

*The GALE*  
ENCYCLOPEDIA  
*of MEDICINE*

SECOND EDITION

# *The GALE* ENCYCLOPEDIA *of MEDICINE* SECOND EDITION

VOLUME

2

C-F

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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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### ORGANIZATION

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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- "When Microbes are on the Menu." *Harvard Health Letter* (Dec. 1994) : 4-5.

## 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>>.

David Kaminstein, MD

## 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
<b>Viruses</b>	
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
<b>Bacteria</b>	
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.

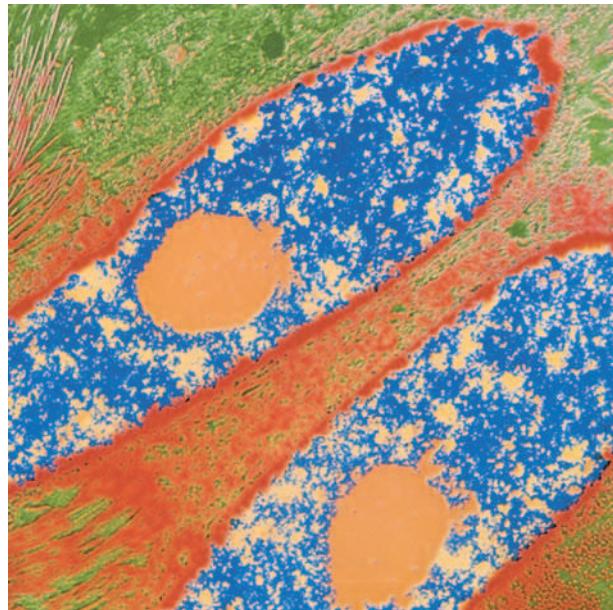
Certain drugs, which are currently being used for treatment, could also be suitable for prevention. For example, the drug tamoxifen (Nolvadex), which has been very effective against breast cancer, is currently being tested by the National Cancer Institute for its ability to prevent cancer. Similarly, 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.

## Resources

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### ORGANIZATIONS

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

Cancer Research Institute (National Headquarters). 681 Fifth Avenue, New York, NY 10022. (800) 992-2623. <<http://www.cancerresearch.org>>.

National Cancer Institute. 9000 Rockville Pike, Building 31, room 10A16, Bethesda, Maryland, 20892. (800) 422-6237. <<http://www.cancer.gov>>.

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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- American Cancer Society. <<http://www.cancer.org>>. National Cancer Institute. <<http://cancerinfo.nci.nih.gov/cancerlit.shtml>>.

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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.

### Resources

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### ORGANIZATIONS

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American Cancer Society. <<http://www.cancer.org>>.

American Pain Society. <<http://www.ampsoc.org>>.

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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.

### **Resources**

#### **PERIODICALS**

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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*

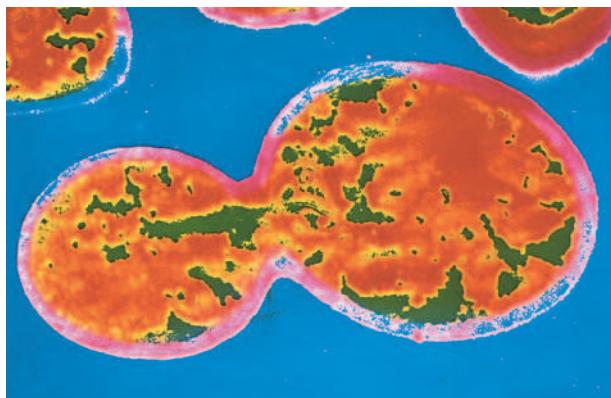
This is usually treated with prescription lozenges or mouthwashes. Some of the most-used prescriptions are nystatin mouthwashes (Nilstat or Nitrostat) and clotrimazole lozenges.

#### *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

### BOOKS

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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

#### BOOKS

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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

### BOOKS

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*National Institute of Diabetes and Digestive and Kidney Disease*. <<http://www.niddk.nih.gov>>.

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# 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

Cooper, Dennis L. "Tumor Markers." In *Cecil Textbook of Medicine*, ed. J. Claude Bennet and Fred Plum. Philadelphia: W. B. Saunders Co., 1996.

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

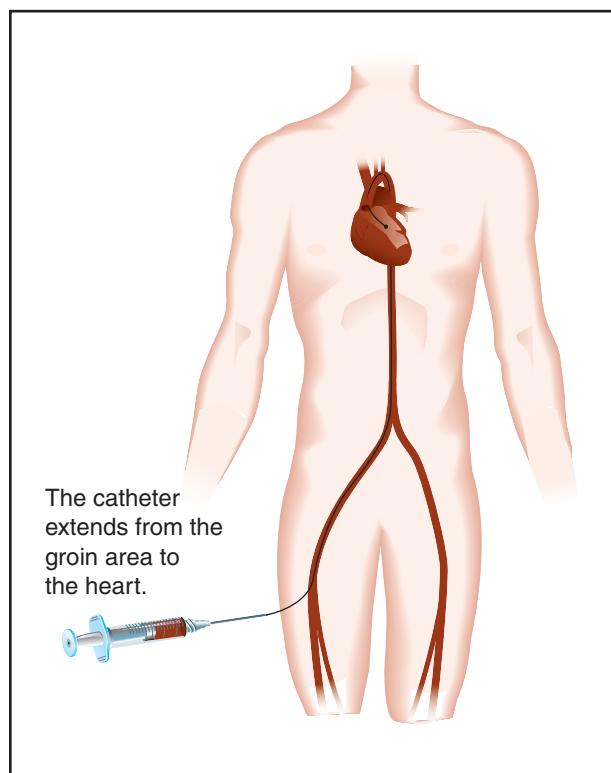
While cardiac catheterization may be performed on an outpatient basis, a patient may require close monitoring following the procedure while remaining in the hospital for at least 24 hours. The patient will be instructed to rest in bed for at least eight hours immediately after the test. If the catheter 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

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- 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**



## 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.

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.)

### 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.

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#### PERIODICALS

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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.

### Resources

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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-

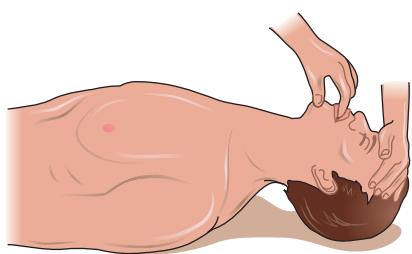


Figure A



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.

## Resources

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### ORGANIZATIONS

- American College of Emergency Physicians. P.O. Box 619911, Dallas, TX 75261-9911. (800) 798-1822 or (972) 550-0911. Fax: (972) 580-2816. <[info@acep.org](mailto:info@acep.org)>. <<http://www.acep.org>>.
- 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](http://www.americanheart.org/Heart_and_Stroke_A_Z_Guide/heim.html)>.
- Heimlich Institute. PO Box 8858, Cincinnati, OH 45208. <[heimlich@iglou.com](mailto: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](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](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**.

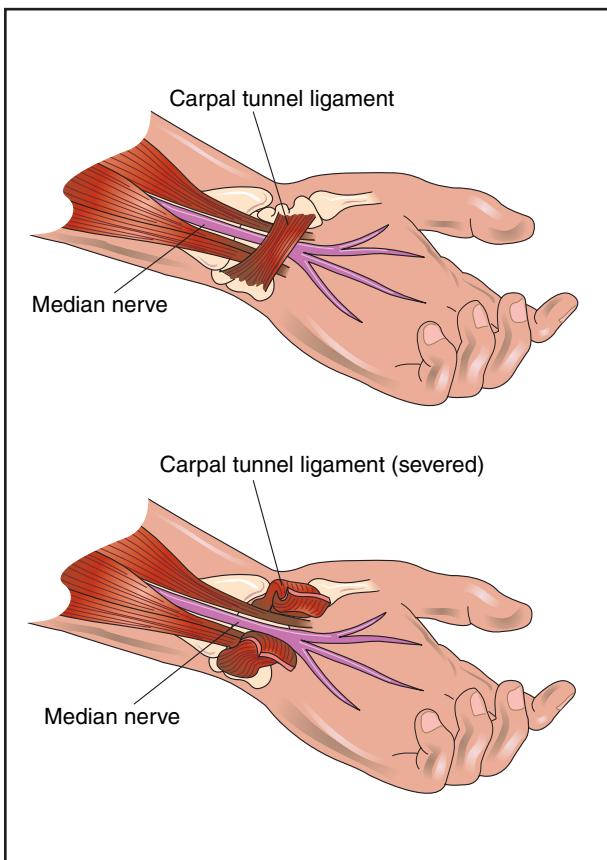
### Description

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.

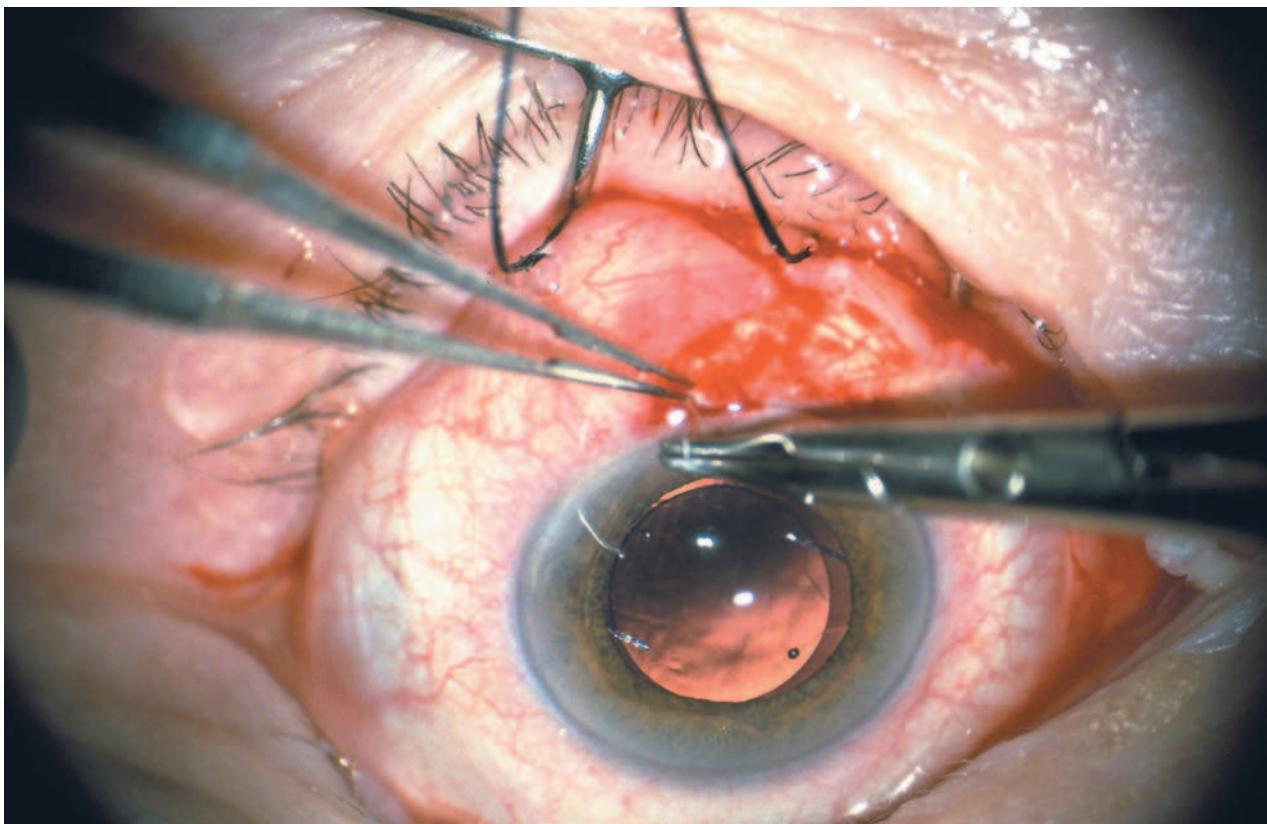
Extracapsular cataract surgery is the removal of the lens where the capsule is left in place. Each year in the United States, over a million cataracts are removed this way.

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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## 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.

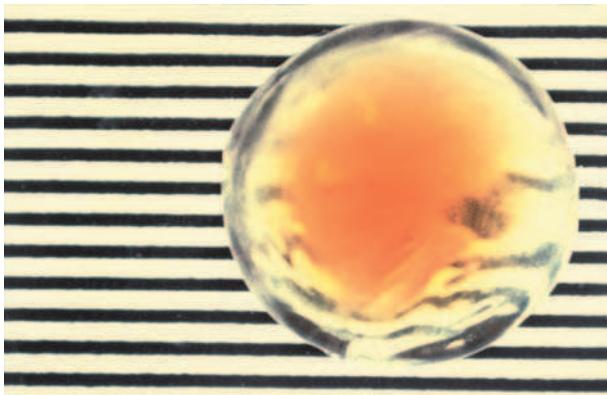
Cataracts in the elderly are so common that they are thought to be a normal part of the aging process. Between the ages of 52 and 64, there is a 50% chance of having a cataract, while at least 70% of those 70 and older are affected. Cataracts associated with aging (senile or age-related cataracts) most 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

### BOOKS

"Cataract." In *Medical-Surgical Nursing: Concepts and Clinical Practice*. 5th ed. Ed. Wilma J. Phipps, et al. St. Louis: Mosby-Year Book, Inc., 1995.

## 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>>.

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## 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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Frisch, Noreen Cavan, and Lawrence E. Frisch. *Psychiatric Mental Health Nursing*. Albany, NY: Delmar Publishers, 1998.

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).

### Resources

#### BOOKS

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Jacobs, David S., et al. *Laboratory Test Handbook*. 4th ed. New York: Lexi-Comp Inc., 1996.

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

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## 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

#### BOOKS

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"Radiofrequency Catheter Ablation for Treatment of Cardiac Arrhythmias." *Medical Letter on Drugs and Therapeutics* 38 (26 Mar. 1996): 409.

#### ORGANIZATIONS

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.

### Resources

#### BOOKS

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#### PERIODICALS

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Pruessner, H. "Detecting Celiac Disease in Your Patients." *American Family Physician* 57 (March 1998): 1023–34.

## 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>>.

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# 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

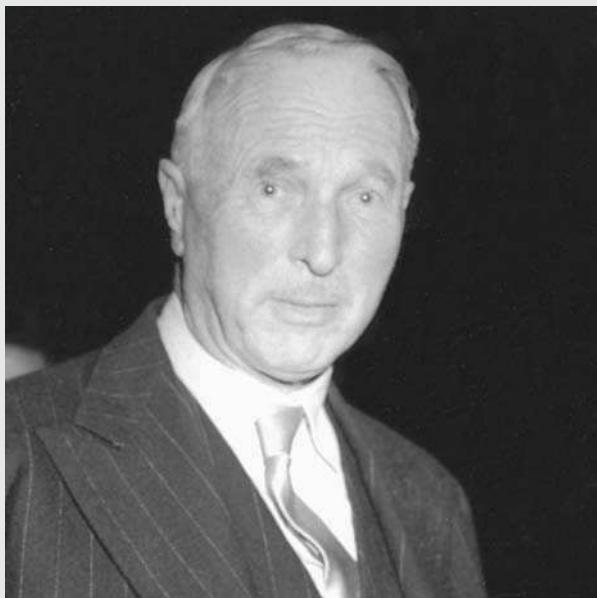
The theory behind cell therapy has been in existence for several hundred years. The first recorded discussion of the concept of cell therapy can be traced to Phillipus Aureolus Paracelsus (1493–1541), a German-Swiss physician and alchemist who wrote in his *Der grossen Wundartzney* ("Great Surgery Book") 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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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.

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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.

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Children and Adults with Attention Deficit Disorders  
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### OTHER

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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

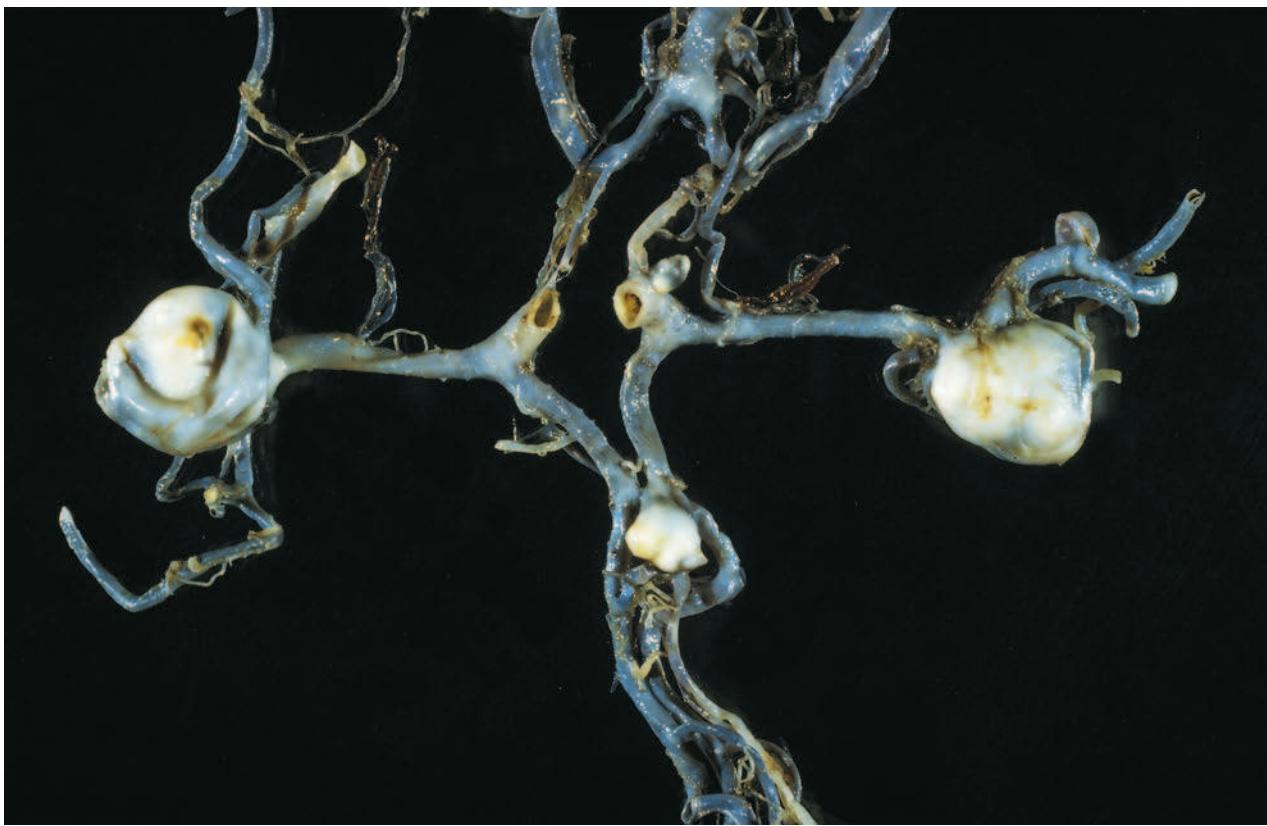
A cerebral aneurysm can occur anywhere in the brain. Aneurysms can have several shapes. The saccular aneurysm, once called a berry aneurysm, resembles a piece of fruit dangling from a branch. Saccular aneurysms are usually found at a branch in the blood vessel where they balloon out by a thin neck. Saccular 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 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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- Stephenson, Joan. "Cerebral Palsy Clues." *The Journal of the American Medical Association* 280 (21 October 1998): 1298.

### ORGANIZATIONS

- 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](mailto: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>>.

## OTHER

"Cerebral Palsy Information Page." *National Institute of Neurological Disorders and Stroke*. <[http://www.ninds.nih.gov/health\\_and\\_medical/pubs/cerebral\\_palsy.htm](http://www.ninds.nih.gov/health_and_medical/pubs/cerebral_palsy.htm)>.

"Cerebral Palsy: Hope Through Research." *National Institute of Neurological Disorders and Stroke*. <[http://www.ninds.nih.gov/health\\_and\\_medical/pubs/cerebral\\_palsytr.htm](http://www.ninds.nih.gov/health_and_medical/pubs/cerebral_palsytr.htm)>.

Scott J. Polzin, MS

## Cerebrospinal fluid (CSF) analysis

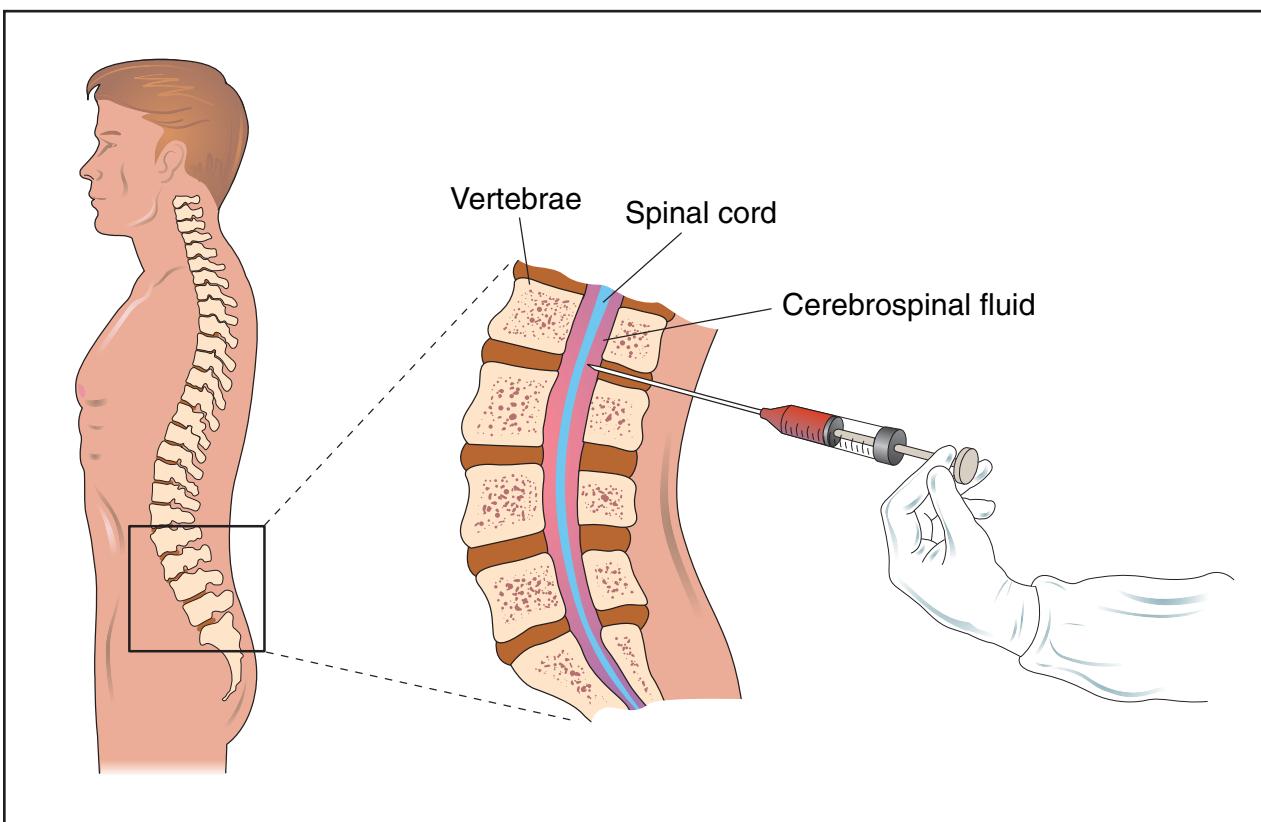
### Definition

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 mmH<sub>2</sub>O
- 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.

## Resources

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## ORGANIZATIONS

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.

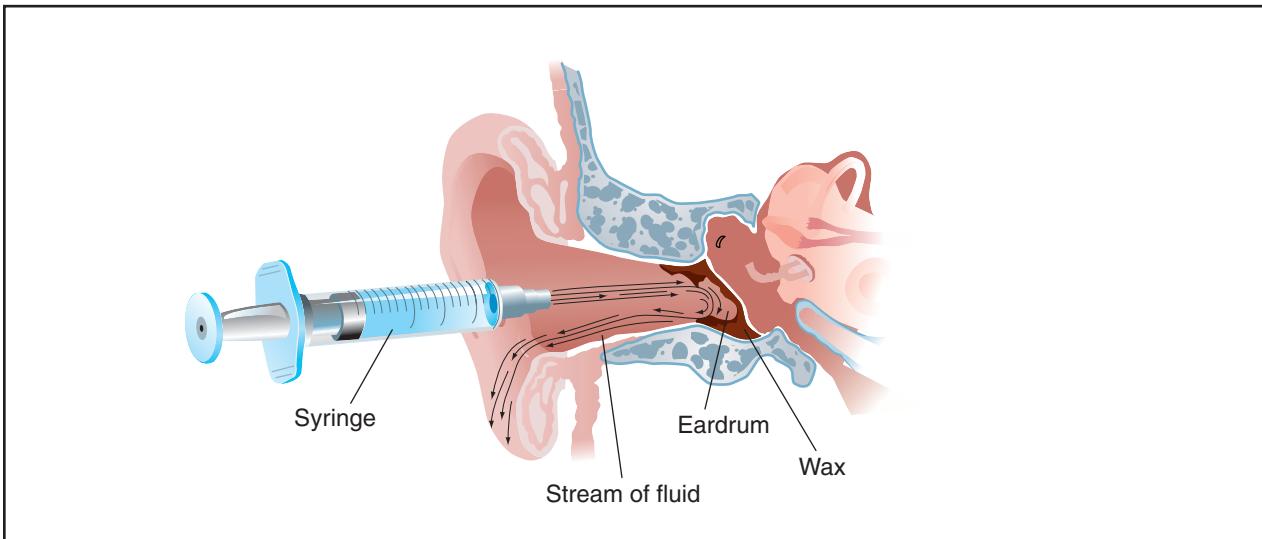
### Causes and symptoms

#### 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

### BOOKS

Jackler, Robert K., MD, and Michael J. Kaplan, MD. "Cerumen Impaction." In "Ear, Nose, & Throat," *Current Medical Diagnosis and Treatment*, 2005, pp. 111–112.

## 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.

### **Resources**

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Runowicz, Carolyn D., Jeanne A. Petrek, and Ted S. Gansler. *Women and Cancer: A Thorough and Compassionate Resource for Patients and their Families*. New York: Villard Books, 1999.

Sweeney, Julia. *God Said "Ha!"* New York: Bantam Books, 1997.

#### **ORGANIZATIONS**

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

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/>>.

#### OTHER

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Lata Cherath  
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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.

### Resources

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#### ORGANIZATIONS

Cancer Information Service. (800) 4-CANCER. <<http://www.rex.nci.nih.gov>>.

Carol A. Turkington

## 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**.

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American Academy of Orthopaedic Surgeons. 6300 North River Road, Rosemont, IL 60018-4262. (800) 346-2267. <<http://www.aaos.org>>.

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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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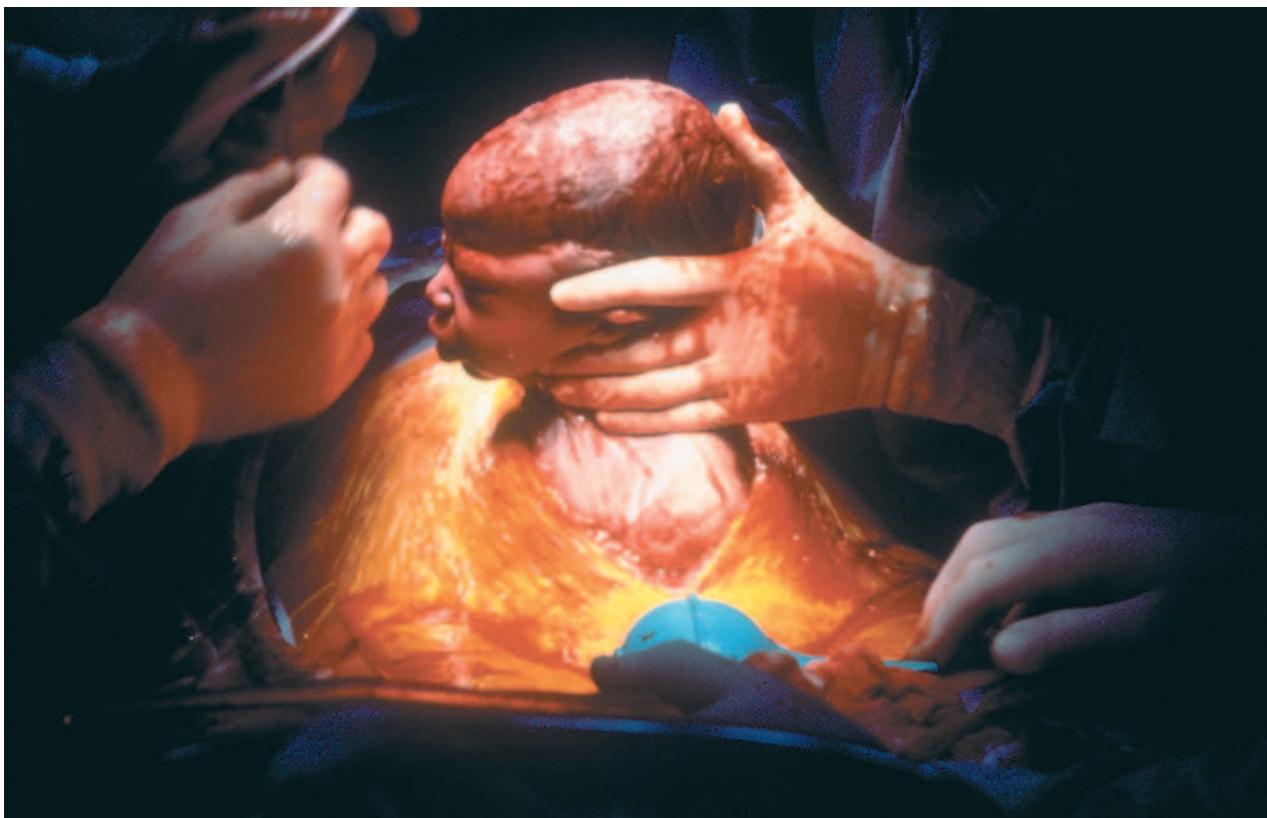
- American College of Obstetricians and Gynecologists. 409 12th Street, SW P.O. Box 96920, Washington, DC 20090-6920. (202) 863-2518. <<http://www.acog.org>>.

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## 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>>.
- United States Department of Health and Human Services. 200 Independence Avenue SW, Washington DC 20201. (202) 619-0257. <<http://www.hhs.gov>>.

### OTHER

- "Cesarean Childbirth." *Perspectives: A Mental Health Magazine*. <<http://mentalhelp.net/perspectives>>.
- "The Cesarean Section FAQ." *Childbirth.org*. 18 July 1998 <<http://www.childbirth.org/section/CSFAQ.html>>.

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

#### BOOKS

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*Centers for Disease Control.* <<http://www.cdc.gov/nccdphe/ddt/ddthome.htm>>.

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.

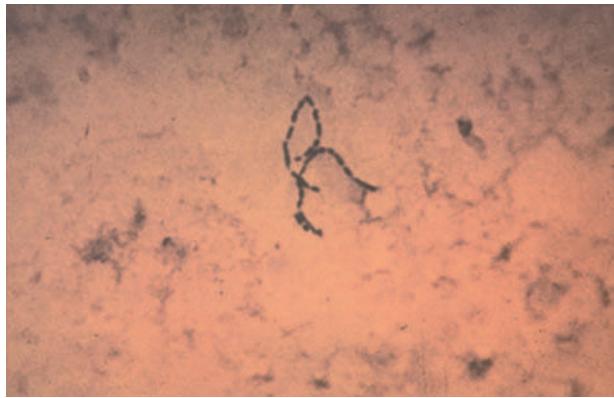
Common locations for chancroid sores (ulcers) in men are the shaft or head of the penis, foreskin, the groove behind the head of the penis, the opening of the penis, and the scrotum. In women, common locations are the labia majora (outer lips), labia minora (inner lips), perianal area (area around the 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.

### Resources

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#### PERIODICALS

DiCarlo, Richard P., and David H. Martin. "The Clinical Diagnosis of Genital Ulcer Disease in Men." *Clinical Infectious Diseases* 25 (Aug. 1997): 292-8.

#### ORGANIZATIONS

Planned Parenthood Federation of America, Inc. 810 Seventh Ave., New York, NY, 10019. (800) 669-0156. <<http://www.plannedparenthood.org>>.

#### 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.

#### **Resources**

##### **BOOKS**

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Mai Tran

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## **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, B<sub>6</sub>, 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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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

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- “GeneClinics.” <[www.geneclinics.org](http://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.

### 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.

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Judith Turner

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.

## Resources

### BOOKS

Dollinger, Malin, et al. *Everyone's Guide to Cancer Therapy: How Cancer is Diagnosed, Treated, and Managed Day to Day*. 3rd ed. Kansas City: Andres & McMeel, 1998.

Drum, David. *Making the Chemotherapy Decision*. Los Angeles: Lowell House, 1996.

McKay, Judith, and Nancee Hirano. *The Chemotherapy Survival Guide*. Oakland, CA: New Harbinger Publications, 1993.

### ORGANIZATIONS

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

### OTHER

"Introduction to Chemotherapy." *OncoLink*. University of Pennsylvania Cancer Center. 1998 <<http://www.oncologink.upenn.edu>>.

"What is Chemotherapy?" *OncoLink*. University of Pennsylvania Cancer Center. 1998 <<http://www.oncologink.upenn.edu>>.

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.

### Resources

#### BOOKS

- “Chest Drainage Therapy.” In *Everything You Need to Know About Medical Treatments*. Springhouse, PA: Springhouse Corp., 1996.
- Current Medical Diagnosis and Treatment*, 1998. 37th ed. Ed. Stephen McPhee, et al. Stamford: Appleton & Lange, 1997.

"Thoracostomy Tube Drainage." In *The Merck Manual of Diagnosis and Therapy*. 16th ed. Ed. Robert Berkow. Rahway, NJ: Merck Research Laboratories, 1992.

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

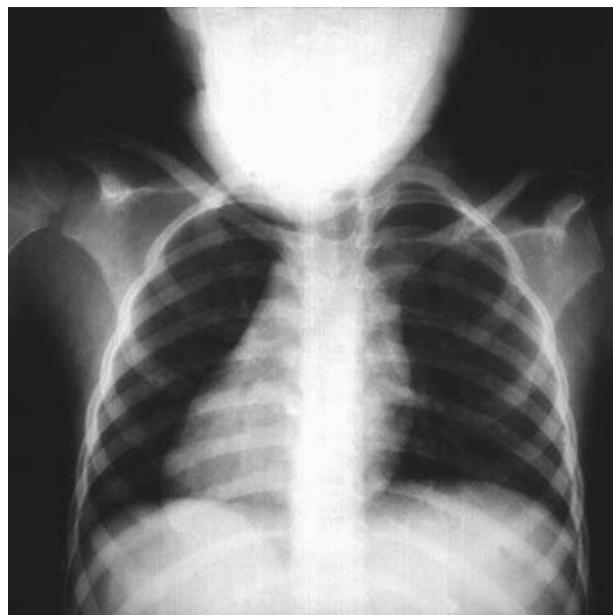
"AARC Clinical Practical Guideline: Postural Drainage." *Respiratory Care* 36 (1991): 1418-1426.

#### 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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 Shapiro, Eugene D., and Phillip S. LaRussa. "Vaccination for Varicella—Just Do It!" *Journal of the American Medical Association* 278 (1997): 1529-1530.

### 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—shrink 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. Anyone 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.

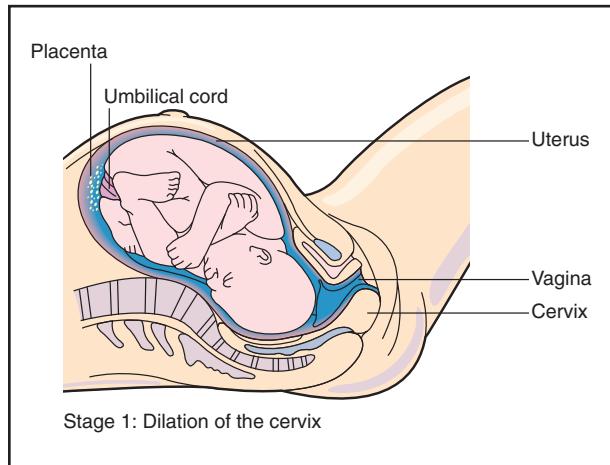
As labor begins, the muscular wall of the uterus begins to contract as the cervix relaxes and expands. As a portion of the amniotic sac surrounding the baby is pushed into the opening, it bursts under the pressure, releasing amniotic fluid. This is called “breaking the bag of waters.”

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.

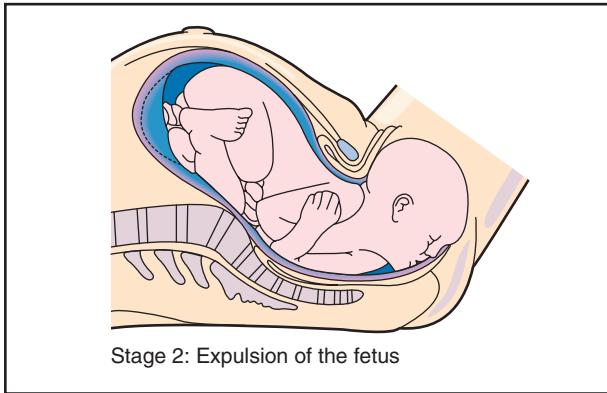
### *Second stage of labor*

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

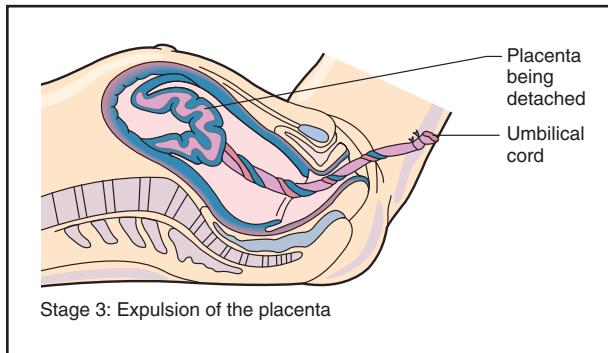
A cesarean section, also called a c-section, is a surgical procedure in which incisions are made through a woman's abdomen and uterus to deliver her baby.

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**.

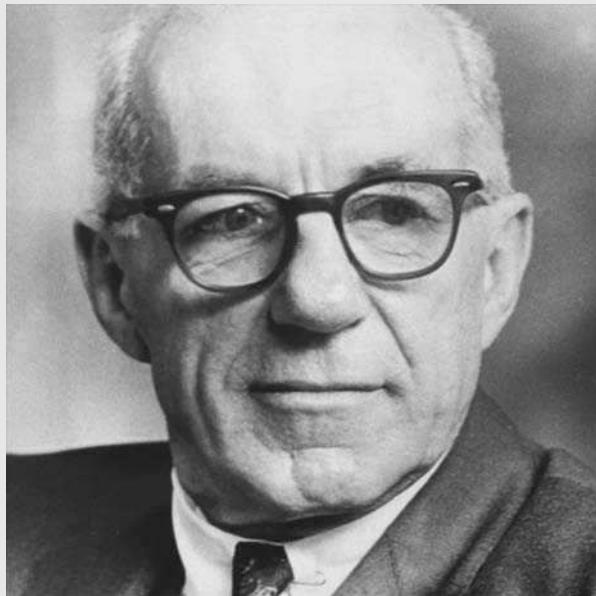
### *Mental health*

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

### BOOKS

- Holtzman, Debra Smiley. *The Panic Proof Parent*. Chicago: NTC/Contemporary, 2000.  
 Pasquariello Jr., Patrick S. ed. *The Children's Hospital of Philadelphia: Book of Pregnancy and Child Care*. New York: John Wiley & Sons, Inc., 1999.  
 White, Linda, and Sunny Mavor. *Kids, Herbs, and Health*. Loveland, CO: Interweave Press, 1998.

### 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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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

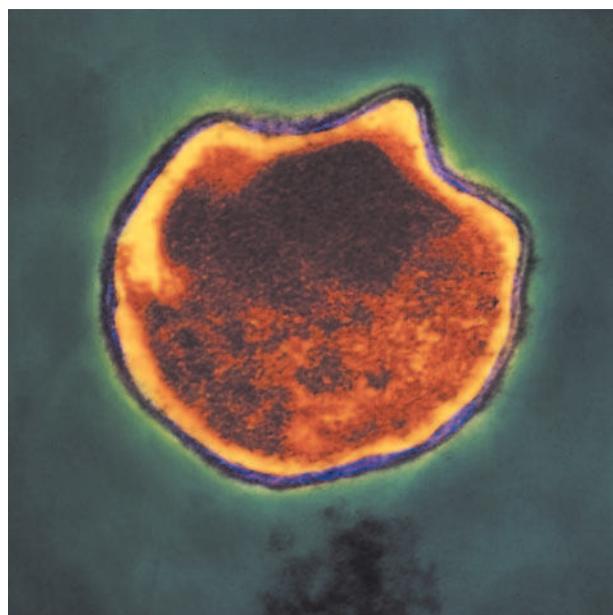
*Chlamydia trachomatis* is a major cause of **sexually transmitted diseases** (called **nongonococcal urethritis** and **pelvic inflammatory disease**). When a woman with an active chlamydial infection gives birth to a baby, the baby may aspirate (suck into his or her lungs) some of the mother's bacteria-laden 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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### ORGANIZATIONS

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

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.

**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.

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

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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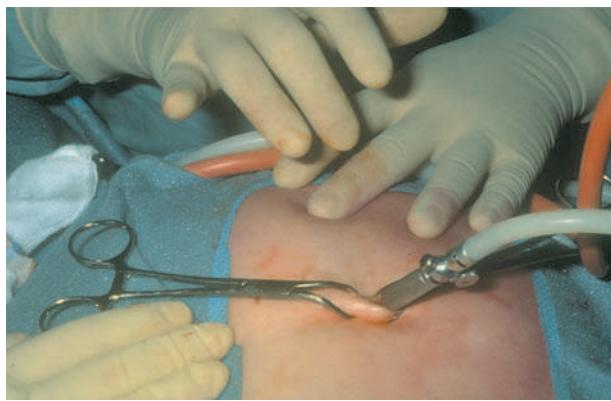
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David Kaminstein, MD



**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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Kathleen D. Wright, RN

## 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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Digestive Disease National Coalition. 507 Capitol Court NE, Suite 200, Washington, DC 20003. (202) 544-7497. <<http://www.ddnc.org>>.  
 National Digestive Diseases Information Clearinghouse. 2 Information Way, Bethesda, MD 20892-3570. (800) 891-5389. <<http://www.niddk.nih.gov/health/digest/nddic.htm>>.

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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.

## Resources

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### PERIODICALS

Pasha T. M., and K. D. Lindor. "Diagnosis and Therapy of Cholestatic Liver Disease." *Medical Clinics of North America* 80 (Sept. 1996): 995-1019.

### ORGANIZATIONS

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>>.

### OTHER

"Hepatic and Biliary Disorders." *The Merck Manual*. 13 Apr. 1998 <<http://www.merck.com>>.

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
<b>Total cholesterol:</b>	
Desirable	<200
Borderline	200 to 240
Undesirable	>240
<b>HDL cholesterol:</b>	
Desirable	>45
Borderline	35 to 45
Undesirable	<35
<b>LDL cholesterol:</b>	
Desirable	<130
Borderline	130 to 160
Undesirable	>160
<b>Ratio of total cholesterol to HDL cholesterol:</b>	
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 B<sub>3</sub>, 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.

### **Resources**

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- Trubo, Richard. *Cholesterol Cures: From Almonds and Antioxidants to Garlic, Golf, Wine and Yogurt*. Emmaus, PA: Rodale Press, 1996.

#### **PERIODICALS**

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- Marandino, Cristin. "The Case for Cholesterol." *Vegetarian Times* (August 1999): 10.
- Schmitt, B.D. "Treating High Cholesterol Levels." *Clinical Reference Systems* (July 1, 1999): 1551.
- VanTyne, Julia, and Davis, Lori. "Drop Your Cholesterol 25 to 100 Points." *Prevention* (November 1999): 110.

#### **ORGANIZATIONS**

- National Cholesterol Education Program. NHLBI Information Center, P.O. Box 30105, Bethesda, MD 20824-0105.  
<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

### BOOKS

Nesto, R. W., and L. Christensen. *Cholesterol-Lowering Drugs: Everything You and Your Family Need to Know*. New York: Morrow, William & Co, 2000.

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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**.

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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

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"Congenital myasthenic syndromes: recent advances."  
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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.

### Resources

#### OTHER

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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.

## Resources

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Anna Rovid Spickler, D.V.M., Ph.D.

Chorionic gonadotropin test see **Human chorionic gonadotropin pregnancy test**

## Chorionic villus sampling

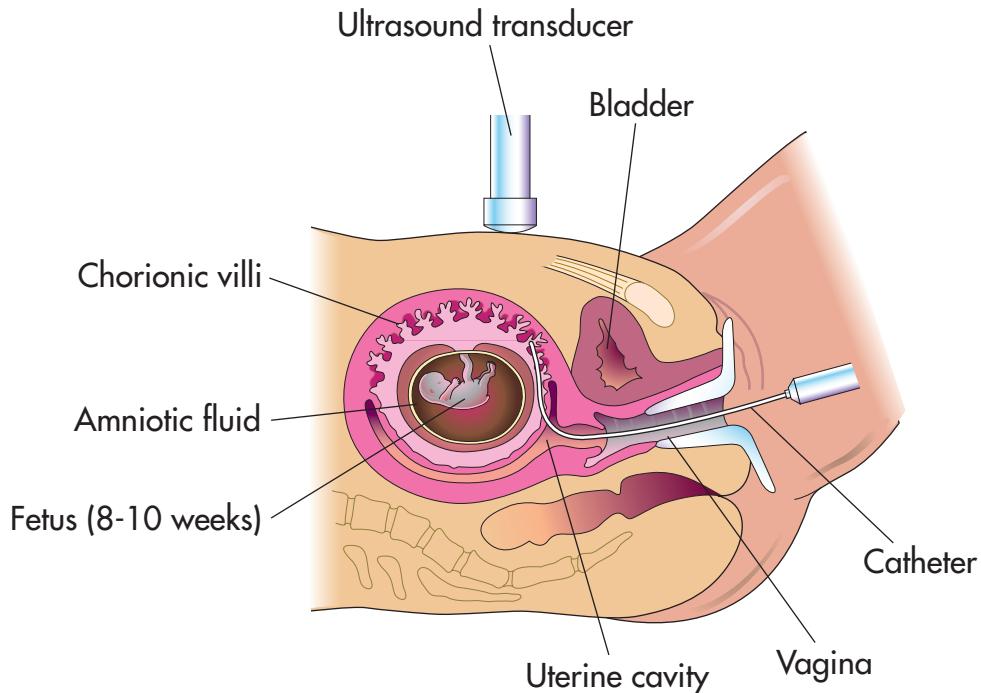
### Definition

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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### OTHER

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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 B<sub>12</sub>, 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.

