely through the passageways, so they become clogged. In the pancreas, clogged passageways prevent secretion of digestive enzymes into the intestine, causing serious impairment of digestion—especially of fat—which may lead to **malnutrition**. Mucus in the lungs may plug the airways, preventing good air exchange and, ultimately, leading to **emphysema**. The mucus is also a rich source of nutrients for bacteria, leading to frequent infections.

INHERITANCE OF CYSTIC FIBROSIS. To understand the inheritance pattern of CF, it is important to realize that genes actually have two functions. First, as noted above, they serve as the blueprint for the production of proteins. Second, they are the material of inheritance: parents pass on characteristics to their children by combining the genes in egg and sperm to make a new individual.

DOROTHY ANDERSEN, MD (1901–1963)



(Library of Congress)

Each person actually has two copies of each gene, including the CFTR gene, in each of their body cells. During sperm and egg production, however, these two copies separate, so that each sperm or egg contains only one copy of each gene. When sperm and egg unite, the newly created cell once again has two copies of each gene.

The two gene copies may be the same or they may be slightly different. For the CFTR gene, for instance, a person may have two normal copies, or one normal and one mutated copy, or two mutated copies. A person with two mutated copies will develop cystic fibrosis. A person with one mutated copy is said to be a carrier. A carrier will not have symptoms of CF, but can pass on the mutated CFTR gene to his/her children.

When two carriers have children, they have a one in four chance of having a child with CF each time they conceive. They have a two in four chance of having a child who is a carrier, and a one in four chance of having a child with two normal CFTR genes.

Approximately one in every 25 Americans of northern-European descent is a carrier of the mutated CF gene, while only one in 17,000 African Americans and one in 30,000 Asian Americans are carriers. Since carriers are symptom-free, very few people will know whether or not they are carriers unless there is a family history of the disease. Two white Americans with no

Dorothy Andersen was born on May 15, 1901, in Asheville, North Carolina. She was the only child of Hans Peter Andersen and the former Mary Louise Mason. Orphaned as a young adult, Andersen put herself through Saint Johnsbury Academy and Mount Holyoke College before enrolling in the Johns Hopkins School of Medicine, from which she received her M.D. in 1926.

Andersen turned instead to medical research as a pathologist at Babies Hospital of the Columbia-Presbyterian Medical Center in New York City, where she stayed for more than 20 years, eventually becoming chief of pathology in 1952. Andersen is probably best known for discovery of cystic fibrosis in 1935. That discovery came about during the postmortem examination of a child who had supposedly died of celiac disease, a nutritional disorder. She searched for similar cases in the autopsy files and in medical literature, eventually realizing that she had found a disease that had never been described and to which she gave the name cystic fibrosis.

family history of CF have a one in 2,500 chance of having a child with CF.

It may seem puzzling that a mutated gene with such harmful consequences would remain so common; one might guess that the high mortality of CF would quickly lead to loss of the mutated gene from the population. Some researchers now believe the reason for the persistence of the CF gene is that carriers, those with only one copy of the gene, are protected from the full effects of **cholera**, a microorganism that infects the intestine, causing intense **diarrhea** and eventual **death by dehydration**. It is believed that having one copy of the CF gene is enough to prevent the full effects of cholera infection, while not enough to cause the symptoms of CF. This so-called "heterozygote advantage" is seen in some other genetic disorders, including sickle-cell anemia.

Symptoms

The most severe effects of cystic fibrosis are seen in two body systems: the gastrointestinal (digestive) system, and the respiratory tract, from the nose to the lungs. CF also affects the sweat glands and male fertility. Symptoms develop gradually, with gastrointestinal symptoms often the first to appear.

GASTROINTESTINAL SYSTEM. Ten to fifteen percent of babies who inherit CF have meconium **ileus** at birth.

Meconium is the first dark stool that a baby passes after birth; ileus is an obstruction of the digestive tract. The meconium of a newborn with meconium ileus is thickened and sticky, due to the presence of thickened mucus from the intestinal glands. Meconium ileus causes abdominal swelling and vomiting, and often requires surgery immediately after birth. Presence of meconium ileus is considered highly indicative of CF. Borderline cases may be misdiagnosed, however, and attributed instead to "milk allergy."