### Resources

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#### ORGANIZATIONS

- American Association for Chronic Fatigue Syndrome. 7 Van Buren St., Albany, NY 12206. (518) 435-1765. <<http://weber.u.washington.edu/~dedra/aacfsl.html>>.
- 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>>.

#### OTHER

- "The Facts about Chronic Fatigue Syndrome." *Centers for Disease Control*. <<http://www.cdc.gov/ncidod/diseases/cfs/facts1.htm>>.

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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.

## Resources

### BOOKS

- Abbas, Abul K., Andrew H. Lichtman, and Jordan S. Pober. *Cellular and Molecular Immunology*. 3rd ed. Philadelphia. W. B. Saunders Co., 1997.  
*Professional Guide to Diseases*. 5th ed. Springhouse, PA: Springhouse Corporation, 1995.

### 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.

## **Resources**

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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.

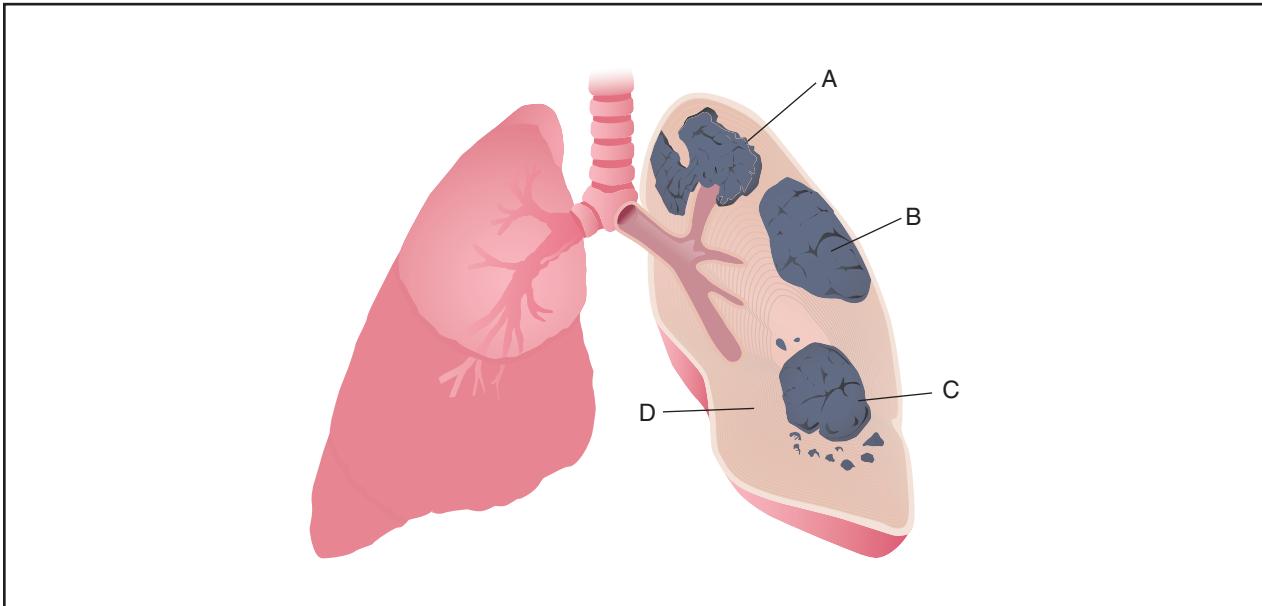
### Causes and symptoms

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: Beta<sub>2</sub> 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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 Cordova, Francis C., and Gerard J. Griner. "Management of Advanced Chronic Obstructive Pulmonary Disease." *Comprehensive Therapy* 23, no. 6: 413-424.  
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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.

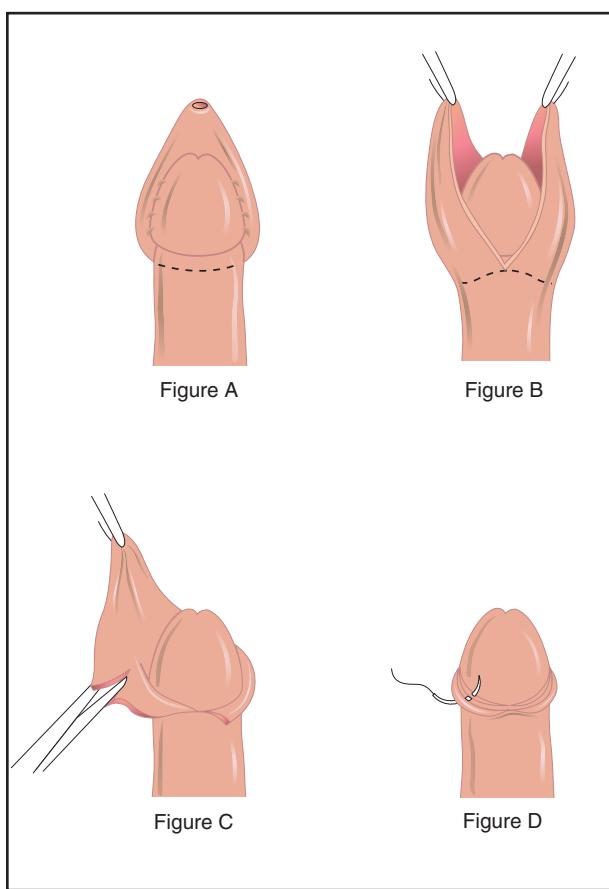
### Purpose

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.

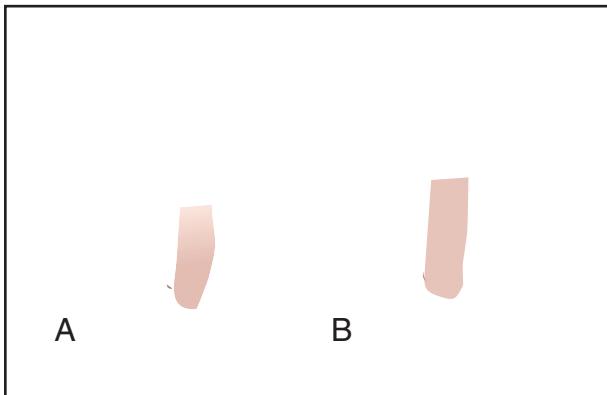
## Precautions

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.

### Resources

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#### OTHER

American Academy of Pediatrics. *New AAP Circumcision Policy Released (Press Release)*. 1 Mar. 1999. <<http://www.aap.org/advocacy/archives/marcircum.htm>>.

Janie F. Franz

## Cirrhosis

### Definition

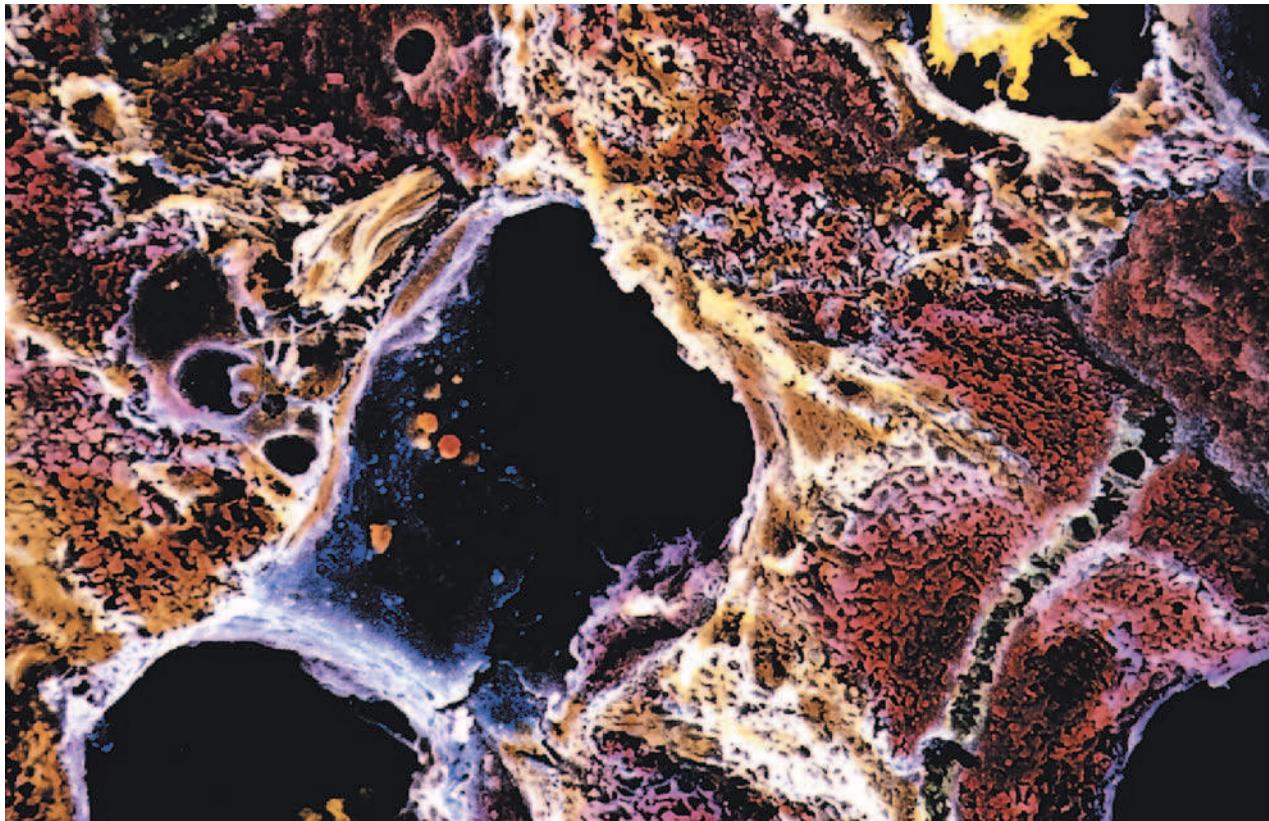
Cirrhosis is a chronic, degenerative disease in which normal liver cells are damaged and are then replaced by scar tissue.

### 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 ( $\alpha_1$ -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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The Editors of Time-Life Books. *The Medical Advisor: The Complete Guide to Alternative and Conventional Treatments*. Alexandria, VA: Time Life, Inc., 1996.

Springhouse Corporation. *Everything You Need to Know About Diseases*. Springhouse, PA: Springhouse Corporation, 1996.

### ORGANIZATIONS

American Liver Foundation. 1425 Pompton Ave., Cedar Grove, NJ 07009. (800) 223-0179. <<http://www.liverfoundation.org>>.

United Network for Organ Sharing. 1100 Boulders Parkway, Suite 500, P.O. Box 13770, Richmond, VA 23225-8770. (804) 330-8500.

### OTHER

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"Cirrhosis of the Liver." *National Institute of Diabetes and Digestive and Kidney Disease*. <<http://www.niddk.nih.gov>>.

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**.

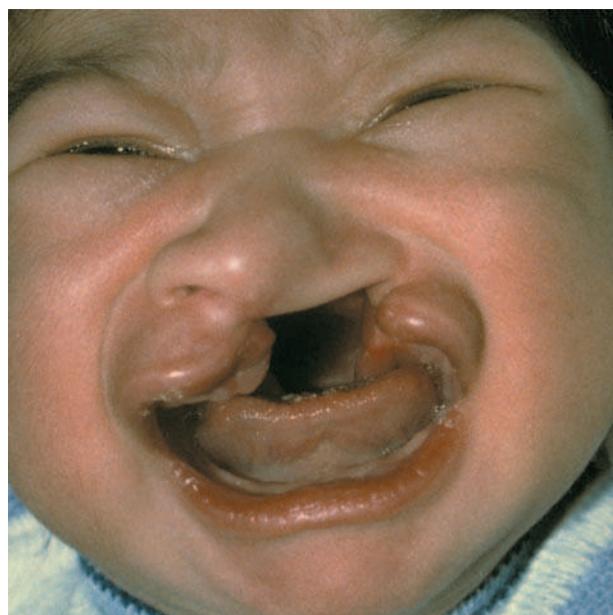
Teeth may also erupt misaligned.

## 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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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.

## Resources

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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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### ORGANIZATIONS

- 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.

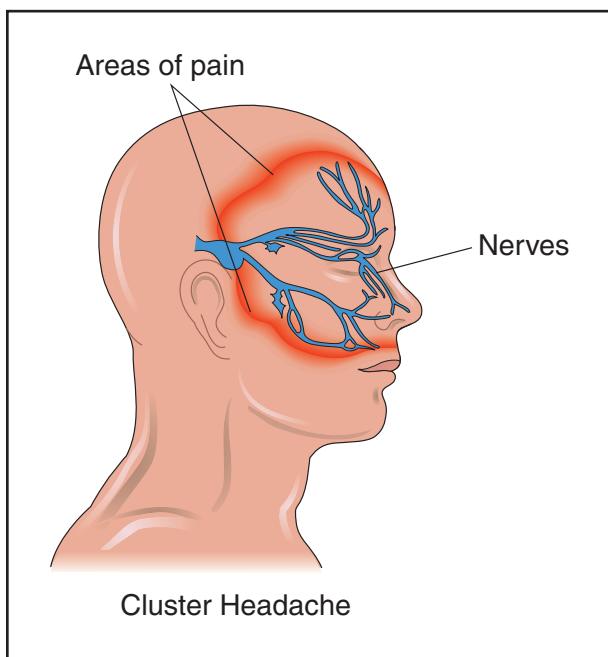
### Causes and symptoms

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 Mantua 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.

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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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## 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](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>>.

### OTHER

Powderly, William G. "Coccidioidomycosis." *Journal of the International Association of Physicians in AIDS Care*. Dec. 1996 <<http://www.iapac.org>>.

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.

### Resources

#### OTHER

- Maigne, Jean-Yves. "Treatment Strategies for Coccydynia." May 7, 2001 <<http://www.coccyx.org/whatisit.htm>>.
- "Treatments for Coccydynia." May 7, 2001 <<http://www.coccyx.org/treatment.htm>>.
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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 signs 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

#### BOOKS

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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.

### PERIODICALS

- Arana-Ward, Marie. "The Pros and Cons of Cochlear Implants." *Newsday*, 3 June 1997, B29.
- Platman, Cheryl. "Hearing aids: A Link to the World." *FDA Consumer* 24 (1 Feb. 1990): 18-23.
- Signor, Roger. "Sound Advice: Cochlear Implants Improve Learning for Deaf Children." *St. Louis Post-Dispatch*, 9 June 1995: 01C.

### 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

### BOOKS

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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.

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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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Taylor, Robert, ed. *Family Medicine Principles and Practice*. New York: Springer-Verlag, 1994.

### ORGANIZATIONS

American Academy of Family Physicians. 8880 Ward Parkway, Kansas City, MO 64114. (816) 333-9700. <<http://www.aafp.org>>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098. (847) 434-4000. <<http://www.aap.org>>.

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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.

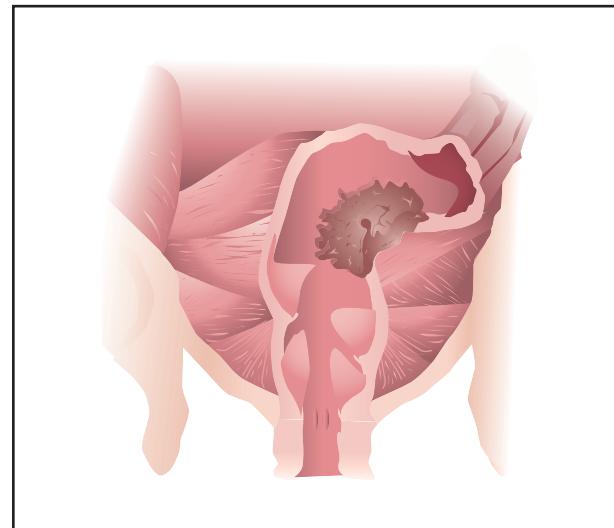
The American Cancer Society has recommended the following screening protocol for those of normal risk over 50 years of age:

- 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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### ORGANIZATIONS

- American Cancer Society. (800) ACS-2345. <<http://www.cancer.org>>.
- Cancer Information Service of the NCI. (1-800-4-CANCER). <<http://www.cancer.gov>>.
- Colon Cancer Alliance. <<http://www.ccalliance.org>>.
- National Cancer Institute Cancer Trials. <<http://cancertrials.nci.nih.gov/system>>. <<http://www.cancertrials.com>>.

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.

## Resources

### BOOKS

Bonk, Melinda, ed. *Alternative Medicine Yellow Pages*. Tiburon, CA: Future Medicine Publishing, Inc., 1994.

## 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

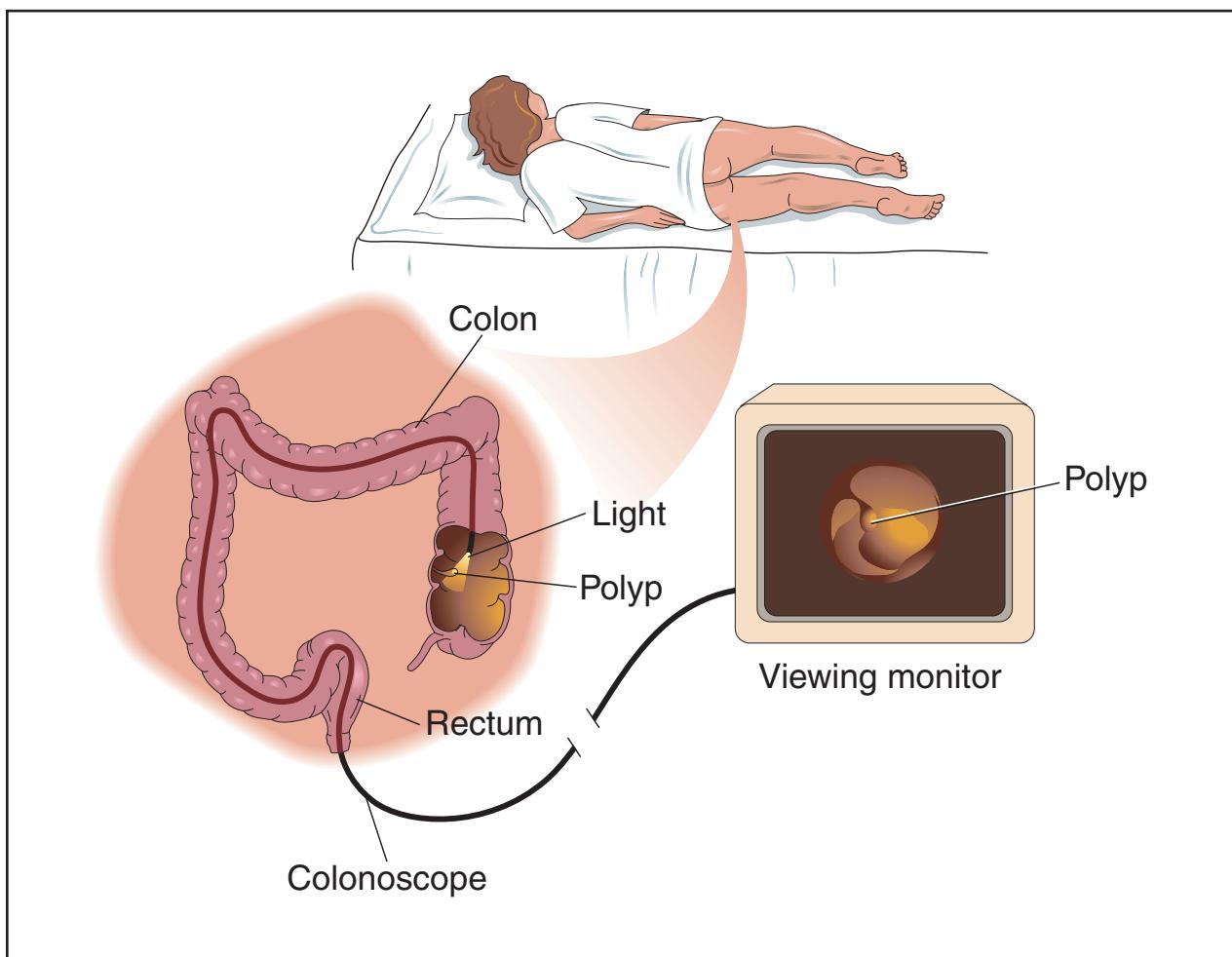
A colonoscopy is generally recommended when the patient complains of rectal bleeding or has a change in bowel habits and other unexplained abdominal symptoms. The test is frequently used to test for colorectal cancer, especially when polyps or tumor-like growths have been detected using the **barium 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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 Sobel, David S., and T. Ferguson. *The People's Book of Medical Tests*. Summit Books, 1985.

### 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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- Achromatopsia Network. C/O Frances Futterman, PO Box 214, Berkeley, CA 94701-0214. <[http://www.achromat.org/how\\_to\\_join.html](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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- "Color Blindness." *Geocities*. <<http://www.geocities.com/Heartland/8833/coloreye.html>>.
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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.

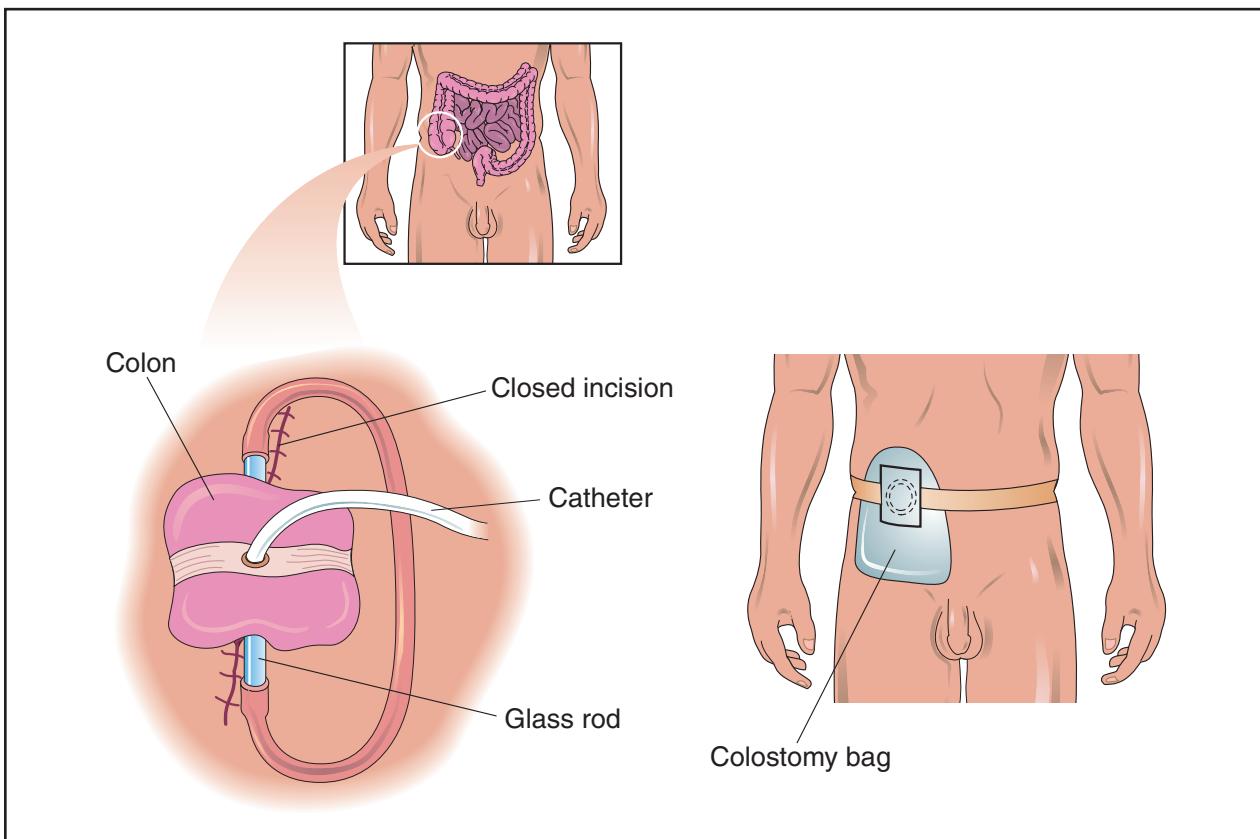
### Purpose

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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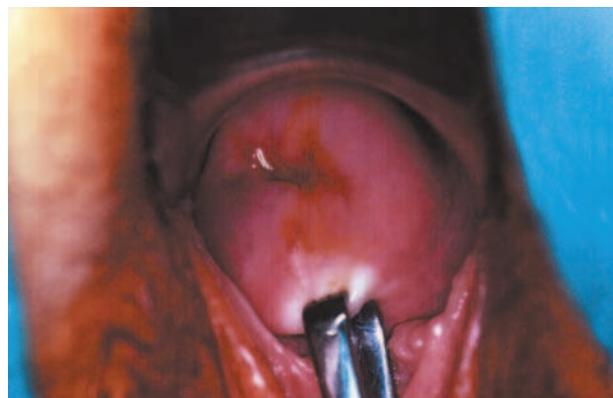
- United Ostomy Association, Inc. (UOA). 19772 MacArthur Blvd., Suite 200, Irvine, CA 92612-2405. (800) 826-0826. <<http://www.uoa.org>>.

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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

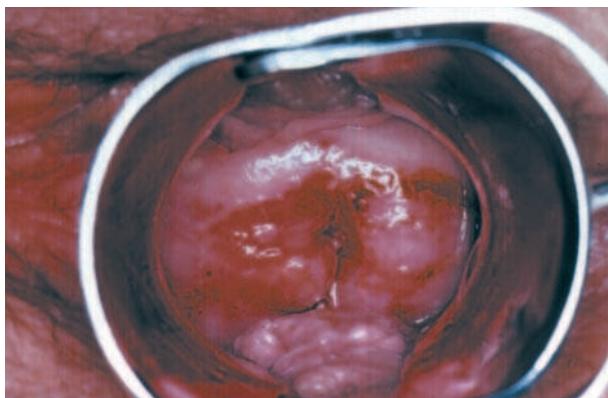
### Definition

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).

## Resources

### BOOKS

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Ryan, Kenneth J., Ross S. Berkowitz, and Robert L. Barbieri. *Kistner's Gynecology*. 6th ed. St. Louis: Mosby, 1995.

### 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.

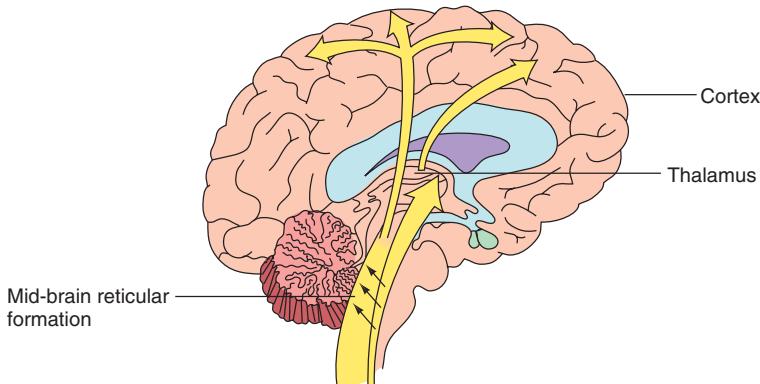
In order to understand the loss of function suffered by a comatose individual, it is necessary to first understand the important characteristics of the conscious state. Consciousness is defined by two fundamental elements: awareness and arousal.

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.

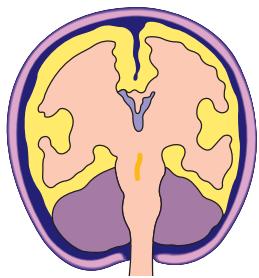
### Causes and symptoms

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

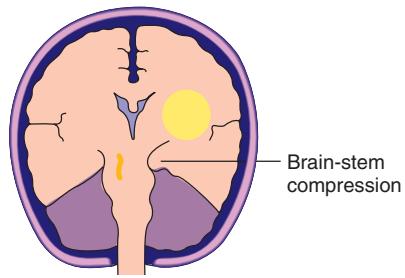
**Diffuse and bilateral damage to the cerebral cortex (relative preservation of brain-stem reflexes)**



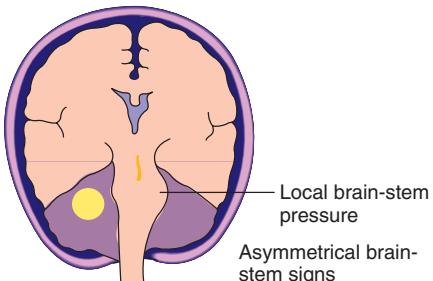
**Possible causes**

- 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)**

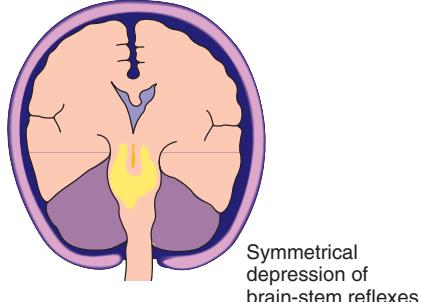


**Structural lesions within this region also resulting in compression of the brain-stem and damage to the reticular activating substance (RAS)**



**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.

## Resources

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*Professional Guide to Diseases*. 5th ed. Springhouse, PA: Springhouse Corporation, 1995.

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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"Complement Deficiency States." *Immune Deficiency and Allied Disorders Newsletter* 2 (June 1996): 1-6.

### ORGANIZATIONS

Immune Deficiency Foundation. 25 W. Chesapeake Ave., Suite 206, Towson, MD 21204. (800) 296-4433. <<http://www.primaryimmune.org>>.

### OTHER

"The Clinical Presentation of the Primary Immunodeficiency Diseases." *International Patient Organization for Patient's with Primary Immunodeficiencies*. <<http://www.ipopi.org>>.

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

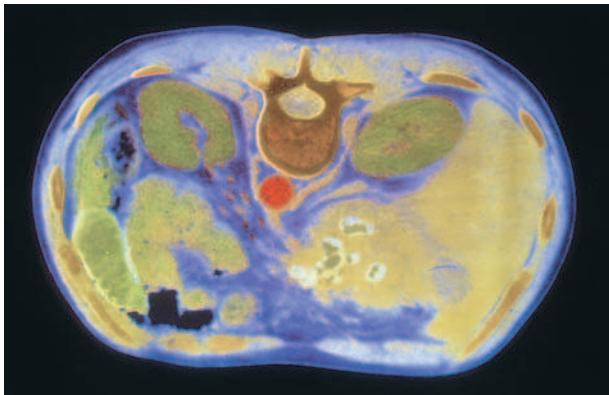
Computed tomography, also called CT scan, CAT scan, or computerized axial tomography, is a combination of focused x-ray beams and computerized production of an image. Introduced in the early 1970s, this radiologic procedure has advanced rapidly and is now widely used, sometimes in the place of standard 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

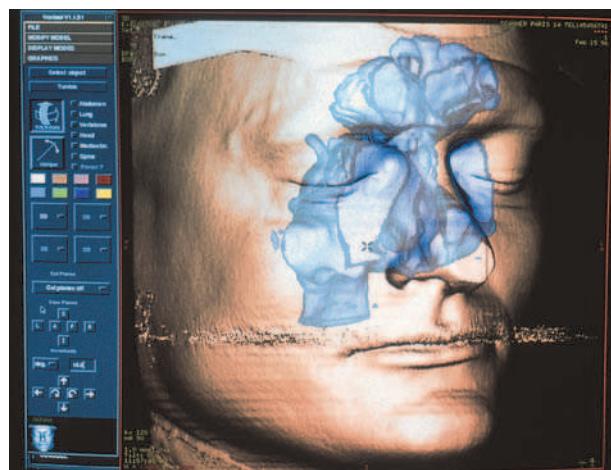
Abnormal results may show different characteristics of tissues within organs. Accumulations of blood or other fluids where they do not belong may be detected. Radiologists can differentiate among types of tumors throughout the body by viewing details of their makeup.

#### 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).

### Resources

#### BOOKS

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### ORGANIZATIONS

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.

## Resources

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### PERIODICALS

Quality Standards Subcommittee, American Academy of Neurology. "Practice Parameter: The Management of Concussion in Sports." *Neurology* 48 (Mar. 1997): 581-585.  
"Sports-Related Recurrent Brain Injuries—United States." *Morbidity and Mortality Weekly Report*, 14 Mar. 1997, 224-227.

### ORGANIZATIONS

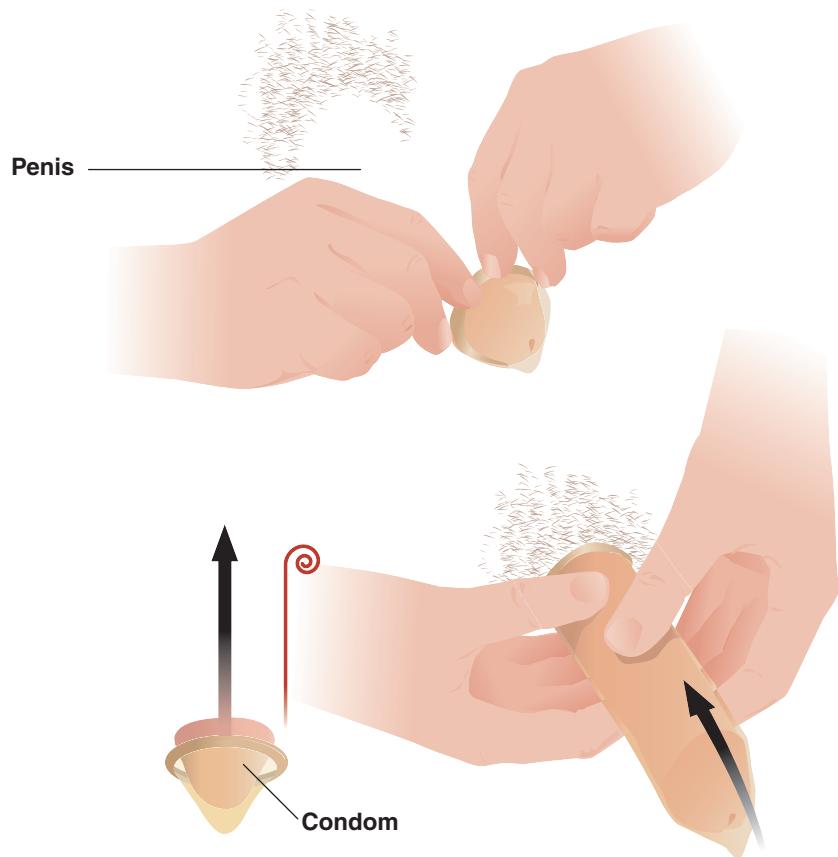
American Academy of Neurology. 1080 Montreal Ave., St. Paul, MN 55116. (612) 695-1940. <<http://www.aan.com>>.

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

### OTHER

“The Condom.” *Sexual Health InfoCenter*. <<http://www.sexhealth.org/infocenter/GuideSS/condoms.htm>>.

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“Spermicides, Condoms and Other Barrier Methods.” *Epigee Birth Control Guide*. <<http://epigee.netministries.org/guide/barrier.html>>.

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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#### ORGANIZATIONS

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.

## Resources

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### ORGANIZATIONS

- American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098. (847) 434-4000.
- National Adrenal Diseases Foundation. 510 Northern Boulevard, Great Neck, NY 11021. (516) 487-4992. <<http://medhlp.netusa.net/www/nadf.htm>>.

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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.

### Resources

#### BOOKS

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International Child Amputee Network. <<http://www.amp-info.net/childamp.htm>>.  
March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (914) 428-7100. <<http://www.modimes.org>>.

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.

## Resources

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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 meningomyelocoele. 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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March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (914) 428-7100. <<http://www.modimes.org>>.

Jeffrey P. Larson, RPT

## 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

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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

#### BOOKS

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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.

## Resources

### BOOKS

Bellenir, Karen, and Peter D. Dresser, eds. *Cardiovascular Diseases and Disorders Sourcebook*. Detroit: Omnigraphics, 1995.

Texas Heart Institute. *Heart Owner's Handbook*. New York: John Wiley and Sons, 1996.

### 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>>.

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.

## Resources

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American Heart Association. 7272 Greenville Ave., Dallas, TX 75231-4596. (214) 373-6300 or (800) 242-8721. [<inquire@heart.org>](mailto:<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.

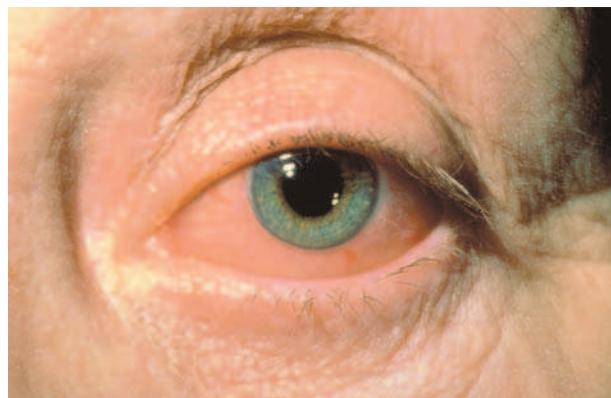
Bacterial conjunctivitis can occur in adults and children and is caused by organisms such as *Staphylococcus*, *Streptococcus*, and *Hemophilus*. Symptoms of bacterial conjunctivitis include a pus-like discharge and crusty eyelids after awakening. Redness of the conjunctiva can be mild to severe and 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

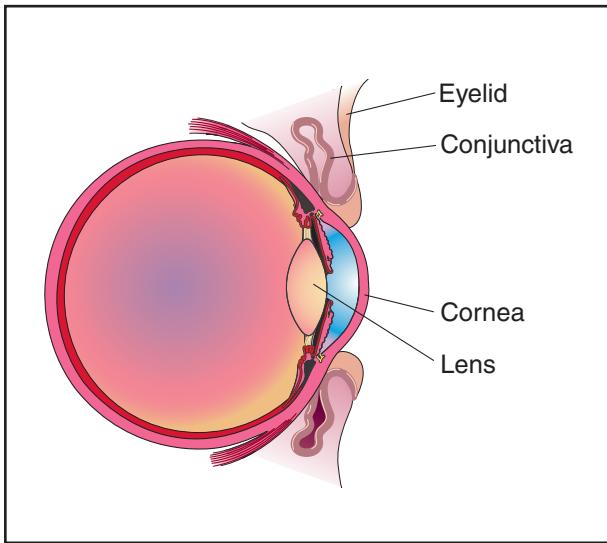
The treatment of conjunctivitis depends on what caused the condition. In all cases, warm compresses applied to the affected eye several times a day may help to reduce discomfort.

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

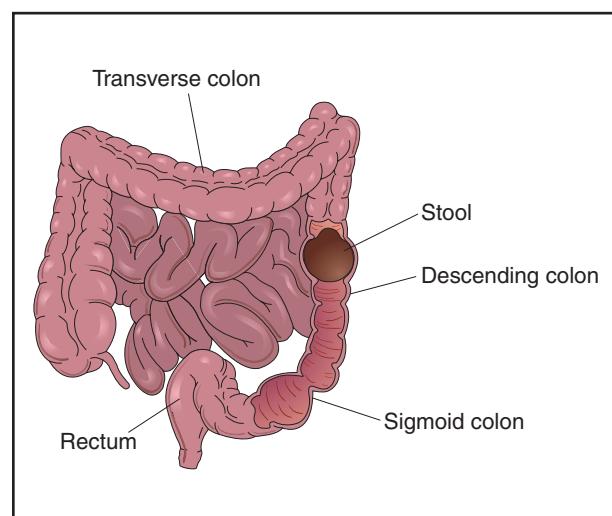
Six drops of rosemary (*Rosmarinus officinalis*) and six drops of thyme (*Thymus* spp.) diluted by 1 oz of almond oil, olive oil, or another carrier oil can relieve constipation when used to massage the abdomen.

### 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

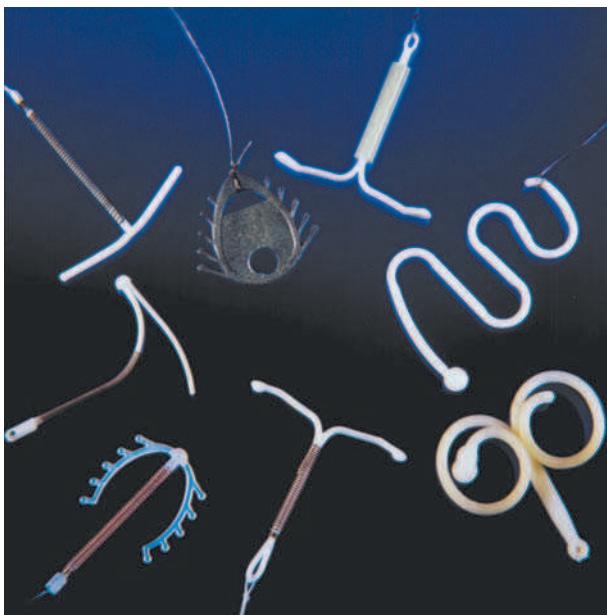
er methods include the **condom**, diaphragm, and cervical cap. The condom is the only form of birth control that also protects against **sexually transmitted diseases**, including HIV (the virus that causes **AIDS**).

- 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.

### Resources

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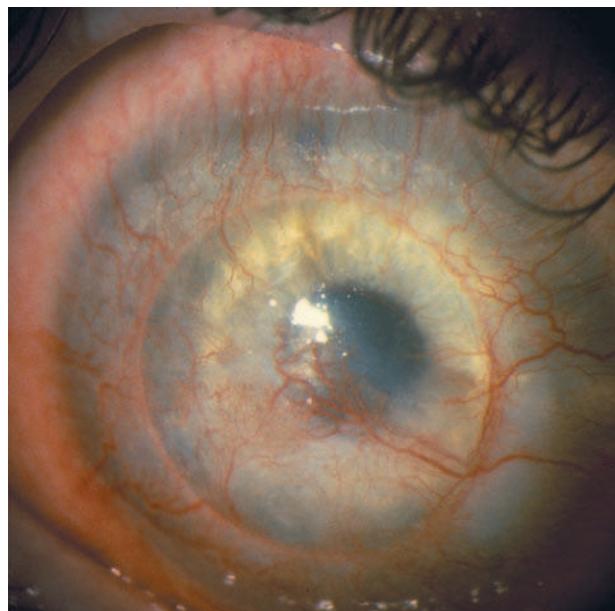
American Heart Association. 7320 Greenville Ave. Dallas, TX 75231. (214) 373-6300. <<http://www.americanheart.org>>.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

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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.)

## 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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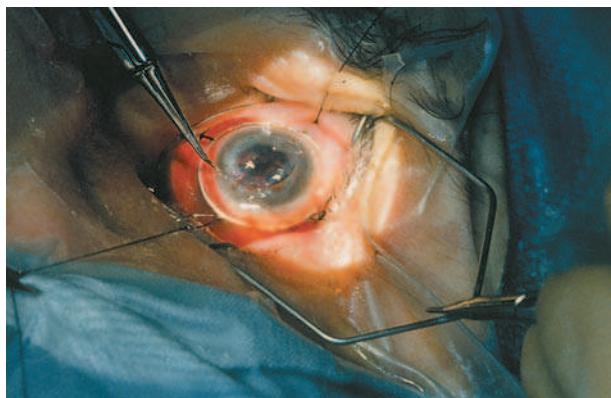
### ORGANIZATIONS

- 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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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.

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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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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

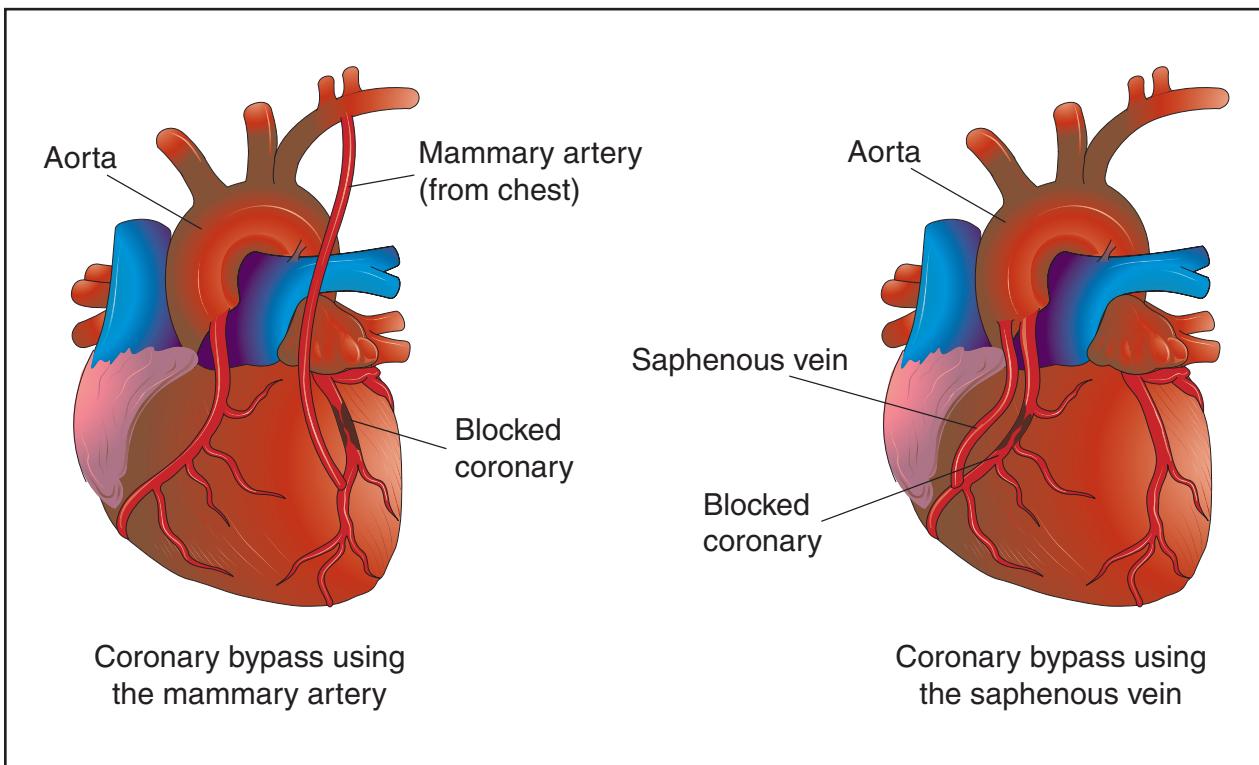
Coronary artery bypass graft surgery should ideally be postponed for three months after a heart attack. Patients should be medically stable before the surgery, if possible.

### 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.

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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**

### **BOOKS**

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#### PERIODICALS

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- "More Evidence for Infection as a Cause of Heart Disease." *Harvard Heart Letter* 7 (Feb. 1997): 6-7.
- "Women and Heart Disease" *Harvard Women's Health Watch* 4 (July 1997): 4-5.
- Zamula, Evelyn. "Balloons to Bypass Bypass Surgery." *FDA Consumer* 22 (1 May 1988): 24-27.

#### 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>>.
- Texas Heart Institute. Heart Information Service. P.O. Box 20345, Houston, TX 77225-0345. <<http://www.tmc.edu/thi>>.

#### OTHER

- "Facts About Coronary Heart Disease." *National Heart, Lung, and Blood Institute*. 27 Nov. 1998. 3 Mar. 1998 <<http://www.nhlbi.nih.gov/index.htm>>.
- "Heart and Stroke 1998 Update." *American Heart Association*. 3 Mar. 1998 <<http://www.americanheart.org>>.

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.

### Resources

#### BOOKS

Garratt, Kirk N., Guy S. Reeder, and David R. Holmes Jr. "Cardiac Catheterization and Angiography." In *Mayo Clinic Practice of Cardiology*, ed. Emilio R. Giuliani, et al. St. Louis: Mosby Publishing, 1996.

*Clinic Practice of Cardiology*, ed. Emilio R. Giuliani, et al. St. Louis: Mosby Publishing, 1996.

#### ORGANIZATIONS

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>>.

Samuel Uretsky, PharmD

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

### BOOKS

- Cahill, Mathew. *Handbook of Diagnostic Tests*. Springhouse, PA: Springhouse Corporation, 1995.  
 Jacobs, David S., et al. *Laboratory Test Handbook*. 4th ed. New York: Lexi-Comp, Inc., 1996.  
 Pagana, Kathleen Deska. *Mosby's Manual of Diagnostic and Laboratory Tests*. St. Louis: Mosby, Inc., 1998.

Janis O. Flores

## 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>>.

Joseph Knight, PA

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

### OTHER

- Day, C. *Costochondritis Web Site*. 2001. <<http://www.geocities.com/Hots/2338/#Frequently%20Asked%20Questions>>.  
 Flowers, L. K., and B. D. Wippermann. "Costochondritis." *eMedicine Journal: Emergency Medicine/Rheumatology*. 23 Feb. 2001. <<http://www.emedicine.com/emerg/topic116.htm>>.

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.

## Resources

### BOOKS

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Schumann, Lorna. "Alterations in Respiratory Function." In *Perspectives on Pathophysiology*, ed. Lee-Ellen Copstead. Philadelphia: W. B. Saunders Co., 1994.

### PERIODICALS

Philp, Elizabeth B. "Chronic Cough." *American Family Physician* 56 (1 Oct. 1997).

### ORGANIZATIONS

National Heart, Lung and Blood Institute. PO Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

Jeffrey P. Larson, RPT

## 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

#### BOOKS

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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

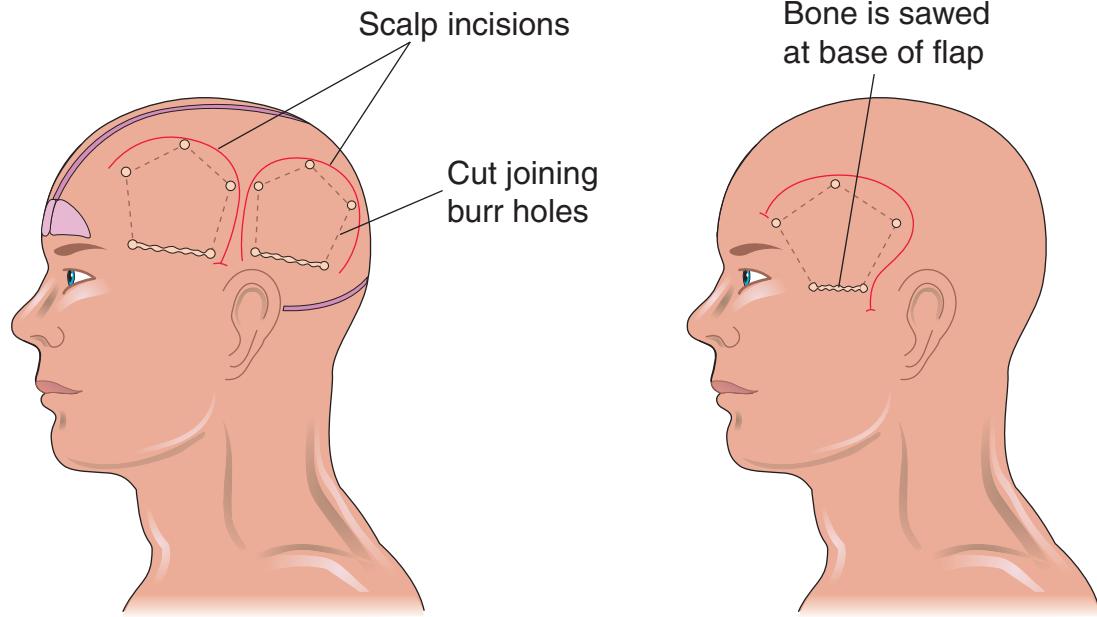


Figure A

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

- Smeltzer, Suzanne, and Brenda Bare. "Management of Patients with Neurological Dysfunction." In *Brunner and Suddarth's Textbook of Medical/Surgical Nursing*. 7th ed. Philadelphia: J. B. Lippincott Co., 1992.
- 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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 Pagana, Kathleen Deska. *Mosby's Manual of Diagnostic and Laboratory Tests*. St. Louis: Mosby, Inc., 1998.

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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 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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#### ORGANIZATIONS

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>>.

#### OTHER

*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- $\alpha$ ). 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- $\alpha$  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.

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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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- 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.

## Resources

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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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### ORGANIZATIONS

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

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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.

### Resources

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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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Michael Sherwin Walston  
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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- 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

#### BOOKS

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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 uninvolved, 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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#### ORGANIZATIONS

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

Michelle Johnson, M.S., J.D.

## 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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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.

## OTHER

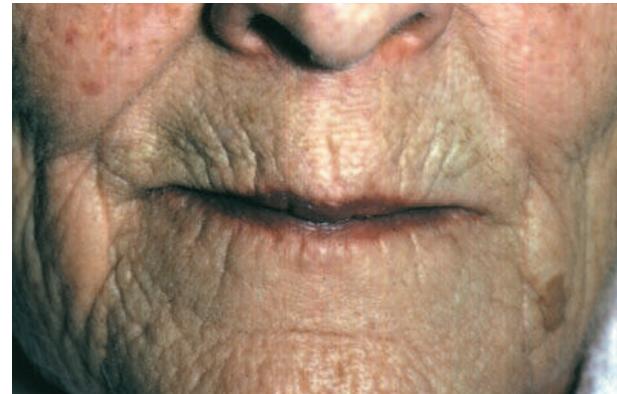
*OMIM Homepage, Online Mendelian Inheritance in Man.*  
<http://www.ncbi.nlm.nih.gov/Omim>.

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.

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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.

## Resources

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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.

### Resources

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#### OTHER

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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.

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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.

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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

ratio of UTIs in children younger than 12 months is 4:1 because of the high rate of **birth defects** in the urinary tract of male infants. In adult life, the male/female ratio of UTIs is 1:50. After age 50, however, the incidence among males increases due to prostate disorders.

### Cystitis in women

Cystitis is a common female problem. It is estimated that 50% of adult women experience at least one episode of dysuria (painful urination); half of these patients have a bacterial UTI. Between 2–5% of women's visits to primary care doctors are for UTI symptoms. About 90% of UTIs in women are uncomplicated but recurrent.

### Cystitis in men

UTIs are uncommon in younger and middle-aged men, but may occur as complications of bacterial infections of the kidney or prostate gland.

### Cystitis in children

In children, cystitis is often caused by congenital abnormalities (present at birth) of the urinary tract. **Vesicoureteral reflux** is a condition in which the child cannot completely empty the bladder. It allows urine to remain in or flow backward (reflux) into the partially empty bladder.

## Causes and symptoms

The causes of cystitis vary according to sex because of the differences in anatomical structure of the urinary tract.

### Females

Most bladder infections in women are so-called ascending infections, which means that they are caused by disease agents traveling upward through the urethra to the bladder. The relative shortness of the female urethra (1.2–2 in in length) makes it easy for bacteria to gain entry to the bladder and multiply. The most common bacteria associated with UTIs in women include *Escherichia coli* (about 80% of cases), *Staphylococcus saprophyticus*, *Klebsiella*, *Enterobacter*, and *Proteus* species. Risk factors for UTIs in women include:

- Sexual intercourse. The risk of infection increases if the woman has multiple partners.
- Use of a diaphragm for **contraception**
- An abnormally short urethra
- Diabetes or chronic **dehydration**
- The absence of a specific enzyme (fucosyltransferase) in vaginal secretions. The lack of this enzyme makes it easier for the vagina to harbor bacteria that cause UTIs.

- Inadequate personal hygiene. Bacteria from fecal matter or vaginal discharges can enter the female urethra because its opening is very close to the vagina and anus.
- History of previous UTIs. About 80% of women with cystitis develop recurrences within two years.

The early symptoms of cystitis in women are dysuria, or **pain** on urination; urgency, or a sudden strong desire to urinate; and increased frequency of urination. About 50% of female patients experience **fever**, pain in the lower back or flanks, **nausea and vomiting**, or shaking chills. These symptoms indicate pyelonephritis, or spread of the infection to the upper urinary tract.

### Males

Most UTIs in adult males are complications of kidney or prostate infections. They are usually associated with a tumor or **kidney stones** that block the flow of urine and are often persistent infections caused by drug-resistant organisms. UTIs in men are most likely to be caused by *E. coli* or another gram-negative bacterium. *S. saprophyticus*, which is the second most common cause of UTIs in women, rarely causes infections in men. Risk factors for UTIs in men include:

- Lack of **circumcision**. The foreskin can harbor bacteria that cause UTIs.
- Urinary catheterization. The longer the period of catheterization, the higher the risk of UTIs.

The symptoms of cystitis and pyelonephritis in men are the same as in women.

### Hemorrhagic cystitis

Hemorrhagic cystitis, which is marked by large quantities of blood in the urine, is caused by an acute bacterial infection of the bladder. In some cases, hemorrhagic cystitis is a side effect of **radiation therapy** or treatment with cyclophosphamide. Hemorrhagic cystitis in children is associated with adenovirus type 11.

## Diagnosis

When cystitis is suspected, the doctor will first examine the patient's abdomen and lower back, to evaluate unusual enlargements of the kidneys or swelling of the bladder. In small children, the doctor will check for fever, abdominal masses, and a swollen bladder.

The next step in diagnosis is collection of a urine sample. The procedure differs somewhat for women and men. Laboratory testing of urine samples can now be performed with dipsticks that indicate immune system responses to infection, as well as with microscopic analysis of samples. Normal human urine is sterile. The

presence of bacteria or pus in the urine usually indicates infection. The presence of hematuria, or blood in the urine, may indicate acute UTIs, kidney disease, kidney stones, inflammation of the prostate (in men), **endometriosis** (in women), or **cancer** of the urinary tract. In some cases, blood in the urine results from athletic training, particularly in runners.

### *Females*

Female patients require a pelvic examination as part of the procedure to obtain urine specimens. The patient lies on an obstetrical table with legs in the stirrups. The doctor first takes a vaginal culture smear. The patient is then asked to void while lying on the table. The first 5–10 ml are collected to test for urethral infection. A midstream urine sample of 200 ml is then collected to test for bladder infection.

In women, a vaginal bacterial count that is higher than those of the two urine samples indicates vaginitis. A high bacterial count in the first urine sample indicates urethritis. A count of more than 104 bacteria CFU/ml (colony forming units per milliliter) in the midstream sample indicates a bladder or kidney infection. A colony is a large number of microorganisms that grow from a single cell within a substance called a culture. A bacterial count can be given in CFU (colony forming units).

### *Males*

In male patients, the doctor will cleanse the opening to the urethra with an antiseptic before collecting the urine sample. The first 10 ml of specimen are collected separately. The patient then voids a midstream sample of 200 ml. Following the second sample, the doctor will massage the patient's prostate and collect several drops of prostatic fluid. The patient then voids a third urine specimen for prostatic culture.

A high bacterial count in the first urine specimen or the prostatic specimens indicates urethritis or prostate infections respectively. A bacterial count greater than 100,000 bacteria CFU/ml in the midstream sample suggests a bladder or kidney infection.

### *Other tests*

Women with recurrent UTIs can be given ultrasound tests of the kidneys and bladder together with a voiding cystourethrogram to test for structural abnormalities. (A cystourethrogram is an x-ray test in which an iodine dye is used to better view the urinary bladder and urethra.) Voiding cystourethrograms are also used to evaluate children with UTIs. In some cases, **computed tomography scans** (CT scans) can be used to evaluate patients for possible cancers in the urinary tract.

## KEY TERMS

**Bacteriuria**—The presence of bacteria in the urine.

**Dysuria**—Painful or difficult urination.

**Hematuria**—The presence of blood in the urine.

**Pyelonephritis**—Bacterial inflammation of the upper urinary tract.

**Urethritis**—Inflammation of the urethra, which is the passage through which the urine moves from the bladder to the outside of the body.

## Treatment

### *Medications*

Uncomplicated cystitis is treated with **antibiotics**. These include penicillin, ampicillin, and amoxicillin; sulfisoxazole or sulfamethoxazole; trimethoprim; nitrofurantoin; **cephalosporins**; or **fluoroquinolones**. (Fluoroquinolones are generally not used in children under 18 years of age.) Treatment for women is short-term; most patients respond within three days. Men do not respond as well to short-term treatment and require seven to 10 days of oral antibiotics for uncomplicated UTIs.

Patients of either sex may be given phenazopyridine or flavoxate to relieve painful urination.

Trimethoprim and nitrofurantoin are preferred for treating recurrent UTIs in women.

Over 50% of older men with UTIs also suffer from infection of the prostate gland. Some antibiotics, including amoxicillin and the cephalosporins, do not affect the prostate gland. Fluoroquinolone antibiotics or trimethoprim are the drugs of choice for these patients.

Patients with pyelonephritis can be treated with oral antibiotics or intramuscular doses of cephalosporins. Medications are given for 10–14 days, and sometimes longer. If the patient requires hospitalization because of high fever and dehydration caused by vomiting, antibiotics can be given intravenously.

### *Surgery*

A minority of women with complicated UTIs may require surgical treatment to prevent recurrent infections. Surgery is also used to treat reflux problems (movement of the urine backwards) or other structural abnormalities in children and anatomical abnormalities in adult males.

## Alternative treatment

Alternative treatment for cystitis may emphasize eliminating all sugar from the diet and drinking lots of water. Drinking unsweetened cranberry juice not only adds fluid, but is also thought to help prevent cystitis by making it more difficult for bacteria to cling to the bladder wall. A variety of herbal therapies are also recommended. Generally, the recommended herbs are antimicrobials, such as garlic (*Allium sativum*), goldenseal (*Hydrastis canadensis*), and bearberry (*Arctostaphylos uva-ursi*), and/or demulcents that soothe and coat the urinary tract, including corn silk and marsh mallow (*Althaea officinalis*).

Homeopathic medicine can also be effective in treating cystitis. Choosing the correct remedy based on the individual's symptoms is always key to the success of this type of treatment. **Acupuncture** and Chinese traditional herbal medicine can also be helpful in treating acute and chronic cases of cystitis.

## Prognosis

### Females

The prognosis for recovery from uncomplicated cystitis is excellent.

### Males

The prognosis for recovery from uncomplicated UTIs is excellent; however, complicated UTIs in males are difficult to treat because they often involve bacteria that are resistant to commonly used antibiotics.

## Prevention

### Females

Women with two or more UTIs within a six-month period are sometimes given prophylactic treatment, usually nitrofurantoin or trimethoprim for three to six months. In some cases the patient is advised to take an antibiotic tablet following sexual intercourse.

Other preventive measures for women include:

- drinking large amounts of fluid
- voiding frequently, particularly after intercourse
- proper cleansing of the area around the urethra

### Males

The primary preventive measure for males is prompt treatment of prostate infections. Chronic **prostatitis** may go unnoticed, but can trigger recurrent UTIs. In addition, males who require temporary catheterization following surgery can be given antibiotics to lower the risk of UTIs.

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Rebecca J. Frey

## Cystometry

### Definition

Cystometry is a test of bladder function in which pressure and volume of fluid in the bladder is measured during filling, storage, and voiding.

### Purpose

The urinary bladder stores urine produced by the kidneys. The main muscle of the bladder wall, the detrusor, relaxes to allow expansion of the bladder during filling. The urethra, the tube through which urine exits, is held closed by a ring of muscle, known as the urethral sphincter. As volume increases, stretching of the detrusor and pressure on the sphincter sends signals to the brain, indicating the need for urination, or voiding. Voluntary relaxation of the sphincter and automatic contractions of the detrusor allow successful and virtually complete voiding.

A cystometry study is performed to diagnose problems with urination, including incontinence, urinary retention, and recurrent urinary tract infections. Urinary difficulties may occur because of weak or hyperactive sphincter or detrusor, or incoordination of their two

activities. Infection of the bladder or urethra may cause incontinence, as can obstruction of the urethra from scar tissue, prostate enlargement, or other benign or cancerous growths. Loss of sensation due to nerve damage can lead to chronic overfilling.

### Precautions

The mild irritation of the urinary tract necessary for insertion of the catheter may occasionally cause flushing, sweating, and nausea.

### Description

The patient begins by emptying the bladder as much as possible. A thin plastic catheter is then slowly inserted into the urethra until it reaches the bladder. Measurements are taken of the residual urine volume and bladder pressure. Pressure measurements may require a rectal probe to account for the contribution of the abdominal muscles to the pressure recording.

The bladder is then gradually filled with either warm water, room temperature water, saline solution, carbon dioxide gas, or a contrast solution for x-ray analysis, depending on the type of study being done. The patient is asked to describe sensations during filling, including temperature sensations and when the first feeling of bladder fullness occurs. Once the bladder is completely full, the patient is asked to begin voiding, and measurements are again made of pressure and volume, as well as flow rate and pressure.

### Preparation

There is no special preparation needed for this test. The patient may be asked to stop taking certain medications in advance of the test, including sedatives, cholinergics, and anticholinergics.

### Aftercare

Cystometry can be somewhat uncomfortable. The patient may wish to reserve an hour or so afterward to recover. Urinary frequency or urgency, and some reddening of the urine, may last for a day. Increasing fluid intake helps to flush out the bladder, but caffeinated, carbonated, or alcoholic beverages are discouraged, because they may irritate the bladder lining. Signs of infection, such as **fever**, **chills**, **low back pain**, or persistent blood in the urine, should be reported to the examining physician.

### Risks

There is a slight risk of infection due to tearing of the urethral lining.

## KEY TERMS

**Detrusor**—Muscle of the bladder wall.

**Sphincter**—Ring of muscle between the bladder and the urethra that functions to close off the urethra.

**Urethra**—Tube that empties urine from the bladder to the exterior of the body.

### Normal results

The normal bladder should not begin contractions during filling and should initially expand without resistance. A feeling of fullness occurs with a volume of 100–200 ml. The adult bladder capacity is 300–500 ml. The sphincter should relax and open when the patient wills it, accompanied by detrusor contractions. During voiding, detrusor contraction should be smooth and lead to a steady urine stream.

### Abnormal results

Inability of the bladder to relax during filling, or low bladder volume, may indicate interstitial **cystitis**, prostate enlargement, or **bladder cancer**. Contraction of the bladder during filling may be due to irritation from infection or cysts, obstruction of the bladder outlet, or neurological disease such as **stroke**, **multiple sclerosis**, or **spinal cord injury**. Diminished sensation may occur with nerve lesions, **peripheral neuropathy**, or chronic overfilling.

### Resources

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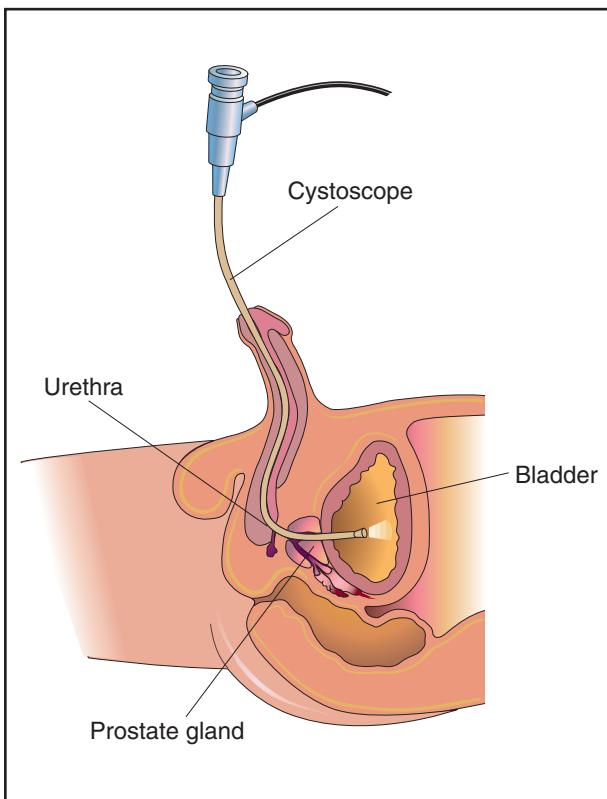
"Cystometrogram, Simple and Complex." *HealthGate Page*. <<http://www.healthgate.com/HealthGate/free/dph/static/dph.0085.shtml>>.

Richard Robinson

## Cystoscopy

### Definition

Cystoscopy (cystourethroscopy) is a diagnostic procedure that is used to look at the bladder (lower urinary



**Cystoscopy** is a diagnostic procedure which is used to view the bladder, collect urine samples, and examine the prostate gland. This procedure also enables biopsies to be taken. The primary instrument used in cystoscopy is the cystoscope, a tube which is inserted through the penis into the urethra, and ultimately into the bladder. (Illustration by Electronic Illustrators Group.)

tract), collect urine samples, and examine the prostate gland. Performed with an optic instrument known as a cystoscope (urethroscope), this instrument uses a lighted tip for guidance to aid in diagnosing urinary tract disease and prostate disease. Performed by a urologist, this surgical test also enables biopsies to be taken or small stones to be removed by way of a hollow channel in the cystoscope.

### Purpose

Categorized as an endoscopic procedure, cystoscopy is used by urologists to examine the entire bladder lining and take biopsies of any areas that look questionable. This test is not used on a routine basis, but may benefit the urologist who is needing further information about a patient who displays the following symptoms or diagnosis:

- blood in the urine (also known as hematuria)
- incontinence or the inability to control urination
- a urinary tract infection

- a urinary tract which display signs of congenital abnormalities
- tumors located in the bladder
- the presence of bladder or **kidney stones**
- a stiffness or strained feeling of the urethra or ureters
- symptoms of an **enlarged prostate**

Blood and urine studies, in addition to x rays of the kidneys, ureters, and bladder, may all occur before a cystoscopy. At the time of surgery, a retrograde pyelogram may also be performed. Additional blood studies may be needed immediately following surgery.

### Precautions

While the cystoscopy procedure is commonly relied upon to gather additional diagnostic information, it is an invasive surgical technique that may involve risks for certain patients. Those who are extremely overweight (obese), smoke, are recovering from a recent illness, or are treating a chronic condition may face additional risks from surgery.

Surgical risk also increases in patients who are currently using certain drugs including antihypertensives; **muscle relaxants**; tranquilizers; sleep inducers; insulin; sedatives; **beta blockers**; or cortisone. Those who use mind-altering drugs also put themselves at increased risk of complications during surgery. The following mind-altering drugs should be avoided: narcotics; psychedelics; hallucinogens; marijuana; sedatives; hypnotics; or **cocaine**.

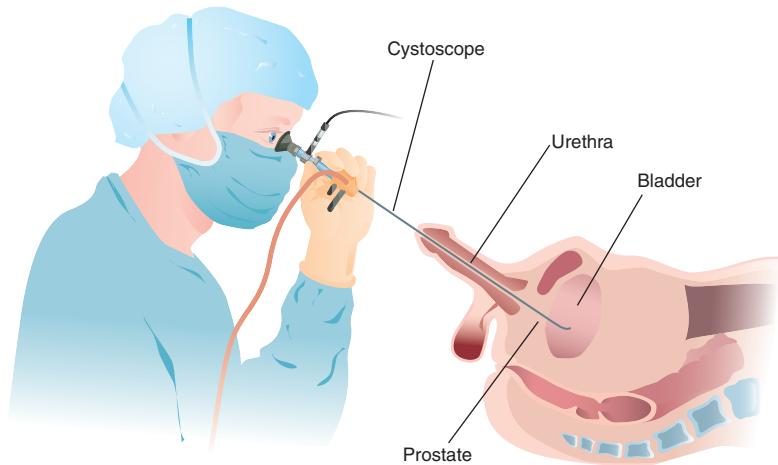
### Description

Depending on the type of information needed from a cystoscopy, the procedure typically takes 10–40 minutes to complete. The patient will be asked to urinate before surgery which allows an accurate measurement of the remaining urine in the bladder. A well lubricated cystoscope is inserted through the urethra into the bladder where a urine sample is taken. Fluid is then pushed in to inflate the bladder and allow the urologist to examine the entire bladder wall.

During an examination, the urologist may take the following steps: remove either bladder or kidney stones; gather tissue samples; and treat any suspicious lesions. In order to perform x-ray studies (retrograde pyelogram), a harmless dye is injected into the ureters by way of a catheter that is passed through the previously placed cystoscope. After completion of all needed tests, the cystoscope is removed.

### Preparation

As a procedure that can be completed in a hospital, doctor's office, or outpatient surgical facility, an injection



**A cystoscope helps the doctor examine the urethra, bladder, and prostate.** (Illustration by Argosy Inc.)

of spinal or general anesthesia may be used prior to a cystoscopy. While this test is typically performed on an outpatient basis, a patient may require up to three days of recovery in the hospital.

### Aftercare

Patients who have undergone a cystoscopy will be instructed to follow these steps to ensure a quick recovery:

- due to soreness or discomfort that may occur in the urethra, especially while urinating, several warm baths a day are recommended to relieve any **pain**
- allow four days for recovery
- blood may appear in the urine—this is common, and soon clears up in one to two days following the procedure
- avoid strenuous **exercise** for a minimum of two weeks following surgery
- sexual relations may continue when the urologist determines that healing is complete
- wait at least two days after surgery before driving

Patients may also be prescribed pain relievers and **antibiotics** following surgery. Minor pain may also be treated with over-the-counter, non-prescription drugs such as **acetaminophen**.

### Risks

As with any surgical procedure, there are some risks involved with a cystoscopy. Complications may include: profuse bleeding; a damaged urethra; a perforated bladder; a urinary tract infection; or an injured penis.

Patients should also contact their physician if they experience any of the following symptoms following surgery: pain, redness, swelling, drainage, or bleeding from the surgical site; signs of infection that may include **headache**, muscle aches, **dizziness** or an overall ill feeling and **fever**; nausea or vomiting; strenuous or painful urination; or symptoms that may result as side-effects from the medication.

### Normal results

A successful cystoscopy includes a thorough examination of the bladder and collection of urine samples for cultures. If no abnormalities are seen, the results are indicated as normal.

### Abnormal results

Cystoscopy allows the urologist to detect inflammation of the bladder lining, prostatic enlargement, or tumors. If these are seen, further evaluation or biopsies may be needed in addition to the removal of some tumors.

### Resources

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## KEY TERMS

**Endoscopy**—Examination of body organs or cavities through the use of an endoscope (a lighted optical instrument used to see inside body cavities), such as a cystoscope used to complete a cystoscopy.

**Retrograde pyelogram**—A pyelography or x-ray technique where radiopaque dye is injected into the kidneys from below, by way of the ureters, allowing further examination of the kidneys.

**Ureter**—The tube that carries urine from the kidney to the bladder, with each kidney having one ureter.

**Urethra**—A passageway from the bladder to the outside for the discharge of urine. In the female, this canal lies between the vagina and clitoris; in the male, the urethra travels through the penis, opening at the tip.

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

Beth A. Kapes

Cystourethroscopy see **Cystoscopy**

Cytomegalic inclusion disease see  
**Cytomegalovirus infection**

## I Cytomegalovirus antibody screening test

### Definition

Cytomegalovirus (CMV) is a common human virus. Antibodies to CMV are evidence of a current or past infection.

### Purpose

Consequences of a CMV infection can be devastating in a pregnant woman, a transplant patient, or a person with human immunodeficiency virus (HIV). Antibody screening helps control the infection risk for these groups.

In a healthy, nonpregnant person, CMV infection is almost never serious. Symptoms, if present, are mild, often resembling **infectious mononucleosis** due to Epstein-Barr virus. Antibody screening distinguishes between these two infections.

### Description

When first exposed to CMV, a person's immune system is triggered and quickly makes antibodies to fight the virus. Antibodies are special proteins designed to attack and destroy foreign material, in this case, the cytomegalovirus.

The test combines a person's serum with a substance to which CMV antibodies attach. This antibody-antigen complex is measured and the amount of original antibody determined. If positive for antibodies, the serum is diluted, or titered, and the test repeated until the serum is so dilute it no longer gives a positive result. The last dilution that gives a positive result is the titer reported.

A test positive for CMV antibodies means the person has been infected with the virus, either currently or in the past; it doesn't mean the person has lifetime immunity. After an infection, this virus, like all members of the herpes virus group, can stay hidden inside a person and cause infection if the person's immune system later weakens and antibody protection decreases. In fact, reactivation of such hidden (or latent) infection is not at all uncommon and usually occurs without symptoms.

Transplant patients and people with weakened immune systems, including those with HIV, are vulnerable to infection from several routes, including from another person, from a donated organ or transfused blood, or from reactivation of a past infection. Before transplant, both the recipient and donor are usually tested for antibodies. A recipient who has never had CMV (negative for antibodies), should not receive an organ from a donor who has had CMV (positive for antibodies). CMV infection can be associated with organ rejection, or can cause illness such as **pneumonia**, hepatitis, or **death**. Similarly, blood is usually screened for CMV antibodies before being transfused into a person with a weakened immune system.

CMV infection is the most common congenital infection (existing at birth). The infection, passed from mother to baby, can cause permanent mental or physical damage, or death. The antibody screening test tells a

woman whether or not she has antibody protection against the virus in case she is exposed during pregnancy.

Tests that measure a specific type of antibody help tell the difference between a current and a past infection. Immunoglobulin M (IgM) antibodies appear at the beginning of an infection and last only weeks. Immunoglobulin G (IgG) antibodies appear 10–14 days later and can last a lifetime. A person suspected of having a current infection should be tested at the beginning of the infection and again 10–14 days later.

The CMV antibody screening test is also called the transplant reaction screening test. Results are usually available the following day.

## Preparation

This test requires 5 mL of blood. Collection of the sample takes only a few minutes.

## 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

A person without previous exposure to CMV will test negative.

## Abnormal results

The presence of antibodies means the person has been infected with CMV, either now or in the past. An antibody titer at least four times higher at the end of the illness than at the beginning, or the presence of IgM antibodies, indicates a recent or current first time infection.

People with weak immune systems may not generate antibodies against CMV. A current infection in a transplant patient or a person with HIV is confirmed with other tests, such as viral culture.

## Resources

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Isada, Carlos M., et al. *Infectious Diseases Handbook*. Hudson: Lexi-Comp Inc., 1995.

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## KEY TERMS

**Antibody**—A special protein built by the body as a defense against foreign material entering the body.

**Cytomegalovirus (CMV)**—A common human virus causing mild or no symptoms in healthy people, but permanent damage or death to an infected fetus, a transplant patient, or a person with HIV.

**Titer**—A dilution of a substance with an exact known amount of fluid. For example, one part of serum diluted with four parts of saline is a titer of 1:4.

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Nancy J. Nordenson

## Cytomegalovirus infection

### Definition

Cytomegalovirus (CMV) is a virus related to the group of herpes viruses. Infection with CMV can cause no symptoms, or can be the source of serious illness in people with weak immune systems. CMV infection is also an important cause of birth defects.

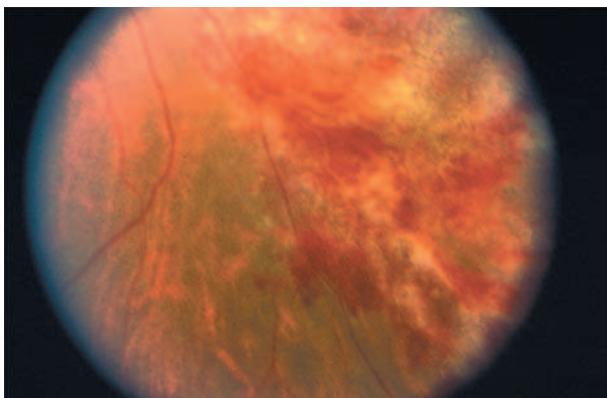
### Description

CMV is an extremely common organism worldwide. It is believed that about 85% of the adult population in the United States have been infected by CMV at some point in their lives. CMV is found in almost all of the body's organs. It is also found in body fluids, including semen, saliva, urine, feces, breast milk, blood, and secretions of the cervix (the narrow, lower section of the uterus).

CMV is also able to cross the placenta (the organ that provides oxygen and nutrients to the unborn baby in the uterus). Because CMV can cross the placental barrier, initial infection in a pregnant woman can lead to infection of the developing baby.

### Causes and symptoms

CMV is passed between people through contact with body fluids. CMV can also be passed on through sexual



**An infected retina of an AIDS patient.** Cytomegaloviruses are herpes viruses that can, among other problems, act as opportunistic infectious agents in suppressed immune systems, a common problem with AIDS sufferers. (Custom Medical Stock Photo. Reproduced by permission.)

contact. Babies can be born infected with CMV, either becoming infected in the uterus (congenital infection) or during birth (from infected cervical secretions).

Like other herpes viruses, CMV remains inactive (dormant) within the body for life after the initial infection. Some of the more serious types of CMV infections occur in people who have been harboring the dormant virus, only to have it reactivate when their immune system is stressed. Immune systems may be weakened because of **cancer chemotherapy**, medications given after organ transplantation, or diseases that significantly lower immune resistance like acquired **immunodeficiency syndrome (AIDS)**.

In a healthy person, initial CMV infection often occurs without symptoms and is rarely noticed. Occasionally, a first-time infection with CMV may cause a mild illness called mononucleosis. Symptoms include swollen glands, liver, and spleen; **fever**; increased white blood cells; **headache**; **fatigue**; and **sore throat**. About 8% of all mononucleosis cases are due to CMV infection. A similar infection, though slightly more serious, may occur two to four weeks after receiving a blood **transfusion** containing CMV.

In people with weakened immune systems, CMV infection can cause more serious and potentially life-threatening illnesses. These illnesses include **pneumonia**, and inflammations of the liver (hepatitis), brain (**encephalitis**), esophagus (esophagitis), large intestine (colitis), and retina of the eye (retinitis).

Babies who contract CMV from their mothers during birth rarely develop any illness from these infections. Infants born prematurely who become CMV infected during birth have a greater chance of complications,

including pneumonia, hepatitis, and decreased blood platelets.

However, an unborn baby is at great risk for serious problems when the mother becomes infected with CMV for the first time while pregnant. About 10% of these babies will be born with obvious problems, including **prematurity**, lung problems, an enlarged liver and spleen, **jaundice**, anemia, low birth weight, small head size, and inflammation of the retina. About 90% of these babies may appear perfectly normal at birth. Unfortunately, about 20% of these babies will later develop severe hearing impairments and **mental retardation**.

### Diagnosis

Body fluids or tissues can be tested to reveal CMV infection. However, this information is not always particularly helpful because CMV stays dormant in the cells for life. Tests to look for special immune cells (antibodies) directed specifically against CMV are useful in proving that a person has been infected with CMV. However, these tests do not give any information regarding when the CMV infection first occurred.

### Treatment

Ganciclovir and foscarnet are both antiviral medications that have been used to treat patients with weak immune systems who develop a serious illness from CMV (including retinitis). As of 1998, research is still being done to try to find useful drugs to treat newborn babies suffering from congenital infection with CMV. **Antiviral drugs** are not used to treat CMV infection in otherwise healthy patients because the drugs have significant side effects that outweigh their benefits.

### Prognosis

Prognosis in healthy people with CMV infection is excellent. About 0.1% of all newborn babies will have serious damage from CMV infection occurring while they were developing in the uterus. About 50% of all transplant patients will develop severe illnesses due to reactivation of dormant CMV infection. These illnesses have a high rate of serious complications and **death**.

### Prevention

Prevention of CMV infection in the normal, healthy person involves good handwashing. Blood products can be screened or treated to insure that they do not contain CMV.

## Resources

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Shelton, Brenda K. "Understanding Cytomegalovirus Infection." *Nursing* 25, no. 1 (Jan. 1995): 32.

### ORGANIZATIONS

Baylor College of Medicine. 1 Baylor Plaza, Houston, TX 77030. (713) 798-4951. <<http://public.bcm.tmc.edu>>.

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

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (914) 428-7100. <<http://www.modimes.org>>.

Rosalyn Carson-DeWitt, MD

# D

D & C see **Dilatation and curettage**

## Dacryocystitis

### Definition

Dacryocystitis is an inflammation of the tear sac (lacrimal sac) at the inner corner of the eye.

### Description

Tears drain into little openings (puncta) in the inner corners of the eyelids. From there, the tears travel through little tube-like structures (canalliculi) to the lacrimal sac. The nasolacrimal ducts then take the tears from the lacrimal sac to the nose. That's why people need to blow their nose when they cry a lot.

Dacryocystitis is usually caused by a blockage of the nasolacrimal duct, which allows fluid to drain into the nasal passages. When the lacrimal sac does not drain, bacteria can grow in the trapped fluid. This condition is most common in infants and people over 40 years old.

### Causes and symptoms

In newborn infants, the nasolacrimal duct may fail to form an opening—a condition called dacryostenosis. The cause of dacryocystitis in adults is usually associated with inflammation and infection in the nasal region. Dacryocystitis can be acute, having a sudden onset, or it can be chronic, with symptoms occurring over the course of weeks or months. Symptoms of acute dacryocystitis can include **pain**, redness, tearing, and swelling at the inner corner of the eye by the nose. In chronic dacryocystitis, the eye area may be swollen, watery or teary, and, when pressure is applied to the area, there may be a discharge of pus or mucus through the punctum.



**Dacryocystitis of the right eye.** The inner corner of the lower lid is bulging from an inflamed tear sac. Blockage of the tear duct causes fluid to be trapped in the tear sac, which becomes infected. (Custom Medical Stock Photo. Reproduced by permission.)

### Diagnosis

Dacryocystitis usually occurs in only one eye. As mentioned, the symptoms can range from watery eyes, pain, swelling, and redness to a discharge of pus when pressure is applied to the area between the bridge of the nose and the inner eyelids. A sample of the pus may be collected on a swab or in a tube for laboratory analysis. The type of antibiotic and treatment may depend on which bacteria is present. In the acute form, a blood test may reveal an elevated white blood cell (WBC) count; with a chronic infection, the WBC count is usually normal. To identify the exact location of the blockage, an x ray can be taken after a dye is injected into the duct in a procedure called dacryocystography.

## KEY TERMS

**Canaliculi**—Also known as lacrimal ducts, these tube-like structures carry the tears from the eyes to the lacrimal sac.

**Cannula**—A narrow tube that can be inserted into a duct.

**Dacryocystography**—An x ray of the tear duct after injection of a dye that is used to help locate a blockage in the duct.

**Dacryocystorhinostomy**—A surgical procedure to drain the tear sac into the nasal passage.

**Dacryostenosis**—Obstruction or narrowing of the nasolacrimal duct. May be present at birth.

**Nasoacral duct**—The tube that carries the tears from the lacrimal sac to the nose.

**Punctum**—Tiny opening at the inner corners of the upper and lower lids. The area for the beginning of tear drainage.

## Prevention

There are no specific recommendations for the prevention of dacryocystitis; however, good hygiene may decrease the chances of infection.

## Resources

### BOOKS

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Eckman, Margaret, and Nancy Priff, eds. “Disorders of the Eyelids and Lacrimal Ducts.” In *Diseases*. 2nd ed. Springhouse, PA: Springhouse Corporation, 1997.

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Altha Roberts Edgren

Dandruff see **Seborrheic dermatitis**

## Treatment

A warm compress applied to the area can help relieve pain and promote drainage. Topical and oral antibiotics may be prescribed if an infection is present. Intravenous antibiotics may be needed if the infection is severe. In some cases, a tiny tube (cannula) is inserted into the tear duct which is then flushed with a sterile salt water solution (sterile saline). If other treatments fail to clear up the symptoms, surgery (dacryocystorhinostomy) to drain the lacrimal sac into the nasal cavity can be performed. In extreme cases, the lacrimal sac will be removed completely.

In infants, gentle massage of the lacrimal sac four times daily for up to nine months can drain the sac and sometimes clear a blockage. As the infant grows, the duct may open by itself. If the duct does not open, it may need to be dilated with a minor surgical procedure.

## Prognosis

Treatment of dacryocystitis with antibiotics is usually successful in clearing the infection that is present. If there is a permanent blockage that prevents drainage, infection may recur and surgery may be required to open the duct. If left untreated, the infected sac can rupture, forming an open, draining sore.

## Death

### Definition

Death is defined as the cessation of all vital functions of the body including the heartbeat, brain activity (including the brain stem), and breathing.

### Description

Death comes in many forms, whether it be expected after a diagnosis of terminal illness or an unexpected accident or medical condition.

### Terminal illness

When a terminal illness is diagnosed, a person, family, friends, and physicians are all able to prepare for the impending death. A terminally ill individual goes through several levels of emotional acceptance while in the process of dying. First, there is denial and **isolation**. This is followed by anger and resentment. Thirdly, a person tries to escape the inevitable. With the realization that death is eminent, most people suffer from depression. Lastly, the reality of death is realized and accepted.

## Causes and symptoms

As of 2001, the two leading causes of death for both men and women in the United States were heart disease and **cancer**. Accidental death was a distant third followed by such problems as **stroke**, chronic lung disorders, **pneumonia**, suicide, **cirrhosis**, **diabetes mellitus**, and murder. The order of these causes of death varies among persons of different age, ethnicity, and gender.

## Diagnosis

In an age of organ transplantation, identifying the moment of death may now involve another life. It thereby takes on supreme legal importance. It is largely due to the need for transplant organs that death has been so precisely defined.

The official signs of death include the following:

- no pupil reaction to light
- no response of the eyes to caloric (warm or cold) stimulation
- no jaw reflex (the jaw will react like the knee if hit with a reflex hammer)
- no gag reflex (touching the back of the throat induces vomiting)
- no response to **pain**
- no breathing
- a body temperature above 86°F (30°C), which eliminates the possibility of resuscitation following cold-water drowning
- no other cause for the above, such as a **head injury**
- no drugs present in the body that could cause apparent death
- all of the above for 12 hours
- all of the above for six hours and a flat-line electroencephalogram (brain wave study)
- no blood circulating to the brain, as demonstrated by **angiography**

Current ability to resuscitate people who have "died" has produced some remarkable stories. Drowning in cold water (under 50°F/10°C) so effectively slows metabolism that some persons have been revived after a half hour under water.

## Treatment

Only recently has there been concerted public effort to address the care of the dying in an effort to improve their comfort and lessen their alienation from those still living. Hospice care represents one of the greatest

## ELISABETH KÜBLER-ROSS (1926- )

Contemporary physician who has become a world authority on the subject of death and after-death states. Born in Switzerland on July 8, 1926, she worked as a country doctor before moving to the United States. During World War II she spent weekends at the Kantonsspital (Cantonai Hospital) in Zürich, where she volunteered to assist escaped refugees. After the war she visited Majdanek concentration camp, where the horrors of the death chambers stimulated in her a desire to help people facing death and to understand the human impulses of love and destruction. She extended her medical background by becoming a practicing psychiatrist. Her formal work with dying patients began in 1965 when she was a faculty member at the University of Chicago. She also conducted research on basic questions concerning life after death at the Manhattan State Hospital, New York. Her studies of death and dying have involved accounts by patients who reported out-of-the-body travel. Her research tends to show that while dying can be painful, death itself is a peaceful condition. Her 1969 text, *On Death and Dying*, was hailed by her colleagues and also became a popular best-seller.

In 1978 Kübler-Ross helped to found Shanti Nilaya (Final Home of Peace), a healing and growth center in Escondido, California. This was an extension of her well-known "Life-Death and Transition" workshops conducted in various parts of the United States and Canada, involving physicians, nurses, social workers, laypeople, and terminally ill patients. Much of Kübler-Ross's later research was directed toward proving the existence of life after death. Her publication *To Live Until We Say Good-bye* (1979) was both praised as a "celebration of life" and criticized as "prettifying" the real situation. She has also dealt with issues such as AIDS and "near death" experiences. In the mid-1980s Shanti Nilaya moved from San Diego County, California, to Head Waters, Virginia, where it continues to offer courses and short- and long-term therapeutic sessions.

advances made in this direction. There has also been a liberalization of the use of narcotics and other drugs for symptomatic relief and improvement in the quality of life for the dying.

### *Living will*

One of the most difficult issues surrounding death in the era of technology is that there is now a choice, not of the event itself, but of its timing. When to die, and more often, when to let a loved one die, is coming within people's power to determine. This is both a blessing and a dilemma. Insofar as the decision can be made

## KEY TERMS

**Angiography**—X rays of blood vessels filled with a contrast agent.

**Caloric testing**—Flushing warm and cold water into the ear stimulates the labyrinth and causes vertigo and nystagmus if all the nerve pathways are intact.

**Electroencephalogram**—Recording of electrical activity in the brain.

**Hospice**—Systematized care of dying persons.

**Living will**—A legal document detailing a person's wishes during the end of life, to be carried out by designated decision makers.

**Stroke**—Interruption of blood flow to a part of the brain with consequent brain damage, also known as a cerebrovascular accident (CVA).

ahead of time, a living will is an attempt to address this dilemma. By outlining the conditions under which one would rather be allowed to die, a person can contribute significantly to that final decision, even if not competent to do so at the time of actual death. The problem is that there are uncertainties surrounding every severely ill person. Each instance presents a greater or lesser chance of survival. The chance is often greater than zero. The best living will follows an intimate discussion with decision makers covering the many possible scenarios surrounding the end of life. This discussion is difficult, for few people like to contemplate their own demise. However, the benefits of a living will are substantial, both to physicians and to loved ones who are faced with making final decisions. Most states have passed living will laws, honoring instructions on artificial **life support** that were made while a person was still mentally competent.

### Euthanasia

Another issue that has received much attention is assisted suicide (euthanasia). In 1997, the State of Oregon placed the issue on the ballot, amid much consternation and dispute. Perhaps the main reason euthanasia has become front page news is because Dr. Jack Kevorkian, a pathologist from Michigan, is one of its most vocal advocates. The issue highlights the many new problems generated by increasing ability to intervene effectively in the final moments of life and unnaturally prolong the process of dying. The public appearance of euthanasia has also stimulated discussion about more compassionate care of the dying.