Other abdominal symptoms are caused by the inability of the pancreas to supply digestive enzymes to the intestine. During normal digestion, as food passes from the stomach into the small intestine, it is mixed with pancreatic secretions which help to break down the nutrients for absorption. While the intestines themselves also provide some digestive enzymes, the pancreas is the major source of enzymes for the digestion of all types of foods, especially fats and proteins.

In CF, thick mucus blocks the pancreatic duct, which is eventually closed off completely by scar tissue formation, leading to a condition known as pancreatic insufficiency. Without pancreatic enzymes, large amounts of undigested food pass into the large intestine. Bacterial action on this rich food source can cause gas and abdominal swelling. The large amount of fat remaining in the feces makes it bulky, oily, and foul-smelling.

Because nutrients are only poorly digested and absorbed, the person with CF is often ravenously hungry, underweight, and shorter than expected for his age. When CF is not treated for a longer period, a child may develop symptoms of malnutrition, including anemia, bloating, and, paradoxically, appetite loss.

Diabetes becomes increasingly likely as a person with CF ages. Scarring of the pancreas slowly destroys those pancreatic cells which produce insulin, producing type I, or insulin-dependent diabetes.

Gall stones affect approximately 10% of adults with CF. Liver problems are less common, but can be caused by the buildup of fat within the liver. Complications of liver enlargement may include internal hemorrhaging, abdominal fluid (**ascites**), spleen enlargement, and liver failure.

Other gastrointestinal symptoms can include a prolapsed rectum, in which part of the rectal lining protrudes through the anus; intestinal obstruction; and rarely, **intussusception**, in which part of the intestinal tube slips over an adjoining part, cutting off blood supply.

Somewhat less than 10% of people with CF do not have gastrointestinal symptoms. Most of these people do not have the delta-F508 mutation, but rather a different

one, which presumably allows at least some of their CFTR proteins to function normally in the pancreas.

RESPIRATORY TRACT. The respiratory tract includes the nose, the throat, the trachea (or windpipe), the bronchi (which branch off from the trachea within each lung), the smaller bronchioles, and the blind sacs called alveoli, in which gas exchange takes place between air and blood.

Swelling of the sinuses within the nose is common in people with CF. This usually shows up on x-ray, and may aid the diagnosis of CF. However, this swelling, called pansinusitis, rarely causes problems, and does not usually require treatment.

Nasal polyps, or growths, affect about one in five people with CF. These growths are not cancerous, and do not require removal unless they become annoying. While nasal polyps appear in older people without CF, especially those with **allergies**, they are rare in children without CF.

The lungs are the site of the most life-threatening effects of CF. The production of a thick, sticky mucus increases the likelihood of infection, decreases the ability to protect against infection, causes inflammation and swelling, decreases the functional capacity of the lungs, and may lead to emphysema. People with CF will live with chronic populations of bacteria in their lungs, and lung infection is the major cause of death for those with CF.

The bronchioles and bronchi normally produce a thin, clear mucus that traps foreign particles including bacteria and viruses. Tiny hair-like projections on the surface of these passageways slowly sweep the mucus along, out of the lungs and up the trachea to the back of the throat, where it may be swallowed or coughed up. This "mucociliary escalator" is one of the principal defenses against lung infection.

The thickened mucus of CF prevents easy movement out of the lungs, and increases the irritation and inflammation of lung tissue. This inflammation swells the passageways, partially closing them down, further hampering the movement of mucus. A person with CF is likely to **cough** more frequently and more vigorously as the lungs attempt to clean themselves out.

At the same time, infection becomes more likely since the mucus is a rich source of nutrients. **Bronchitis**, bronchiolitis, and **pneumonia** are frequent in CF. The most common infecting organisms are the bacteria *Staphylococcus aureus*, *Haemophilus influenzae*, and *Pseudomonas aeruginosa*. A small percentage of people with CF have infections caused by *Burkholderia cepacia*, a bacterium which is resistant to most current **antibiotics** (*Burkholderia cepacia* was formerly known as

Pseudomonas cepacia.) The fungus *Aspergillus fumigatus* may infect older children and adults.

The body's response to infection is to increase mucus production; white blood cells fighting the infection thicken the mucus even further as they break down and release their cell contents. These white blood cells also provoke more inflammation, continuing the downward spiral that marks untreated CF.

As mucus accumulates, it can plug up the smaller passageways in the lungs, decreasing functional lung volume. Getting enough air can become difficult; tiredness, **shortness of breath**, and intolerance of exercise become more common. Because air passes obstructions more easily during inhalation than during exhalation, over time, air becomes trapped in the smallest chambers of the lungs, the alveoli. As millions of alveoli gradually expand, the chest takes on the enlarged, barrel-shaped appearance typical of emphysema.

For unknown reasons, recurrent respiratory infections lead to "digital clubbing," in which the last joint of the fingers and toes becomes slightly enlarged.

SWEAT GLANDS. The CFTR protein helps to regulate the amount of salt in sweat. People with CF have sweat that is much saltier than normal, and measuring the saltiness of a person's sweat is the most important diagnostic test for CF. Parents may notice that their infants taste salty when they kiss them. Excess salt loss is not usually a problem except during prolonged exercise or heat. While most older children and adults with CF compensate for this extra salt loss by eating more salty foods, infants and young children are in danger of suffering its effects (such as heat prostration), especially during summer. Heat prostration is marked by lethargy, weakness, and loss of appetite, and should be treated as an emergency condition.

FERTILITY. Ninety-eight percent of men with CF are sterile, due to complete obstruction or absence of the vas deferens, the tube carrying sperm out of the testes. While boys and men with CF form normal sperm and have normal levels of sex hormones, sperm are unable to leave the testes, and fertilization is not possible. Most women with CF are fertile, though they often have more trouble getting pregnant than women without CF. In both boys and girls, **puberty** is often delayed, most likely due to the effects of poor **nutrition** or chronic lung infection. Women with good lung health usually have no problems with **pregnancy**, while those with ongoing lung infection often do poorly.

Diagnosis

The decision to test a child for cystic fibrosis may be triggered by concerns about recurring gastrointestinal or

KEY TERMS

Carrier—A person with one copy of a defective gene, who does not have the disease it causes, but can pass along the defective gene to offspring.

CFTR—Cystic fibrosis transmembrane conductance regulator, the protein responsible for regulating chloride movement across cells in some tissues. When a person has two defective copies of the CFTR gene, cystic fibrosis is the result.

Emphysema—A pathological accumulation of air in organs or tissues; term especially applied to the condition when in the lungs.

Mucociliary escalator—The coordinated action of tiny projections on the surfaces of cells lining the respiratory tract, which moves mucus up and out of the lungs.

Mucolytic—An agent that dissolves or destroys mucin, the chief component of mucus.

Pancreatic insufficiency—Reduction or absence of pancreatic secretions into the digestive system due to scarring and blockage of the pancreatic duct.

respiratory symptoms, or salty sweat. A child born with meconium ileus will be tested before leaving the hospital. Families with a history of CF may wish to have all children tested, especially if there is a child who already has the disease. Some hospitals now require routine screening of newborns for CF.

Sweat test

The sweat test is both the easiest and most accurate test for CF. In this test, a small amount of the drug pilocarpine is placed on the skin. A very small electrical current is then applied to the area, which drives the pilocarpine into the skin. The drug stimulates sweating in the treated area. The sweat is absorbed onto a piece of filter paper, and is then analyzed for its salt content. A person with CF will have salt concentrations that are one-and-one-half to two times greater than normal. The test can be done on persons of any age, including newborns, and its results can be determined within an hour. Virtually every person who has CF will test positively on it, and virtually everyone who does not will test negatively.

Genetic testing

The discovery of the CFTR gene in 1989 allowed the development of an accurate genetic test for CF.

Genes from a small blood or tissue sample are analyzed for specific mutations; presence of two copies of the mutated gene confirms the diagnosis of CF in all but a very few cases. However, since there are so many different possible mutations, and since testing for all of them would be too expensive and time-consuming, a negative gene test cannot rule out the possibility of CF.

Couples planning a family may decide to have themselves tested if one or both have a family history of CF. Prenatal **genetic testing** is possible through **amniocentesis**. Many couples who already have one child with CF decide to undergo prenatal screening in subsequent pregnancies, and use the results to determine whether to terminate the pregnancy. Siblings in these families are also usually tested, both to determine if they will develop CF, and to determine if they are carriers, to aid in their own family planning. If the sibling has no symptoms, determining his carrier status is often delayed until his teen years or later, when he is closer to needing the information to make decisions.

Newborn screening

Some states now require screening of newborns for CF, using a test known as the IRT test. This is a blood test which measures the level of immunoreactive trypsinogen, which is generally higher in babies with CF than those without it. This test gives many false positive results immediately after birth, and so requires a second test several weeks later. A second positive result is usually followed by a sweat test.