## Prevention

**Autopsy** after death is a way to precisely determine a cause of death. The word autopsy is derived from Greek meaning to see with one's own eyes. A pathologist extensively examines a body and submits a detailed report to an attending physician. Although an autopsy can do nothing for an individual after death, it can benefit the family and, in some cases, medical science. Hereditary disorders and disease may be found. This knowledge could be used to prevent illness in other family members. Information culled from an autopsy can be used to further medical research. The link between **smoking** and lung cancer was confirmed from data gathered through autopsy. Early information about **AIDS** was also compiled through autopsy reports.

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### ORGANIZATIONS

- American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211-2672. (913) 906-6000. <<http://www.aafp.org>>.  
 American Medical Association. 515 N. State Street, Chicago, IL 60610. (312) 464-5000. <<http://www.ama-assn.org>>.

American Society of Clinical Pathologists. 2100 West Harrison Street, Chicago, IL 60612. (312) 738-1336. <<http://www.ascp.org/index.asp>>.

College of American Pathologists. 325 Waukegan Road, Northfield, IL 60093. (800) 323-4040. <<http://www.cap.org>>.

Hospice Foundation of America. 2001 S St. NW Suite 300, Washington, DC 20009. (800) 854-3402. <<http://www.hospicefoundation.org>>.

#### OTHER

*American Association of Retired Persons.* <<http://www.aarp.org>>. *Association for Death Education and Counseling.*

<<http://www.adec.org>>.

*Death and Dying Grief Support.* <<http://www.death-dying.com>>.

*National Center for Health Statistics.* <<http://www.cdc.gov/nchs>>.

L. Fleming Fallon, Jr., MD, DrPH

## Debridement

### Definition

Debridement is the process of removing non-living tissue from pressure ulcers, **burns**, and other **wounds**.

### Purpose

Debridement speeds the healing of pressure ulcers, burns, and other wounds. Wounds that contain non-living (necrotic) tissue take longer to heal. The necrotic tissue may become colonized with bacteria, producing an unpleasant odor. Though the wound is not necessarily infected, the bacteria can cause inflammation and strain the body's ability to fight infection. Necrotic tissue may also hide pockets of pus called abscesses. Abscesses can develop into a general infection that may lead to **amputation** or **death**.

### Precautions

Not all wounds need debridement. Sometimes it is better to leave a hardened crust of dead tissue, called an eschar, than to remove it and create an open wound, particularly if the crust is stable and the wound is not inflamed. Before performing debridement, the physician will take a medical history with attention to factors that might complicate healing, such as medications being taken and **smoking**. The physician will also note the cause of the wound and the ways it has been treated. Some ulcers and other wounds occur in places where blood flow is impaired, for example, the foot ulcers that can accompany **diabetes mellitus**. In such cases, the physician or nurse

may decide not to debride the wound because blood flow may be insufficient for proper healing.

### Description

In debridement, dead tissue is removed so that the remaining living tissue can adequately heal. Dead tissue exposed to the air will form a hard black crust, called an eschar. Deeper tissue will remain moist and may appear white, or yellow and soft, or flimsy. The four major debridement techniques are surgical, mechanical, chemical, and autolytic.

#### Surgical debridement

Surgical debridement (also known as sharp debridement) uses a scalpel, scissors, or other instrument to cut dead tissue from a wound. It is the quickest and most efficient method of debridement. It is the preferred method if there is rapidly developing inflammation of the body's connective tissues (**cellulitis**) or a more generalized infection (**sepsis**) that has entered the bloodstream. The procedure can be performed at a patient's bedside. If the target tissue is deep or close to another organ, however, or if the patient is experiencing extreme **pain**, the procedure may be done in an operating room. Surgical debridement is generally performed by a physician, but in some areas of the country an advance practice nurse or physician assistant may perform the procedure.

The physician will begin by flushing the area with a saline (salt water) solution, and then will apply a topical anesthetic gel to the edges of the wound to minimize pain. Using a forceps to grip the dead tissue, the physician will cut it away bit by bit with a scalpel or scissors. Sometimes it is necessary to leave some dead tissue behind rather than disturb living tissue. The physician may repeat the process again at another session.

#### Mechanical debridement

In mechanical debridement, a saline-moistened dressing is allowed to dry overnight and adhere to the dead tissue. When the dressing is removed, the dead tissue is pulled away too. This process is one of the oldest methods of debridement. It can be very painful because the dressing can adhere to living as well as nonliving tissue. Because mechanical debridement cannot select between good and bad tissue, it is an unacceptable debridement method for clean wounds where a new layer of healing cells is already developing.

#### Chemical debridement

Chemical debridement makes use of certain enzymes and other compounds to dissolve necrotic tissue. It is more

## KEY TERMS

**Eschar**—A hardened black crust of dead tissue that may form over a wound.

**Pressure ulcer**—Also known as a decubitus ulcer, pressure ulcers are open wounds that form whenever prolonged pressure is applied to skin covering bony outcrops of the body. Patients who are bedridden are at risk of developing pressure ulcers. Pressure ulcers are commonly known as bedsores.

**Sepsis**—A severe systemic infection in which bacteria have entered the blood stream.

selective than mechanical debridement. In fact, the body makes its own enzyme, collagenase, to break down collagen, one of the major building blocks of skin. A pharmaceutical version of collagenase is available and is highly effective as a debridement agent. As with other debridement techniques, the area first is flushed with saline. Any crust of dead tissue is etched in a cross-hatched pattern to allow the enzyme to penetrate. A topical antibiotic is also applied to prevent introducing infection into the bloodstream. A moist dressing is then placed over the wound.

### *Autolytic debridement*

Autolytic debridement takes advantage of the body's own ability to dissolve dead tissue. The key to the technique is keeping the wound moist, which can be accomplished with a variety of dressings. These dressings help to trap wound fluid that contains growth factors, enzymes, and immune cells that promote wound healing. Autolytic debridement is more selective than any other debridement method, but it also takes the longest to work. It is inappropriate for wounds that have become infected.

### **Preparation**

The physician or nurse will begin by assessing the need for debridement. The wound will be examined, frequently by inserting a gloved finger into the wound to estimate the depth of dead tissue and evaluate whether it lies close to other organs, bone, or important body features. The area may be flushed with a saline solution before debridement begins, and a topical anesthetic gel or injection may be applied if surgical or mechanical debridement is being performed.

### **Aftercare**

After surgical debridement, the wound will be packed with a dry dressing for a day to control bleeding.

Afterward, moist dressings are applied to promote wound healing. Moist dressings are also used after mechanical, chemical, and autolytic debridement. Many factors contribute to wound healing, which frequently can take considerable time. Debridement may need to be repeated.

### **Risks**

It is possible that underlying tendons, blood vessels or other structures will be damaged during the examination of the wound and during surgical debridement. Surface bacteria may also be introduced deeper into the body, causing infection.

### **Normal results**

Removal of dead tissue from pressure ulcers and other wounds speeds healing. Although these procedures cause some pain, they are generally well tolerated by patients and can be managed more aggressively. It is not uncommon to debride a wound again in a subsequent session.

### **Resources**

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#### **ORGANIZATIONS**

American Academy of Wound Management. 1255 23rd St., NW, Washington, DC 20037. (202) 521-0368. <<http://www.aawm.org>>.

Wound Care Institute. 1100 N.E. 163rd Street, Suite #101, North Miami Beach, FL 33162. (305) 919-9192. <<http://woundcare.org>>.

Richard H. Camer

## Decompression sickness

### **Definition**

Decompression sickness (DCS) is a dangerous and occasionally lethal condition caused by nitrogen bubbles that form in the blood and other tissues of scuba divers who surface too quickly.

## Description

According to the Divers Alert Network (DAN), a worldwide organization devoted to safe-diving research and promotion, less than 1% of divers fall victim to DCS or the rarer bubble problem called **gas embolism**, **air embolism**, or arterial gas embolism (AGE). A study of the United States military community in Okinawa, where tens of thousands of sport and military dives are made each year, identified 84 DCS and 10 AGE cases in 1989–95, including nine deaths. This translated into estimates of one case in every 7,400 dives and one **death** in every 76,900 dives. DCS symptoms can be quite mild, however, and many cases certainly go unnoticed by divers.

At times the terminology adopted by writers on DCS can be confusing. Some substitute the term decompression illness (DCI) for DCS. Others treat DCI as a label encompassing both DCS and AGE. An older term for DCS is caisson disease, coined in the nineteenth century when it was discovered that bridge construction crews working at the bottom of lakes and rivers in large pressurized enclosures (caissons) were experiencing joint **pain** (a typical DCS symptom) on returning to the surface.

## Causes and symptoms

The air we breathe is mostly a mixture of two gases, nitrogen (78%) and oxygen (21%). Unlike oxygen, nitrogen is a biologically inert gas, meaning that it is not metabolized (converted into other substances) by the body. For this reason, most of the nitrogen we inhale is expelled when we exhale, but some is dissolved into the blood and other tissues. During a dive, however, the lungs take in more nitrogen than usual. This happens because the surrounding water pressure is greater than the air pressure at sea level (twice as great at 33 ft [10 m], for instance). As the water pressure increases, so does the pressure of the nitrogen in the compressed air inhaled by the diver. Because increased pressure causes an increase in gas density, the diver takes in more nitrogen with each breath than he or she would at sea level. Instead of being exhaled, however, the extra nitrogen safely dissolves into the tissues, where it remains until the diver begins his or her return to the surface (under some circumstances the extra nitrogen can cause **nitrogen narcosis**, but that condition is distinct from DCS). On the way up, decompression occurs (in other words, the water pressure drops), and with the change in pressure, the extra nitrogen gradually diffuses out of the tissues and is delivered by the bloodstream to the lungs, which expel it from the body. If the diver surfaces too quickly, however, potentially dangerous nitrogen bubbles can form in the tissues and cause DCS. These bubbles can compress nerves, obstruct arter-

ies, veins, and lymphatic vessels, and trigger harmful chemical reactions in the blood. The precise reasons for bubble formation remain unclear.

How much extra nitrogen enters the tissues varies with the dive's depth and duration. Dive tables prepared by the U.S. Navy and other organizations specify how long most divers can safely remain at a particular depth. If the dive table limits are exceeded, the diver must pause on the way up to allow the nitrogen to diffuse into the bloodstream without forming bubbles; these pauses are called decompression stops, and are carefully calibrated. DCS can occur, however, even when a diver obeys safe diving rules. In such cases, the predisposing factors include **fatigue**, **obesity**, **dehydration**, **hypothermia**, and recent alcohol use. As well, people who fly or travel to high-altitude locations without letting 12–24 hours pass after their last dive are at risk for DCS as well because their bodies undergo further decompression. This is true even when flying in commercial aircraft. Many travelers are unaware that to save money on fuel the cabin pressure in commercial aircraft is set much lower than the pressure at sea level. At 30,000 ft (9,144 m), for instance, cabin pressure is usually equivalent to the pressure at 7,000–8,000 ft (2,133–2,438 m) above sea level, a safe setting for everyone but recent divers. Exactly how long a diver should wait before flying or traveling to a high-altitude location depends on how much diving he or she has done and other considerations. If there is uncertainty about the appropriate waiting period, the sensible course of action is to let the full 24 hours pass.

Because the nitrogen bubbles that cause DCS can affect any of the body's tissues, including the blood, bones, nerves, and muscles, many kinds of symptoms are possible. Symptoms can appear minutes after a diver surfaces, and in about 80% of cases do so within eight hours. Pain is often the only symptom; this is sometimes called the bends, although many people incorrectly use that term as a synonym for DCS itself. The pain, which ranges from mild to severe, is usually limited to the joints, but can be felt anywhere. Severe **itching** (pruritis), **skin rashes**, and skin mottling (cutis marmorata) are other possible symptoms. All of these are sometimes classified as manifestations of type 1 or "mild" DCS. Type 2 or "serious" DCS can lead, among other things, to **paralysis**, brain damage, heart attacks, and death. Many DCS victims, however, experience both type 1 and type 2 symptoms.

## Diagnosis

Diagnosis requires taking a medical history (questioning the patient about his or her health and recent activities) and conducting a **physical examination**.

## KEY TERMS

**Gas embolism**—The presence of a gas bubble in the bloodstream that obstructs circulation.

**Hyperbaric chamber**—A sealed compartment in which air pressure is gradually increased and then gradually decreased, allowing nitrogen bubbles to shrink and the nitrogen to safely diffuse out of body tissue.

**Lymphatic vessels**—Vessels that carry a fluid called lymph from the tissues to the bloodstream.

**Nitrogen narcosis**—Also called “rapture of the deep,” the condition is caused by increased nitrogen pressure at depth and is characterized by symptoms similar to alcohol intoxication.

### Treatment

DCS is treated by giving the patient oxygen and placing him or her in a hyperbaric chamber, an enclosure in which the air pressure is first gradually increased and then gradually decreased. This shrinks the bubbles and allows the nitrogen to safely diffuse out of the tissues. Hyperbaric chamber facilities exist throughout the United States. No matter how mild one's symptoms may appear, immediate transportation to a facility is essential. Treatment is necessary even if the symptoms clear up before the facility is reached, because bubbles may still be in the bloodstream and pose a threat. DAN maintains a list of facilities and a 24-hour hotline that can provide advice on handling DCS and other diving emergencies.

### Prognosis

DCS sufferers who undergo chamber treatment within a few hours of symptom onset usually enjoy a full recovery. If treatment is delayed the consequences are less predictable, although many people have been helped even after several days have passed. A 1992 DAN report on diving accidents indicated that full recovery following chamber treatment was immediate for about 50% of divers. Some people, however, suffer numbness, tingling, or other symptoms that last weeks, months, or even a lifetime. In the Okinawa study, six of the 94 patients experienced “long-lasting” symptoms even after repeated chamber treatments.

### Prevention

The obvious way to minimize the risk of falling victim to DCS is to follow the rules on safe diving and air

travel after a dive. People who are obese, suffer from lung or heart problems, or are otherwise in poor health should not dive. And because the effect of nitrogen diffusion on the fetus remains unknown, diving while pregnant is not recommended.

### Resources

#### BOOKS

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Clenney, Timothy L., and Lorenz F. Lassen. “Recreational Scuba Diving Injuries.” *American Family Physician* (Apr. 1996): 1761+.

#### ORGANIZATIONS

American College of Hyperbaric Medicine. P.O. Box 25914-130, Houston, Texas 77265. (713) 528-0657. <<http://www.hyperbaricmedicine.org>>.

Divers Alert Network. The Peter B. Bennett Center, 6 West Colony Place, Durham, NC 27705. (800) 446-2671. <<http://www.diversaltnetwork.org>>.

Undersea and Hyperbaric Medical Society. 10531 Metropolitan Ave., Kensington, MD 20895. (301) 942-2980. <<http://www.uhms.org>>.

Howard Baker

## Decongestants

### Definition

Decongestants are medicines used to relieve nasal congestion (stuffy nose).

### Purpose

A congested or stuffy nose is a common symptom of colds and **allergies**. This congestion results when membranes lining the nose become swollen. Decongestants relieve the swelling by narrowing the blood vessels that supply the nose. This reduces the blood supply to the swollen membranes, causing the membranes to shrink.

These medicines do not cure colds or reverse the effects of histamines—chemicals released as part of the

allergic reaction. They will not relieve all of the symptoms associated with colds and allergies, only the stuffiness.

When considering whether to use a decongestant for cold symptoms, keep in mind that most colds go away with or without treatment and that taking medicine is not the only way to relieve a stuffy nose. Drinking hot tea or broth or eating chicken soup may help. There are also adhesive strips can be placed on the nose to help widen the nasal passages, making breathing through the nasal passages a bit easier when congestion is present.

## Precautions

Decongestant nasal sprays and nose drops may cause a problem called rebound congestion if used repeatedly over several days. When this happens, the nose remains stuffy or gets worse with every dose. The only way to stop the cycle is to stop using the drug. The stuffiness should then go away within about a week. Anyone who shows signs of severe rebound congestion should also contact his or her physician.

Do not use decongestant nasal sprays for more than three days. Decongestants taken by mouth should not be used for more than seven days. If the congestion has not gone away in this time, or if the symptoms are accompanied by **fever**, call a physician.

Do not use a decongestant nasal spray after the product's expiration date. If the product has become cloudy or discolored, throw it away and do not use it. Do not share droppers or spray bottles with anyone else, as this could spread infection. Do not let droppers and bottle tips touch countertops or other surfaces.

Some decongestants cause drowsiness. People 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.

In general, older people may be more sensitive to the effects of decongestants and may need to take lower doses to avoid side effects. People in this age group should not take long-acting (extended release) forms of decongestants unless they have previously taken a short-acting form with no ill effects.

Children may also be more sensitive to the effects of decongestants. Before giving any decongestant to a child, check the package label carefully. Some of these medicines are too strong for use in children. Serious side effects are possible if they are given large amounts of these drugs or if they swallow nose drops, nasal spray or eye drops. If this happens, call a physician or poison center immediately.

## Special conditions

People with certain medical conditions or who are taking certain other medicines can have problems if they take decongestants. Before taking these drugs, be sure to let the physician know about any of these conditions:

**ALLERGIES.** Anyone who has had unusual reactions to decongestants in the past should let his or her physician know before these drugs or any similar drugs are prescribed. The physician should also be told about any allergies to foods, dyes, preservatives, or other substances.

**PREGNANCY.** In studies of laboratory animals, some decongestants have had unwanted effects on fetuses. However, it is not known whether such effects also occur in people. Women who are pregnant or who plan to become pregnant should check with their physicians before taking decongestants.

**BREASTFEEDING.** Some decongestants pass into breast milk and may have unwanted effects on nursing babies whose mothers take the drugs. Women who are breastfeeding should check with their physicians before using decongestants. If they need to take the medicine, it may be necessary to bottle feed the baby with formula while taking it.

**OTHER MEDICAL CONDITIONS.** Anyone with heart or blood vessel disease, high blood pressure, diabetes, **enlarged prostate**, or overactive thyroid should not take decongestants unless under a physician's supervision. The medicine can increase blood sugar in people with diabetes. It can be especially dangerous in people with high blood pressure, as it may increase blood pressure.

Before using decongestants, people with any of these medical problems should make sure their physicians are aware of their conditions:

- glaucoma
- history of mental illness

Decongestants may have a variety of side effects, and may also interact with other medications the patient is taking.

## Side effects

### DECONGESTANT NASAL SPRAYS AND NOSE DROPS.

The most common side effects from decongestant nasal sprays and nose drops are sneezing and temporary burning, stinging, or dryness. These effects are usually temporary and do not need medical attention. If any of the following side effects occur after using a decongestant nasal spray or nose drops, stop using the medicine immediately and call the physician:

- increased blood pressure
- **headache**
- fast, slow, or fluttery heartbeat
- nervousness
- **dizziness**
- nausea
- sleep problems

**DECONGESTANTS TAKEN BY MOUTH.** The most common side effects of decongestants taken by mouth are nervousness, restlessness, excitability, dizziness, drowsiness, headache, nausea, weakness, and sleep problems. Anyone who has these symptoms while taking decongestants should stop taking them immediately.

Patients who have these symptoms while taking decongestants should call the physician immediately:

- increased blood pressure
- fast, irregular, or fluttery heartbeat
- severe headache
- tightness or discomfort in the chest
- breathing problems
- fear or anxiety
- **hallucinations**
- trembling or shaking
- convulsions (seizures)
- pale skin
- painful or difficult urination

Other side effects may occur. Anyone who has unusual symptoms after taking a decongestant should get in touch with his or her physician.

#### *Interactions with other medicines*

Decongestants may interact with a variety of other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. Do not take decongestants at the same time as these drugs:

- Monoamine oxidase inhibitors (MAO inhibitors) such as phenelzine (Nardil) or tranylcypromine (Parnate), used to treat conditions including depression and **Parkinson's disease**. Do not take decongestants at the same time as a MAO inhibitor or within two weeks of stopping treatment with an MAO inhibitor unless a physician approves.
- Other products containing the same or other decongestants.

- Caffeine.

In addition, anyone who takes decongestants should let the physician know all other medicines he or she is taking. Among the drugs that may interact with decongestants are:

- tricyclic antidepressants such as imipramine (Tofranil) or desipramine (Norpramin)
- the antidepressant maprotiline (Ludiomil)
- amantadine (Symmetrel)
- amphetamines
- medicine to relieve **asthma** or other breathing problems
- methylphenidate (Ritalin)
- appetite suppressants
- other medicine for colds, sinus problems, hay fever or other allergies
- beta-blockers such as atenolol (Tenormin) and propranolol (Inderal)
- digitalis glycosides, used to treat heart conditions

The list above does not include every drug that may interact with decongestants. Be sure to check with a physician or pharmacist before combining decongestants with any other prescription or nonprescription (over-the-counter) medicine.

#### **Description**

Decongestants are sold in many forms, including tablets, capsules, caplets, gelcaps, liqui-caps, liquids, nasal sprays, and nose drops. These drugs are sometimes combined with other medicines in cold and allergy products designed to relieve several symptoms. Some decongestant products require a physician's prescription, but there are also many nonprescription (over-the-counter) products. Ask a physician or pharmacist about choosing an appropriate decongestant.

Commonly used decongestants include oxymetazoline (Afrin and other brands) and pseudoephedrine (Sudafed, Actifed, and other brands). The decongestant oxymetazoline is also used in some eye drops to relieve redness and **itching**.

The recommended dosage depends on the drug. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage, and always take the medicine exactly as directed. If using nonprescription (over-the-counter) types, follow the directions on the package label or ask a pharmacist for assistance. Never take larger or more frequent doses, and do not take the drug for longer than directed.

## KEY TERMS

- Fetus**—A developing baby inside the womb.
- Hallucination**—A false or distorted perception of objects, sounds, or events that seems real. Hallucinations usually result from drugs or mental disorders.

## Risks

Anyone considering taking a decongestant should take a close look at the labels of any already in their medicine cabinet. In 2000, the Food and Drug Administration prohibited over-the-counter sales of medicines containing the decongestant phenylpropanolamine. The medicine is associated with an increased risk of **stroke** in people ages 18 to 49, especially women. Many cold remedies contained this medicine. Contact a pharmacist if there is any question about the ingredients in a medication. Over-the-counter remedies containing phenylpropanolamine should be discarded.

## Normal results

The desired result when taking decongestants is the short-term relief of nasal congestion.

## Resources

### PERIODICALS

- “An Ingredient Under Fire: Drugmakers are Jittery After an FDA Panel Ruling.” *Newsweek* (October 30, 2000): 59.
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Deanna M. Swartout-Corbeil, RN

Decubitus ulcers see **Bedsores**

## Deep vein thrombosis

### Definition

Deep vein thrombosis (DVT) is a blood clot in a major vein, usually in the legs and/or pelvis.

## Description

Deep vein thrombosis is a common but difficult to detect illness that can be fatal if not treated effectively. According to the American Heart Association, more than two million Americans develop deep vein thrombosis annually. An estimated 600,000 of these develop **pulmonary embolism**, a potentially fatal complication where the blood clots break off and form pulmonary emboli, plugs that block the lung arteries. Sixty thousand people die of pulmonary **embolism** each year. Deep vein thrombosis is also called venous thromboembolism, **thrombophlebitis** or phlebothrombosis.

Deep vein thrombosis is a major complication in patients who have had **orthopedic surgery** or pelvic, abdominal, or **thoracic surgery**. Patients with **cancer** and other chronic illnesses (including **congestive heart failure**), as well as those who have suffered a recent myocardial infarction, are also at high risk for developing DVT. Deep vein thrombosis can be chronic, with recurrent episodes.

## Causes and symptoms

Deep vein thrombosis is caused by blood clots in blood vessels that form in veins where blood flow is sluggish or has been disturbed, in pockets in the calf's deep veins, or in veins that have been traumatized. Symptoms include swelling and tenderness of the calf or thigh, and possibly warmth. Only 23–50% of patients experience symptoms, so it's often “silent.” Some individuals and families have underlying clotting tendencies that can be tested for.

## Diagnosis

Deep vein thrombosis can be detected through **venography** and radionuclide venography, **Doppler ultrasonography**, and impedance plethysmography. Venography is the most accurate test, but it is not used much, because it is often painful, expensive, exposes the patient to radiation, and can cause reactions and complications. Venography identifies the location, extent, and degree of attachment of the blood clots, and enables the condition of the deep leg veins to be assessed. A contrast solution is injected into a foot vein through a catheter. The physician observes the movement of the solution through the vein with a fluoroscope while a series of x rays are taken. Venography takes 30–45 minutes and can be done in a physician's office, a laboratory, or a hospital. Radionuclide venography, in which a radioactive isotope is injected, is occasionally used, especially if a patient has had reactions to contrast solutions.



This illustration features a dissected human lower leg showing clot formation (thrombosis) along the length of a vein.  
(Custom Medical Stock Photo. Reproduced by permission.)

Doppler ultrasonography is usually the preferred procedure for detecting deep vein thrombosis. This technique uses sound waves to measure blood flow through leg veins and arteries. A blood pressure cuff is wrapped around the patient's ankle and a transducer with gel on it is placed over pulse points of the foot and lower leg. High-frequency sounds bounce off the soft tissue, and the echoes are converted into images on a monitor. It is very accurate in detecting clots above the knee that can become pulmonary embolisms. Usually performed in a physician's office or hospital outpatient diagnostic center, Doppler ultrasound usually takes 30–45 minutes.

Impedance plethysmography records changes in blood volume and vessel resistance. A blood pressure cuff is wrapped around the leg above the knee, four electrodes are placed near the knee and the ankle, and the cuff is inflated. How efficiently the veins return to normal is measured. Performed in a physician's office, it takes about 15 minutes.

## KEY TERMS

**Pulmonary embolism**—An obstruction of a blood vessel in the lungs, usually caused by a blood clot that blocks a coronary artery. Pulmonary embolism can be very serious and, in some cases, fatal.

**Thrombosis**—The development of a blood clot inside a blood vessel.

## Treatment

Deep vein thrombosis can be treated with drug therapy, bed rest, and gradient elastic stockings. Medications include anticoagulants that "thin" blood to prevent further growth of blood clots, as well as clot-dissolving drugs. Heparin is a common injectable anticoagulant, and is usually followed by coumadin tablets for at least three months. Bed rest with the patient's legs elevated is necessary until the condition improves. Gradient elastic stockings should then be worn, and standing for long periods of time avoided. In some cases, a filter is placed in the major vein (the inferior vena cava) to trap emboli or clots before they get to the heart and lungs.

## Alternative treatment

Deep vein thrombosis can be life-threatening and must be treated with conventional medical therapies. However, there are alternative therapies that can be used in conjunction with emergency treatments to dissolve the clot that help support the body and prevent recurrence. A trained alternative health care practitioner should be consulted due to the severity of this condition.

## Prognosis

In many cases, deep vein thrombosis can be successfully treated if diagnosed early.

## Prevention

Deep vein thrombosis can be prevented through prophylactic anticoagulant drugs and venous stasis prevention with gradient elastic stockings and intermittent pneumatic compression of the legs. High-risk patients often need to remain on anticoagulants like Coumadin indefinitely.

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Lori De Milto

Deer-fly fever see **Tularemia**



**Defibrillation by paddles.** (Photograph by Patricia Barber, RBP, Custom Medical Stock Photo. Reproduced by permission.)

defibrillation is crucial to the patient's survival, the American Heart Association has called for the integration of defibrillation into an effective emergency cardiac care system. The system should include early access, early **cardiopulmonary resuscitation**, early defibrillation, and early advanced cardiac care.

Defibrillators deliver a brief electric shock to the heart, which enables the heart's natural pacemaker to regain control and establish a normal heart rhythm. The defibrillator is an electronic device with electrocardiogram leads and paddles. During defibrillation, the paddles are placed on the patient's chest, caregivers stand back, and the electric shock is delivered. The patient's pulse and heart rhythm are continually monitored. Medications to treat possible causes of the abnormal heart rhythm may be administered. Defibrillation continues until the patient's condition stabilizes or the procedure is ordered to be discontinued.

Early defibrillators, about the size and weight of a car battery, were used primarily in ambulances and hospitals. The American Heart Association now advocates public access defibrillation; this calls for placing automated external defibrillators (AEDs) in police vehicles, airplanes, and at public events, etc. The AEDs are smaller, lighter, less expensive, and easier to use than the early defibrillators. They are computerized to provide simple, verbal instructions to the operator and to make it impossible to deliver a shock to a patient whose heart is not fibrillating. The placement of AEDs is likely to expand to many public locations.

## Defibrillation

### Definition

Defibrillation is a process in which an electronic device sends an electric shock to the heart to stop an extremely rapid, irregular heartbeat, and restore the normal heart rhythm.

### Purpose

Defibrillation is performed to correct life-threatening fibrillations of the heart, which could result in cardiac arrest. It should be performed immediately after identifying that the patient is experiencing a cardiac emergency, has no pulse, and is unresponsive.

### Precautions

Defibrillation should not be performed on a patient who has a pulse or is alert, as this could cause a lethal heart rhythm disturbance or cardiac arrest. The paddles used in the procedure should not be placed on a woman's breasts or over a pacemaker.

### Description

Fibrillations cause the heart to stop pumping blood, leading to brain damage and/or cardiac arrest. About 10% of the ability to restart the heart is lost with every minute that the heart stays in fibrillation. **Death** can occur in minutes unless the normal heart rhythm is restored through defibrillation. Because immediate

### Preparation

After help is called for, cardiopulmonary resuscitation (**CPR**) is begun and continued until the caregivers arrive and set up the defibrillator. Electrocardiogram

## KEY TERMS

**Cardiac arrest**—A condition in which the heart stops functioning. Fibrillation can lead to cardiac arrest if not corrected quickly.

**Fibrillation**—Very rapid contractions or twitching of small muscle fibers in the heart.

**Pacemaker**—A surgically implanted electronic device that sends out electrical impulses to regulate a slow or erratic heartbeat.

leads are attached to the patient's chest. Gel or paste is applied to the defibrillator paddles, or two gel pads are placed on the patient's chest. The caregivers verify lack of a pulse, and select a charge.

### Aftercare

After defibrillation, the patient's cardiac status, breathing, and vital signs are monitored until he or she is stable. Typically, this monitoring takes place after the patient has been removed to an intensive care or cardiac care unit in a hospital. An electrocardiogram and **chest x ray** are taken. The patient's skin is cleansed to remove gel or paste, and, if necessary, ointment is applied to **burns**. An intravenous line provides additional medication, as needed.

### Risks

Skin burns from the defibrillator paddles are the most common complication of defibrillation. Other risks include injury to the heart muscle, abnormal heart rhythms, and blood clots.

### Resources

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Lori De Milto

Definitive cancer therapy see **Cancer therapy, definitive**

Degenerative arthritis see **Osteoarthritis**

## Dehydration

### Definition

Dehydration is the loss of water and salts essential for normal body function.

### Description

Dehydration occurs when the body loses more fluid than it takes in. This condition can result from illness; a hot, dry climate; prolonged exposure to sun or high temperatures; not drinking enough water; and overuse of **diuretics** or other medications that increase urination. Dehydration can upset the delicate fluid-salt balance needed to maintain healthy cells and tissues.

Water accounts for about 60% of a man's body weight. It represents about 50% of a woman's weight. Young and middle-aged adults who drink when they're thirsty do not generally have to do anything more to maintain their body's fluid balance. Children need more water because they expend more energy, but most children who drink when they are thirsty get as much water as their systems require.

### Age and dehydration

Adults over the age of 60 who drink only when they are thirsty probably get only about 90% of the fluid they need. Developing a habit of drinking only in response to the body's thirst signals raises an older person's risk of becoming dehydrated. Seniors who have relocated to areas where the weather is warmer or drier than the climate they are accustomed to are even likelier to become dehydrated unless they make it a practice to drink even when they are not thirsty.

Dehydration in children usually results from losing large amounts of fluid and not drinking enough water to replace the loss. This condition generally occurs in children who have stomach flu characterized by vomiting and **diarrhea**, or who can not or will not take enough fluids to compensate for excessive losses associated with **fever** and sweating of acute illness. An infant can become dehydrated only hours after becoming ill. Dehydration is a major cause of infant illness and **death** throughout the world.

### *Types of dehydration*

Mild dehydration is the loss of no more than 5% of the body's fluid. Loss of 5–10% is considered moderate dehydration. Severe dehydration (loss of 10–15% of body fluids) is a life-threatening condition that requires immediate medical care.

### *Complications of dehydration*

When the body's fluid supply is severely depleted, **hypovolemic shock** is likely to occur. This condition, which is also called physical collapse, is characterized by pale, cool, clammy skin; rapid heartbeat; and shallow breathing.

Blood pressure sometimes drops so low it can not be measured, and skin at the knees and elbows may become blotchy. **Anxiety**, restlessness, and thirst increase. After the patient's temperature reaches 107°F (41.7°C) damage to the brain and other vital organs occurs quickly.

### **Causes and symptoms**

Strenuous activity, excessive sweating, high fever, and prolonged vomiting or diarrhea are common causes of dehydration. So are staying in the sun too long, not drinking enough fluids, and visiting or moving to a warm region where it doesn't often rain. Alcohol, **caffeine**, and diuretics or other medications that increase the amount of fluid excreted can cause dehydration.

Reduced fluid intake can be a result of:

- appetite loss associated with acute illness
- excessive urination (polyuria)
- nausea
- bacterial or viral infection or inflammation of the pharynx (pharyngitis)
- inflammation of the mouth caused by illness, infection, irritation, or vitamin deficiency (**stomatitis**)

Other conditions that can lead to dehydration include:

- disease of the adrenal glands, which regulate the body's water and salt balance and the function of many organ systems
- diabetes mellitus
- eating disorders
- kidney disease
- chronic lung disease

An infant who does not wet a diaper in an eight-hour period is dehydrated. The soft spot on the baby's head (fontanel) may be depressed. Symptoms of dehydration at any age include cracked lips, dry or sticky mouth, lethargy, and sunken eyes. A person who is dehydrated cries without shedding tears and does not urinate very often. The skin is less elastic than it should be and is slow to return to its normal position after being pinched.

Dehydration can cause confusion, **constipation**, discomfort, drowsiness, fever, and thirst. The skin turns pale and cold, the mucous membranes lining the mouth and nose lose their natural moisture. The pulse sometimes races and breathing becomes rapid. Significant fluid loss can cause serious neurological problems.

### **Diagnosis**

The patient's symptoms and medical history usually suggest dehydration. **Physical examination** may reveal shock, rapid heart rate, and/or low blood pressure. Laboratory tests, including blood tests (to check electrolyte levels) and urine tests (e.g., urine specific gravity and creatinine), are used to evaluate the severity of the problem. Other laboratory tests may be ordered to determine the underlying condition (such as diabetes or an adrenal gland disorder) causing the dehydration.

### **Treatment**

Increased fluid intake and replacement of lost electrolytes are usually sufficient to restore fluid balances in patients who are mildly or moderately dehydrated. For individuals who are mildly dehydrated, just drinking plain water may be all the treatment that is needed. Adults who need to replace lost electrolytes may drink sports beverages (e.g., Gatorade or Recharge) or consume a little additional salt. Parents should follow label instructions when giving children Pedialyte or other commercial products recommended to relieve dehydration. Children who are dehydrated should receive only clear fluids for the first 24 hours.

A child who is vomiting should sip one or two teaspoons of liquid every 10 minutes. A child who is less

than a year old and who is not vomiting should be given one tablespoon of liquid every 20 minutes. A child who is more than one year old and who is not vomiting should take two tablespoons of liquid every 30 minutes. A baby who is being breast-fed should be given clear liquids for two consecutive feedings before breastfeeding is resumed. A bottle-fed baby should be given formula diluted to half its strength for the first 24 hours after developing symptoms of dehydration.

In order to accurately calculate fluid loss, it's important to chart weight changes every day and keep a record of how many times a patient vomits or has diarrhea. Parents should note how many times a baby's diaper must be changed.

Children and adults can gradually return to their normal diet after they have stopped vomiting and no longer have diarrhea. Bland foods should be reintroduced first, with other foods added as the digestive system is able to tolerate them. Milk, ice cream, cheese, and butter should not be eaten until 72 hours after symptoms have disappeared.

#### **Medical care**

Severe dehydration can require hospitalization and intravenous fluid replacement. If an individual's blood pressure drops enough to cause or threaten the development of shock, medical treatment is usually required. A doctor should be notified whenever an infant or child exhibits signs of dehydration or a parent is concerned that a stomach virus or other acute illness may lead to dehydration.

A doctor should also be notified if:

- a child less than three months old develops a fever higher than 100°F (37.8°C)
- a child more than three months old develops a fever higher than 102°F (38.9°C)
- symptoms of dehydration worsen
- an individual urinates very sparingly or does not urinate at all during a six-hour period
- dizziness, listlessness, or excessive thirst occur
- a person who is dieting and using diuretics loses more than 3 lb (1.3 kg) in a day or more than 5 lb (2.3 kg) a week

When treating dehydration, the underlying cause must also be addressed. For example, if dehydration is caused by vomiting or diarrhea, medications may be prescribed to resolve these symptoms. Patients who are dehydrated due to diabetes, kidney disease, or adrenal gland disorders must receive treatment for these conditions as well as for the resulting dehydration.

## **KEY TERMS**

**Electrolytes**—Mineral salts, such as sodium and potassium, dissolved in body fluid.

#### **Alternative treatment**

Gelatin water can be substituted for electrolyte-replacement solutions. It is made by diluting a 3-oz package in a quart of water or by adding one-quarter teaspoon of salt and a tablespoon of sugar to a pint of water.

#### **Prognosis**

Mild dehydration rarely results in complications. If the cause is eliminated and lost fluid is replaced, mild dehydration can usually be cured in 24–48 hours.

Vomiting and diarrhea that continue for several days without adequate fluid replacement can be fatal. The risk of life-threatening complications is greater for young children and the elderly. However, dehydration that is rapidly recognized and treated has a good outcome.

#### **Prevention**

Patients who are vomiting or who have diarrhea can prevent dehydration by drinking enough fluid for their urine to remain the color of pale straw. Ensuring that patients always drink adequate fluids during an illness will help prevent dehydration. Infants and young children with diarrhea and vomiting can be given electrolyte solutions such as Pedialyte to help prevent dehydration. People who are not ill can maintain proper fluid balance by drinking several glasses of water before going outside on a hot day. It is also a good idea to avoid coffee and tea, which increase body temperature and water loss.

Patients should know whether any medication they are taking can cause dehydration and should get prompt medical care to correct any underlying condition that increases the risk of dehydration.

Other methods of preventing dehydration and ensuring adequate fluid intake include:

- eating more soup at mealtime
- drinking plenty of water and juice at mealtime and between meals
- keeping a glass of water nearby when working or relaxing

## Resources

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Maureen Haggerty

**Delavirdine** see **Non-nucleoside reverse transcriptase inhibitors**

by placing a series of adhesive patches on the skin containing potential allergens, or allergy-causing substances.

- To assess the vitality of the T cell response as part of the evaluation of immune system health in infection, **cancer**, immune disorders, pre-transplantation screening, **aging**, and **malnutrition**. DHT can help predict survival in immunocompromised patients, and evaluate the success of restorative therapy. Antigens used for these tests must be ones the patient has been exposed to before, and, therefore, include inactivated antigens from common infectious agents to which the patient might have been exposed, such as **mumps**, *Candida albicans*, **tetanus** toxoid, and trichophyton (a skin fungus).

## Precautions

No special precautions are necessary for most patients. Those with known hypersensitivity to certain skin irritants should alert the clinician performing the test. Some commercial preparations of fungal antigens contain mercury, a source of irritation to some patients.

## Description

The most accurate TB test is the Mantoux test, in which a small amount of TB antigen is injected into the skin. The area is examined 48–72 hours after the injection.

In the patch test, 20–30 adhesive patches are usually placed on the upper back. The patches are kept in place and the area is kept dry for 48 hours. The patches are then removed, and the skin is examined 24 hours afterward, and possibly again a day or more following that. Patch testing is usually performed following a patient complaint of skin irritation from an unknown substance. Testing may suggest several candidates; identifying the right one requires careful review of the patient’s possible exposure.

The test of overall T cell responsiveness is performed with several injections. Each area injected is circled and marked. Results are read 48 hours after the injection.

## Preparation

No special preparation is necessary.

## KEY TERMS

**Allergen**—A foreign substance that provokes an immune reaction in some sensitive people but not in most others.

**Anaphylaxis**—An exaggerated, life-threatening hypersensitivity reaction to a previously encountered antigen.

**Antibody**—An immune system protein made to fight infection.

**Antigen**—A foreign substance detected that provokes an immune reaction.

### Aftercare

Patches should be kept dry. Injection sites may be washed, but excessive rubbing should be avoided. Patches and injection sites may become reddened or irritated. If a patch causes severe **itching** or discomfort, the patient should remove it immediately.

### Risks

DHT is quite safe for virtually all people. There is no risk of infection from the agents injected, since they are purified antigens, not whole organisms. Life threatening, hypersensitive reactions (**anaphylaxis**) are a very small risk; patients should notify the administering physician immediately if signs of **wheezing**, swelling, or diffuse redness of the skin develops.

### Normal results

Absence of exposure to TB is indicated by absent or very little skin reaction; redness or hardness smaller than 5 mm (about 0.25 in) is considered normal for a person not exposed or infected with TB.

Patch test sites should be normal or only slightly red.

T cell responsiveness tests should be positive; that is, the injected areas should be reddened and hard. Two affected areas of 2 mm or more is considered a positive result.

### Abnormal results

TB exposure is indicated by a reaction of 10 mm or more. The degree of redness is not important. A 5–10 mm area could indicate exposure if there is an underlying risk to TB.

Patch test areas that become reddened and irritated indicate reaction to the substance in the patch.

Absence of any reaction to injected areas indicates lack of T cell responsiveness, a condition called anergy. T cell anergy is seen in immune deficiency diseases including **AIDS**, some cases of infectious diseases, malignancies, immunosuppressive therapy (including corticosteroid treatment), some autoimmune diseases, malnutrition, major surgery, and some viral immunizations.

### Resources

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Richard Robinson

## Delirium

### Definition

Delirium is a state of mental confusion that develops quickly and usually fluctuates in intensity.

### Description

Delirium is a syndrome, or group of symptoms, caused by a disturbance in the normal functioning of the brain. The delirious patient has a reduced awareness of and responsiveness to the environment, which may be manifested as disorientation, incoherence, and memory disturbance. Delirium is often marked by **hallucinations**, **delusions**, and a dream-like state.

Delirium affects at least one in 10 hospitalized patients, and is a common part of many terminal illnesses. Delirium is more common in the elderly than in the general population. While it is not a specific disease itself, patients with delirium usually fare worse than those with the same illness who do not have delirium.

### Causes and symptoms

#### Causes

There are a large number of possible causes of delirium. Metabolic disorders are the single most common cause, accounting for 20–40% of all cases. This type of delirium, termed “metabolic encephalopathy,” may result from organ failure, including liver or kidney failure. Other metabolic causes include **diabetes mellitus**, **hyperthyroidism** and **hypothyroidism**, vitamin defi-

ciencies, and imbalances of fluids and electrolytes in the blood. Severe **dehydration** can also cause delirium.

Drug intoxication (“intoxication confusional state”) is responsible for up to 20% of delirium cases, either from side effects, overdose, or deliberate ingestion of a mind-altering substance. Medicinal drugs with delirium as a possible side effect or result of overdose include:

- anticholinergics, including atropine, scopolamine, chlorpromazine (an antipsychotic), and diphenhydramine (an antihistamine)
- sedatives, including **barbiturates**, **benzodiazepines**, and ethanol (drinking alcohol)
- antidepressant drugs
- anticonvulsant drugs
- nonsteroidal anti-inflammatory drugs (NSAIDs), including ibuprofen and **acetaminophen**
- corticosteroids, including prednisone
- anticancer drugs, including methotrexate and procarbazine
- lithium
- cimetidine
- antibiotics
- L-dopa

Delirium may result from ingestion of legal or illegal psychoactive drugs, including:

- ethanol (drinking alcohol)
- marijuana
- LSD (**lysergic acid diethylamide**) and other hallucinogens
- amphetamines
- cocaine
- opiates, including heroin and morphine
- PCP (phencyclidine)
- inhalants

Drug withdrawal may also cause delirium. Delirium tremens, or “DTs,” may occur during alcohol withdrawal after prolonged or intense consumption. Withdrawal symptoms are also possible from many of the psychoactive prescription drugs.

Poisons may cause delirium (“toxic encephalopathy”), including:

- solvents, such as gasoline, kerosene, turpentine, benzene, and alcohols
- carbon monoxide
- refrigerants (Freon)

## KEY TERMS

**Dementia**—A loss of mental ability severe enough to interfere with functioning. While dementia and delirium have some of the same symptoms, dementia has a much slower onset.

**Electroencephalogram (EEG)**—A chart of the brain wave patterns picked up by electrodes placed on the scalp. This is useful for diagnosing central nervous system disorders.

**Encephalopathy**—A brain dysfunction or disorder.

- heavy metals, such as lead, mercury, and arsenic
- insecticides, such as Parathion and Sevin
- mushrooms, such as *Amanita* species
- plants such as jimsonweed (*Datura stramonium*) and morning glory (*Ipomoea* spp.)
- animal venoms

Other causes of delirium include:

- infection
- fever
- head trauma
- epilepsy
- brain hemorrhage or infarction
- brain tumor
- low blood oxygen (hypoxemia)
- high blood carbon dioxide (hypercapnia)
- post-surgical complication.

### Symptoms

The symptoms of delirium come on quickly, in hours or days, in contrast to those of **dementia**, which develop much more slowly. Delirium symptoms typically fluctuate through the day, with periods of relative calm and lucidity alternating with periods of florid delirium. The hallmark of delirium is a fluctuating level of consciousness. Symptoms may include:

- decreased awareness of the environment
- confusion or disorientation, especially of time
- memory impairment, especially of recent events
- hallucinations
- illusions and misinterpreted stimuli
- increased or decreased activity level

- mood disturbance, possibly including **anxiety**, euphoria or depression
- language or speech impairment

## Diagnosis

Delirium is diagnosed through the medical history and recognition of symptoms during **mental status examination**. The most important part of diagnosis is determining the cause of the delirium. Tests may include blood and urine analysis for levels of drugs, fluids, electrolytes, and blood gases, and to test for infection; lumbar puncture ("spinal tap") to test for central nervous system infection; x ray, **computed tomography scans** (CT), or **magnetic resonance imaging** (MRI) scans to look for tumors, hemorrhage, or other brain abnormalities; thyroid tests; **electroencephalography** (EEG); **electrocardiography** (ECG); and possibly others as dictated by the likely cause.