Treatment

There is no cure for CF. Treatment has advanced considerably in the past several decades, increasing both the life span and the quality of life for most people affected by CF. Early diagnosis is important to prevent malnutrition and infection from weakening the young child. With proper management, many people with CF engage in the full range of school and sports activities.

Nutrition

People with CF usually require high-calorie **diets** and vitamin supplements. Height, weight, and growth of a person with CF are monitored regularly. Most people with CF need to take pancreatic enzymes to supplement or replace the inadequate secretions of the pancreas. Tablets containing pancreatic enzymes are taken with every meal; depending on the size of the tablet and the meal, as many as 20 tablets may be needed. Because of incomplete absorption even with pancreatic enzymes, a person with CF needs to take in about 30% more food

than a person without CF. Low-fat diets are *not* recommended except in special circumstances, since fat is a source of both essential fatty acids and abundant calories.

Some people with CF cannot absorb enough nutrients from the foods they eat, even with specialized diets and enzymes. For these people, tube feeding is an option. Nutrients can be introduced directly into the stomach through a tube inserted either through the nose (a nasogastric tube) or through the abdominal wall (a **gastrostomy** tube). A jejunostomy tube, inserted into the small intestine, is also an option. Tube feeding can provide nutrition at any time, including at night while the person is sleeping, allowing constant intake of high-quality nutrients. The feeding tube may be removed during the day, allowing normal meals to be taken.

Respiratory health

The key to maintaining respiratory health in a person with CF is regular monitoring and early treatment. Lung function tests are done frequently to track changes in functional lung volume and respiratory effort. Sputum samples are analyzed to determine the types of bacteria present in the lungs. Chest x rays are usually taken at least once a year. Lung scans, using a radioactive gas, can show closed off areas not seen on the x ray. Circulation in the lungs may be monitored by injection of a radioactive substance into the bloodstream.

People with CF live with chronic bacterial colonization; that is, their lungs are constantly host to several species of bacteria. Good general health, especially good nutrition, can keep the immune system healthy, which decreases the frequency with which these colonies begin an infection, or attack on the lung tissue. Exercise is another important way to maintain health, and people with CF are encouraged to maintain a program of regular exercise.

In addition, clearing mucus from the lungs helps to prevent infection; and mucus control is an important aspect of CF management. Postural drainage is used to allow gravity to aid the mucociliary escalator. For this technique, the person with CF lies on a tilted surface with head downward, alternately on the stomach, back, or side, depending on the section of lung to be drained. An assistant thumps the rib cage to help loosen the secretions. A device called a "flutter" offers another way to loosen secretions: it consists of a stainless steel ball in a tube. When a person exhales through it, the ball vibrates, sending vibrations back through the air in the lungs. Some special breathing techniques may also help clear the lungs.

Several drugs are available to prevent the airways from becoming clogged with mucus. **Bronchodilators** and theophyllines open up the airways; steroids reduce inflammation; and mucolytics loosen secretions. Acetyl-

cysteine (Mucomyst) has been used as a mucolytic for many years but is not prescribed frequently now, while DNase (Pulmozyme) is a newer product gaining in popularity. DNase breaks down the DNA from dead white blood cells and bacteria found in thick mucus.

People with CF may pick up bacteria from other CF patients. This is especially true of *Burkholderia cepacia*, which is not usually found in people without CF. While the ideal recommendation from a health standpoint might be to avoid contact with others who have CF, this is not usually practical (since CF clinics are a major site of care), nor does it meet the psychological and social needs of many people with CF. At a minimum, CF centers recommend avoiding prolonged close contact between people with CF, and scrupulous hygiene, including frequent hand washing. Some CF clinics schedule appointments on different days for those with and without *B. cepacia* colonies.

Some doctors choose to prescribe antibiotics only during infection, while others prefer long-term antibiotic treatment against *S. aureus*. The choice of antibiotic depends on the particular organism or organisms found. Some antibiotics are given as aerosols directly into the lungs. Antibiotic treatment may be prolonged and aggressive.

Supplemental oxygen may be needed as lung disease progresses. **Respiratory failure** may develop, requiring temporary use of a ventilator to perform the work of breathing.

Lung transplantation has become increasingly common for people with CF, although the number of people who receive them is still much lower than those who want them. Transplantation is not a cure, however, and has been likened to trading one disease for another. Long-term immunosuppression is required, increasing the likelihood of other types of infection. About 50% of adults and more than 80% of children who receive lung transplants live longer than two years. Liver transplants are also done for CF patients whose livers have been damaged by fibrosis.

Long-term use of ibuprofen has been shown to help some people with CF, presumably by reducing inflammation in the lungs. Close medical supervision is necessary, however, since the effective dose is high and not everyone benefits. Ibuprofen at the required doses interferes with kidney function, and together with aminoglycoside antibiotics, may cause kidney failure.

A number of experimental treatments are currently the subject of much research. Some evidence indicates that aminoglycoside antibiotics may help overcome the genetic defect in some CF mutations, allowing the protein to be made normally. While promising, these results would apply to only about 5% of those with CF.

Gene therapy is currently the most ambitious approach to curing CF. In this set of techniques, non-defective copies of the CFTR gene are delivered to affected cells, where they are taken up and used to create the CFTR protein. While elegant and simple in theory, gene therapy has met with a large number of difficulties in trials so far, including immune resistance, very short duration of the introduced gene, and inadequately widespread delivery.

Alternative treatment

In homeopathic medicine, the symptoms of the disease would be addressed to enhance the quality of life for the person with cystic fibrosis. Treating the cause of CF, because of the genetic basis for the disease, is not possible. Homeopathic medicine seeks to treat the whole person, however, and in CF, this approach might include:

- mucolytics to help thin mucus
- supplementation of pancreatic enzymes to assist in digestion
- respiratory symptoms can be addressed to open lung passages
- hydrotherapy techniques to help ease the respiratory symptoms and help the body eliminate
- immune enhancements can help prevent the development of secondary infections
- dietary enhancements and adjustments are used to treat digestive and nutritional problems

Prognosis

People with CF may lead relatively normal lives with the control of symptoms. The possible effect of pregnancy on the health of a woman with CF requires careful consideration before beginning a family as do issues of longevity and their children's status as carriers. Although most men with CF are functionally sterile, new procedures for removing sperm from the testes are being tried, and may offer more men the chance to become fathers.

Approximately half of people with CF live past the age of 30. Because of better and earlier treatment, a person born today with CF is expected, on average, to live to age 40.

Prevention

Adults with a family history of cystic fibrosis may obtain a genetic test of their carrier status for purposes of family planning. Prenatal testing is also available. There is currently no known way to prevent development of CF in a person with two defective gene copies.

Resources

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ORGANIZATIONS

Cystic Fibrosis Foundation. 6931 Arlington Road, Bethesda, MD 20814. (800) 344-4823. <<http://www.cff.org>>.

OTHER

CysticFibrosis.com. <<http://www.cysticfibrosis.com>>.

Richard Robinson

Cystinuria

Definition

Cystinuria is an inborn error of amino acid transport that results in the defective absorption by the kidneys of the amino acid called cystine. The name means “cystine in the urine.”

Description

Cystine is an amino acid. Amino acids are organic compounds needed by the body to make proteins and for many normal functions. When the kidneys don't absorb cystine, this compound builds up in the urine. When the amount of cystine in the urine exceeds its solubility (the greatest amount that can be dissolved), crystals form. As the amount of cystine continues to increase in the urine, the number of crystals also increases. When very large numbers of cystine crystals form, they clump together into what is called a stone.

Causes and symptoms

Cystinuria is a rare disease that occurs when people inherit an abnormal gene from their parents. This disease occurs in differing degrees of severity in people who have inherited either one or two abnormal genes. Humans have two copies of each gene. When both are abnormal, the condition is called homozygous for the disease. When one copy is normal and the other is abnormal, the condition is called heterozygous for the disease. Persons with one abnormal gene can have a milder form of cystinuria that rarely results in the formation of stones.

Severe cystinuria occurs when people are homozygous for the disease. For these individuals, the kidneys may excrete as much as 30 times the normal amount of

cystine. Research has shown that this condition is caused by mutations on chromosome number two (humans have 23 pairs of chromosomes).

A person who has inherited cystinuria may have other abnormal bodily functions. In addition to excess levels of the amino acid cystine, high amounts of the amino acids lysine, arginine, and ornithine are found in the urine. This condition indicates that these amino acids are not being reabsorbed by the body.