## Treatment

Treatment of delirium begins with recognizing and treating the underlying cause. Delirium itself is managed by reducing disturbing stimuli, or providing soothing ones; use of simple, clear language in communication; and reassurance, especially from family members. Physical restraints may be needed if the patient is a danger to himself or others, or if he insists on removing necessary medical equipment such as intravenous lines or monitors. Sedatives or **antipsychotic drugs** may be used to reduce anxiety, hallucinations, and delusions.

## Prognosis

Persons with delirium usually have a worse prognosis for the underlying disease than the person without delirium. Nonetheless, those without terminal illness usually recover from delirium. They may not, however, regain all their original cognitive abilities, and may be left with some permanent impairments, including **fatigue**, irritability, difficulty concentrating, or mood changes.

## Prevention

Prevention of delirium is focused on treating or avoiding its underlying causes. The most preventable forms are those induced by drugs. Strategies for reducing delirium include following prescriptions, consulting the prescribing physician immediately if symptoms occur, and consulting the physician before discontinuing the drug, even if it has been ineffective; avoiding intoxication with legal or illegal drugs, and seeking professional

assistance before suddenly discontinuing an addictive drug such as alcohol or heroin; maintaining good **nutrition**, which promotes general health and can minimize the likelihood of delirium from alcohol intoxication and withdrawal; and avoiding exposure to solvents, insecticides, heavy metals, or biological poisons in the home or workplace.

## Resources

### BOOKS

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Richard Robinson

Delta virus hepatitis see **Hepatitis D**

# Delusions

## Definition

A delusion is an unshakable belief in something untrue. These irrational beliefs defy normal reasoning, and remain firm even when overwhelming proof is presented to dispute them. Delusions are often accompanied by **hallucinations** and/or feelings of **paranoia**, which act to strengthen confidence in the delusion. Delusions are distinct from culturally or religiously based beliefs that may be seen as untrue by outsiders.

## Description

Delusions are a common symptom of several mood and personality-related mental illnesses, including **schizoaffective disorder**, **schizophrenia**, shared psychotic disorder, major depressive disorder, and **bipolar disorder**. They are also the major feature of delusional disorder. Individuals with delusional disorder suffer from long-term, complex delusions that fall into one of six categories: persecutory, grandiose, jealousy, erotomanic, somatic, or mixed. There are also delusional disorders such as **dementia** that clearly have organic or physical causes.

### Persecutory

Individuals with persecutory delusional disorder are plagued by feelings of paranoia and an irrational yet unshakable belief that someone is plotting against them, or out to harm them.

### *Grandiose*

Individuals with grandiose delusional disorder have an over-inflated sense of self-worth. Their delusions center on their own importance, such as believing that they have done or created something of extreme value or have a “special mission.”

### *Jealousy*

Jealous delusions are unjustified and irrational beliefs that an individual’s spouse or significant other has been unfaithful.

### *Eerotomaniac*

Individuals with erotomaniac delusional disorder believe that another person, often a stranger, is in love with them. The object of their affection is typically of a higher social status, sometimes a celebrity. This type of delusional disorder may lead to stalking or other potentially dangerous behavior.

### *Somatic*

Somatic delusions involve the belief that something is physically wrong with the individual. The delusion may involve a medical condition or illness or a perceived deformity. This condition differs from **hypochondriasis** in that the deformity is perceived as a fixed condition not a temporary illness.

### *Mixed*

Mixed delusions are those characterized by two or more of persecutory, grandiose, jealousy, erotomaniac, or somatic themes.

### **Causes and symptoms**

Some studies have indicated that delusions may be generated by abnormalities in the limbic system, the portion of the brain on the inner edge of the cerebral cortex that is believed to regulate emotions. The exact source of delusions has not been conclusively found, but potential causes include genetics, neurological abnormalities, and changes in brain chemistry. Delusions are also a known possible side effect of drug use and abuse (e.g., amphetamines, **cocaine**, PCP).

### **Diagnosis**

Patients with delusional symptoms should undergo a thorough **physical examination** and patient history to rule out possible organic causes (such as dementia). If a psychological cause is suspected, a mental health professional will typically conduct an interview with the

## KEY TERMS

**Hallucinations**—False or distorted sensory experiences that appear to be real perceptions.

**Paranoia**—An unfounded or exaggerated distrust of others.

**Shared psychotic disorder**—Also known as folie à deux; shared psychotic disorder is an uncommon disorder in which the same delusion is shared by two or more individuals.

patient and administer one of several clinical inventories, or tests, to evaluate mental status.

### **Treatment**

Delusions that are symptomatic of delusional disorder should be treated by a psychologist and/or psychiatrist. Though **antipsychotic drugs** are often not effective, antipsychotic medication such as thioridazine (Mellaril), haloperidol (Haldol), chlorpromazine (Thorazine), clozapine (Clozaril), or risperidone (Risperdal) may be prescribed, and cognitive therapy or psychotherapy may be attempted.

If an underlying condition such as schizophrenia, depression, or drug abuse is found to be triggering the delusions, an appropriate course of medication and/or psychosocial therapy is employed to treat the primary disorder. The medication, typically, will include an antipsychotic agent.

### **Prognosis**

Delusional disorder is typically a chronic condition, but with appropriate treatment, a remission of delusional symptoms occurs in up to 50% of patients. However, because of their strong belief in the reality of their delusions and a lack of insight into their condition, individuals with this disorder may never seek treatment, or may be resistant to exploring their condition in psychotherapy.

### **Resources**

#### **BOOKS**

American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*. 4th ed. Washington, DC: American Psychiatric Press, Inc., 1994.

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Siegel, Ronald K. *Whispers: The Voices of Paranoia*. New York: Crown, 1994.

**PERIODICALS**

Manschreck, Theo C. "Delusional Disorder: The Recognition and Management of Paranoia." *Journal of Clinical Psychiatry* 57, supplement 3 (1996): 32-38.

**ORGANIZATIONS**

American Psychiatric Association. 1400 K Street NW, Washington DC 20005. (888) 357-7924. <<http://www.psych.org>>.

American Psychological Association (APA). 750 First St. NE, Washington, DC 20002-4242. (202) 336-5700. <<http://www.apa.org>>.

National Alliance for the Mentally Ill (NAMI). Colonial Place Three, 2107 Wilson Blvd., Ste. 300, Arlington, VA 22201-3042. (800) 950-6264. <<http://www.nami.org>>.

National Institutes of Mental Health (NIMH). 6001 Executive Boulevard, Rm. 8184, MSC 9663

Paula Anne Ford-Martin

## Dementia

### Definition

Dementia is a loss of mental ability severe enough to interfere with normal activities of daily living, lasting more than six months, not present since birth, and not associated with a loss or alteration of consciousness.

### Description

Dementia is a group of symptoms caused by gradual **death** of brain cells. The loss of cognitive abilities that occurs with dementia leads to impairments in memory, reasoning, planning, and personality. While the overwhelming number of people with dementia are elderly, it is not an inevitable part of **aging**. Instead, dementia is caused by specific brain diseases. **Alzheimer's disease** (AD) is the most common cause, followed by vascular or multi-infarct dementia.

The prevalence of dementia has been difficult to determine, partly because of differences in definition among different studies, and partly because there is some normal decline in functional ability with age. Dementia affects 5–8% of all people between ages 65 and 74, and up to 20% of those between 75 and 84. Estimates for dementia in those 85 and over range from 30–47%. Between two and four million Americans have AD; that number is expected to grow to as many as 14 million by the middle of the twenty-first century as the population as a whole ages.

The cost of dementia can be considerable. While most people with dementia are retired and do not suffer income losses from their disease, the cost of care is often

enormous. Financial burdens include lost wages for family caregivers, medical supplies and drugs, and home modifications to ensure safety. Nursing home care may cost several thousand dollars a month or more. The psychological cost is not as easily quantifiable but can be even more profound. The person with dementia loses control of many of the essential features of his life and personality, and loved ones lose a family member even as they continue to cope with the burdens of increasing dependence and unpredictability.

### Causes and symptoms

#### Causes

Dementia is usually caused by degeneration in the cerebral cortex, the part of the brain responsible for thoughts, memories, actions and personality. Death of brain cells in this region leads to the cognitive impairment which characterizes dementia.

The most common cause of dementia is AD, accounting for half to three quarters of all cases. The brain of a person with AD becomes clogged with two abnormal structures, called neurofibrillary tangles and senile plaques. Neurofibrillary tangles are twisted masses of protein fibers inside nerve cells, or neurons. Senile plaques are composed of parts of neurons surrounding a group of proteins called beta-amyloid deposits. Why these structures develop is unknown. Current research indicates possible roles for inflammation, blood flow restriction, and toxic molecular fragments known as free radicals. Several genes have been associated with higher incidences of AD, although the exact role of these genes is still unknown.

Vascular dementia is estimated to cause from 5–30% of all dementias. It occurs from decrease in blood flow to the brain, most commonly due to a series of small strokes (multi-infarct dementia). Other cerebrovascular causes include: **vasculitis** from **syphilis**, **Lyme disease**, or **systemic lupus erythematosus**; **subdural hematoma**; and **subarachnoid hemorrhage**. Because of the usually sudden nature of its cause, the symptoms of vascular dementia tend to begin more abruptly than those of Alzheimer's dementia. Symptoms may progress stepwise with the occurrence of new strokes. Unlike AD, the incidence of vascular dementia is lower after age 75.

Other conditions which may cause dementia include:

- AIDS
- Parkinson's disease
- Lewy body disease
- Pick's disease

- Huntington's disease
- Creutzfeldt-Jakob disease
- brain tumor
- **hydrocephalus**
- head trauma
- multiple sclerosis
- prolonged abuse of alcohol or other drugs
- vitamin deficiency: thiamin, niacin, or B<sub>12</sub>
- hypothyroidism
- hypercalcemia

### Symptoms

Dementia is marked by a gradual impoverishment of thought and other mental activities. Losses eventually affect virtually every aspect of mental life. The slow progression of dementia is in contrast with **delirium**, which involves some of the same symptoms, but has a very rapid onset and fluctuating course with alteration in the level of consciousness. However, delirium may occur with dementia, especially since the person with dementia is more susceptible to the delirium-inducing effects of many types of drugs.

Symptoms include:

- Memory losses. Memory loss is usually the first symptom noticed. It may begin with misplacing valuables such as a wallet or car keys, then progress to forgetting appointments, where the car was left, and the route home, for instance. More profound losses follow, such as forgetting the names and faces of family members.
- Impaired abstraction and planning. The person with dementia may lose the ability to perform familiar tasks, to plan activities, and to draw simple conclusions from facts.
- Language and comprehension disturbances. The person may be unable to understand instructions, or follow the logic of moderately complex sentences. Later, he or she may not understand his or her own sentences, and have difficulty forming thoughts into words.
- Poor judgment. The person may not recognize the consequences of his or her actions or be able to evaluate the appropriateness of behavior. Behavior may become ribald, overly-friendly, or aggressive. Personal hygiene may be ignored.
- Impaired orientation ability. The person may not be able to identify the time of day, even from obvious visual clues; or may not recognize his or her location, even if familiar. This disability may stem partly from losses of memory and partly from impaired abstraction.

- Decreased attention and increased restlessness. This may cause the person with dementia to begin an activity and quickly lose interest, and to wander frequently. Wandering may cause significant safety problems, when combined with disorientation and memory losses. The person may begin to cook something on the stove, then become distracted and wander away while it is cooking.
- Personality changes and **psychosis**. The person may lose interest in once-pleasurable activities, and become more passive, depressed, or anxious. **Delusions**, suspicion, **paranoia**, and **hallucinations** may occur later in the disease. Sleep disturbances may occur, including **insomnia** and sleep interruptions.

### Diagnosis

Since dementia usually progresses slowly, diagnosing it in its early stages can be difficult. Several office visits over several months or more may be needed. Diagnosis begins with a thorough physical exam and complete medical history, usually including comments from family members or caregivers. A family history of either AD or cerebrovascular disease may provide clues to the cause of symptoms. Simple tests of mental function, including word recall, object naming, and number-symbol matching, are used to track changes in the person's cognitive ability.

Depression is common in the elderly and can be mistaken for dementia; therefore, ruling out depression is an important part of the diagnosis. Distinguishing dementia from the mild normal cognitive decline of advanced age is also critical. The medical history includes a complete listing of drugs being taken, since a number of drugs can cause dementia-like symptoms.

Determining the cause of dementia may require a variety of medical tests, chosen to match the most likely etiology. Cerebrovascular disease, hydrocephalus, and tumors may be diagnosed with x-rays, CT or MRI scans, and vascular imaging studies. Blood tests may reveal nutritional deficiencies or hormone imbalances.

### Treatment

Treatment of dementia begins with treatment of the underlying disease, where possible. The underlying causes of nutritional, hormonal, tumor-caused and drug-related dementias may be reversible to some extent. Treatment for stroke-related dementia begins by minimizing the risk of further strokes, through **smoking cessation**, **aspirin** therapy, and treatment of **hypertension**, for instance. There are no therapies that can reverse the progression of AD. Aspirin, estrogen,

vitamin E, and selegiline are currently being evaluated for their ability to slow the rate of progression.

Care for a person with dementia can be difficult and complex. The patient must learn to cope with functional and cognitive limitations, while family members or other caregivers assume increasing responsibility for the person's physical needs. In progressive dementias such as AD, the person may ultimately become completely dependent. Education of the patient and family early on in the disease progression can help them anticipate and plan for inevitable changes.

Symptoms of dementia may be treated with a combination of psychotherapy, environmental modifications, and medication. Drug therapy can be complicated by forgetfulness, especially if the prescribed drug must be taken several times daily.

Behavioral approaches may be used to reduce the frequency or severity of problem behaviors, such as aggression or socially inappropriate conduct. Problem behavior may be a reaction to frustration or overstimulation; understanding and modifying the situations that trigger it can be effective. Strategies may include breaking down complex tasks, such as dressing or feeding, into simpler steps, or reducing the amount of activity in the environment to avoid confusion and agitation. Pleasurable activities, such as crafts, games, and music, can provide therapeutic stimulation and improve mood.

Modifying the environment can increase safety and comfort while decreasing agitation. Home modifications for safety include removal or lock-up of hazards such as sharp knives, dangerous chemicals, and tools. Child-proof latches or Dutch doors may be used to limit access as well. Lowering the hot water temperature to 120°F (48.9°C) or less reduces the risk of scalding. Bed rails and bathroom safety rails can be important safety measures, as well. Confusion may be reduced with simpler decorative schemes and presence of familiar objects. Covering or disguising doors (with a mural, for example) may reduce the tendency to wander. Positioning the bed in view of the bathroom can decrease incontinence.

Two drugs, tacrine (Cognex) and donepezil (Aricept), are commonly prescribed for AD. These drugs inhibit the breakdown of acetylcholine in the brain, prolonging its ability to conduct chemical messages between brain cells. They provide temporary improvement in cognitive functions for about 40% of patients with mild to moderate AD. Hydergine is sometimes prescribed as well, though it is of questionable benefit for most patients.

Psychotic symptoms, including paranoia, delusions, and hallucinations, may be treated with **antipsychotic drugs**, such as haloperidol, chlorpromazine, risperidone,

and clozapine. Side effects of these drugs can be significant. **Antianxiety drugs** such as Valium may improve behavioral symptoms, especially agitation and **anxiety**, although BuSpar has fewer side effects. The anticonvulsant carbamazepine is also sometimes prescribed for agitation. Depression is treated with antidepressants, usually beginning with **selective serotonin reuptake inhibitors** (SSRIs) such as Prozac or Paxil, followed by **monoamine oxidase inhibitors** or tricyclic antidepressants. **Electroconvulsive therapy** may be appropriate for some patients with severe depression who are unresponsive to drug therapy. In general, medications should be administered very cautiously to demented patients, in the lowest possible effective doses, to minimize side effects. Supervision of taking medications is generally required.

Long-term institutional care may be needed for the person with dementia, as profound cognitive losses often precede death by a number of years. Early planning for the financial burden of nursing home care is critical. Useful information about financial planning for long-term care is available through the Alzheimer's Association.

Family members or others caring for a person with dementia are often subject to extreme **stress**, and may develop feelings of anger, resentment, guilt, and hopelessness, in addition to the sorrow they feel for their loved one and for themselves. Depression is an extremely common consequence of being a full-time caregiver for a person with dementia. Support groups can be an important way to deal with the stress of caregiving. The location and contact numbers for caregiver support groups are available from the Alzheimer's Association; they may also be available through a local social service agency or the patient's physician. Medical treatment for depression may be an important adjunct to group support.

## Alternative treatment

Several drugs are currently being tested for their ability to slow the progress of AD. These include acetyl-l-carnitine, which acts on the cellular energy structures known as mitochondria; propentofylline, which may aid circulation; milameline, which acts similarly to tacrine and donezepil; and ginkgo extract.

Ginkgo extract, derived from the leaves of the *Ginkgo biloba* tree, interferes with a circulatory protein called platelet activating factor. It also increases circulation and oxygenation to the brain. Ginkgo extract has been used for many years in China and is widely prescribed in Europe for treatment of circulatory problems. A 1997 study of patients with dementia seemed to show that ginkgo extract could improve their symptoms, though the study was criticized for certain flaws in its method.

## KEY TERMS

**Donepezil**—A drug commonly prescribed for Alzheimer's disease that provides temporary improvement in cognitive functions for some patients with mild-to-moderate forms of the disease.

**Ginkgo extract**—Made from the leaves of the *Ginkgo biloba* tree, this extract, used in other countries to treat circulatory problems, may improve the symptoms of patients with dementia.

**Neurofibrillary tangles**—Abnormal structures, composed of twisted masses of protein fibers within nerve cells, found in the brains of persons with Alzheimer's disease.

**Senile plaques**—Abnormal structures, composed of parts of nerve cells surrounding protein deposits, found in the brains of persons with Alzheimer's disease.

**Tacrine**—A drug commonly prescribed for Alzheimer's disease that provides temporary improvement in cognitive functions for some patients with mild-to-moderate forms of the disease.

## Prognosis

The prognosis for dementia depends on the underlying disease. On average, people with Alzheimer's disease live eight years past their diagnosis, with a range from one to 20 years. Vascular dementia is usually progressive, with death from stroke, infection, or heart disease.

## Prevention

There is no known way to prevent Alzheimer's disease, although several of the drugs under investigation may reduce its risk or slow its progression. The risk of developing multi-infarct dementia may be reduced by reducing the risk of stroke.

## Resources

### BOOKS

- Jacques, Alan. *Understanding Dementia*. New York: Churchill Livingstone, 1992.  
 Mace, Nancy L., and Peter V. Rabins. *The 36-Hour Day*. Baltimore: John Hopkins University Press, 1995.

### ORGANIZATIONS

- Alzheimer's Association. 919 North Michigan Ave., Suite 1000, Chicago, IL 60611. (800) 272-3900. <<http://www.alz.org>>.

Richard Robinson

Demyelinating disease see **Multiple sclerosis**

## Dengue fever

### Definition

Dengue fever is a disease caused by one of a number of viruses that are carried by mosquitoes. These mosquitoes then transmit the virus to humans.

### Description

The virus that causes dengue fever is called an arbovirus, which stands for arthropod-borne virus. Mosquitoes are a type of arthropod. In a number of regions, mosquitoes carry this virus and are responsible for passing it along to humans. These regions include the Middle East, the far East, Africa, and the Caribbean Islands. In these locations, the dengue fever arbovirus is endemic, meaning that the virus naturally and consistently lives in that location. The disease only shows up in the United States sporadically.

In order to understand how dengue fever is transmitted, several terms need to be defined. The word "host" means an animal (including a human) that can be infected with a particular disease. The word "vector" means an organism that can carry a particular disease-causing agent (like a virus or bacteria) without actually developing the disease. The vector can then pass the virus or bacteria on to a new host.

Many of the common illnesses in the United States (including the **common cold**, many viral causes of **diarrhea**, and **influenza** or "flu") are spread because the viruses that cause these illnesses can be passed directly from person to person. However, dengue fever cannot be passed directly from one infected person to another. Instead, the virus responsible for dengue fever requires an intermediate vector, a mosquito, that carries the virus from one host to another. The mosquito that carries the arbovirus responsible for dengue fever is the same type of mosquito that can transmit other diseases, including **yellow fever**. This mosquito is called *Aedes egypti*. The most common victims are children younger than 10 years of age.

### Causes and symptoms

Dengue fever can occur when a mosquito carrying the arbovirus bites a human, passing the virus on to the new host. Once in the body, the virus travels to various glands where it multiplies. The virus can then enter the

bloodstream. The presence of the virus within the blood vessels, especially those feeding the skin, causes changes to these blood vessels. The vessels swell and leak. The spleen and lymph nodes become enlarged, and patches of liver tissue die. A process called disseminated intravascular coagulation (DIC) occurs, where chemicals responsible for clotting are used up and lead to a risk of severe bleeding (hemorrhage).

After the virus has been transmitted to the human host, a period of incubation occurs. During this time (lasting about five to eight days) the virus multiplies. Symptoms of the disease appear suddenly and include **high fever**, chills, **headache**, eye **pain**, red eyes, enlarged lymph nodes, a red flush to the face, lower back pain, extreme weakness, and severe aches in the legs and joints.

This initial period of illness lasts about two–three days. After this time, the fever drops rapidly and the patient sweats heavily. After about a day of feeling relatively well, the patient's temperature increases again, although not as much as the first time. A rash of small red bumps begins on the arms and legs, spreading to the chest, abdomen, and back. It rarely affects the face. The palms of the hands and the soles of the feet become swollen and turn bright red. The characteristic combination of fever, rash, and headache are called the “dengue triad.” Most people recover fully from dengue fever, although weakness and **fatigue** may last for several weeks. Once a person has been infected with dengue fever, his or her immune system keeps producing cells that prevent reinfection for about a year.

More severe illness may occur in some people. These people may be experiencing dengue fever for the first time. However, in some cases a person may have already had dengue fever at one time, recovered, and then is reinfected with the virus. In these cases, the first infection teaches the immune system to recognize the presence of the arbovirus. When the immune cells encounter the virus during later infections, the immune system over-reacts. These types of illnesses, called dengue hemorrhagic fever (DHF) or dengue shock syndrome (DSS), involve more severe symptoms. Fever and headache are the first symptoms, but the other initial symptoms of dengue fever are absent. The patient develops a **cough**, followed by the appearance of small purplish spots (petechiae) on the skin. These petechiae are areas where blood is leaking out of the vessels. Large bruised areas appear as the bleeding worsens and abdominal pain may be severe. The patient may begin to vomit a substance that looks like coffee grounds. This is actually a sign of bleeding into the stomach. As the blood vessels become more damaged, they leak more and continue to increase in diameter (dilate), causing a decrease in blood flow to all tissues of the body. This state of low

blood flow is called shock. Shock can result in damage to the body's organs (especially the heart and kidneys) because low blood flow deprives them of oxygen.

## Diagnosis

Diagnosis should be suspected in endemic areas whenever a high fever goes on for two to seven days, especially if accompanied by a bleeding tendency. Symptoms of shock should suggest the progression of the disease to DSS.

The arbovirus causing dengue fever is one of the few types of arbovirus that can be isolated from the serum of the blood. The serum is the fluid in which blood cells are suspended. Serum can be tested because the phase in which the virus travels throughout the bloodstream is longer in dengue fever than in other arboviral infections. A number of tests are used to look for reactions between the patient's serum and laboratory-produced antibodies. Antibodies are special cells that recognize the markers (or antigens) present on invading organisms. During these tests, antibodies are added to a sample of the patient's serum. Healthcare workers then look for reactions that would only occur if viral antigens were present in that serum.

## Treatment

There is no treatment available to shorten the course of dengue fever, DHF, or DSS. Medications can be given to lower the fever and to decrease the pain of muscle aches and headaches. Fluids are given through a needle in a vein to prevent **dehydration**. Blood transfusions may be necessary if severe hemorrhaging occurs. Oxygen should be administered to patients in shock.

## Prognosis

The prognosis for uncomplicated dengue fever is very good, and almost 100% of patients fully recover. However, as many as 6–30% of all patients die when DHF occurs. The **death** rate is especially high among the youngest patients (under one year old). In places where excellent medical care is available, very close monitoring and immediate treatment of complications lowers the death rate among DHF and DSS patients to about 1%.

## Prevention

Prevention of dengue fever means decreasing the mosquito population. Any sources of standing water (buckets, vases, etc.) where the mosquitoes can breed must be eliminated. Mosquito repellent is recommended for those areas where dengue fever is endemic. To help break the cycle of transmission, sick patients should be

## KEY TERMS

**Endemic**—Naturally and consistently present in a certain geographical region.

**Host**—The organism (such as a monkey or human) in which another organism (such as a virus or bacteria) is living.

**Vector**—A carrier organism (such as a fly or mosquito) that delivers a virus (or other agent of infection) to a host.

placed in bed nets so that mosquitoes cannot bite them and become arboviral vectors.

## Resources

### BOOKS

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### PERIODICALS

Farley, Dixie. "Treating Tropical Diseases." *FDA Consumer* (Jan./Feb. 1997): 26+.

### ORGANIZATIONS

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

Rosalyn Carson-DeWitt, MD

Dental caries see **Tooth decay**

Dental cavity see **Tooth decay**

Dental hygiene see **Oral hygiene**

Dental injuries see **Dental trauma**

## Dental trauma

### Definition

Dental trauma is injury to the mouth, including teeth, lips, gums, tongue, and jawbones. The most common dental trauma is a broken or lost tooth.

### Description

Dental trauma may be inflicted in a number of ways: contact sports, motor vehicle accidents, fights, falls, eating hard foods, drinking hot liquids, and other such mishaps. As oral tissues are highly sensitive, injuries to the mouth are typically very painful. Dental trauma should receive prompt treatment from a dentist.

### Causes and symptoms

Soft tissue injuries, such as a "fat lip," a burned tongue, or a cut inside the cheek, are characterized by **pain**, redness, and swelling with or without bleeding. A broken tooth often has a sharp edge that may cut the tongue and cheek. Depending on the position of the fracture, the tooth may or may not cause **toothache** pain. When a tooth is knocked out (eviscerated), the socket is swollen, painful, and bloody. A jawbone may be broken if the upper and lower teeth no longer fit together properly (**malocclusion**), or if the jaws have pain with limited ability to open and close (mobility), especially around the temporomandibular joint (TMJ).

### Diagnosis

Dental trauma is readily apparent upon examination. Dental x rays may be taken to determine the extent of the damage to broken teeth. More comprehensive x rays are needed to diagnose a broken jaw.

### Treatment

Soft tissue injuries may require only cold compresses to reduce swelling. Bleeding may be controlled with direct pressure applied with clean gauze. Deep lacerations and punctures may require stitches. Pain may be managed with **aspirin** or **acetaminophen** (Tylenol, Aspirin Free Excedrin) or ibuprofen (Motrin, Advil).

Treatment of a broken tooth will vary depending on the severity of the fracture. For immediate first aid, the injured tooth and surrounding area should be rinsed gently with warm water to remove dirt, then covered with a cold compress to reduce swelling and ease pain. A dentist should examine the injury as soon as possible. Any pieces from the broken tooth should be saved and brought along.

If a piece of the outer tooth has chipped off, but the inner core (pulp) is undisturbed, the dentist may simply smooth the rough edges or replace the missing section with a small composite filling. In some cases, a fragment of broken tooth may be bonded back into place. If enough tooth is missing to compromise the entire tooth structure, but the pulp is not permanently damaged, the tooth will require a protective coverage with a gold or

porcelain crown. If the pulp has been seriously damaged, the tooth will require **root canal treatment** before it receives a crown. A tooth that is vertically fractured or fractured below the gumline will require root canal treatment and protective restoration. A tooth which no longer has enough remaining structure to retain a crown may have to be extracted (surgically removed).

When a permanent tooth has been knocked out, it may be saved with prompt action. The tooth must be found immediately after it has been lost. It should be picked up by the natural crown (the top part covered by hard enamel). It must not be handled by the root. If the tooth is dirty, it may be gently rinsed under running water. It should never be scrubbed, and it should never be washed with soap, toothpaste, mouthwash, or other chemicals. The tooth should not be dried or wrapped in a tissue or cloth. It must be kept moist at all times.

The tooth may be placed in a clean container of milk, cool water with or without a pinch of salt, or in saliva. If possible, the patient and the tooth should be brought to the dentist within 30 minutes of the tooth loss. Rapid action improves the chances of successful re-implantation; however, it is possible to save a tooth after 30 minutes, if the tooth has been kept moist and handled properly.

The body usually rejects re-implantation of a primary (baby) tooth. In this case, the empty socket is treated as a soft tissue injury and monitored until the permanent tooth erupts.

A broken jaw must be set back into its proper position and stabilized with wires while it heals. Healing may take six weeks or longer, depending on the patient's age and the severity of the fracture.

### Alternative treatment

There is no substitute for treatment by a dentist or other medical professional. There are, however, homeopathic remedies and herbs that can be used simultaneously with dental care and throughout the healing process. Homeopathic arnica (*Arnica montana*) should be taken as soon as possible after the injury to help the body deal with the trauma. Repeating a dose several times daily for the duration of healing is also useful. Homeopathic hypericum (*Hypericum perforatum*) can be taken if nerve pain is involved, especially with a **tooth extraction** or root canal. Homeopathic comfrey (*Symphytum officinale*) may be helpful in treating pain due to broken jaw bones, but should only be used after the bones have been reset. Calendula (*Calendula officinalis*) and plantain (*Plantago major*) can be used as a mouth rinse to enhance tissue healing. These herbs should not be used with deep lacerations that need to heal from the inside first.

### KEY TERMS

**Crown**—1 The natural part of the tooth covered by enamel. 2 A restorative crown is a protective shell that fits over a tooth.

**Eruption**—The process of a tooth breaking through the gum tissue to grow into place in the mouth.

**Evulsion**—The forceful, and usually accidental, removal of a tooth from its socket in the bone.

**Extraction**—The surgical removal of a tooth from its socket in the bone.

**Malocclusion**—A problem in the way the upper and lower teeth fit together in biting or chewing.

**Pulp**—The soft innermost layer of a tooth containing blood vessels and nerves.

**Root canal treatment**—The process of removing diseased or damaged pulp from a tooth, then filling and sealing the pulp chamber and root canals.

**Temporomandibular joint (TMJ)**—The jaw joint formed by the mandible (lower jaw bone) moving against the temporal (temple and side) bone of the skull.

### Prognosis

When dental trauma receives timely attention and proper treatment, the prognosis for healing is good. As with other types of trauma, infection may be a complication, but a course of antibiotics is generally effective.

### Prevention

Most dental trauma is preventable. Car seat belts should always be worn, and young children should be secured in appropriate car seats. Homes should be monitored for potential tripping and slipping hazards. Child-proofing measures should be taken, especially for toddlers. In addition to placing gates across stairs and padding sharp table edges, electrical cords should be tucked away. Young children may receive severe oral burns from gnawing on live power cords.

Everyone who participates in contact sports should wear a mouthguard to avoid dental trauma. Athletes in football, ice hockey, wrestling, and boxing commonly wear mouthguards. The mandatory use of mouthguards in football prevents about 200,000 oral injuries annually. Mouthguards should also be worn along with helmets in noncontact sports such as skateboarding, in-line skating,

and bicycling. An athlete who does not wear a mouthguard is 60 times more likely to sustain dental trauma than one who does. Any activity involving speed, an increased chance of falling, and potential contact with a hard piece of equipment has the likelihood of dental trauma that may be prevented or substantially reduced in severity with the use of mouthguards.

## Resources

### ORGANIZATIONS

American Academy of Pediatric Dentistry. 211 East Chicago Ave., Ste. 700, Chicago, IL 60611-2616. (312) 337-2169. <<http://www.aapd.org>>.

American Association of Endodontists. 211 East Chicago Ave., Ste. 1100, Chicago, IL 60611-2691. (800) 872-3636. <<http://www.aae.org>>.

American Association of Oral and Maxillofacial Surgeons. 9700 West Bryn Mawr Ave., Rosemont, IL 60018-5701. (847) 678-6200. <<http://www.aaoms.org>>.

American Dental Association. 211 E. Chicago Ave., Chicago, IL 60611. (312) 440-2500. <<http://www.ada.org>>.

Donald Gardner Barstow

Depersonalization disorder see **Dissociative disorders**



**A physician inserts a contraceptive implant under the skin of a woman's arm.** (Photo Researchers, Inc. Reproduced by permission.)

released. The mucus in the cervix (opening into the uterus or womb) becomes thicker, making it difficult for the sperm to enter. Depo-Provera and Lunelle also cause the lining of the uterus to become thinner, making implantation of a fertilized egg unlikely.

An injection of Depo-Provera or Lunelle must be given within the first five days of a normal period. Depo-Provera provides protection against pregnancy for three months, while Lunelle provides similar protection for one month. Ovulation (release of a mature egg) typically occurs within 60 days of the last injection of Lunelle, about twice as fast after use of Depo-Provera. Also, because Lunelle is a combined hormone contraceptive as opposed to progestin-only Depo-Provera and Norplant, it is less likely to cause irregular or absent menstruation.

Norplant capsules contain a synthetic hormone that is slowly released over a period of up to five years. It functions like Depo-Provera in that it prevents the ovaries from producing ova (eggs) and also results in thicker mucus in the cervix, which prevents the sperm from passing through the cervix. Norplant can be inserted at any time.

## Preparation

The woman being considered for Depo-Provera or Lunelle will have a pelvic and breast examination, a **Pap test** (a microscopic examination of cell samples taken from the cervix), blood pressure check, weight check, and a review of her medical history. Women who have **diabetes mellitus**, major depression, blood clotting problems, liver disease, or weight problems should use these methods only under strict medical supervision. Depo-Provera or Lunelle should not be used if the woman is pregnant, has unexplained vaginal bleeding,

## Depo-Provera/Norplant

### Definition

Norplant is a long-acting hormone that is inserted under the skin and prevents conception for up to five years. Depo-Provera is also a hormone, but is administered by intramuscular injection and provides protection against **pregnancy** for three months. Lunelle is another injectable contraceptive that is administered monthly (every 28 to 30 days); it was approved by the Food and Drug Administration (FDA) in October 2000. The hormone in Norplant and Depo-Provera is progestin, a synthetic hormone similar to one found naturally in a woman's body; Lunelle contains the hormones progestin and estrogen.

### Purpose

The purpose of these hormones is to prevent pregnancy; they are about 99% effective in achieving this goal. No hormonal contraceptive methods provide protection from **AIDS** or other **sexually transmitted diseases**.

Depo-Provera and Lunelle are given as an injection and work in several ways to prevent conception. First, the egg (ovum) is prevented from maturing and being

## KEY TERMS

**Hormone**—A chemical produced in a gland or organ and transported by the blood to another area of the body where it produces a specific effect.

**Pap test**—A microscopic examination of cell samples taken from the cervix.

suffers from severe liver disease, has **breast cancer**, or has a history of blood clots or **stroke**.

Individuals who select Norplant will receive the same basic **physical examination**. If approved for this method, a site of implantation will be selected (usually the inside of the upper arm), and the area prepared for minor surgery. The skin will be washed with soap and water, and an antiseptic, such as iodine solution, will be applied. The physician will use a local anesthetic to numb the area, a small incision will be made, the six Norplant capsules will be inserted, and the incision sewn up (sutured). Protection against pregnancy normally begins within 24 hours. If necessary, the implants can be removed in 15–20 minutes. Norplant should not be used by women who are pregnant, have blood clotting problems, or have unexplained vaginal bleeding. Advantages include light periods with less cramping and decreased anemia. This form of birth control may also be protective against **endometrial cancer**.

Because Depo-Provera and Norplant use only the hormone progestin, they may provide an alternative for women who can not use estrogen-containing birth control pills. One benefit of Lunelle, however, is that its effects wear off more quickly than Depo-Provera, an important factor in the event that a woman has serious side effects or wants to become pregnant.

### Risks

The most common side effects associated with Depo-Provera and Lunelle are yellowing of the skin, **headache**, nervousness, **dizziness**, abdominal **pain**, hair loss, rash, increase in the number of migraine headaches, increased or decreased interest in sexual intercourse, the development of dark spots on the skin, depression, and weakness. Danger signs that need to be reported immediately include weight gain, heavy vaginal bleeding, frequent urination, blurred vision, **fainting**, severe abdominal pain, and coughing up blood. Because the effects of Depo-Provera may last up to 12 weeks, it may take a longer time for women trying to conceive to become pregnant after discontinuing the injections.

The main reactions to Norplant include headache, weight gain, irregular periods or no period at all, breast tenderness, **acne**, gain or loss of facial hair, color changes of the skin over the area of insertion, and **ovarian cysts**. The doctor should be notified immediately of lumps in the breast, heavy vaginal bleeding, yellowing of the skin or eyes, or infection of the incision. Women who use Norplant are discouraged from **smoking**.

### Normal results

These hormone contraceptive methods normally result in a success rate of 99%.

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Donald Gardner Barstow

Depression see **Bipolar disorder**;  
**Postpartum depression**

## Depressive disorders

### Definition

Depression or depressive disorders (unipolar depression) are mental illnesses characterized by a profound and persistent feeling of sadness or despair and/or a loss of interest in things that were once pleasurable. Disturbance in sleep, appetite, and mental processes are a common accompaniment.

## Description

Everyone experiences feelings of unhappiness and sadness occasionally. But when these depressed feelings start to dominate everyday life and cause physical and mental deterioration, they become what are known as depressive disorders. Each year in the United States, depressive disorders affect an estimated 17 million people at an approximate annual direct and indirect cost of \$53 billion. One in four women is likely to experience an episode of severe depression in her lifetime, with a 10–20% lifetime prevalence, compared to 5–10% for men. The average age a first depressive episode occurs is in the mid-20s, although the disorder strikes all age groups indiscriminately, from children to the elderly.

There are two main categories of depressive disorders: major depressive disorder and dysthymic disorder. Major depressive disorder is a moderate to severe episode of depression lasting two or more weeks. Individuals experiencing this major depressive episode may have trouble sleeping, lose interest in activities they once took pleasure in, experience a change in weight, have difficulty concentrating, feel worthless and hopeless, or have a preoccupation with **death** or suicide. In children, the major depression may appear as irritability.

While major depressive episodes may be acute (intense but short-lived), dysthymic disorder is an ongoing, chronic depression that lasts two or more years (one or more years in children) and has an average duration of 16 years. The mild to moderate depression of dysthymic disorder may rise and fall in intensity, and those afflicted with the disorder may experience some periods of normal, non-depressed mood of up to two months in length. Its onset is gradual, and dysthymic patients may not be able to pinpoint exactly when they started feeling depressed. Individuals with dysthymic disorder may experience a change in sleeping and eating patterns, low self-esteem, **fatigue**, trouble concentrating, and feelings of hopelessness.

Depression can also occur in **bipolar disorder**, an affective mental illness that causes radical emotional changes and mood swings, from manic highs to depressive lows. The majority of bipolar individuals experience alternating episodes of **mania** and depression.

## Causes and symptoms

The causes behind depression are complex and not yet fully understood. While an imbalance of certain neurotransmitters—the chemicals in the brain that transmit messages between nerve cell—is believed to be key to depression, external factors such as upbringing (more so in dysthymia than major depression) may be as impor-

### Signs Of Mental Depression

Depressed mood  
Lack of interest or pleasure in daily activities  
Significant weight loss (without dieting) or weight gain  
Difficulty sleeping or excessive sleeping  
Loss of energy  
Feelings of worthlessness or guilt  
Difficulty in making decisions  
Restlessness  
Recurrent thoughts of death

tant. For example, it is speculated that, if an individual is abused and neglected throughout childhood and adolescence, a pattern of low self-esteem and negative thinking may emerge. From that, a lifelong pattern of depression may follow.

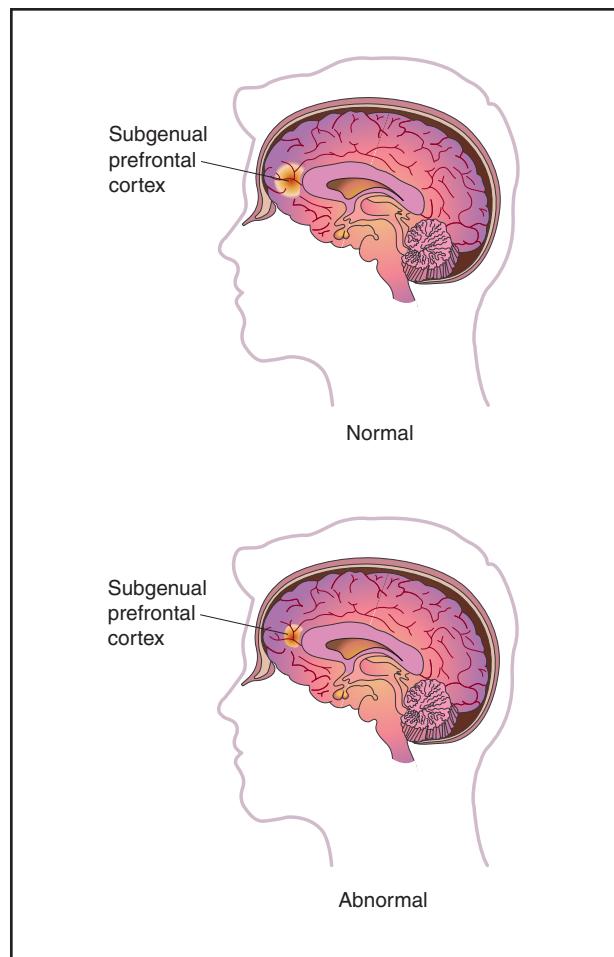
Heredity does seem to play a role in who develops depressive disorders. Individuals with major depression in their immediate family are up to three times more likely to have the disorder themselves. It would seem that biological and genetic factors may make certain individuals pre-disposed or prone to depressive disorders, but environmental circumstances may often trigger the disorder.

External stressors and significant life changes, such as chronic medical problems, death of a loved one, divorce or estrangement, **miscarriage**, or loss of a job, can also result in a form of depression known as adjustment disorder. Although periods of adjustment disorder usually resolve themselves, occasionally they may evolve into a major depressive disorder.

### *Major depressive episode*

Individuals experiencing a major depressive episode have a depressed mood and/or a diminished interest or pleasure in activities. Children experiencing a major depressive episode may appear or feel irritable rather than depressed. In addition, five or more of the following symptoms will occur on an almost daily basis for a period of at least two weeks:

- Significant change in weight.
- **Insomnia** or hypersomnia (excessive sleep).
- Psychomotor agitation or retardation.
- Fatigue or loss of energy.
- Feelings of worthlessness or inappropriate guilt.
- Diminished ability to think or to concentrate, or indecisiveness.
- Recurrent thoughts of death or suicidal and/or suicide attempts.



Recent scientific research has indicated that the size of the subgenual prefrontal cortex of the brain (located behind the bridge of the nose) may be a determining factor in hereditary depressive disorders. (Illustration by Electronic Illustrators Group.)

### Dysthymic disorder

Dysthymia commonly occurs in tandem with other psychiatric and physical conditions. Up to 70% of dysthymic patients have both dysthymic disorder and major depressive disorder, known as double depression. Substance abuse, panic disorders, **personality disorders**, social **phobias**, and other psychiatric conditions are also found in many dysthymic patients. Dysthymia is prevalent in patients with certain medical conditions, including **multiple sclerosis**, **AIDS**, **hypothyroidism**, **chronic fatigue syndrome**, **Parkinson's disease**, diabetes, and post-cardiac transplantation. The connection between dysthymic disorder and these medical conditions is unclear, but it may be related to the way the medical condition and/or its pharmacological treatment affects neurotransmitters. Dysthymic disorder can lengthen or complicate the recovery of patients also suffering from medical conditions.

Along with an underlying feeling of depression, people with dysthymic disorder experience two or more of the following symptoms on an almost daily basis for a period for two or more years (most suffer for five years), or one year or more for children:

- under or overeating
- insomnia or hypersomnia
- low energy or fatigue
- low self-esteem
- poor concentration or trouble making decisions
- feelings of hopelessness

### Diagnosis

In addition to an interview, several clinical inventories or scales may be used to assess a patient's mental status and determine the presence of depressive symptoms. Among these tests are: the Hamilton Depression Scale (HAM-D), Child Depression Inventory (CDI), Geriatric Depression Scale (GDS), Beck Depression Inventory (BDI), and the Zung Self-Rating Scale for Depression. These tests may be administered in an outpatient or hospital setting by a general practitioner, social worker, psychiatrist, or psychologist.

### Treatment

Major depressive and dysthymic disorders are typically treated with antidepressants or psychosocial therapy. Psychosocial therapy focuses on the personal and interpersonal issues behind depression, while antidepressant medication is prescribed to provide more immediate relief for the symptoms of the disorder. When used together correctly, therapy and antidepressants are a powerful treatment plan for the depressed patient.

#### Antidepressants

**Selective serotonin reuptake inhibitors** (SSRIs) such as fluoxetine (Prozac) and sertraline (Zoloft) reduce depression by increasing levels of serotonin, a neurotransmitter. Some clinicians prefer SSRIs for treatment of dysthymic disorder. **Anxiety**, **diarrhea**, drowsiness, **headache**, sweating, nausea, poor sexual functioning, and insomnia are all possible side effects of SSRIs.

Tricyclic antidepressants (TCAs) are less expensive than SSRIs, but have more severe side-effects, which may include persistent **dry mouth**, **sedation**, **dizziness**, and cardiac **arrhythmias**. Because of these side effects, caution is taken when prescribing TCAs to elderly patients. TCAs include amitriptyline (Elavil), imipramine (Tofranil), and nortriptyline (Aventyl, Pamelor). A 10-day supply of TCAs can be lethal if ingested all at once, so these

drugs may not be a preferred treatment option for patients at risk for suicide.

**Monoamine oxidase inhibitors (MAOIs)** such as tranylcypromine (Parnate) and phenelzine (Nardil) block the action of monoamine oxidase (MAO), an enzyme in the central nervous system. Patients taking MAOIs must cut foods high in tyramine (found in aged cheeses and meats) out of their diet to avoid potentially serious hypertensive side effects.

Heterocyclics include bupropion (Wellbutrin) and trazodone (Desyrel). Bupropion should not be prescribed to patients with a **seizure disorder**. Side effects of the drug may include agitation, anxiety, confusion, tremor, dry mouth, fast or irregular heartbeat, headache, low blood pressure, and insomnia. Because trazodone has a sedative effect, it is useful in treating depressed patients with insomnia. Other possible side effects of trazodone include dry mouth, gastrointestinal distress, dizziness, and headache.