When excess cystine crystals clump together to form a stone, the stone can block portions of the interior of the kidney or the tube (the ureter) that connects the kidney to the urinary bladder. These cystine stones can be painful, and depending upon where the stone becomes trapped, the **pain** can be felt in the lower back or the abdomen. **Nausea and vomiting** can also occur, and patients may sometimes feel the need to urinate often. Cystine stones can also cause blood in the urine. When the urinary tract is blocked by a stone, urinary tract infections or kidney failure may result.

Diagnosis

Small stones (called “silent”) often do not cause any symptoms, although they can be detected by an x ray. Large stones are often painful and easily noticed by the patient. Blood in the urine can also mean that a stone has formed.

When the urine contains extremely high amounts of cystine, yellow-brown hexagonal crystals are visible when a sample is examined under the microscope. Urine samples can also be mixed with chemicals that change color when high levels of cystine are present. When the compound nitroprusside is added to urine that has been made alkaline by the addition of ammonia, the urine specimen turns red if it contains excess cystine.

Treatment

No treatment can decrease cystine excretion. The best treatment for cystinuria is to prevent stones from forming. Stones can be prevented by drinking enough liquid each day (about 5–7 qts) to produce at least 8 pts of urine, thus keeping the concentration of cystine in the urine low. Because a person doesn't drink throughout the night, less urine is produced, and the likelihood of stone formation increases. This risk can be minimized by drinking water or other liquids just before going to bed.

Drug treatments

In addition to drinking large amounts of fluids, it is helpful to make the urine more alkaline. Cystine dis-

solves more easily in alkaline urine. To increase urine alkalinity, a person may take sodium bicarbonate and acetazolamide. Penicillamine, a drug that increases the solubility of cystine, may be prescribed for patients who do not respond well to other therapies. This drug must be used with caution, however, because it can cause serious side effects or allergic reactions. For those unable to take penicillamine, another drug, alpha-mercaptopropionyl-glycine (Thiola), may be prescribed.

Surgical treatments

Most stones can be removed from the body by normal urination, helped by drinking large amounts of water. Large stones that cannot be passed this way must be removed by surgical procedures.

Large stones can be surgically removed by having a device called a ureteroscope placed into the urethra, up through the bladder and into the ureter, where the trapped stone can be seen and removed. Another method involves using sound-wave energy aimed from outside the body to break the large stone into small pieces that can be passed by urination. This external technique is called extracorporeal shock-wave lithotripsy (ESWL).

For large stones in the kidney, a procedure called percutaneous nephrolithotomy may be used. In this procedure, the surgeon makes a small incision in the back over the kidney. An instrument called a nephroscope is inserted through the incision into the kidney. The surgeon uses the nephroscope to locate and remove the stone. If the stone is very large, it may be broken up into smaller pieces by an ultrasonic or other kind of probe before removal.

Prognosis

As many as 50% of patients who have had surgical treatment for a kidney stone will have another stone within five years if no medicines are used to treat this condition.

Prevention

Cystinuria is a genetic disorder that currently cannot be prevented.

Resources

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KEY TERMS

Alkaline—A solution is considered alkaline if it contains fewer hydrogen atoms than pure water.

Amino acid—An organic compound made of an amino group (containing nitrogen and hydrogen) and a carboxylic acid group. Amino acids are an essential part of protein molecules.

Nephroscope—An instrument made of a light source in a tube. The tube is inserted into the kidney through an incision in the back and used to locate kidney stones. The stones are broken up with high frequency sound waves and removed by suction through the scope.

Nitroprusside—A compound that is used in laboratory tests to identify large amounts of cystine in urine samples.

Uretoscope—A tube-shaped device inserted into the body through the urinary system that allows objects to be both seen and grasped for removal.

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ORGANIZATIONS

- Cystinuria Support Network. 21001 NE 36th St., Redmond, WA 98053. (425) 868-2996. <<http://www.cystinuria.com>>. National Kidney Foundation. 30 East 33rd St., New York, NY 10016. (800) 622-9010. <<http://www.kidney.org>>.

Dominic De Bellis, PhD

Cystitis

Definition

Cystitis is defined as inflammation of the urinary bladder. **Urethritis** is an inflammation of the urethra, which is the passageway that connects the bladder with the exterior of the body. Sometimes cystitis and urethritis are referred to collectively as a lower urinary tract infection, or UTI. Infection of the upper urinary tract involves the spread of bacteria to the kidney and is called **pyelonephritis**.

Description

The frequency of bladder infections in humans varies significantly according to age and sex. The male/female