#### *Psychosocial therapy*

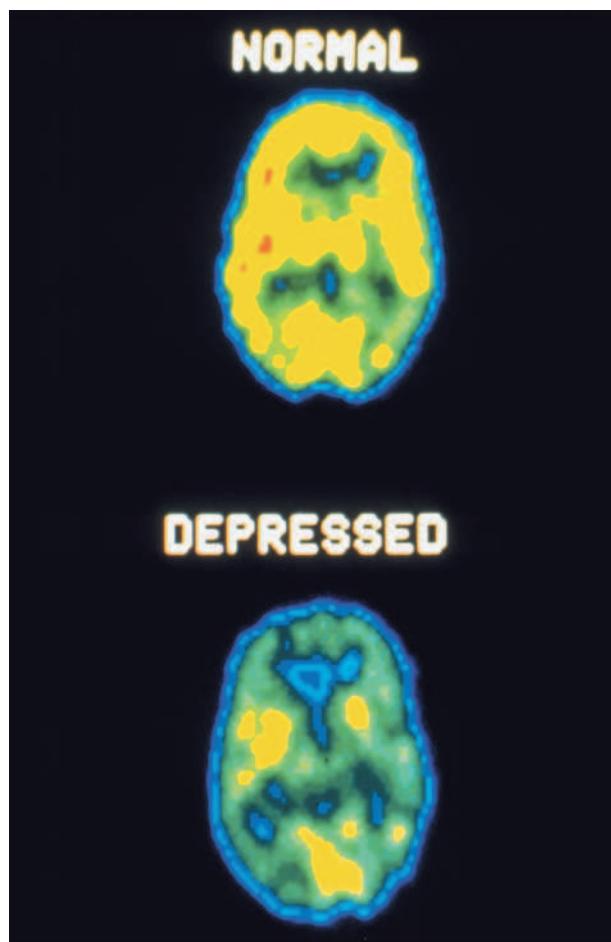
Psychotherapy explores an individual's life to bring to light possible contributing causes of the present depression. During treatment, the therapist helps the patient to become self-aware of his or her thinking patterns and how they came to be. There are several different subtypes of psychotherapy, but all have the common goal of helping the patient develop healthy problem solving and coping skills.

**Cognitive-behavioral therapy** assumes that the patient's faulty thinking is causing the current depression and focuses on changing the depressed patient's thought patterns and perceptions. The therapist helps the patient identify negative or distorted thought patterns and the emotions and behavior that accompany them, and then retrains the depressed individual to recognize the thinking and react differently to it.

#### *Electroconvulsive therapy*

ECT, or **electroconvulsive therapy**, is usually employed after all therapy and pharmaceutical treatment options have been explored. However, it is sometimes used early in treatment when severe depression is present and the patient refuses oral medication, or when the patient is becoming dehydrated, extremely suicidal, or psychotic.

The treatment consists of a series of electrical pulses that move into the brain through electrodes on the patient's head. ECT is given under general anesthesia and patients are administered a muscle relaxant to prevent convulsions. Although the exact mechanisms behind the success of ECT therapy are not known, it is believed that the electrical current modifies the electrochemical



Positron emission tomography (PET) scans comparing a normal brain with that of someone with a depressed mental disorder. (Photo Researchers, Inc. Reproduced by permission.)

processes of the brain, consequently relieving depression. Headaches, muscle soreness, nausea, and confusion are possible side effects immediately following an ECT procedure. Memory loss, typically transient, has also been reported in ECT patients.

#### **Alternative treatment**

**St. John's wort** (*Hypericum perforatum*) is used throughout Europe to treat depressive symptoms. Unlike traditional prescription antidepressants, this herbal anti-depressant has few reported side effects. Some users may experience high blood pressure, headaches, stiff neck, nausea, and vomiting. As of early 1998, United States clinical trials organized by the National Institute of Mental Health were still in the planning phase. Its efficacy in severe depression is very uncertain.

Homeopathic treatment can also be very therapeutic in treating depression. Good **nutrition**, proper sleep,

## KEY TERMS

**Hypersomnia**—The need to sleep excessively; a symptom of dysthymic and major depressive disorder.

**Neurotransmitter**—A chemical in the brain that transmits messages between neurons, or nerve cells. Changes in the levels of certain neurotransmitters, such as serotonin, norepinephrine, and dopamine, are thought to be related to depressive disorders.

**Psychomotor agitation**—Disturbed physical and mental processes (e.g., fidgeting, wringing of hands, racing thoughts); a symptom of major depressive disorder.

**Psychomotor retardation**—Slowed physical and mental processes (e.g., slowed thinking, walking, and talking); a symptom of major depressive disorder.

**exercise**, and full engagement in life are very important to a healthy mental state.

### Prognosis

Untreated or improperly treated depression is the number one cause of suicide in the United States. Proper treatment relieves symptoms in 80–90% of depressed patients. After each major depressive episode, the risk of recurrence climbs significantly—50% after one episode, 70% after two episodes, and 90% after three episodes. For this reason, patients need to be aware of the symptoms of recurring depression and may require long-term maintenance treatment of antidepressants and/or therapy.

### Prevention

Patient education in the form of therapy or self-help groups is crucial for training patients with depressive disorders to recognize symptoms of depression and to take an active part in their treatment program. Extended maintenance treatment with antidepressants may be required in some patients to prevent relapse. Early intervention with children with depression is effective in arresting development of more severe problems.

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American Psychological Association (APA). 750 First St. NE, Washington, DC 20002-4242. (202) 336-5700. <<http://www.apa.org>>.

National Alliance for the Mentally Ill (NAMI). Colonial Place Three, 2107 Wilson Blvd., Ste. 300, Arlington, VA 22201-3042. (800) 950-6264. <<http://www.nami.org>>.

National Depressive and Manic-Depressive Association (NDMDA). 730 N. Franklin St., Suite 501, Chicago, IL 60610. (800) 826-3632. <<http://www.ndmda.org>>.

National Institute of Mental Health. Mental Health Public Inquiries, 5600 Fishers Lane, Room 15C-05, Rockville, MD 20857. (888) 826-9438. <<http://www.nimh.nih.gov>>.

Paula Anne Ford-Martin

Dermabrasion see **Skin resurfacing**

## Dermatitis

### Definition

Dermatitis is a general term used to describe inflammation of the skin.

### Description

Most types of dermatitis are characterized by an itchy pink or red rash.

**Contact dermatitis** is an allergic reaction to something that irritates the skin and is manifested by one or more lines of red, swollen, blistered skin that may itch or

seep. It usually appears within 48 hours after touching or brushing against a substance to which the skin is sensitive. The condition is more common in adults than in children.

Contact dermatitis can occur on any part of the body, but it usually affects the hands, feet, and groin. Contact dermatitis usually does not spread from one person to another, nor does it spread beyond the area exposed to the irritant unless affected skin comes into contact with another part of the body. However, in the case of some irritants, such as poison ivy, contact dermatitis can be passed to another person or to another part of the body.

Stasis dermatitis is characterized by scaly, greasy looking skin on the lower legs and around the ankles. Stasis dermatitis is most apt to affect the inner side of the calf.

Nummular dermatitis, which is also called nummular eczematous dermatitis or nummular eczema, generally affects the hands, arms, legs, and buttocks of men and women older than 55 years of age. This stubborn inflamed rash forms circular, sometimes itchy, patches and is characterized by flares and periods of inactivity.

**Atopic dermatitis** is characterized by **itching**, scaling, swelling, and sometimes blistering. In early childhood it is called infantile eczema and is characterized by redness, oozing, and crusting. It is usually found on the face, inside the elbows, and behind the knees.

**Seborrheic dermatitis** may be dry or moist and is characterized by greasy scales and yellowish crusts on the scalp, eyelids, face, external surfaces of the ears, underarms, breasts, and groin. In infants it is called "cradle cap."

### Causes and symptoms

Allergic reactions are genetically determined, and different substances cause contact dermatitis to develop in different people. A reaction to resin produced by poison ivy, poison oak, or poison sumac is the most common source of symptoms. It is, in fact, the most common allergy in this country, affecting one of every two people in the United States.

Flowers, herbs, and vegetables can also affect the skin of some people. **Burns** and **sunburn** increase the risk of dermatitis developing, and chemical irritants that can cause the condition include:

- chlorine
- cleansers
- detergents and soaps
- fabric softeners
- glues used on artificial nails
- perfumes
- topical medications



**Dermatitis on hands and fingers.** (Custom Medical Stock Photo. Reproduced by permission.)

Contact dermatitis can develop when the first contact occurs or after years of use or exposure.

Stasis dermatitis, a consequence of poor circulation, occurs when leg veins can no longer return blood to the heart as efficiently as they once did. When that happens, fluid collects in the lower legs and causes them to swell. Stasis dermatitis can also result in a rash that can break down into sores known as stasis ulcers.

The cause of nummular dermatitis is not known, but it usually occurs in cold weather and is most common in people who have dry skin. Hot weather and **stress** can aggravate this condition, as can the following:

- allergies
- fabric softeners
- soaps and detergents
- wool clothing
- bathing more than once a day

Atopic dermatitis can be caused by allergies, **asthma**, or stress, and there seems to be a genetic predisposition for atopic conditions. It is sometimes caused by an allergy to nickel in jewelry.

Seborrheic dermatitis (for which there may also be a genetic predisposition) is usually caused by overproduction of the oil glands. In adults it can be associated with **diabetes mellitus** or gold allergy. In infants and adults it may be caused by a biotin deficiency.

### Diagnosis

The diagnosis of dermatitis is made on the basis of how the rash looks and its location. The doctor may scrape off a small piece of affected skin for microscopic examination or direct the patient to discontinue use of any potential irritant that has recently come into contact with

## KEY TERMS

**Allergic reaction**—An inappropriate or exaggerated genetically determined reaction to a chemical that occurs only on the second or subsequent exposures to the offending agent, after the first contact has sensitized the body.

**Corticosteroid**—A group of synthetic hormones that are used to prevent or reduce inflammation. Toxic effects may result from rapid withdrawal after prolonged use or from continued use of large doses.

**Patch test**—A skin test that is done to identify allergens. A suspected substance is applied to the skin. After 24–48 hours, if the area is red and swollen, the test is positive for that substance. If no reaction occurs, another substance is applied. This is continued until the patient experiences an allergic reaction where the irritant was applied to the skin.

**Rash**—A spotted, pink or red skin eruption that may be accompanied by itching and is caused by disease, contact with an allergen, food ingestion, or drug reaction.

**Ulcer**—An open sore on the skin, resulting from tissue destruction, that is usually accompanied by redness, pain, or infection.

the affected area. Two weeks after the rash disappears, the patient may resume use of the substances, one at a time, until the condition recurs. Eliminating the substance most recently added should eliminate the irritation.

If the origin of the irritation has still not been identified, a dermatologist may perform one or more patch tests. This involves dabbing a small amount of a suspected irritant onto skin on the patient's back. If no irritation develops within a few days, another patch test is performed. The process continues until the patient experiences an allergic reaction at the spot where the irritant was applied.

### Treatment

Treating contact dermatitis begins with eliminating or avoiding the source of irritation. Prescription or over-the-counter corticosteroid creams can lessen inflammation and relieve irritation. Creams, lotions, or ointments not specifically formulated for dermatitis can intensify the irritation. Oral **antihistamines** are sometimes recommended to alleviate itching, and **antibiotics** are prescribed if the rash becomes infected. Medications taken by mouth to relieve symptoms of dermatitis can make skin red and scaly and cause hair loss.

Patients who have a history of dermatitis should remove their rings before washing their hands. They should use bath oils or glycerine-based soaps and bathe in lukewarm saltwater.

Patting rather than rubbing the skin after bathing and thoroughly massaging lubricating lotion or nonprescription cortisone creams into still-damp skin can soothe red, irritated nummular dermatitis. Highly concentrated cortisone preparations should not be applied to the face, armpits, groin, or rectal area. Periodic medical monitoring is necessary to detect side effects in patients who use such preparations on **rashes** covering large areas of the body.

Coal-tar salves can help relieve symptoms of nummular dermatitis that have not responded to other treatments, but these ointments have an unpleasant odor and stain clothing.

Patients who have stasis dermatitis should elevate their legs as often as possible and sleep with a pillow between the lower legs.

Tar or zinc paste may also be used to treat stasis dermatitis. Because these compounds must remain in contact with the rash for as long as two weeks, the paste and bandages must be applied by a nurse or a doctor.

Coal-tar shampoos may be used for seborrheic dermatitis that occurs on the scalp. Sun exposure after the use of these shampoos should be avoided because the risk of sunburn of the scalp is increased.

### Alternative treatment

Some herbal therapies can be useful for skin conditions. Among the herbs most often recommended are:

- Burdock root (*Arctium lappa*)
- Calendula (*Calendula officinalis*) ointment
- Chamomile (*Matricaria recutita*) ointment
- Cleavers (*Galium* spp.)
- Evening primrose oil (*Oenothera biennis*)
- Nettles (*Urtica dioica*)

Contact dermatitis can be treated botanically and homeopathically. Grindelia (*Grindelia* spp.) and sassafras (*Sassafras albidum*) can help when applied topically. Determining the source of the problem and eliminating it is essential. Oatmeal baths are very helpful in relieving the itch. Bentonite clay packs or any mud pack draws the fluid out, and helps dry up the lesions. Cortisone creams are not recommended.

Stasis dermatitis should be treated by a trained practitioner. This condition responds well to topical herbal therapies, however, the cause must also be addressed.

Selenium-based shampoos, topical applications of flax oil and/or olive oil, and biotin supplementation are among the therapies recommended for seborrheic dermatitis.

## Prognosis

Dermatitis is often chronic, but symptoms can generally be controlled.

## Prevention

Contact dermatitis can be prevented by avoiding the source of irritation. If the irritant cannot be avoided completely, the patient should wear gloves and other protective clothing whenever exposure is likely to occur.

Immediately washing the exposed area with soap and water can stem allergic reactions to poison ivy, poison oak, or poison sumac, but because soaps can dry the skin, patients susceptible to dermatitis should use them only on the face, feet, genitals, and underarms.

Clothing should be loose fitting and 100% cotton. New clothing should be washed in dye-free, unscented detergent before being worn.

Injury to the lower leg can cause stasis dermatitis to ulcerate (form open sores). If stasis ulcers develop, a doctor should be notified immediately.

**Yoga** and other relaxation techniques may help prevent atopic dermatitis caused by stress.

Avoidance of sweating may aid in preventing seborrheic dermatitis.

A patient who has dermatitis should also notify a doctor if any of the following occurs:

- fever develops
- skin oozes or other signs of infection appear
- symptoms do not begin to subside after seven days' treatment
- he/she comes into contact with someone who has a wart, **cold sore**, or other viral skin infection

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Maureen Haggerty

Dermatophyte infections see **Ringworm**

## DES exposure

### Definition

DES (diethylstilbestrol) is a hormone that was prescribed for pregnant women in the 1950s and early 1960s. Many years later, doctors discovered that the daughters of the women who received DES were at high risk for a variety of problems, including **infertility**, **premature labor**, and **cancer** of the vagina and cervix.

### Description

In the 1950s and early 1960s, several drug companies claimed that DES (diethylstilbestrol) could prevent miscarriages. DES is a synthetic hormone, related to estrogen. Since up to 20% of all pregnancies end in **miscarriage**, this seemed like an important breakthrough and DES was prescribed for many women who had bleeding in early **pregnancy**. Ultimately, it was found to have no effect on miscarriages and the practice of prescribing DES was stopped in the 1960s. Almost 10 years later, the daughters of women who had taken DES during pregnancy began to develop unusual symptoms.

Doctors discovered that when these young women reached their teens, they were at higher risk for a variety of problems, including:

- clear cell adenocarcinoma of the vagina and cervix
- infertility
- premature labor and other problems in pregnancy

### Causes and symptoms

DES has affected a very specific group of women. These are women who were exposed to DES in utero before 18 weeks of pregnancy. In other words, their mothers must have taken DES within the first four to five months of pregnancy. It is now known that the female reproductive organs are formed during that time. DES

## KEY TERMS

**Cervix**—The opening at the bottom of the uterus.

**Colposcopy**—A special examination of the cervix using a magnifying scope. This is a procedure that can be done in the doctor's office.

**Fallopian tubes**—The tubes that carry the ovum (egg) from the ovary to the uterus.

**Pap smear**—A screening test for precancerous and cancerous cells on the cervix. This simple test is done during a routine pelvic exam and involves scraping cells from the cervix.

appears to interfere with proper growth and development of the uterus, cervix, vagina, and fallopian tubes.

In the early 1970s, there was an increase in a rare form of cancer, clear cell adenocarcinoma of the vagina and cervix. Up until that time, doctors had seen these cancers only in elderly women. Suddenly, young women who had the disease appeared.

This was so unusual that researchers studied these women to see if they had anything in common. After a great deal of questioning and examination, it was found that they all had one factor in common. All of the young women had been exposed to DES in utero in the early weeks of pregnancy.

Today, it is difficult to imagine how shocking this discovery was. Doctors had only recently recognized that medications and exposure to chemicals during pregnancy could cause **birth defects**. This was a birth defect that had gone undetected for almost two decades.

Since then, doctors have studied DES daughters very carefully. Fortunately, the risk of clear cell adenocarcinoma is actually quite low. In fact, it appears that if a DES daughter has not developed this cancer by age 30, she will not develop it. Since all DES daughters are now over age 30, there should be no further cases related to DES exposure. However, there are a number of other symptoms and problems associated with DES exposure.

- Cervix and vagina. DES daughters often have distinctive changes of the cervix and vagina that can be seen during a **pelvic exam**. These changes include a cervical hood (a vaginal fold draped over the cervix), cockscomb cervix (an abnormally shaped cervix), and adenosis (glandular cells normally located within the cervix that appear on the outside of the cervix and in the vagina).

- Fallopian tubes. Some DES daughters have fallopian tube abnormalities that lead to infertility.
- Uterus. Many DES daughters have a uterus that is abnormal in size and shape. The classic sign is the T-shaped uterus. In the normal uterus, the cavity (hollow space inside) is rounded. In a T-shaped uterus, the cavity is reduced to a thin T. The abnormal shape of the inside of the uterus makes it harder for a woman to get pregnant and leads to a higher risk of premature labor and birth.

## Diagnosis

Women who have been exposed to DES should have a pelvic exam at least once a year. In addition to the usual pelvic exam and Pap smear, DES daughters should also have Pap smears of the vagina and, if possible, **colposcopy**. During colposcopy, the doctor looks at the cervix and vagina through a special magnifying scope. In this way, tiny areas of abnormal cells can be seen. This procedure is easily performed in the doctor's office.

When DES daughters get pregnant, they may be at high risk for premature labor and birth and should be monitored very carefully.

Not all women who were exposed to DES develop problems in pregnancy. However, if problems like infertility or miscarriage do occur, the doctor may recommend a special x-ray test to check the woman's fallopian tubes and uterus. This special test is called a hysterosalpingogram.

## Treatment

There is no treatment for the abnormalities of the fallopian tubes and uterus caused by DES exposure. Fortunately, there are treatments that can help with infertility and premature labor. Clear cell adenocarcinoma of the vagina or cervix must be treated with surgery and, possibly, **chemotherapy**.

## Resources

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Detached retina see **Retinal detachment**

## Detoxification

### Definition

Detoxification is one of the more widely used treatments and concepts in alternative medicine. It is based on the principle that illnesses can be caused by the accumulation of toxic substances (toxins) in the body. Eliminating existing toxins and avoiding new toxins are essential parts of the healing process. Detoxification utilizes a variety of tests and techniques.

### Purpose

Detoxification is helpful for those patients suffering from many chronic diseases and conditions, including **allergies**, **anxiety**, **arthritis**, **asthma**, chronic infections, depression, diabetes, headaches, heart disease, **high cholesterol**, low blood sugar levels, digestive disorders, mental illness, and **obesity**. It is helpful for those with conditions that are influenced by environmental factors, such as **cancer**, as well as for those who have been exposed to high levels of toxic materials due to accident or occupation. Detoxification therapy is useful for those suffering from allergies or immune system problems that conventional medicine is unable to diagnose or treat, including **chronic fatigue syndrome**, environmental illness/multiple chemical sensitivity, and **fibromyalgia**. Symptoms for those suffering these conditions may include unexplained **fatigue**, increased allergies, hypersensitivity to common materials, intolerance to certain foods and **indigestion**, aches and pains, low grade **fever**, headaches, **insomnia**, depression, sore throats, sudden weight loss or gain, lowered resistance to infection, general malaise, and disability. Detoxification can be used as a beneficial preventative measure and as a tool to increase overall health, vitality, and resistance to disease.

### Description

#### Origins

Detoxification methods of healing have been used for thousands of years. **Fasting**, is one of the oldest therapeutic practices in medicine. Hippocrates, the ancient Greek known as the “Father of Western medicine,” recommended fasting as a means for improving health. **Ayurvedic medicine**, a traditional healing system that has developed over thousands of years, utilizes detoxification methods to treat many chronic conditions and to prevent illness.

Detoxification treatment has become one of the cornerstones of alternative medicine. Conventional medicine notes that environmental factors can play a significant role

in many illnesses. Environmental medicine is a field that studies exactly how those environmental factors influence disease. Conditions such as asthma, cancer, chronic fatigue syndrome, **multiple chemical sensitivity**, and many others are strongly influenced by exposure to toxic or allergenic substances in the environment. The United States Centers for Disease Control estimate that over 80% of all illnesses have environmental and lifestyle causes.

Detoxification has also become a prominent treatment as people have become more aware of environmental pollution. It is estimated that one in every four Americans suffers from some level of **heavy metal poisoning**. Heavy metals, such as lead, mercury, cadmium, and arsenic, are by-products of industry. Synthetic agriculture chemicals, many of which are known to cause health problems, are also found in food, air, and water. American agriculture uses nearly 10 lb (4.5 kg) of pesticides per person on the food supply each year. These toxins have become almost unavoidable. Pesticides that are used only on crops in the southern United States have been found in the tissue of animals in the far north of Canada. DDT, a cancer-causing insecticide that has been banned for decades, is still regularly found in the fatty tissue of animals, birds, and fish, even in extremely remote regions such as the North Pole.

The problem of toxins in the environment is compounded because humans are at the top of the food chain and are more likely to be exposed to an accumulation of toxic substances in the food supply. For instance, pesticides and herbicides are sprayed on grains that are then fed to farm animals. Toxic substances are stored in the fatty tissue of those animals. In addition, those animals are often injected with synthetic hormones, **antibiotics**, and other chemicals. When people eat meat products, they are exposed to the full range of chemicals and additives used along the entire agricultural chain. Detoxification specialists call this build up of toxins *bioaccumulation*. They assert that the bioaccumulation of toxic substances over time is responsible for many physical and mental disorders, especially ones that are increasing rapidly (like asthma, cancer, and mental illness). As a result, detoxification therapies are increasing in importance and popularity.

Toxins in the body include heavy metals and various chemicals such as pesticides, pollutants, and food additives. Drugs and alcohol have toxic effects in the body. Toxins are produced as normal by-products in the intestines by the bacteria that break down food. The digestion of protein also creates toxic by-products in the body.

The body has natural methods of detoxification. Individual cells get detoxified in the lymph and circulatory system. The liver is the principle organ of detoxifi-

cation, assisted by the kidneys and intestines. Toxins can be excreted from the body by the kidneys, bowels, skin, and lungs. Detoxification treatments become necessary when the body's natural detoxification systems become overwhelmed. This can be caused by long-term effects of improper diet, **stress**, overeating, sedentary lifestyles, illness, and poor health habits in general. When a build up of toxic substances in the body creates illness, it's called toxemia. Some people's digestive tracts become unable to digest food properly, due to years of overeating and **diets** that are high in fat and processed foods and low in fiber (the average American diet). When this happens, food cannot pass through the digestive tract efficiently. Instead of being digested properly or eliminated from the bowel, food can literally rot inside the digestive tract and produce toxic by-products. This state is known as toxic colon syndrome or intestinal toxemia.

Detoxification therapies try to activate and assist the body's own detoxification processes. They also try to eliminate additional exposure to toxins and strengthen the body and immune system so that toxic imbalances won't occur in the future.

#### *Testing for toxic substances*

Detoxification specialists use a variety of tests to determine the causes contributing to toxic conditions. These causes include infections, allergies, addictions, toxic chemicals, and digestive and organ dysfunction. Blood, urine, stool, and hair analyses, as well as **allergy tests**, are used to measure a variety of bodily functions that may indicate problems. Detoxification therapists usually have access to laboratories that specialize in sophisticated diagnostic tests for toxic conditions.

People who have toxemia are often susceptible to infection because their immune systems are weakened. Infections can be caused by parasites, bacteria, viruses, and a common yeast. Therapists will screen patients for underlying infections that may be contributing to illness.

Liver function is studied closely with blood and urine tests because the liver is the principle organ in the body responsible for removing toxic compounds. When the liver detoxifies a substance from the body, it does so in two phases. Tests are performed that indicate where problems may be occurring in these phases, which may point to specific types of toxins. Blood and urine tests can also be completed that screen for toxic chemicals such as PCBs (environmental poisons), formaldehyde (a common preservative), pesticides, and heavy metals. Another useful blood test is a test for zinc deficiency, which may reveal heavy metal **poisoning**. Hair analysis is used to test for heavy metal levels in the body. Blood and urine tests check immune system activity, and hor-

mone levels can also indicate specific toxic compounds. A 24-hour urine analysis, where samples are taken around the clock, allows therapists to determine the efficiency of the digestive tract and kidneys. Together with stool analysis, these tests may indicate toxic bowel syndrome and digestive system disorders. Certain blood and urine tests may point to nutritional deficiencies and proper recovery diets can be designed for patients as well.

Detoxification therapists may also perform extensive allergy and hypersensitivity tests. Intradermal (between layers of the skin) and sublingual (under the tongue) allergy tests are used to determine a patient's sensitivity to a variety of common substances, including formaldehyde, auto exhaust, perfume, tobacco, chlorine, jet fuel, and other chemicals.

Food allergies require additional tests because these allergies often cause reactions that are delayed for several days after the food is eaten. The RAST (radioallergosorbent test) is a blood test that determines the level of antibodies (immunoglobulins) in the blood after specific foods are eaten. The cytotoxic test is a blood test that determines if certain substances affect blood cells, including foods and chemicals. The ELISA-ACT (enzyme-linked immunoserological assay activated cell test) is considered to be one of the most accurate tests for allergies and hypersensitivity to foods, chemicals, and other agents. Other tests for food allergies are the elimination and rotation diets, in which foods are systematically evaluated to determine the ones that are causing problems.

Detoxification therapists usually interview and counsel patients closely to determine and correct lifestyle, occupational, psychological, and emotional factors that may also be contributing to illness.

#### *Detoxification therapies*

Detoxification therapists use a variety of healing techniques after a diagnosis is made. The first step is to eliminate a patient's exposure to all toxic or allergenic substances. These include heavy metals, chemicals, radiation (from x rays, power lines, cell phones, computer screens, and microwaves), smog, polluted water, foods, drugs, **caffeine**, alcohol, perfume, excess noise, and stress. If mercury poisoning has been determined, the patient will be advised to have mercury fillings from the teeth removed, preferably by a holistic dentist.

Specific treatments are used to stimulate and assist the body's detoxification process. Dietary change is immediately enacted, eliminating allergic and unhealthy foods, and emphasizing foods that assist detoxification and support healing. Detoxification diets are generally low in fat, high in fiber, and vegetarian with a raw food emphasis. Processed foods, alcohol, and caffeine are

## Common Herbs Used For Detoxification

Antibiotics	Anticatarrhals (Help Eliminate Mucus)	Blood Cleaners
Clove	Boneset	Burdock root
Echinacea	Echinacea	Dandelion root
Eucalyptus	Garlic	Echinacea
Garlic	Goldenseal root	Oregon grape root
Myrrh	Hyssop	Red clover blossoms
Prickly ash bark	Sage	Yellow dock root
Propolis		
Wormwood	Yarrow	
Diaphoretics/Skin Cleaners	Diuretics	Laxatives
Boneset	Cleavers	Buckthorn
Burdock root	Corn silk	Cascara sagrada
Cayenne pepper	Horsetail	Dandelion root
Elder flowers	Juniper berries	Licorice root
Ginger root	Parsley leaf	Rhubarb root
Goldenseal root	Uva ursi	Senna leaf
Peppermint	Yarrow dock	Yellow dock
Oregon grape root		
Yellow dock		

avoided. Nutritional supplements such as **vitamins, minerals**, antioxidants, amino acids, and essential fatty acids are often prescribed. Spirulina is a sea algae that is frequently given to assist in eliminating heavy metals. Lipotropic agents are certain vitamins and nutrients that promote the flow of bile and fat from the liver.

Many herbal supplements are used in detoxification therapies as well. Milk thistle extract, called silymarin, is one of the more potent herbs for detoxifying the liver. Naturopathy, Ayurvedic medicine, and **traditional Chinese medicine** (TCM) recommend numerous herbal formulas for detoxification and immune strengthening. If infections or parasites have been found, these are treated with herbal formulas and, in difficult cases, antibiotics.

For toxic bowel syndrome and digestive tract disorders, herbal **laxatives** and high fiber foods such as psyllium seeds may be given to cleanse the digestive tract and promote elimination. Colonics are used to cleanse the lower intestines. Digestive enzymes are prescribed to improve digestion, and acidophilus and other friendly bacteria are reintroduced into the system with nutritional supplements.

Fasting is another major therapy in detoxification. Fasting is one of the quickest ways to promote the elimination of stored toxins in the body and to prompt the healing process. People with severe toxic conditions are supervised closely during fasting because the number of toxins in the body temporarily increases as they are being released.

**Chelation therapy** is used by detoxification specialists to rid the body of heavy metals. Chelates are particular substances that bind to heavy metals and speed their

elimination. Homeopathic remedies have also been shown to be effective for removing heavy metals.

Sweating therapies can also detoxify the body because the skin is a major organ of elimination. Sweating helps release those toxins that are stored in the subcutaneous (under the skin) fat cells. Saunas, therapeutic baths, and **exercise** are some of these treatments. Body therapies may also be prescribed, including **massage therapy, acupressure, shiatsu**, manual lymph drainage, and **polarity therapy**. These body therapies seek to improve circulatory and structural problems, reduce stress, and promote healing responses in the body. Mind/body therapies such as psychotherapy, counseling, and stress management techniques may be used to heal the psychological components of illness and to help patients overcome their negative patterns contributing to illness.

### Practitioners and treatment costs

The costs of detoxification therapies can vary widely, depending on the number of tests and treatments required. Detoxification treatments can be lengthy and involved since illnesses associated with toxic conditions usually develop over many years and may not clear up quickly. Detoxification treatments may be lengthy because they often strive for the holistic healing of the body, mind, and emotions.

Practitioners may be conventionally trained medical doctors with specialties in environmental medicine or interests in alternative treatment. The majority of detoxification therapists are alternative practitioners, such as naturopaths, homeopaths, ayurvedic doctors, or traditional Chinese doctors. Insurance coverage varies, depending

## KEY TERMS

**Allergen**—A foreign substance, such as mites in house dust or animal dander, that when inhaled, causes the airways to narrow and produces symptoms of asthma.

**Antibody**—A protein, also called immunoglobulin, produced by immune system cells to remove antigens (the foreign substances that trigger the immune response).

**Fibromyalgia**—A condition of debilitating pain, among other symptoms, in the muscles and the myofascia (the thin connective tissue that surrounds muscles, bones, and organs).

**Hypersensitivity**—The state where even a tiny amount of allergen can cause severe allergic reactions.

**Multiple chemical sensitivity**—A condition characterized by severe and crippling allergic reactions to commonly used substances, particularly chemicals. Also called environmental illness.

on the practitioner and the treatment involved. Consumers should review their individual insurance policies regarding treatment coverage.

### Preparations

Patients can assist diagnosis and treatment by keeping detailed diaries of their activities, symptoms, and contact with environmental factors that may be affecting their health. Reducing exposure to environmental toxins and making immediate dietary and lifestyle changes may speed the detoxification process.

### Side effects

During the detoxification process, patients may experience side effects of fatigue, malaise, aches and pains, emotional duress, **acne**, headaches, allergies, and symptoms of colds and flu. Detoxification specialists claim that these negative side effects are part of the healing process. These reactions are sometimes called *healing crises*, which are caused by temporarily increased levels of toxins in the body due to elimination and cleansing.

### Research and general acceptance

Although environmental medicine is gaining more respect within conventional medicine, detoxification

treatment is scarcely mentioned by the medical establishment. The research that exists on detoxification is largely testimonial, consisting of individual personal accounts of healing without statistics or controlled scientific experiments. In the alternative medical community, detoxification is an essential and widely accepted treatment for many illnesses and chronic conditions.

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*Journal of Occupational and Environmental Medicine*. 1114 N. Arlington Heights Rd., Arlington Heights, IL 60004. (847) 818-1800.

#### ORGANIZATIONS

- American Holistic Medical Association. 4101 Lake Boone Trail, Suite 201, Raleigh, NC 27607.  
 Cancer Prevention Coalition. 2121 West Taylor St., Chicago, IL 60612. (312) 996-2297. <<http://www.preventcancer.com>>.  
 Center for Occupational and Environmental Medicine. 7510 Northforest Dr., North Charleston, SC 29420. (843) 572-1600. <<http://www.coem.com>>.  
 Northeast Center for Environmental Medicine. P.O. Box 2716, Syracuse, NY 13220. (800) 846-ONUS.  
 Northwest Center for Environmental Medicine. 177 NE 102nd St., Portland, OR 97220. (503) 561-0966.

#### OTHER

- A Citizens Toxic Waste Manual*. Greenpeace USA, 1436 U St. NW, Washington, DC 20009. (202) 462-1177.

Douglas Dupler

## Deviated septum

### Definition

The nasal septum is a thin structure, separating the two sides of the nose. If it is not in the middle of the nose, then it is deviated.

## Description

The nasal septum is composed of two parts. Toward the back of the head the nasal septum is rigid bone, but further forward the bone becomes cartilage. With one finger in each nostril this cartilage can easily be bent back and forth. If the nasal septum is sufficiently displaced to one side, it will impede the flow of air and mucus through the nose. This condition, called a deviated septum, can cause symptoms and disease.

## Causes and symptoms

A deviated septum can be a simple variation in normal structure or the result of a broken nose. Any narrowing of the nasal passageway that it causes will threaten the drainage of secretions from the sinuses, which must pass through the nose. It is a general rule of medicine that when flow is obstructed, whether it is mucus from the sinuses or bile from the gall bladder, infection results. People with **allergic rhinitis** (hay fever) are at greater risk of obstruction because their nasal passageways are already narrowed by the swollen membranes lining them. The result is **sinusitis**, which can be acute and severe or chronic and lingering.

## Diagnosis

It is easy to see that a septum is deviated. It is more difficult to determine if that deviation needs correction. It is common for a patient to complain that he/she can breathe through only one nostril. Then the diagnosis is easy. A deviated septum may also contribute to **snoring**, **sleep apnea**, and other breathing disorders.

## Treatment

The definitive treatment is surgical repositioning of the septum, accomplished by breaking it loose and fixing it in a proper place while it heals. **Decongestants** like pseudoephedrine or phenylpropanolamine will shrink the membranes and thereby enlarge the passages. **Antihistamines**, nasal cortisone spray, and other allergy treatments may also be temporarily beneficial.

## Alternative treatment

As a palliative, saline drops and sprays are very helpful in loosening mucus in the obstructed side and preventing drying in the other side, where all the air blows. Hot peppers, such as jalapenos, can produce enough tears and discharge to flush out a stopped-up nose. An even more effective treatment is called a nasal lavage, often done using a small pot with a spout. Saline solution is poured into one nostril and allowed to flow out the other nostril.



**A close-up of person with a deviated septum.** (Custom Medical Stock Photo. Reproduced by permission.)

Then, the process is repeated in reverse. These therapies are all useful to take care of symptoms, but do not correct the problem. Nasospecific, a procedure where a deflated balloon is inserted in the nostril and inflated to a large enough degree to adjust the septal deviation, can be an alternative to surgery. A trained practitioner in the nasospecific procedure is necessary.

## Prognosis

Surgical repair is curative and carries little risk. Chronic infection can be painful and lead to complications until it is resolved. If there is continued obstruction, the infection will very likely return.

## Prevention

Avoidance of virus colds, airborne dusts, air pollution, and known allergens will minimize the irritation and swelling of the membranes lining the nasal passages.

## KEY TERMS

**Allergen**—Any substance that irritates people sensitive (allergic) to it.

**Allergic rhinitis**—Swelling and inflammation of the nasal membranes caused by sensitivity to airborne matter like pollen or cat hair.

**Saline**—A salt solution in water. Normal saline has the same salt concentration as the body, 0.9%.

**Sinuses**—The nasal sinuses, air-filled cavities surrounding the eyes and nose, like the nose itself are lined with mucus-producing membranes. They provide cleansing to the nose, resonance to the voice, and structure to the face.

**Sinusitis**—Infection of the sinuses.

**Sleep apnea**—A condition in which breathing is temporarily interrupted during sleep. It leads to high blood pressure, sleepiness, and a variety of other problems.

### Resources

#### BOOKS

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J. Ricker Polsdorfer, MD

Dextromethorphan see **Cough suppressants**

## ■ Diabetes insipidus

### Definition

Diabetes insipidus (DI) is a disorder that causes the patient to produce tremendous quantities of urine. The massively increased urine output is usually accompanied by intense thirst.

### Description

The balance of fluid within the body is maintained through a number of mechanisms. One important chemical involved in fluid balance is called antidiuretic hormone (ADH). ADH is produced by the pituitary, a small gland located at the base of the brain. In a healthy person and under normal conditions, ADH is continuously

released. ADH influences the amount of fluid that the kidneys reabsorb into the circulatory system and the amount of fluid that the kidneys pass out of the body in the form of urine.

Production of ADH is regulated by the osmolality of the circulating blood. Osmolality refers to the concentration of dissolved chemicals (such as sodium, potassium, and chloride; together called solute) circulating in the fluid base of the blood (plasma). When there is very little fluid compared to the concentration of solute, the pituitary will increase ADH production. This tells the kidneys to retain more water and to decrease the amount of urine produced. As fluid is retained, the concentration of solute will normalize. At other times, when the fluid content of the blood is high in comparison to the concentration of solute, ADH production will decrease. The kidneys are then free to pass an increased amount of fluid out of the body in the urine. Again, this will allow the plasma osmolality to return to normal.

Diabetes insipidus occurs when either the amount of ADH produced by the pituitary is below normal (central DI), or the kidneys' ability to respond to ADH is defective (nephrogenic DI). In either case, a person with DI will pass extraordinarily large quantities of urine, sometimes reaching 10 or more liters each day. At the same time, the patient's blood will be very highly concentrated, with low fluid volume and high concentrations of solute.

DI occurs on average when a person is about 24 years old, and occurs more frequently in males than in females.

### Causes and symptoms

DI may run in families. The cause of this type of DI is unknown. Other times, central DI can be caused by:

- an injury to the head
- brain surgery
- cancers that have spread to the pituitary gland (most commonly occurring with **breast cancer**)
- sarcoidosis (or other related disorders), causing destruction of the pituitary gland
- any condition or illness that causes decreased oxygen delivery to the brain
- the use of certain medications that decrease ADH production (like the antiseizure drug phenytoin)
- the excessive use of alcohol

Central DI may also occur in women who are pregnant or have just given birth, and in patients with **AIDS** who have suffered certain types of brain infections. Nephrogenic DI sometimes occurs in patients who are

taking the medication lithium, patients who have high levels of blood calcium, and patients who are pregnant.

DI is easily confused with an entirely unrelated disorder, psychogenic polydipsia. Polydipsia refers to drinking large amounts of water. Psychogenic polydipsia is a psychiatric problem that makes a person drink huge quantities of water uncontrollably.

Symptoms of DI include extreme thirst and the production of tremendous quantities of urine. Patients with DI typically drink huge amounts of water, and usually report a specific craving for cold water. When the amount of water passed in the urine exceeds the patient's ability to drink ample replacement water, the patient may begin to suffer from symptoms of **dehydration**. These symptoms include weakness, **fatigue**, **fever**, low blood pressure, increased heart rate, **dizziness**, and confusion. If left untreated, the patient could lapse into unconsciousness and die.

## Diagnosis

Diagnosis should be suspected in any patient with sudden increased thirst and urination. Laboratory examination of urine will reveal very dilute urine, made up mostly of water with no solute. Examination of the blood will reveal very concentrated blood, high in solute and low in fluid volume.

A water deprivation test may be performed. This test requires a patient to stop all fluid intake. The patient is weighed just before the test begins, and urine is collected and examined hourly. The test is stopped when:

- the patient has lost more than 5% of his or her original body weight
- the patient has reached certain limits of low blood pressure and increased heart rate
- the urine is no longer changing significantly from one sample to the next in terms of solute concentration

The next step of the test involves injecting a synthetic form of ADH, with one last urine sample examined 60 minutes later. Comparing plasma and urine osmolality allows the doctor to diagnose either central DI, nephrogenic DI, partial DI, or psychogenic polydipsia.

## Treatment

A number of medications can be given to decrease the quantity of fluid passed out into the urine. These include vasopressin (Pitressin) injected and desmopressin acetate (DDAVP) inhaled through the nose. Other medications that may be given include some antidiuretic drugs (chlorpropamide, clofibrate, carbamazepine). Patients with nephrogenic DI, however, will also require

## KEY TERMS

**Concentration**—Refers to the amount of solute present in a solution, compared to the total amount of solvent.

**Dilute**—A solution that has comparatively more fluid in it, relative to the quantity of solute.

**Osmolality**—A measure of the solute-to-solvent concentration of a solution.

**Solute**—Solid substances that are dissolved in liquid in order to make a solution.

special **diets** that restrict the amount of solute taken in. These patients are also treated with a type of medication called a thiazide diuretic.

## Prognosis

Uncomplicated diabetes insipidus is controllable with adequate intake of water and most patients can lead normal lives.

## Resources

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## Diabetes mellitus

### Definition

Diabetes mellitus is a condition in which the pancreas no longer produces enough insulin or when cells stop responding to the insulin that is produced, so that

glucose in the blood cannot be absorbed into the cells of the body. Symptoms include frequent urination, lethargy, excessive thirst, and hunger. The treatment includes changes in diet, oral medications, and in some cases, daily injections of insulin.

## Description

Diabetes mellitus is a chronic disease that causes serious health complications including renal (kidney) failure, heart disease, **stroke**, and blindness. Approximately 14 million Americans (about 5% of the population) have diabetes. Unfortunately, as many as one-half are unaware that they have it.

## Background

Every cell in the human body needs energy in order to function. The body's primary energy source is glucose, a simple sugar resulting from the digestion of foods containing carbohydrates (sugars and starches). Glucose from the digested food circulates in the blood as a ready energy source for any cells that need it. Insulin is a hormone or chemical produced by cells in the pancreas, an organ located behind the stomach. Insulin bonds to a receptor site on the outside of cell and acts like a key to open a doorway into the cell through which glucose can enter. Some of the glucose can be converted to concentrated energy sources like glycogen or fatty acids and saved for later use. When there is not enough insulin produced or when the doorway no longer recognizes the insulin key, glucose stays in the blood rather entering the cells.

The body will attempt to dilute the high level of glucose in the blood, a condition called hyperglycemia, by drawing water out of the cells and into the bloodstream in an effort to dilute the sugar and excrete it in the urine. It is not unusual for people with undiagnosed diabetes to be constantly thirsty, drink large quantities of water, and urinate frequently as their bodies try to get rid of the extra glucose. This creates high levels of glucose in the urine.

At the same time that the body is trying to get rid of glucose from the blood, the cells are starving for glucose and sending signals to the body to eat more food, thus making patients extremely hungry. To provide energy for the starving cells, the body also tries to convert fats and proteins to glucose. The breakdown of fats and proteins for energy causes acid compounds called ketones to form in the blood. Ketones will also be excreted in the urine. As ketones build up in the blood, a condition called ketoacidosis can occur. This condition can be life threatening if left untreated, leading to **coma** and **death**.

## *Types of diabetes mellitus*

Type I diabetes, sometimes called juvenile diabetes, begins most commonly in childhood or adolescence. In this form of diabetes, the body produces little or no insulin. It is characterized by a sudden onset and occurs more frequently in populations descended from Northern European countries (Finland, Scotland, Scandinavia) than in those from Southern European countries, the Middle East, or Asia. In the United States, approximately three people in 1,000 develop Type I diabetes. This form is also called insulin-dependent diabetes because people who develop this type need to have daily injections of insulin.

Brittle diabetics are a subgroup of Type I where patients have frequent and rapid swings of blood sugar levels between hyperglycemia (a condition where there is too much glucose or sugar in the blood) and **hypoglycemia** (a condition where there is abnormally low levels of glucose or sugar in the blood). These patients may require several injections of different types of insulin during the day to keep the blood sugar level within a fairly normal range.

The more common form of diabetes, Type II, occurs in approximately 3–5% of Americans under 50 years of age, and increases to 10–15% in those over 50. More than 90% of the diabetics in the United States are Type II diabetics. Sometimes called age-onset or adult-onset diabetes, this form of diabetes occurs most often in people who are overweight and who do not **exercise**. It is also more common in people of Native American, Hispanic, and African-American descent. People who have migrated to Western cultures from East India, Japan, and Australian Aboriginal cultures are also more likely to develop Type II diabetes than those who remain in their original countries.

Type II is considered a milder form of diabetes because of its slow onset (sometimes developing over the course of several years) and because it can usually be controlled with diet and oral medication. The consequences of uncontrolled and untreated Type II diabetes, however, are just as serious as those for Type I. This form is also called noninsulin-dependent diabetes, a term that is somewhat misleading. Many people with Type II diabetes can control the condition with diet and oral medications, however, insulin injections are sometimes necessary if treatment with diet and oral medication is not working.

Another form of diabetes called **gestational diabetes** can develop during **pregnancy** and generally resolves after the baby is delivered. This diabetic condition develops during the second or third trimester of pregnancy in about 2% of pregnancies. The condition is usually treated by diet, however, insulin injections may

be required. These women who have diabetes during pregnancy are at higher risk for developing Type II diabetes within 5–10 years.

Diabetes can also develop as a result of pancreatic disease, **alcoholism**, **malnutrition**, or other severe illnesses that stress the body.

## Causes and symptoms

### Causes

The causes of diabetes mellitus are unclear, however, there seem to be both hereditary (genetic factors passed on in families) and environmental factors involved. Research has shown that some people who develop diabetes have common genetic markers. In Type I diabetes, the immune system, the body's defense system against infection, is believed to be triggered by a virus or another microorganism to destroy the cells in the pancreas that produce insulin. In Type II diabetes, age, **obesity**, and family history of diabetes play a role.

In Type II diabetes, the pancreas may produce enough insulin, however, cells have become resistant to the insulin produced and it may not work as effectively. Symptoms of Type II diabetes can begin so gradually that a person may not know that they have it. Early signs are lethargy, extreme thirst, and frequent urination. Other symptoms may include sudden weight loss, slow wound healing, urinary tract infections, gum disease, or blurred vision. It is not unusual for Type II diabetes to be detected while a patient is seeing a doctor about another health concern that is actually being caused by the yet undiagnosed diabetes.

Individuals who are at high risk of developing Type II diabetes mellitus include people who:

- are obese (more than 20% above their ideal body weight)
- have a relative with diabetes mellitus
- belong to a high-risk ethnic population (African-American, Native American, Hispanic, or Native Hawaiian)
- have been diagnosed with gestational diabetes or have delivered a baby weighing more than 9 lbs (4 kg)
- have high blood pressure (140/90 mmHg or above)
- have a high density lipoprotein cholesterol level less than or equal to 35 mg/dL and/or a triglyceride level greater than or equal to 250 mg/dL
- have had impaired glucose tolerance or impaired **fasting** glucose on previous testing

Several common medications can impair the body's use of insulin, causing a condition known as secondary diabetes. These medications include treatments for high



**Wrinkled, dehydrated skin of a person in a diabetic coma.** Untreated diabetes mellitus results in elevated blood glucose levels, causing a variety of symptoms that can culminate in a diabetic coma. (Photo Researchers, Inc. Reproduced by permission.)

blood pressure (furosemide, clonidine, and thiazide diuretics), drugs with hormonal activity (**oral contraceptives**, thyroid hormone, progestins, and glucocorticoids), and the anti-inflammation drug indomethacin. Several drugs that are used to treat **mood disorders** (such as **anxiety** and depression) can also impair glucose absorption. These drugs include haloperidol, lithium carbonate, phenothiazines, tricyclic antidepressants, and adrenergic agonists. Other medications that can cause diabetes symptoms include isoniazid, nicotinic acid, cimetidine, and heparin.

### Symptoms

Symptoms of diabetes can develop suddenly (over days or weeks) in previously healthy children or adolescents, or can develop gradually (over several years) in overweight adults over the age of 40. The classic symptoms include feeling tired and sick, frequent urination, excessive thirst, excessive hunger, and weight loss.

Ketoacidosis, a condition due to **starvation** or uncontrolled diabetes, is common in Type I diabetes. Ketones are acid compounds that form in the blood when the body breaks down fats and proteins. Symptoms include abdominal **pain**, vomiting, rapid breathing, extreme lethargy and drowsiness. Patients with ketoacidosis will also have a sweet breath odor. Left untreated, this condition can lead to coma and death.

With Type II diabetes, the condition may not become evident until the patient presents for medical treatment for some other condition. A patient may have heart disease, chronic infections of the gums and urinary tract, blurred vision, numbness in the feet and legs, or slow-healing **wounds**. Women may experience genital **itching**.

## KEY TERMS

**Cataracts**—A condition where the lens of the eye becomes cloudy.

**Diabetic peripheral neuropathy**—A condition where the sensitivity of nerves to pain, temperature, and pressure is dulled particularly in the legs and feet.

**Diabetic retinopathy**—A condition where the tiny blood vessels to the retina, the tissues that sense light at the back of the eye, are damaged, leading to blurred vision, sudden blindness, or black spots, lines, or flashing light in the field of vision.

**Glaucoma**—A condition where pressure within the eye causes damage to the optic nerve, which sends visual images to the brain.

**Hyperglycemia**—A condition where there is too much glucose or sugar in the blood.

**Hypoglycemia**—A condition where there is too little glucose or sugar in the blood.

**Insulin**—A hormone or chemical produced by the pancreas, insulin is needed by cells of the body in order to use glucose (sugar), the body's main source of energy.

**Ketoacidosis**—A condition due to starvation or uncontrolled Type I diabetes. Ketones are acid compounds that form in the blood when the body breaks down fats and proteins. Symptoms include abdominal pain, vomiting, rapid breathing, extreme tiredness, and drowsiness.

**Kidney dialysis**—A process where blood is filtered through a dialysis machine to remove waste products that would normally be removed by the kidneys. The filtered blood is then circulated back into the patient. This process is also called renal dialysis.

**Pancreas**—A gland located behind the stomach that produces insulin.

## Diagnosis

Diabetes is suspected based on symptoms. Urine and blood tests can be used to confirm a diagnosis of diabetes based on the amount of glucose. Urine tests can also detect ketones and protein in the urine that may help diagnose diabetes and assess how well the kidneys are functioning. These tests can also be used to monitor the disease once the patient is on a standardized diet, oral medications, or insulin.

### Urine tests

Clinistix and Diastix are paper strips or dipsticks that change color when dipped in urine. The test strip is compared to a chart which shows the amount of glucose in the urine based on the change in color. The level of glucose in the urine lags behind the level of glucose in the blood. Testing the urine with a test stick, paper strip, or tablet that changes color when sugar is present is not as accurate as blood testing, however it can give a fast and simple reading.

Ketones in the urine can be detected using similar types of dipstick tests (Acetest or Ketostix). Ketoacidosis can be a life-threatening situation in Type I diabetics, so having a quick and simple test to detect ketones can assist in establishing a diagnosis sooner.

Another dipstick test can determine the presence of protein or albumin in the urine. Protein in the urine can

indicate problems with kidney function and can be used to track the development of renal failure. A more sensitive test for urine protein uses radioactively tagged chemicals to detect microalbuminuria, small amounts of protein in the urine, that may not show up on dipstick tests.

### Blood tests

**FASTING GLUCOSE TEST.** Blood is drawn from a vein in the patient's arm after a period at least eight hours when the patient has not eaten, usually in the morning before breakfast. The red blood cells are separated from the sample and the amount of glucose is measured in the remaining plasma. A plasma level of 7.8 mmol/L (200 mg/L) or greater can indicate diabetes. The fasting glucose test is usually repeated on another day to confirm the results.

**POSTPRANDIAL GLUCOSE TEST.** Blood is taken right after the patient has eaten a meal.

**ORAL GLUCOSE TOLERANCE TEST.** Blood samples are taken from a vein before and after a patient drinks a thick, sweet syrup of glucose and other sugars. In a non-diabetic, the level of glucose in the blood goes up immediately after the drink and then decreases gradually as insulin is used by the body to metabolize, or absorb, the sugar. In a diabetic, the glucose in the blood goes up and stays high after drinking the sweetened liquid. A plasma glucose level of 11.1 mmol/L (200 mg/dL) or higher at two hours after drinking the syrup and at one other point during the two-hour test period confirms the diagnosis of diabetes.

A diagnosis of diabetes is confirmed if there are symptoms of diabetes and a plasma glucose level of at least 11.1 mmol/L, a fasting plasma glucose level of at least 7 mmol/L; or a two-hour plasma glucose level of at least 11.1 mmol/L during an oral glucose tolerance test.

Home blood glucose monitoring kits are available so patients with diabetes can monitor their own levels. A small needle or lancet is used to prick the finger and a drop of blood is collected and analyzed by a monitoring device. Some patients may test their blood glucose levels several times during a day and use this information to adjust their doses of insulin.

## Treatment

There is currently no cure for diabetes; the condition, however, can be managed so that patients can live a relatively normal life. Treatment of diabetes focuses on two goals: keeping blood glucose within normal range and preventing the development of long-term complications. Careful monitoring of diet, exercise, and blood glucose levels are as important as the use of insulin or oral medications in preventing complications of diabetes.

### Dietary changes

Diet and moderate exercise are the first treatments implemented in diabetes. For many Type II diabetics, weight loss may be an important goal in helping them to control their diabetes. A well-balanced, nutritious diet provides approximately 50–60% of calories from carbohydrates, approximately 10–20% of calories from protein, and less than 30% of calories from fat. The number of calories required by an individual depends on their age, weight, and activity level. The calorie intake also needs to be distributed over the course of the entire day so surges of glucose entering the blood system are kept to a minimum.

Keeping track of the number of calories provided by different foods can become complicated, so patients are usually advised to consult a nutritionist or dietitian. An individualized, easy to manage diet plan can be set up for each patient. Both the American Diabetes Association and the American Dietetic Association recommend **diets** based on the use of food exchange lists. Each food exchange contains a known amount of calories in the form of protein, fat, or carbohydrate. A patient's diet plan will consist of a certain number of exchanges from each food category (meat or protein, fruits, breads and starches, vegetables, and fats) to be eaten at meal times and as snacks. Patients have flexibility in choosing which foods they eat as long as they stick with the number of exchanges prescribed.

For many Type II diabetics, weight loss is an important factor in controlling their condition. The food

exchange system, along with a plan of moderate exercise, can help them lose excess weight and improve their overall health.

### Oral medications

Oral medications are available to lower blood glucose in Type II diabetics. The drugs first prescribed for Type II diabetes are in a class of compounds called sulfonylureas and include tolbutamide, tolazamide, acetohexamide, and chlorpropamide. Newer drugs in the same class are now available and include glyburide, glimeperide, and glipizide. The way that these drugs work is not well understood, however, they seem to stimulate cells of the pancreas to produce more insulin. New medications that are available to treat diabetes include metformin, acarbose, and troglitzone. The choice of the right medication depends in part on the individual patient profile. All drugs have side effects that may make them inappropriate for particular patients. Some for example, may stimulate weight gain or cause stomach irritation, so they may not be the best treatment for someone who is already overweight or who also has stomach ulcers. While these medications are an important aspect of treatment for Type II diabetes, they are not a substitute for a well planned diet and moderate exercise. Oral medications are not effective for Type I diabetes, in which the patient produces little or no insulin.

### Insulin

Patients with Type I diabetes need daily injections of insulin to help their bodies use glucose. The amount and type of insulin required depends on the height, weight, age, food intake, and activity level of the individual diabetic patient. Some patients with Type II diabetes may need to use insulin injections if their diabetes cannot be controlled with diet, exercise, and oral medication. Injections are given subcutaneously, that is, just under the skin, using a small needle and syringe. Injection sites can be anywhere on the body where there is looser skin, including the upper arm, abdomen, or upper thigh.

Purified human insulin is most commonly used, however, insulin from beef and pork sources are also available. Insulin may be given as an injection of a single dose of one type of insulin once a day. Different types of insulin can be mixed and given in one dose or split into two or more doses during a day. Patients who require multiple injections over the course of a day may be able to use an insulin pump that administers small doses of insulin on demand. The small battery-operated pump is worn outside the body and is connected to a needle that is inserted into the abdomen. Pumps can be programmed to inject small doses of insulin at various times during the day, or the patient may be able to adjust the insulin doses to coincide with meals and exercise.

Regular insulin is fast-acting and starts to work within 15–30 minutes, with its peak glucose-lowering effect about two hours after it is injected. Its effects last for about four to six hours. NPH (neutral protamine Hagedorn) and Lente insulin are intermediate-acting, starting to work within one to three hours and lasting up to 18–26 hours. Ultra-lente is a long-acting form of insulin that starts to work within four to eight hours and lasts 28–36 hours.

Hypoglycemia, or low blood sugar, can be caused by too much insulin, too little food (or eating too late to coincide with the action of the insulin), alcohol consumption, or increased exercise. A patient with symptoms of hypoglycemia may be hungry, cranky, confused, and tired. The patient may become sweaty and shaky. Left untreated, the patient can lose consciousness or have a seizure. This condition is sometimes called an insulin reaction and should be treated by giving the patient something sweet to eat or drink like a candy, sugar cubes, juice, or another high sugar snack.

### Surgery

Transplantation of a healthy pancreas into a diabetic patient is a successful treatment, however, this transplant is usually done only if a kidney transplant is performed at the same time. Although a pancreas transplant is possible, it is not clear if the potential benefits outweigh the risks of the surgery and drug therapy needed.

### Alternative treatment

Since diabetes can be life-threatening if not properly managed, patients should not attempt to treat this condition without medical supervision. A variety of alternative therapies can be helpful in managing the symptoms of diabetes and supporting patients with the disease. **Acupuncture** can help relieve the pain associated with **diabetic neuropathy** by stimulation of certain points. A qualified practitioner should be consulted. Herbal remedies may also be helpful in managing diabetes. Although there is no herbal substitute for insulin, some herbs may help adjust blood sugar levels or manage other diabetic symptoms. Some options include:

- fenugreek (*Trigonella foenum-graecum*) has been shown in some studies to reduce blood insulin and glucose levels while also lowering cholesterol
- bilberry (*Vaccinium myrtillus*) may lower blood glucose levels, as well as helping to maintain healthy blood vessels
- garlic (*Allium sativum*) may lower blood sugar and cholesterol levels
- onions (*Allium cepa*) may help lower blood glucose levels by freeing insulin to metabolize it

- cayenne pepper (*Capsicum frutescens*) can help relieve pain in the peripheral nerves (a type of diabetic neuropathy)
- ginkgo (*Ginkgo biloba*) may maintain blood flow to the retina, helping to prevent diabetic retinopathy

Any therapy that lowers stress levels can also be useful in treating diabetes by helping to reduce insulin requirements. Among the alternative treatments that aim to lower stress are **hypnotherapy**, **biofeedback**, and **meditation**.

### Prognosis

Uncontrolled diabetes is a leading cause of blindness, end-stage renal disease, and limb amputations. It also doubles the risks of heart disease and increases the risk of stroke. Eye problems including **cataracts**, **glaucoma**, and diabetic retinopathy are also more common in diabetics.

**Diabetic peripheral neuropathy** is a condition where nerve endings, particularly in the legs and feet become less sensitive. Diabetic foot ulcers are a particular problem since the patient does not feel the pain of a blister, callous, or other minor injury. Poor blood circulation in the legs and feet contribute to delayed wound healing. The inability to sense pain along with the complications of delayed wound healing can result in minor injuries, blisters, or callouses becoming infected and difficult to treat. In cases of severe infection, the infected tissue begins to break down and rot away. The most serious consequence of this condition is the need for **amputation** of toes, feet, or legs due to severe infection.

Heart disease and kidney disease are common complications of diabetes. Long-term complications may include the need for **kidney dialysis** or a kidney transplant due to kidney failure.

Babies born to diabetic mothers have an increased risk of **birth defects** and distress at birth.

### Prevention

Research continues on ways to prevent diabetes and to detect those at risk for developing diabetes. While the onset of Type I diabetes is unpredictable, the risk of developing Type II diabetes can be reduced by maintaining ideal weight and exercising regularly. The physical and emotional stress of surgery, illness, pregnancy, and alcoholism can increase the risks of diabetes, so maintaining a healthy lifestyle is critical to preventing the onset of Type II diabetes and preventing further complications of the disease.

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### ORGANIZATIONS

- American Diabetes Association. 1701 North Beauregard Street, Alexandria, VA 22311. (800) 342-2383. <<http://www.diabetes.org>>.
- American Dietetic Association. 216 W. Jackson Blvd., Chicago, IL 60606-6995. (312) 899-0040. <<http://www.eatright.org>>.
- Juvenile Diabetes Foundation. 120 Wall St., 19th Floor, New York, NY 10005. (800) 533-2873. <<http://www.jdf.org>>.
- National Diabetes Information Clearinghouse. 1 Information Way, Bethesda, MD 20892-3560. (800) 860-8747. <<http://www.niddk.nih.gov/health/diabetes/ndic.htm>>.
- 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>>.

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Altha Roberts Edgren

Diabetic control index see **Glycosylated hemoglobin test**

## Diabetic foot infections

### Definition

Diabetic foot infections are infections that can develop in the skin, muscles, or bones of the foot as a result of the nerve damage and poor circulation that is associated with diabetes.

### Description

People who have diabetes have a greater-than-average chance of developing foot infections. Because a person who has diabetes may not feel foot **pain** or discomfort, problems can remain undetected until **fever**, weakness, or other signs of systemic infection appear. As a result, even minor irritations occur more often, heal more slowly, and are more likely to result in serious health problems.

With diabetes, foot infections occur more frequently because the disease causes nervous system changes and poor circulation. Because the nerves that control sweating no longer work, the skin of the feet can become very dry and cracked, and calluses tend to occur more frequently and build up faster. If not trimmed regularly, these calluses can turn into open sores or ulcers. Because diabetic nerve damage can cause a loss of sensation (**neuropathy**), if the feet are not regularly inspected, an ulcer can quickly become infected and, if not treated, may result in the **death of tissue (gangrene)** or **amputation**.

The risk of infection is greatest for people who are over the age of 60 and for those who have one or more of the following:

- poorly controlled diabetes
- foot ulcers
- laser treatment for changes in the retina
- kidney or vascular disease
- loss of sensation (neuropathy)

### Causes and symptoms

Bacteria can cause an infection through small cracks (**fissures**) that can develop in the dry skin around the heel and on other parts of the foot or through corns, calluses, blisters, hangnails, or ulcers. If not treated, the bacterial infection can destroy skin, tissue, and bone or spread throughout the body.

Common sites of diabetic foot infections include the following:

- blisters, corns, or callouses that bleed beneath the skin
- bunions, hammertoes, or other abnormalities in the bones of the foot



**Persons with diabetes often suffer from foot ulcers, as shown above.** (Custom Medical Stock Photo. Reproduced by permission.)

- scar tissue that has grown over the site of an earlier infection
- foot ulcers caused by pressure, nerve damage, or poor circulation (Ulcers occur most often over the ball of the foot, on the bottom of the big toe, or on the sides of the foot due to poorly fitting shoes.)
- injuries that tear or puncture the skin.

## Diagnosis

A physician who specializes in the treatment of the foot (podiatrist) or the doctor who normally treats the patient's diabetes will treat the infection. An x ray of the foot will be taken to determine whether the bone has become infected. A sample from the wound will be cultured to identify the organism that is causing the infection so that the appropriate antibiotic can be selected.

## Treatment

From the results of the culture, the appropriate antibiotic will be prescribed. Any dead or infected tissue will be surgically removed and, if necessary, a cast and/or special shoes may be used to protect the area. In addition, the patient will be instructed to keep off their feet. If the ulcer does not heal, the physician may perform surgery to increase blood flow to the foot. It is also important for the patient to practice good diabetes control and keep blood glucose levels from getting too high.

## Alternative treatment

**Acupuncture** and vitamin C can boost the body's infection-fighting ability. A variety of other **vitamins** and herbs may improve general health and diabetes control. Because diabetes is a potentially deadly disease, it can be

## KEY TERMS

**Fissure**—A deep crack.

**Neuropathy**—An abnormality of the nerves outside the brain and spinal cord.

**Ulcer**—A sore or lesion.

dangerous to try alternative approaches without a doctor's approval or without consulting a trained practitioner of alternative medicine.

## Prognosis

Without proper treatment, diabetic foot infections can lead to serious illness, gangrene, amputation, and even death if the infection spreads throughout the body. If treated properly and the patient practices good **foot care**, the prognosis is generally optimistic.

## Prevention

There are many things that a diabetic individual can do to prevent the occurrence of foot infections, including the following:

- control blood glucose and do not allow it to get too high
- avoid **smoking**
- keep blood pressure and cholesterol under control
- exercise to stimulate blood flow
- keep feet clean, dry, and warm
- check your feet every day for blisters, scratches, and skin that is hard, broken, inflamed or that feels hot or cold when touched
- after bathing, carefully dry feet and apply thin coat of petroleum jelly or hand cream to prevent dry skin from cracking
- use a pumice stone and emery board to trim calluses
- do not neglect an ulcer, should one develop

## Resources

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Maureen Haggerty

## Diabetic ketoacidosis

**Definition**

Diabetic ketoacidosis is a dangerous complication of **diabetes mellitus** in which the chemical balance of the body becomes far too acidic.

**Description**

Diabetic ketoacidosis (DKA) always results from a severe insulin deficiency. Insulin is the hormone secreted by the body to lower the blood sugar levels when they become too high. Diabetes mellitus is the disease resulting from the inability of the body to produce or respond properly to insulin, required by the body to convert glucose to energy. In childhood diabetes, DKA complications represent the leading cause of **death**, mostly due to the accumulation of abnormally large amounts of fluid in the brain (cerebral **edema**). DKA combines three major features: hyperglycemia, meaning excessively high blood sugar levels hyperketonemia, meaning an overproduction of ketones by the body; and acidosis, meaning that the blood has become too acidic.

Insulin deficiency is responsible for all three conditions: the body glucose goes largely unused since most cells are unable to transport glucose into the cell without the presence of insulin; this condition makes the body use stored fat as an alternative source instead of the unavailable glucose for energy, a process that produces acidic ketones, which build up because they require insulin to be broken down. The presence of excess ketones in the bloodstream in turn causes the blood to become more acidic than the body tissues, which creates a toxic condition.

**Causes and symptoms**

DKA is most commonly seen in individuals with type I diabetes, under 19 years of age and is usually caused by the interruption of their insulin treatment or by acute infection or trauma. A small number of people with type II diabetes also experience ketoacidosis, but this is rare given the fact that type II diabetics still produce some insulin naturally. When DKA occurs in type II patients, it is usually caused by a decrease in food intake and an increased insulin deficiency due to hyperglycemia.

Some common DKA symptoms include:

- high blood sugar levels
- frequent urination (polyuria) and thirst
- fatigue and lethargy
- nausea
- vomiting
- abdominal pain
- fruity odor to breath
- rapid, deep breathing
- muscle stiffness or aching
- coma

**Diagnosis**

Diagnosis requires the demonstration of hyperglycemia, hyperketonemia, and acidosis. DKA is established if the patient's urine or blood is strongly positive for glucose and ketones. Normal glucose levels in a non-diabetic person on average range from 80–110 mg/dL. A person with diabetes will typically fluctuate outside those parameters. DKA glucose levels exceed 250 mg/dL and can reach 400 to 800 mg/dL. A low serum bicarbonate level (usually below 15 mEq/L) is also present, indicative of acidosis.

A blood test or **urinalysis** can quickly determine the concentration of glucose in the bloodstream. Test strips are available to patients commercially can submerge in urine to detect the presence or concentration of ketones.

**Treatment**

Ketoacidosis is treated under medical supervision and usually in a hospital setting.

Basic treatment includes:

- administering insulin to correct the hyperglycemia and hyperketonemia

## KEY TERMS

**Acidosis**—A condition that causes the pH of the blood to drop and become more acidic.

**Diabetes mellitus**—Disease characterized by the inability of the body to produce or respond properly to insulin, which is required by the body to convert glucose to energy.

**Edema**—The presence of abnormally large amounts of fluid in the intercellular tissue spaces of the body.

**Glucose**—The type of sugar found in the blood.

**Hyperglycemia**—Condition characterized by excessively high levels of glucose in the blood, and occurs when the body does not have enough insulin or cannot use the insulin it does have to turn glucose into energy. Hyperglycemia is often indicative of diabetes that is out of control.

**Hyperketonemia**—Condition characterized by an overproduction of ketones by the body.

**Hypoglycemia**—Lower than normal levels of glucose in the blood.

**Hypokalemia**—A deficiency of potassium in the blood.

**Insulin**—A hormone secreted by the pancreas in response to high blood sugar levels that induces hypoglycemia. Insulin regulates the body's use of

glucose and the levels of glucose in the blood by acting to open the cells so that they can intake glucose.

**Ketones**—Poisonous acidic chemicals produced by the body when fat instead of glucose is burned for energy. Breakdown of fat occurs when not enough insulin is present to channel glucose into body cells.

**Lactic acidosis**—A serious condition caused by the build up of lactic acid in the blood, causing it to become excessively acidic. Lactic acid is a by-product of glucose metabolism.

**Metabolism**—The sum of all chemical reactions that occur in the body resulting in growth, transformation of foodstuffs into energy, waste elimination and other bodily functions.

**Polyuria**—Excessive secretion of urine.

**Type I diabetes**—Also called juvenile diabetes. Type I diabetes typically begins early in life. Affected individuals have a primary insulin deficiency and must take insulin to stay alive.

**Type II diabetes**—Type II diabetes is the most common form of diabetes and usually appears in middle aged adults. It is often associated with obesity and may be delayed or controlled with diet and exercise.

- Replacing fluids intravenously lost through excessive urination and vomiting
- Balancing electrolytes to re-establish the chemical equilibrium of the blood and prevent potassium deficiency (**hypokalemia**) during treatment
- Treatment for any associated bacterial infection

### Prognosis

With proper medical attention, DKA is almost always successfully treated. The DKA mortality rate is about 10%. Coma on admission adversely affects the prognosis. The major causes of death are circulatory collapse, hypokalemia, infection, and cerebral edema.

### Prevention

Once diabetes has been diagnosed, prevention measures to avoid DKA include regular monitoring of blood glucose, administration of insulin, and lifestyle maintenance. Glucose monitoring is especially important dur-

ing periods of **stress**, infection, and trauma when glucose concentrations typically increase as a response to these situations. Ketone tests should also be performed during these periods or when glucose is elevated.

### Resources

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#### ORGANIZATIONS

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Juvenile Diabetes Foundation. 120 Wall St., New York, NY 10005. (800) 533-CURE. <<http://www.jdf.org/>>.

National Institute of Diabetes and Digestive and Kidney Disorders (NIDDK). 31 Center Drive, MSC 2560, Bethesda, MD 20892-2560. <<http://www.niddk.nih.gov>>.

Gary Gilles

## Diabetic neuropathy

### Definition

Diabetic neuropathy is a nerve disorder caused by **diabetes mellitus**. Diabetic neuropathy may be diffuse, affecting several parts of the body, or focal, affecting a specific nerve and part of the body.

### Description

The nervous system consists of two major divisions: the central nervous systems (CNS) which includes the brain, the cranial nerves, and the spinal cord, and the peripheral nervous system (PNS) which includes the nerves that link the CNS with the sensory organs, muscles, blood vessels, and glands of the body. These peripheral nerves are either motor, meaning that they are involved in motor activity such as walking, or sensory, meaning that they carry sensory information back to the CNS. The PNS also works with the CNS to regulate involuntary (autonomic) processes such as breathing, heartbeat, blood pressure, etc.

There are two types of diffuse diabetic neuropathy that affect different nervous system functions. Diffuse **peripheral neuropathy** primarily affects the limbs, damaging the nerves of the feet and hands. Autonomic neuropathy is the other form of diffuse neuropathy and it affects the heart and other internal organs.

Focal—or localized—diabetic neuropathy affects specific nerves, most commonly in the torso, leg, or head.

Diabetic neuropathy can lead to muscular weakness, loss of feeling or sensation, and loss of autonomic functions such as digestion, erection, bladder control and sweating among others.

The longer a person has diabetes, the more likely the development of one or more forms of neuropathy. Approximately 60–70% of patients with diabetes have neuropathy, but only about 5% will experience painful symptoms.

### Causes and symptoms

The exact cause of diabetic neuropathy is not known. Researchers believe that the process of nerve damage is related to high glucose concentrations in the blood that could cause chemical changes in nerves, disrupting their ability to effectively send messages. High blood glucose is also known to damage the blood vessels that carry oxygen and other nutrients to the nerves. In addition, some people may have a genetic predisposition to develop neuropathy.

There is a wide range of symptoms associated with diabetic neuropathy, and they depend on which nerves and parts of the body affected and also on the type of neuropathy present. Some patients have very mild symptoms, while others are severely disabled.

Common symptoms of diffuse peripheral neuropathy include:

- numbness and feelings of tingling or burning
- insensitivity to **pain**
- needle-like jabs of pain
- extreme sensitivity to touch
- loss of balance and coordination

Common symptoms of diffuse autonomic neuropathy include:

- impaired urination and sexual function
- bladder infections
- stomach disorders, due to the impaired ability of the stomach to empty (gastric stasis)
- nausea, vomiting, bloating
- dizziness, lightheadedness, **fainting** spells
- loss of appetite

Common symptoms of focal neuropathy include:

- pain in the front of a thigh
- severe pain in the lower back
- pain in the chest or stomach
- ache behind an eye
- double vision
- paralysis on one side of the face

In severe diabetic neuropathy loss of sensation can lead to injuries that are unnoticed, progressing to infections, ulceration, and possibly **amputation**.

### Diagnosis

The diagnosis of neuropathy is based on the symptoms that present during a physical exam. Pain assessment is usually the first step. Patients may have more than one type of pain, and the history helps the doctor determine whether a the pain has a neuropathic cause.

The exam may include:

- a screening test for lost sensation
- nerve conduction studies to check the flow of electric current through a nerve
- electromyography (EMG) to see how well muscles respond to electrical impulses transmitted by nearby nerves.

## KEY TERMS

**Central nervous system (CNS)**—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.

**Diabetes mellitus**—Disease characterized by the inability of the body to produce or respond properly to insulin, required by the body to convert glucose to energy.

**Glucose**—The type of sugar found in the blood.

**Peripheral nervous system (PNS)**—One of the two major divisions of the nervous system. PNS nerves link the central nervous system with sensory organs, muscles, blood vessels, and glands.

- ultrasound to show how the bladder and other parts of the urinary tract are functioning
- sometimes a nerve biopsy may be performed.

Specialists who treat diabetic neuropathy include:

- neurologists: specialists in nervous system disorders
- urologists: specialists in urinary tract disorder
- gastroenterologists: specialists in digestive disorders
- podiatrists: specialists in caring for the feet

### Treatment

Treatment of diabetic neuropathy is usually focused on treating the symptoms associated with the neuropathy and addressing the underlying cause by improving the control of blood sugar levels, which may heal the early stages of neuropathy.

There is no cure for the permanent nerve damage caused by neuropathy. To help control pain, the choice of proven drug therapies has broadened during the past decade. Pain medication, such as the topical skin cream capsaicin, is usually no stronger than codeine because of the potential for **addiction** with long-term use of such drugs. Four main classes of drugs are available for **pain management**, alone or in combination: tricyclic antidepressants (Imipramine, Nortriptyline), narcotic **analgesics** (Morphine), anticonvulsants (Carbamazepine, Gabapentin), and antiarrhythmics.

### Prognosis

Early stage diabetic neuropathy can usually be reversed with good glucose control. Once nerve damage has occurred it cannot be reversed. The prognosis is largely dependent on the management of the underlying condition, diabetes, which may halt the progression of the neuropathy and improve symptoms. Recovery, if it occurs, is slow.

### Prevention

Tight glucose control and the avoidance of alcohol and cigarettes help protect nerves from damage.

### Resources

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Juvenile Diabetes Foundation. 120 Wall St., New York, NY 10005. (800) 533-CURE. <<http://www.jdf.org/>>.

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Gary Gilles

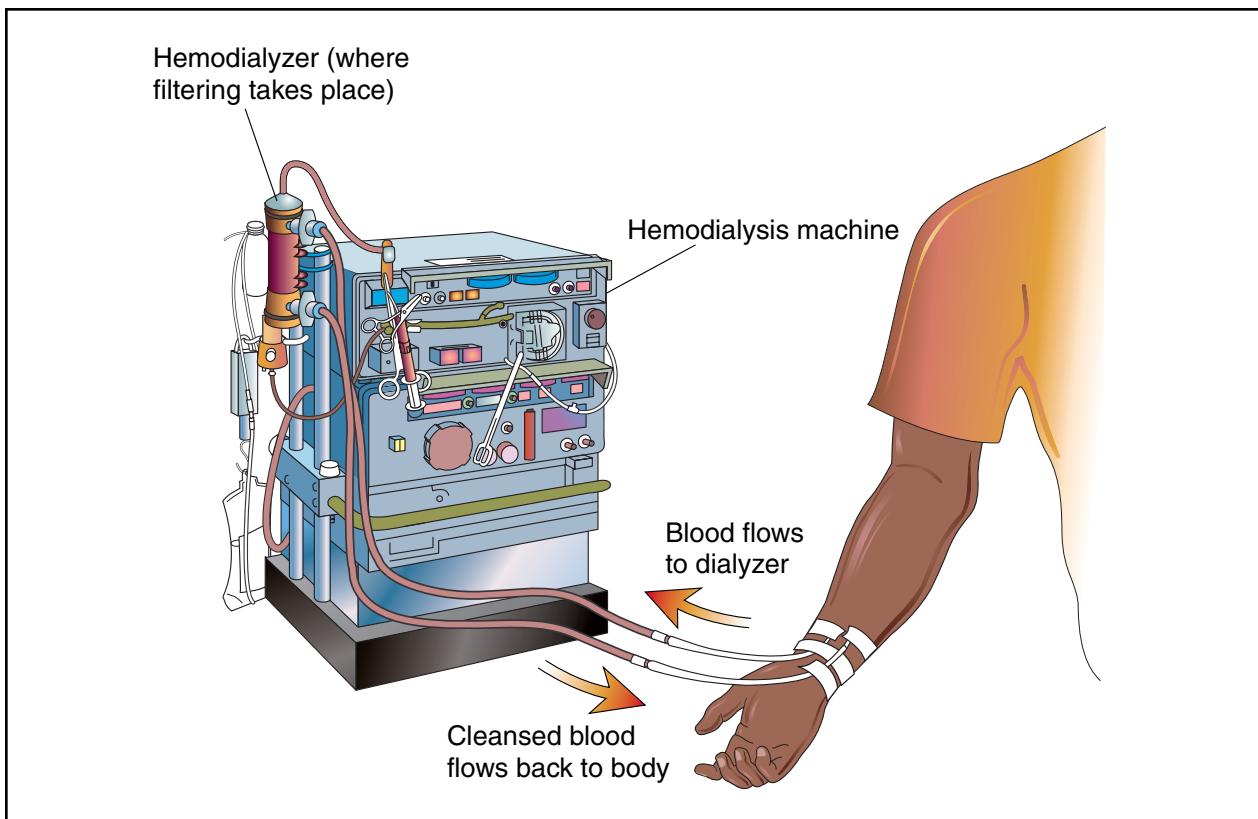
## Dialysis, kidney

### Definition

Dialysis treatment replaces the function of the kidneys, which normally serve as the body's natural filtration system. Through the use of a blood filter and a chemical solution known as dialysate, the treatment removes waste products and excess fluids from the bloodstream, while maintaining the proper chemical balance of the blood. There are two types of dialysis treatment : hemodialysis and peritoneal dialysis.

### Purpose

Dialysis can be used in the treatment of patients suffering from **poisoning** or overdose, in order to quickly remove drugs from the bloodstream. Its most prevalent application, however, is for patients with temporary or permanent kidney failure. For patients with end-stage renal disease (ESRD), whose kidneys are no longer capable of adequately removing fluids and wastes from their body or



**Hemodialysis** is the most frequently prescribed type of dialysis treatment in the United States. This treatment involves circulating the patient's blood outside of the body through a dialysis circuit. The blood is filtered and cleansed inside the hemodialyzer and returned to the body. (Illustration by Electronic Illustrators Group.)

of maintaining the proper level of certain kidney-regulated chemicals in the bloodstream, dialysis is the only treatment option available outside of **kidney transplantation**. In 1996 in the United States, over 200,000 people underwent regular dialysis treatments to manage their ESRD.

### Precautions

Blood pressure changes associated with hemodialysis may pose a risk for patients with heart problems. Peritoneal dialysis may be the preferred treatment option in these cases.

Peritoneal dialysis is not recommended for patients with abdominal adhesions or other abdominal defects, such as a **hernia**, that might compromise the efficiency of the treatment. It is also not recommended for patients who suffer frequent bouts of diverticulitis, an inflammation of small pouches in the intestinal tract.

### Description

There are two types of dialysis treatment: hemodialysis and peritoneal dialysis:

### Hemodialysis

Hemodialysis is the most frequently prescribed type of dialysis treatment in the United States. The treatment involves circulating the patient's blood outside of the body through an extracorporeal circuit (ECC), or dialysis circuit. Two needles are inserted into the patient's vein, or access site, and are attached to the ECC, which consists of plastic blood tubing, a filter known as a dialyzer (artificial kidney), and a dialysis machine that monitors and maintains blood flow and administers dialysate. Dialysate is a chemical bath that is used to draw waste products out of the blood.

Since the 1980s, the majority of hemodialysis treatments in the United States have been performed with hollow fiber dialyzers. A hollow fiber dialyzer is composed of thousands of tube-like hollow fiber strands encased in a clear plastic cylinder several inches in diameter. There are two compartments within the dialyzer (the blood compartment and the dialysate compartment). The membrane that separates these two compartments is semipermeable. This means that it allows the passage of certain sized molecules across it, but prevents the pas-

## KEY TERMS

**Access site**—The vein tapped for vascular access in hemodialysis treatments. For patients with temporary treatment needs, access to the bloodstream is gained by inserting a catheter into the subclavian vein near the patient's collarbone. Patients in long-term dialysis require stronger, more durable access sites, called fistulas or grafts, that are surgically-created.

**Dialysate**—A chemical bath used in dialysis to draw fluids and toxins out of the bloodstream and supply electrolytes and other chemicals to the bloodstream.

**Dialysis prescription**—The general parameters of dialysis treatment that vary according to each patient's individual needs. Treatment length, type of dialyzer and dialysate used, and rate of ultrafiltration are all part of the dialysis prescription.

**Dialyzer**—An artificial kidney usually composed of hollow fiber that is used in hemodialysis to eliminate waste products from the blood and remove excess fluids from the bloodstream.

**Erythropoietin**—A hormone produced by the kidneys that stimulates the production of red blood cells by bone marrow.

**ESRD**—End-stage renal disease; chronic or permanent kidney failure.

**Extracorporeal circuit (ECC)**—The path the hemodialysis patient's blood takes outside of the body. It typically consists of plastic tubing, a hemodialysis machine, and a dialyzer.

**Hematocrit (Hct) level**—A measure of red blood cells.

**Peritoneum**—The abdominal cavity; the peritoneum acts as a blood filter in peritoneal dialysis.

sage of other, larger molecules. As blood is pushed through the blood compartment in one direction, suction or vacuum pressure pulls the dialysate through the dialysate compartment in a countercurrent, or opposite direction. These opposing pressures work to drain excess fluids out of the bloodstream and into the dialysate, a process called ultrafiltration.

A second process called diffusion moves waste products in the blood across the membrane into the dialysate compartment, where they are carried out of the body. At the same time, electrolytes and other chemicals

in the dialysate solution cross the membrane into the blood compartment. The purified, chemically-balanced blood is then returned to the body.

Most hemodialysis patients require treatment three times a week, for an average of three to four hours per dialysis "run." Specific treatment schedules depend on the type of dialyzer used and the patient's current physical condition. While the treatment prescription and regimen is usually overseen by a nephrologist (a doctor that specializes in the kidney), dialysis treatments are typically administered by a nurse or patient care technician in outpatient clinics known as dialysis centers, or in hospital-based dialysis units. In-home hemodialysis treatment is also an option for some patients, although access to this type of treatment may be limited by financial and lifestyle factors. An investment in equipment is required and another person in the household should be available for support and assistance with treatments.

### *Peritoneal dialysis*

In peritoneal dialysis, 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:

- Continuous ambulatory peritoneal dialysis (CAPD). A continuous treatment that is self-administered and requires no machine. The patient inserts fresh dialysate solution into the abdominal cavity, waits four to six hours, and removes the used solution. The solution is immediately replaced with fresh dialysate. A bag attached to the catheter is worn under clothing.
- Continuous cyclic peritoneal dialysis (CCPD). An overnight treatment that uses a machine to drain and refill the abdominal cavity, CCPD takes 10–12 hours per session.
- Intermittent peritoneal dialysis (IPD). This hospital-based treatment is performed several times a week. A machine administers and drains the dialysate solution, and sessions can take up to 24 hours.

Peritoneal dialysis is often the treatment option of choice in infants and children, whose small size can make vascular (through a vein) access difficult to maintain. Peritoneal dialysis can also be done outside of a clinical setting, which is more conducive to regular school attendance.

## Preparation

Patients are weighed immediately before and after each hemodialysis treatment to assess their fluid retention. Blood pressure and temperature are taken and the patient is assessed for physical changes since their last dialysis run. Regular blood tests monitor chemical and waste levels in the blood. Prior to treatment, patients are typically administered a dose of heparin, an anticoagulant that prevents blood clotting, to ensure the free flow of blood through the dialyzer and an uninterrupted dialysis run for the patient.

## Aftercare

Both hemodialysis and peritoneal dialysis patients need to be vigilant about keeping their access sites and catheters clean and infection-free during and between dialysis runs.

Dialysis is just one facet of a comprehensive treatment approach for ESRD. Although dialysis treatment is very effective in removing toxins and fluids from the body, there are several functions of the kidney it cannot mimic, such as regulating high blood pressure and red blood cell production. Patients with ESRD need to watch their diet and fluid intake carefully and take medications as prescribed to manage their disease.

## Risks

Many of the risks and side effects associated with dialysis are a combined result of both the treatment and the poor physical condition of the ESRD patient. Dialysis patients should always report side effects to their healthcare provider.

### Anemia

**Hematocrit** (Hct) levels, a measure of red blood cells, are typically low in ESRD patients. This deficiency is caused by a lack of the hormone erythropoietin, which is normally produced by the kidneys. The problem is elevated in hemodialysis patients, who may incur blood loss during hemodialysis treatments. Epoetin alfa, or EPO (sold under the trade name Epogen), a hormone therapy, and intravenous or oral iron supplements are used to manage anemia in dialysis patients.

### Cramps, nausea, vomiting, and headaches

Some hemodialysis patients experience cramps and flu-like symptoms during treatment. These can be caused by a number of factors, including the type of dialysate used, composition of the dialyzer membrane, water quality in the dialysis unit, and the ultrafiltration rate of the

treatment. Adjustment of the dialysis prescription often helps alleviate many symptoms.

### Hypotension

Because of the **stress** placed on the cardiovascular system with regular hemodialysis treatments, patients are at risk for **hypotension**, a sudden drop in blood pressure. This can often be controlled by medication and adjustment of the patient's dialysis prescription.

### Infection

Both hemodialysis and peritoneal dialysis patients are at risk for infection. Hemodialysis patients should keep their access sites clean and watch for signs of redness and warmth that could indicate infection. Peritoneal dialysis patients must follow the same precautions with their catheter. **Peritonitis**, an infection of the peritoneum, causes flu-like symptoms and can disrupt dialysis treatments if not caught early.

### Infectious diseases

Because there is a great deal of blood exposure involved in dialysis treatment, a slight risk of contracting **hepatitis B** and **hepatitis C** exists. The hepatitis B **vaccination** is recommended for most hemodialysis patients. As of 1997, there has only been one documented case of HIV being transmitted in a United States dialysis unit to a staff member, and no documented cases of HIV ever being transmitted between dialysis patients in the United States. The strict standards of **infection control** practiced in modern hemodialysis units makes the chance of contracting one of these diseases very small.

## Normal results

Puffiness in the patient related to **edema**, or fluid retention, may be relieved after dialysis treatment. The patient's overall sense of physical well-being may also be improved. Because dialysis is an ongoing treatment process for many patients, a baseline for normalcy can be difficult to gauge.

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- American Association of Kidney Patients. 100 S. Ashley Dr., #280, Tampa, FL 33602. (800) 749-2257. <<http://www.aakp.org>>.
- American Kidney Fund. 6110 Executive Boulevard, Rockville, MD 20852. (800) 638-8299. <<http://216.248.130.102/Default.htm>>.
- 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

## Diaper rash

### Definition

**Dermatitis** of the buttocks, genitals, lower abdomen, or thigh folds of an infant or toddler is commonly referred to as diaper rash.

### Description

The outside layer of skin normally forms a protective barrier that prevents infection. One of the primary

causes of dermatitis in the diaper area is prolonged skin contact with wetness. Under these circumstances, natural oils are stripped away, the outer layer of skin is damaged, and there is increased susceptibility to infection by bacteria or yeast.

Diaper rash is a term that covers a broad variety of skin conditions that occur on the same area of the body. Some babies are more prone to diaper rash than others.

### Causes and symptoms

Frequently a flat, red rash is caused by simple chafing of the diaper against tender skin, initiating a friction rash. This type of rash is not seen in the skin folds. It may be more pronounced around the edges of the diaper, at the waist and leg bands. The baby generally doesn't appear to experience much discomfort. Sometimes the chemicals or detergents in the diaper are contributing factors and may result in **contact dermatitis**. These **rashes** should clear up easily with proper attention. Ignoring the condition may lead to a secondary infection that is more difficult to resolve.

Friction of skin against itself can cause a rash in the baby's skin folds, called intertrigo. This rash appears as reddened areas that may ooze and is often uncomfortable when the diaper is wet. Intertrigo can also be found on other areas of the body where there are deep skin folds that tend to trap moisture.

**Seborrheic dermatitis** is the diaper area equivalent of cradle cap. It is scaly and greasy in appearance and may be worse in the folds of the skin.

Yeast, or candidal dermatitis, is the most common infectious cause of diaper rash. The affected areas are raised and quite red with distinct borders, and satellite lesions may occur around the edges. Yeast is part of the normal skin flora, and is often an opportunistic invader when simple diaper rash is untreated. It is particularly common after treatment with **antibiotics**, which kill the good bacteria that normally keep the yeast population in check. Usual treatments for diaper rash will not clear it up. Repeated or difficult to resolve episodes of yeast infection may warrant further medical attention, since this is sometimes associated with diabetes or immune problems.

Another infectious cause of diaper rash is **impetigo**. This bacterial infection is characterized by blisters that ooze and crust.

### Diagnosis

The presence of **skin lesions** in the diaper area means that the baby has diaper rash. However, there are several types of rash that may require specific treatment

in order to heal. It is useful to be able to distinguish them by appearance as described above.

A baby with a rash that does not clear up within two to three days or a rash with blisters or bleeding should be seen by a healthcare professional for further evaluation.

## Treatment

Antibiotics are generally prescribed for rashes caused by bacteria, particularly impetigo. This may be a topical or oral formulation, depending on the size of the area involved and the severity of the infection.

Over-the-counter antifungal creams, such as Lotrimin, are often recommended to treat a rash resulting from yeast. If topical treatment is not effective, an oral antifungal may be prescribed.

Mild steroid creams, such as 0.5–1% hydrocortisone, can be used for seborrheic dermatitis and sometimes intertrigo. Prescription strength creams may be needed for short-term treatment of more stubborn cases.

## Alternative treatment

Good diaper hygiene will prevent or clear up many simple cases of diaper rash. Diapers should be checked very frequently and changed as soon as they are wet or soiled. Good air circulation is also important for healthy skin. Babies should have some time without wearing a diaper, and a waterproof pad can be used to protect the bed or other surface. Rubber pants, or other occlusive fabrics, should not be used over the diaper area. Some cloth-like disposable diapers promote better air circulation than plastic-type diapers. It may be necessary for mothers to experiment with diaper types to see if the baby's skin reacts better to cloth or disposable ones. If disposable diapers are used, the baby's skin may react differently to various brands. If the baby is wearing cloth diapers, they should be washed in a mild detergent and double rinsed.

The diaper area should be cleaned with something mild, even plain water. Some wipes contain alcohol or chemicals that can be irritating for some babies. Plain water may be the best cleansing substance when there is a rash. Using warm water in a spray bottle (or giving a quick bath) and then lightly patting the skin dry can produce less skin trauma than using wipes. In the event of suspected yeast, a tablespoon of cider vinegar can be added to a cup of warm water and used as a cleansing solution. This is dilute enough that it should not burn, but acidifies the skin pH enough to hamper the yeast growth.

Barrier ointments can be valuable to treat rashes. Those that contain zinc oxide are especially effective. These creams and ointments protect already irritated skin



**Baby with severe diaper rash.** (Custom Medical Stock Photo. Reproduced by permission.)

from the additional insult of urine and stool, particularly if the baby has **diarrhea**. Cornstarch powder may be used on rashes that are moist, such as impetigo.

## Nutrition

What the baby eats can make a difference in stool frequency and acidity. Typically, breast-fed babies will have fewer problems with rashes. When adding a new food to the diet, the baby should be observed closely to see whether rashes are produced around the baby's mouth or anus. If this occurs, the new food should be discontinued.

Babies who are taking antibiotics are more likely to get rashes due to yeast. To help bring the good bacterial counts back to normal, *Lactobacillus bifidus* can be added to the diet. It is available in powder form from most health food stores.

## Herbal treatment

Some herbal preparations can be useful for diaper rash. Calendula reduces inflammation, tightens tissues, and disinfects. It has been recommended for seborrheic dermatitis as well as for general inflammation of the skin. The ointment should be applied at each diaper change. Chickweed ointment can also be soothing for irritated skin and may be applied once or twice daily.

## Prognosis

Treated appropriately, diaper rash will resolve fairly quickly if there is no underlying health problem or skin disease.

## Prevention

Frequent diaper changes are important to keep the skin dry and healthy. Application of powders and oint-

## KEY TERMS

**Dermatitis**—Inflammation of the skin.

ments is not necessary when there is no rash. Finding the best combination of cleansing and diapering products for the individual baby will also help to prevent diaper rash.

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Judith Turner

# Diaphragm (birth control)

### Definition

Diaphragms are dome-shaped barrier methods of **contraception** that block sperm from entering the uterus. They are made of latex (rubber) and formed like a shallow cup. Since vaginas vary in size, each patient will need to be fitted by a doctor or nurse with a diaphragm that conforms to the shape and contour of the vagina as well as the strength of the muscles in the vaginal walls. Diaphragms must be used with spermicidal cream or jelly. The device should cause no discomfort, and neither the woman nor her partner should feel that it is there.

### Purpose

The purpose of a diaphragm is to prevent access to the womb (uterus) by the sperm and thus prevent conception. The level of effectiveness is about 95%.

### Precautions

Each client will undergo a **physical examination** and a Pap smear. If these are normal, the physician will fit the patient for the device and give instructions on how to insert, remove, and clean the object. She will also be taught the signs and symptoms of potential complications.

### Description

Prior to insertion, the inside of the dome and the rim are covered with a thick layer (perhaps a tablespoon) of a spermicide that is compatible with the diaphragm being used. The domed area covers the opening into the uterus (cervix) and keeps the spermicide in place. As a result, any sperm that might get under the diaphragm will be destroyed.

Diaphragms may be inserted two to three hours prior to intercourse, and must be left in place for six to eight hours following sexual relations. During this time the woman may not swim, bathe, or douche, but she may shower. If she desires to have intercourse again before the six to eight hours have passed, the diaphragm should not be removed. Instead, an applicator full of spermicide should be deposited into the vagina.

A diaphragm will last for a year or more. It should be examined weekly for holes. This can be done by holding it up to the light or filling it with water.

### Preparation

Before inserting the diaphragm, the woman should empty her bladder and wash her hands with soap and water. The device should be checked for leaks by filling it with water or holding it up to the light. A spermicidal jelly is then applied to the inside and outside, and especially around the rim. While standing with one foot elevated on a chair or step, lying down, or squatting, the woman folds the diaphragm inward toward the middle and inserts it into the vagina as far as it will go.

### Aftercare

When removed, the diaphragm should be washed with a mild soap and water. After being dried, it can be dusted with corn starch before being returned to its container. The diaphragm should always be stored away from sunlight and heat in a cool, dry place. It should not be washed with harsh or perfumed soaps or used with perfumed powders because either of these substances can damage the diaphragm.

### Risks

Although rare, wearing the diaphragm longer than the recommended time can result in **toxic shock syndrome**. The signs and symptoms of this serious illness include sudden onset of high **fever**, vomiting, **diarrhea**, **dizziness**, faintness, weakness, aching muscles and joints, and rash. The doctor must be notified immediately if any of these conditions appear. An allergic reaction to the spermicide or the material from which the device is

## KEY TERMS

**Spermicide**—A substance that kills sperm.

**Toxic shock syndrome**—An uncommon, but potentially fatal, disease that has been associated with the use of diaphragms and vaginal tampons. The symptoms include high fever, vomiting, and diarrhea.

made is also possible. Diaphragm use is also associated with an increased risk of bladder infections.

It should be noted that the diaphragm can become dislodged during intercourse, which could result in an unwanted **pregnancy**. To ensure a secure fit, a woman should be examined for a refitting if she gains or loses more than 10 lbs (4.5 kg), or after she gives birth.

### Normal results

Consumers can expect an efficiency rate of about 95% in preventing pregnancy. Using a male **condom** in conjunction with the diaphragm decreases the potential for pregnancy. Diaphragms provide no protection against **AIDS** or other **sexually transmitted diseases**.

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Donald G. Barstow, RN

Diaphragmatic hernia see **Hernia**

## Diarrhea

### Definition

To most individuals, diarrhea means an increased frequency or decreased consistency of bowel movements; however, the medical definition is more exact than this. In many developed countries, the average number of bowel movements is three per day. However, researchers have found that diarrhea best correlates with an increase in stool weight; stool weights above 10 oz (300 g) per day generally indicates diarrhea. This is mainly due to excess water, which normally makes up 60–85% of fecal matter. In this way, true diarrhea is distinguished from diseases that cause only an increase in the number of bowel movements (hyperdefecation), or incontinence (involuntary loss of bowel contents).

Diarrhea is also classified by physicians into acute, which lasts one to two weeks, and chronic, which continues for longer than 23 weeks. Viral and bacterial infections are the most common causes of acute diarrhea.

### Description

In many cases, acute infectious diarrhea is a mild, limited annoyance. However, worldwide acute infectious diarrhea has a huge impact, causing over five million deaths per year. While most deaths are among children under five years of age in developing nations, the impact, even in developed countries, is considerable. For example, over 250,000 individuals are admitted to hospitals in the United States each year because of one of these episodes. Rapid diagnosis and proper treatment can prevent much of the suffering associated with these devastating illnesses.

Chronic diarrhea also has a considerable effect on health, as well as on social and economic well being. Patients with **celiac disease**, inflammatory bowel disease, and other prolonged diarrheal illnesses develop nutritional deficiencies that diminish growth and immunity. They affect social interaction and result in the loss of many working hours.

### Causes and symptoms

Diarrhea occurs because more fluid passes through the large intestine (colon) than that organ can absorb. As a rule, the colon can absorb several times more fluid than is required on a daily basis. However, when this reserve capacity is overwhelmed, diarrhea occurs.

Diarrhea is caused by infections or illnesses that either lead to excess production of fluids or prevent absorption of fluids. Also, certain substances in the colon,

such as fats and bile acids, can interfere with water absorption and cause diarrhea. In addition, rapid passage of material through the colon can also do the same.

Symptoms related to any diarrheal illness are often those associated with any injury to the gastrointestinal tract, such as **fever**, nausea, vomiting, and abdominal **pain**. All or none of these may be present depending on the disease causing the diarrhea. The number of bowel movements can vary—up to 20 or more per day. In some patients, blood or pus is present in the stool. Bowel movements may be difficult to flush (float) or contain undigested food material.

The most common causes of acute diarrhea are infections (the cause of **traveler's diarrhea**), **food poisoning**, and medications. Medications are a frequent and often over-looked cause, especially **antibiotics** and **antacids**. Less often, various sugar free foods, which sometimes contain poorly absorbable materials, cause diarrhea.

Chronic diarrhea is frequently due to many of the same things that cause the shorter episodes (infections, medications, etc.); symptoms just last longer. Some infections can become chronic. This occurs mainly with parasitic infections (such as *Giardia*) or when patients have altered immunity (**AIDS**).

The following are the more usual causes of chronic diarrhea:

- AIDS
- colon **cancer** and other bowel tumors
- endocrine or hormonal abnormalities (thyroid, **diabetes mellitus**, etc.)
- food allergy
- inflammatory bowel disease (**Crohn's disease** and **ulcerative colitis**)
- lactose intolerance
- malabsorption syndromes (celiac and Whipple's disease)
- other (alcohol, microscopic colitis, radiation, surgery)

### **Complications**

The major effects of diarrhea are **dehydration**, **malnutrition**, and weight loss. Signs of dehydration can be hard to notice, but increasing thirst, **dry mouth**, weakness or lightheadedness (particularly if worsening on standing), or a darkening/decrease in urination are suggestive. Severe dehydration leads to changes in the body's chemistry and could become life-threatening. Dehydration from diarrhea can result in kidney failure, neurological symptoms, arthritis, and skin problems.

### **Diagnosis**

Most cases of acute diarrhea never need diagnosis or treatment, as many are mild and produce few problems. But patients with fever over 102°F (38.9°C), signs of dehydration, bloody bowel movements, severe abdominal pain, known immune disease, or prior use of antibiotics need prompt medical evaluation.

When diagnostic studies are needed, the most useful are **stool culture** and examination for parasites; however these are often negative and a cause cannot be found in a large number of patients. The earlier cultures are performed, the greater the chance of obtaining a positive result. For those with a history of antibiotic use in the preceding two months, stool samples need to be examined for the toxins that cause **antibiotic-associated colitis**. Tests are also available to check stool samples for microscopic amounts of blood and for cells that indicate severe inflammation of the colon. Examination with an endoscope is sometimes helpful in determining severity and extent of inflammation. Tests to check changes in blood chemistry (potassium, magnesium, etc.) and a complete **blood count** (CBC) are also often performed.

Chronic diarrhea is quite different, and most patients with this condition will receive some degree of testing. Many exams are the same as for an acute episode, as some infections and parasites cause both types of diarrhea. A careful history to evaluate medication use, dietary changes, family history of illnesses, and other symptoms is necessary. Key points in determining the seriousness of symptoms are weight loss of over 10 lb (4.5 kg), blood in the stool, and nocturnal diarrhea (symptoms that awaken the patient from sleep).

Both prescription and over-the-counter medications can contain additives, such as lactose and sorbitol, that will produce diarrhea in sensitive individuals. Review of **allergies** or skin changes may also point to a cause. Social history may indicate if **stress** is playing a role or identify activities which can be associated with diarrhea (for example, diarrhea that occurs in runners).

A combination of stool, blood, and urine tests may be needed in the evaluation of chronic diarrhea; in addition a number of endoscopic and x-ray studies are frequently required.

### **Treatment**

Treatment is ideally directed toward correcting the cause; however, the first aim should be to prevent or treat dehydration and nutritional deficiencies. The type of fluid and nutrient replacement will depend on whether oral feedings can be taken and the severity of fluid loss-

es. Oral rehydration solution (ORS) or intravenous fluids are the choices; ORS is preferred if possible.

A physician should be notified if the patient is dehydrated, and if oral replacement is suggested then commercial (Pedialyte and others) or homemade preparations can be used. The World Health Organization (WHO) has provided this easy recipe for home preparation, which can be taken in small frequent sips:

- Table salt—3/4 tsp
- Baking powder—1 tsp
- Orange juice—1 c
- Water—1 qt (1 L)

When feasible, food intake should be continued even in those with acute diarrhea. A physician should be consulted as to what type and how much food is permitted.

Anti-motility agents (loperamide, diphenoxylate) are useful for those with chronic symptoms; their use is limited or even contraindicated in most individuals with acute diarrhea, especially in those with high fever or bloody bowel movements. They should not be taken without the advice of a physician.

Other treatments are available, depending on the cause of symptoms. For example, the bulk agent psyllium helps some patients by absorbing excess fluid and solidifying stools; cholestyramine, which binds bile acids, is effective in treating bile salt induced diarrhea. Low fat diets or more easily digestible fat is useful in some patients. New antidiarrheal drugs that decrease excessive secretion of fluid by the intestinal tract is another approach for some diseases. Avoidance of medications or other products that are known to cause diarrhea (such as lactose) is curative in some, but should be discussed with a physician.

### Alternative treatment

It is especially important to find the cause of diarrhea, since stopping diarrhea when it is the body's way of eliminating something foreign is not helpful and can be harmful in the long run.

One effective alternative approach to preventing and treating diarrhea involves oral supplementation of aspects of the normal flora in the colon with the yeasts *Lactobacillus acidophilus*, *L. bifidus*, or *Saccharomyces boulardii*. In clinical settings, these "biotherapeutic" agents have repeatedly been helpful in the resolution of diarrhea, especially antibiotic-associated diarrhea. Their effectiveness is also supported by the results of a research study published in the *Journal of the American Medical Association* in 1996.

### KEY TERMS

**Anti-motility medications**—Medications such as loperamide (Imodium), diphenoxylate (Lomotil), or medications containing codeine or narcotics that decrease the ability of the intestine to contract. These can worsen the condition of a patient with dysentery or colitis.

**Colitis**—Inflammation of the colon.

**Endoscope**—An endoscope, as used in the field of gastroenterology, is a thin flexible tube that uses a lens or miniature camera to view various areas of the gastrointestinal tract. Both diagnosis, through biopsies or other means, and therapeutic procedures can be done with this instrument.

**Endoscopy**—The performance of an exam using an endoscope is known generally as endoscopy.

**Lactose intolerance**—An inability to properly digest milk and dairy products.

**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.

**Steatorrhea**—Excessive amounts of fat in the feces.

Nutrient replacement also plays a role in preventing and treating episodes of diarrhea. Zinc especially appears to have an effect on the immune system, and deficiency of this mineral can lead to chronic diarrhea. Also, zinc replacement improves growth in young patients. Plenty of fluids, especially water, should be taken by individuals suffering from diarrhea to prevent dehydration. The BRAT diet also can be useful in helping to resolve diarrhea. This diet limits food intake to bananas, rice, applesauce, and toast. These foods provide soluble and insoluble fiber without irritation. If the toast is slightly burnt, the charcoal can help sequester toxins and pull them from the body.

Acute homeopathic remedies can be very effective for treating diarrhea especially in infants and young children.

### Prognosis

Prognosis is related to the cause of the diarrhea; for most individuals in developed countries, a bout of acute, infectious diarrhea is at best uncomfortable. However, in

both industrialized and developing areas, serious complications and **death** can occur.

For those with chronic symptoms, an extensive number of tests are usually necessary to make a proper diagnosis and begin treatment; a specific diagnosis is found in 90% of patients. In some, however, no specific cause is found and only treatment with bulk agents or anti-motility agents is indicated.

## Prevention

Proper hygiene and food handling techniques will prevent many cases. Traveler's diarrhea can be avoided by use of Pepto-Bismol and/or antibiotics, if necessary. The most important action is to prevent the complications of dehydration.

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Avenue Appia 20, 1211 Geneva 27, Switzerland. (+00 41 22) 791 21 11. <<http://www.who.int>>.

## OTHER

- "Directory of Digestive Diseases Organizations for Patients." *National Institute of Diabetes and Digestive and Kidney Disease*. <<http://www.niddk.nih.gov>>.
- "A Neglected Modality for the Treatment and Prevention of Selected Intestinal and Vaginal Infections." *JAMA*. <<http://pubs.ama-assn.org>>.
- Selected publications and documents on diarrhoeal diseases (including cholera). *World Health Organization (WHO)*. <<http://www.who.ch/chd/pub/cdd/cddpub.htm>>.

David Kaminstein, MD

**Diazep** see **Benzodiazepines**

**Diclofenac** see **Nonsteroidal anti-inflammatory drugs**

**Dicyclomine** see **Antispasmodic drugs**

**Didanosine** see **Antiretroviral drugs**

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## Diets

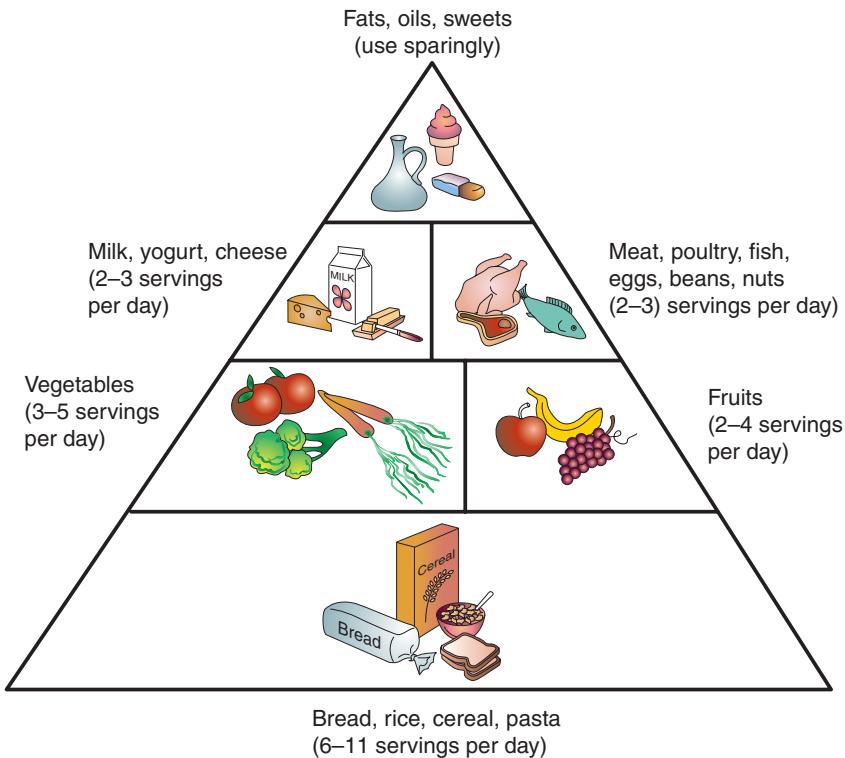
### Definition

Humans may alter their usual eating habits for many reasons, including weight loss, disease prevention or treatment, removing toxins from the body, or to achieve a general improvement in physical and mental health. Others adopt special diets for religious reasons. In the case of some vegetarians and vegans, dietary changes are made out of ethical concerns for the rights of animals.

### Purpose

People who are moderately to severely overweight can derive substantial health benefits from a weight-loss diet. A weight reduction of just 10–20 (4.5–9.1 kg) can result in reduced cholesterol levels and lower blood pressure. Weight-related health problems include heart disease, diabetes, high blood pressure, and high levels of blood sugar and cholesterol.

In individuals who are not overweight, dietary changes may also be useful in the prevention or treatment of a range of ailments including acquired immunodeficiency syndrome (**AIDS**), **cancer**, **osteoporosis**, inflammatory bowel disease, chronic pulmonary disease,



**Suggested daily food servings.** (Illustration by Electronic Illustrators Group.)

renal disease, **Parkinson's disease**, seizure disorders, and food **allergies** and intolerances.

## Description

### Origins

The practice of altering diet for special reasons has existed since antiquity. For example, Judaism has included numerous dietary restrictions for thousands of years. One ancient Jewish sect, the Essenes, is said to have developed a primitive **detoxification** diet aimed at preparing the bodies, minds, and spirits of its members for the coming of a "messiah" who would deliver them from their Roman captors. Preventative and therapeutic diets became quite popular during the late twentieth century. Books promoting the latest dietary plan continue to make the bestseller lists, although not all of the information given is considered authoritative.

The idea of a healthful diet is to provide all of the calories and nutrients needed by the body for optimal performance, at the same time ensuring that neither nutritional deficiencies nor excesses occur. Diet plans that claim to accomplish those objectives are so numer-

ous they are virtually uncountable. These diets employ a variety of approaches, including the following:

- **Fixed-menu:** Offers little choice to the dieter. Specifies exactly which foods will be consumed. Easy to follow, but may be considered "boring" to some dieters.
- **Formula:** Replaces some or all meals with a nutritionally balanced liquid formula or powder.
- **Exchange-type:** Allows the dieter to choose between selected foods from each food group.
- **Flexible:** Doesn't concern itself with the overall diet, simply with one aspect such as fat or energy.

Diets may also be classified according to the types of foods they allow. For example, an omnivorous diet consists of both animal and plant foods, whereas a lacto-ovo-vegetarian diet permits no animal flesh, but does include eggs, milk, and dairy products. A vegan diet is a stricter form of **vegetarianism** in which eggs, cheese, and other milk products are prohibited.

A third way of classifying diets is according to their purpose: religious, weight-loss, detoxification, lifestyle-related, or aimed at prevention or treatment of a specific disease.

## Precautions

Dieters should be cautious about plans that severely restrict the size of food portions, or that eliminate entire food groups from the diet. It is highly probable that they will become discouraged and drop out of such programs. The best diet is one that can be maintained indefinitely without ill effects, that offers sufficient variety and balance to provide everything needed for good health, and that is considerate of personal food preferences.

Low-fat diets are not recommended for children under the age of two. Young children need extra fat to maintain their active, growing bodies. Fat intake may be gradually reduced between the ages of two and five, after which it should be limited to a maximum of 30% of total calories through adulthood. Saturated fat should be restricted to no more than 10% of total calories.

Weight-loss dieters should be wary of the “yo-yo” effect that occurs when numerous attempts are made to reduce weight using high-risk, quick-fix diets. This continued “cycling” between weight loss and weight gain can slow the basal metabolic rate and can sometimes lead to eating disorders. The dieter may become discouraged and frustrated by this success/failure cycle. The end result of “yo-yo” dieting is that it becomes more difficult to maintain a healthy weight.

Caution should also be exercised about weight-loss diets that require continued purchases of special prepackaged foods. Not only do these tend to be costly and over-processed, they may also prevent dieters from learning the food-selection and preparation skills essential to maintenance of weight loss. Further, dieters should consider whether they want to carry these special foods to work, restaurants, or homes of friends.

Concern has been expressed about weight-loss diet plans that do not include **exercise**, considered essential to long-term weight management. Some diets and supplements may be inadvisable for patients with special conditions or situations.

Certain fad diets purporting to be official diets of groups such as the American Heart Association and the Mayo Clinic are in no way endorsed by those institutions. Patients thinking of starting such a diet should check with the institution to ensure its name has not been misappropriated by an unscrupulous practitioner.

## Side effects

A wide range of side effects (some quite serious) can result from special diets, especially those that are nutritionally unbalanced. Further problems can arise if the dieter is taking high doses of dietary supplements. Food

is essential to life, and improper **nutrition** can result in serious illness or **death**.

## Research and general acceptance

It is agreed among traditional and complementary practitioners that many patients could substantially benefit from improved eating habits. Specialized diets have proved effective against a wide variety of conditions and diseases. However, dozens of unproved but widely publicized “fad diets” emerge each year, prompting widespread concerns about their usefulness, cost to the consumer, and their safety.

## Resources

### ORGANIZATIONS

American Dietetic Association. 216 West Jackson Blvd., Chicago, IL 60606-6995. (312) 899-0040. <<http://www.eatright.org>>.

David Helwig

# Diffuse esophageal spasm

## Definition

Diffuse esophageal spasm is a term used to define an uncoordinated or spastic esophagus.

## Description

The esophagus is a muscular tube that actively transports food from the throat to the stomach by rhythmic contractions known as peristalsis. The actual mechanism and anatomy are quite complex, involving three distinct segments and allowing a person to swallow even when upside-down. Diffuse esophageal spasm describes a condition where the entire esophagus is spastic—along its entire length, the muscular activity is increased and uncoordinated. The name corkscrew esophagus describes perfectly the appearance of this disorder on x rays.

X rays may reveal a slightly different appearance and result in the designation rosary bead esophagus, but the cause is still diffuse spasm, and the two entities behave in the same way.

## Causes and symptoms

The cause appears to be disruption of the complex system of nerves that coordinates the muscular activity. The result is difficulty swallowing (dysphagia) and **pain** that feels like a **heart attack** and can involve the entire chest, jaw, and arms.

## KEY TERMS

**Contrast agent**—A substance that produces shadows on x rays.

**Manometry**—Measurement of pressure.

**Peristalsis**—Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that propel the contents along.

### Diagnosis

Swallowing problems usually call for esophagograms. In the x ray department, the patient is given a contrast agent to drink. During swallowing, x rays record the passage of the agent down the esophagus and into the stomach. Instead of a straight tube with well-coordinated waves of contraction, the resulting x rays show a writhing organ resembling a giant corkscrew.

Another test that is used in many disorders of esophageal motility is manometry. Pressures inside the esophagus are measured every inch or so using a balloon device that is passed all the way down to the stomach. The result is a precise record of its activity that yields a specific diagnosis.

### Treatment

Soft and liquid foods pass more easily than solid pieces. Medications of several types are helpful—nifedipine, hydralazine, isoproterenol, and nitrates being the most successful. Several other treatments have uncertain results. For severe cases, relief is obtained two-thirds of the time by cutting the muscles along the entire length of the esophagus. This is a major surgical procedure.

### Prognosis

This condition does not go away, nor is treatment entirely satisfactory. Patients need to be careful of what they eat and continue on medication if a beneficial one is found. Fortunately, the condition does not get progressively worse as time passes.

### Resources

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J. Ricker Polsdorfer, MD

## DiGeorge syndrome

### Definition

DiGeorge syndrome (also called congenital thymic hypoplasia, or third and fourth pharyngeal pouch syndrome) is a birth defect that is caused by an abnormal chromosome and affects the baby's immune system. The syndrome is marked by absence or underdevelopment of the thymus and parathyroid glands. It is named for the pediatrician who first described it in 1965.

### Description

The prevalence of DiGeorge syndrome is debated; the estimates range from 1:4000 to 1:6395. Because the symptoms caused by the chromosomal abnormality vary somewhat from patient to patient, the syndrome probably occurs much more often than was previously thought. DiGeorge syndrome is sometimes described as one of the "CATCH 22" disorders, so named because of their characteristics—cardiac defects, abnormal facial features, thymus underdevelopment, cleft palate, and hypocalcemia—caused by a deletion of several genes in chromosome 22. The specific facial features associated with DiGeorge syndrome include low-set ears, wide-set eyes, a small jaw, and a short groove in the upper lip. The male/female ratio is 1:1. The syndrome appears to be equally common in all racial and ethnic groups.

### Causes and symptoms

DiGeorge syndrome is caused either by inheritance of a defective chromosome 22 or by a new defect in chromosome 22 in the fetus. The type of defect that is involved is called deletion. A deletion occurs when the genetic material in the chromosomes does not recombine properly during the formation of sperm or egg cells. The deletion means that several genes from chromosome 22 are missing in DiGeorge syndrome patients. According to a 1999 study, 6% of children with DiGeorge syndrome inherited the deletion from a parent, while 94% had a new deletion. Other conditions that are associated with

DiGeorge syndrome are diabetes (a condition where the pancreas no longer produces enough insulin) in the mother and **fetal alcohol syndrome** (a pattern of **birth defects** and learning and behavioral problems affecting individuals whose mothers consumed alcohol during pregnancy).

The loss of the genes in the deleted material means that the baby's third and fourth pharyngeal pouches fail to develop normally during the twelfth week of pregnancy. This developmental failure results in a completely or partially absent thymus gland and parathyroid glands. In addition, 74% of fetuses with DiGeorge syndrome have severe heart defects. The child is born with a defective immune system and an abnormally low level of calcium in the blood.

These defects usually become apparent within 48 hours of birth. The infant's heart defects may lead to **heart failure**, or there may be seizures and other evidence of a low level of calcium in the blood (**hypocalcemia**).

## Diagnosis

Diagnosis of DiGeorge syndrome can be made by ultrasound examination around the eighteenth week of pregnancy, when abnormalities in the development of the heart or the palate can be detected. Another technique that is used to diagnose the syndrome before birth is called fluorescence in situ hybridization, or FISH. This technique uses DNA probes from the DiGeorge region on chromosome 22. FISH can be performed on cell samples obtained by **amniocentesis** as early as the fourteenth week of pregnancy. It confirms about 95% of cases of DiGeorge syndrome.

If the mother has not had prenatal testing, the diagnosis of DiGeorge syndrome is sometimes suggested by the child's facial features at birth. In other cases, the doctor makes the diagnosis during heart surgery when he or she notices the absence or abnormal location of the thymus gland. The diagnosis can be confirmed by blood tests for calcium, phosphorus, and parathyroid hormone levels, and by the sheep cell test for immune function.

## Treatment

### Hypocalcemia

Hypocalcemia in DiGeorge patients is unusually difficult to treat. Infants are usually given calcium and vitamin D by mouth. Severe cases have been treated by transplantation of fetal thymus tissue or bone marrow.

### Heart defects

Infants with life-threatening heart defects are treated surgically.

### Defective immune function

Children with DiGeorge syndrome should be kept on low-phosphorus **diets** and kept away from crowds or other sources of infection. They should not be immunized with vaccines made from live viruses or given **corticosteroids**.

## Prognosis

The prognosis is variable; many infants with DiGeorge syndrome die from overwhelming infection, seizures, or heart failure within the first year. Advances in heart surgery indicate that the prognosis is most closely linked to the severity of the heart defects and the partial presence of the thymus gland. In most children who survive, the number of T cells, a type of white blood cell, in the blood rises spontaneously as they mature. Survivors are likely to be mentally retarded, however, and to have other developmental difficulties, including psychiatric problems in later life.

## Prevention

**Genetic counseling** is recommended for parents of children with DiGeorge syndrome because the disorder can be detected prior to birth. Although most children with DiGeorge syndrome did not inherit the chromosome deletion from their parents, they have a 50% chance of passing the deletion on to their own children.

Because of the association between DiGeorge syndrome and fetal alcohol syndrome, pregnant women should avoid drinking alcoholic beverages.

## Resources

### BOOKS

"Common Multiple Congenital Anomaly Syndromes." In *Neonatology: Management, Procedures, On-Call Problems, Diseases and Drugs*, ed. Tricia Lacy Gomella, et al. Norwalk, CT: Appleton & Lange, 1994.

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### ORGANIZATIONS

Canadian 22q Group. 320 Cote Street Antoine, West Montreal, Quebec H3Y 2J4.

## KEY TERMS

**Deletion**—A genetic abnormality in which a segment of a chromosome is lost. DiGeorge syndrome is caused by a deletion on human chromosome 22.

**Fetal alcohol syndrome**—A cluster of birth defects that includes abnormal facial features and mental retardation, caused by the mother's consumption of alcoholic beverages during pregnancy.

**Fluorescence in situ hybridization (FISH)**—A technique for diagnosing DiGeorge syndrome before birth by analyzing cells obtained by amniocentesis with DNA probes. FISH is about 95% accurate.

**Hypocalcemia**—An abnormally low level of calcium in the blood.

**Hypoplasia**—A deficiency or underdevelopment of a tissue or body structure.

**T cells**—A type of white blood cell produced in the thymus gland. T cells are an important part of the immune system. Infants born with an underdeveloped or absent thymus do not have a normal level of T cells in their blood.

Chromosome Deletion Outreach, Inc. P.O. Box 724, Boca Raton, FL 33429-0724. (888) 236-6680.  
International DiGeorge/VCF Support Network, c/o Family Voices of New York. 46 1/2 Clinton Avenue, Cortland, NY 13045. (607) 753-1250.

Rebecca J. Frey, PhD

Digital rectal examination see **Rectal examination**

## Digitalis drugs

### Definition

Digitalis drugs are medicines made from a type of foxglove plant (*Digitalis purpurea*) that have a stimulating effect on the heart.

### Purpose

Digitalis drugs are used to treat heart problems such as congestive **heart failure** and irregular heartbeat.

These medicines help make the heart stronger and more efficient. This, in turn, improves blood circulation and helps relieve the swelling of the hands and ankles that is common in people with heart problems.

### Description

Digitalis drugs, also known as digitalis glycosides, are available only with a physician's prescription. They are sold in tablet, capsule, liquid, and injectable forms. Commonly used digitalis drugs are digitoxin (Crystodigin) and digoxin (Lanoxin).

### Recommended dosage

The recommended dosage is different for each patient. The physician who prescribes the medicine will determine the correct dose. Taking exactly the right amount of medicine and taking it exactly as directed are very important. Never take larger or more frequent doses. During treatment with a digitalis heart medicine, the physician will monitor blood levels of the drug and will decide whether the dose needs to be changed. Patients should never change the dose of this medicine unless told to do so by their physicians.

### Precautions

Seeing a physician regularly while taking digitalis drugs is very important. The physician will check to make sure the medicine is working as it should and will make any necessary changes in dosage or in instructions for taking the medicine.

Patients taking digitalis drugs should learn to take their pulse and should check it regularly while under treatment with this medicine. Changes in pulse rate, rhythm, or force could be signs of side effects.

Do not stop taking this medicine suddenly without checking with the physician who prescribed it. This could cause a serious change in heart function.

Digitalis drugs are responsible for many accidental poisonings in children. Keep this medicine out of the reach of children.

Be alert to the signs of overdose. Overdosing is a serious concern with digitalis drugs, because the amount of medicine that most people need to help their heart problems is very close to the amount that can cause problems from overdose. If any of these signs of overdose occur, check with a physician as soon as possible:

- loss of appetite
- nausea
- vomiting



**Digitalis purpurea.** (Photo Researcher, Inc. Reproduced by permission.)

- pain in the lower stomach
- diarrhea
- extreme tiredness or weakness
- extremely slow or irregular heartbeat (or fast heartbeat in children)
- blurred vision or other vision changes
- drowsiness
- confusion or depression
- headache
- fainting

Anyone who is taking digitalis drugs should be sure to tell the health care professional in charge before having any surgical or dental procedures or receiving emergency treatment. Physicians may advise people taking digitalis drugs to wear or carry medical identification indicating that they are taking this medicine.

Patients need to be very careful not to accidentally take this medicine in place of another medicine that

looks similar. Patients who are taking other medicines that look like their digitalis medicine should ask their pharmacists for suggestions on how to avoid mix-ups.

Anyone who has had unusual reactions to digitalis drugs in the past 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.

Women who are pregnant or breastfeeding or who may become pregnant should check with their physicians before using digitalis drugs.

Older people may be especially sensitive to the effects of digitalis drugs, which may increase the chance of overdose.

Before using digitalis drugs, people with any of the following medical problems should make sure their physicians are aware of their conditions:

- heart disease
- heart rhythm problems
- severe lung disease
- kidney disease
- liver disease
- thyroid disease

### Side effects

Side effects are rare with this medicine. Check with a physician as soon as possible if a skin rash, **hives**, or any other unusual or troublesome symptoms occur. Watch for signs of overdose.

### Interactions

Digitalis drugs may interact with a number of other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. For example:

- Taking digitalis drugs with other heart medicines, amphetamines, or diet pills could increase the risk of heart rhythm problems.
- Calcium channel blockers, used to treat high blood pressure, may cause higher than usual levels of digitalis drugs in the body that could lead to symptoms of overdose as covered in the above section.
- Diuretics (water pills) or other medicines that lower the amount of potassium in the body may increase the side effects of digitalis drugs.
- Medicines that increase the amount of potassium in the body may raise the risk of serious heart rhythm problems when taken with digitalis drugs.

- Diarrhea medicine or cholesterol-lowering drugs such as cholestyramine (Questran) and colestipol (Colestid) may keep digitalis medicines from being absorbed into the body. To prevent this problem, digitalis drugs should be taken several hours before or after taking these medicines.

The list above does not include every drug that may interact with digitalis drugs. Be sure to check with a physician or pharmacist before taking any other prescription or nonprescription (over-the-counter) medicine.

In addition, a diet high in fiber may interfere with the effects of digitalis drugs by preventing the medicine from being absorbed into the body. To avoid this problem, eat high fiber foods (such as bran products, whole wheat bread, and fresh fruits and vegetables) several hours before or after taking digitalis medicine.

Nancy Ross-Flanigan

Digoxin see **Digitalis drugs; Antiarrhythmic drugs**

## Dilatation and curettage

### Definition

Dilatation and curettage (D & C) is a gynecological procedure in which the lining of the uterus (endometrium) is scraped away.

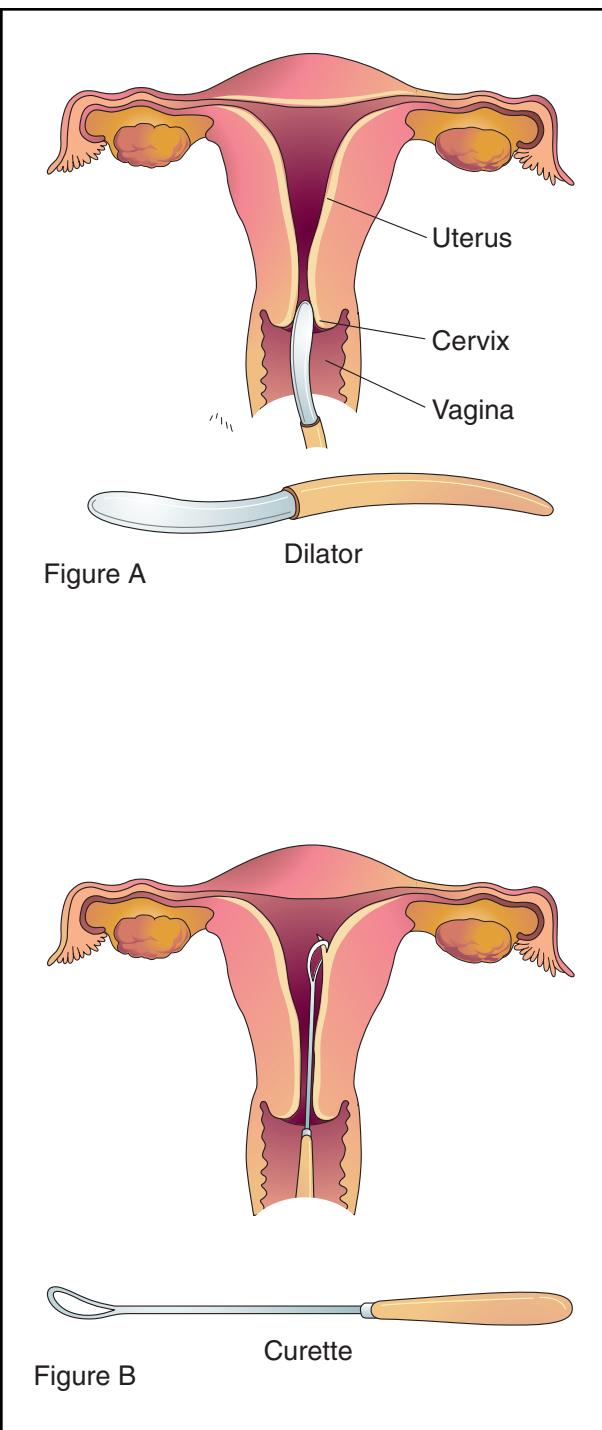
### Purpose

D & C is commonly used to obtain tissue for microscopic evaluation to rule out **cancer**. D & C may also be used to diagnose and treat heavy menstrual bleeding, and to diagnose endometrial polyps and **uterine fibroids**. A D & C can be used as a treatment as well, to remove **pregnancy** tissue after a **miscarriage**, incomplete abortion, or **childbirth**. Endometrial polyps may be removed, and sometimes benign uterine tumors (fibroids) may be scraped away. D & C can also be used as an early abortion technique up to 16 weeks.

### Description

D & C is usually performed under general anesthesia, although local or epidural anesthesia can also be used. A local lessens risk and costs, but the woman will feel cramping during the procedure. The type of anesthesia used often depends upon the reason for the D & C.

In the procedure (which takes only minutes to perform), the doctor inserts an instrument to hold open the



When performing a D & C, the physician inserts a speculum to separate and hold the vaginal walls, then stretches open the cervix with a dilator. Once the cervix is dilated, the physician will insert a curette into the uterus and scrapes away small portions of the uterine lining for laboratory analysis. (Illustration by Electronic Illustrators Group.)

## KEY TERMS

**Endometrial polyps**—A growth in the lining of the uterus (endometrium) that may cause bleeding and can develop into cancer.

**Epidural anesthesia**—A type of anesthesia that is injected into the epidural space of the spinal cord to numb the nerves leading to the lower half of the body.

**Uterine fibroid**—A noncancerous tumor of the uterus that can range from the size of a pea to the size of a grapefruit. Small fibroids require no treatment, but those causing serious symptoms may need to be removed.

vaginal walls, and then stretches the opening of the uterus to the vagina (the cervix) by inserting a series of tapering rods, each thicker than the previous one, or by using other specialized instruments. This process of opening the cervix is called dilation.

Once the cervix is dilated, the physician inserts a spoon-shaped surgical device called a curette into the uterus. The curette is used to scrape away the uterine lining. One or more small tissue samples from the lining of the uterus or the cervical canal are sent for analysis by microscope to check for abnormal cells.

Although simpler, less expensive techniques such as a vacuum aspiration are quickly replacing the D & C as a diagnostic method, it is still often used to diagnose and treat a number of conditions.

### Preparation

Because opening the cervix can be painful, sedatives may be given before the procedure begins. Deep breathing and other relaxation techniques may help ease cramping during cervical dilation.

### Aftercare

A woman who has had a D & C performed in a hospital can usually go home the same day or the next day. Many women experience backache and mild cramps after the procedure, and may pass small blood clots for a day or so. Vaginal staining or bleeding may continue for several weeks.

Most women can resume normal activities almost immediately. Patients should avoid sexual intercourse, douching, and tampon use for at least two weeks to pre-

vent infection while the cervix is closing and to allow the endometrium to heal completely.

### Risks

The primary risk after the procedure is infection. Signs of infection include:

- fever
- heavy bleeding
- severe cramps
- foul-smelling vaginal discharge

A woman should report any of these symptoms to her doctor, who can treat the infection with antibiotics before it becomes serious.

D & C is a surgical operation, which carries certain risks associated with general anesthesia. Rare complications include puncture of the uterus (which usually heals on its own) or puncture of the bowel or bladder (which require further surgery to repair).

### Normal results

Removal of the uterine lining causes no side effects, and may be beneficial if the lining has thickened so much that it causes heavy periods. The uterine lining soon grows again normally, as part of the menstrual cycle.

### Resources

#### BOOKS

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Carol A. Turkington

Dilated cardiomyopathy see **Congestive cardiomyopathy**

Diltiazem see **Calcium channel blockers**

Dilution test see **Kidney function tests**

Diphenhydramine see **Antihistamines**

## Diphtheria

### Definition

Diphtheria is a potentially fatal, contagious disease that usually involves the nose, throat, and air passages,

but may also infect the skin. Its most striking feature is the formation of a grayish membrane covering the tonsils and upper part of the throat.

## Description

Like many other upper respiratory diseases, diphtheria is most likely to break out during the winter months. At one time it was a major childhood killer, but it is now rare in developed countries because of widespread immunization. Since 1988, all confirmed cases in the United States have involved visitors or immigrants. In countries that do not have routine immunization against this infection, the mortality rate varies from 1.5–25%.

Persons who have not been immunized may get diphtheria at any age. The disease is spread most often by droplets from the coughing or sneezing of an infected person or carrier. The incubation period is two to seven days, with an average of three days. It is vital to seek medical help at once when diphtheria is suspected, because treatment requires emergency measures for adults as well as children.

## Causes and symptoms

The symptoms of diphtheria are caused by toxins produced by the diphtheria bacillus, *Corynebacterium diphtheriae* (from the Greek for “rubber membrane”). In fact, toxin production is related to infections of the bacillus itself with a particular bacteria virus called a phage (from bacteriophage; a virus that infects bacteria). The intoxication destroys healthy tissue in the upper area of the throat around the tonsils, or in open **wounds** in the skin. Fluid from the dying cells then coagulates to form the telltale gray or grayish green membrane. Inside the membrane, the bacteria produce an exotoxin, which is a poisonous secretion that causes the life-threatening symptoms of diphtheria. The exotoxin is carried throughout the body in the bloodstream, destroying healthy tissue in other parts of the body.

The most serious complications caused by the exotoxin are inflammations of the heart muscle (**myocarditis**) and damage to the nervous system. The risk of serious complications is increased as the time between onset of symptoms and the administration of antitoxin increases, and as the size of the membrane formed increases. The myocarditis may cause disturbances in the heart rhythm and may culminate in **heart failure**. The symptoms of nervous system involvement can include seeing double (diplopia), painful or difficult swallowing, and slurred speech or loss of voice, which are all indications of the exotoxin’s effect on nerve functions. The exotoxin may also cause severe swelling in the neck (“bull neck”).

The signs and symptoms of diphtheria vary according to the location of the infection:

### Nasal

Nasal diphtheria produces few symptoms other than a watery or bloody discharge. On examination, there may be a small visible membrane in the nasal passages. Nasal infection rarely causes complications by itself, but it is a public health problem because it spreads the disease more rapidly than other forms of diphtheria.

### Pharyngeal

Pharyngeal diphtheria gets its name from the pharynx, which is the part of the upper throat that connects the mouth and nasal passages with the voice box. This is the most common form of diphtheria, causing the characteristic throat membrane. The membrane often bleeds if it is scraped or cut. It is important not to try to remove the membrane because the trauma may increase the body’s absorption of the exotoxin. Other signs and symptoms of pharyngeal diphtheria include mild **sore throat**, **fever** of 101–102°F (38.3–38.9°C), a rapid pulse, and general body weakness.

### Laryngeal

Laryngeal diphtheria, which involves the voice box or larynx, is the form most likely to produce serious complications. The fever is usually higher in this form of diphtheria (103–104°F or 39.4–40°C) and the patient is very weak. Patients may have a severe **cough**, have difficulty breathing, or lose their voice completely. The development of a “bull neck” indicates a high level of exotoxin in the bloodstream. Obstruction of the airway may result in respiratory compromise and **death**.

### Skin

This form of diphtheria, which is sometimes called cutaneous diphtheria, accounts for about 33% of diphtheria cases. It is found chiefly among people with poor hygiene. Any break in the skin can become infected with diphtheria. The infected tissue develops an ulcerated area and a diphtheria membrane may form over the wound but is not always present. The wound or ulcer is slow to heal and may be numb or insensitive when touched.

### Diagnosis

Because diphtheria must be treated as quickly as possible, doctors usually make the diagnosis on the basis of the visible symptoms without waiting for test results.

In making the diagnosis, the doctor examines the patient’s eyes, ears, nose, and throat in order to rule out

other diseases that may cause fever and sore throat, such as **infectious mononucleosis**, a sinus infection, or **strep throat**. The most important single symptom that suggests diphtheria is the membrane. When a patient develops skin infections during an outbreak of diphtheria, the doctor will consider the possibility of cutaneous diphtheria and take a smear to confirm the diagnosis.

### Laboratory tests

The diagnosis of diphtheria can be confirmed by the results of a culture obtained from the infected area. Material from the swab is put on a microscope slide and stained using a procedure called Gram's stain. The diphtheria bacillus is called Gram-positive because it holds the dye after the slide is rinsed with alcohol. Under the microscope, diphtheria bacilli look like beaded rod-shaped cells, grouped in patterns that resemble Chinese characters. Another laboratory test involves growing the diphtheria bacillus on a special material called Loeffler's medium.

### Treatment

Diphtheria is a serious disease requiring hospital treatment in an intensive care unit if the patient has developed respiratory symptoms. Treatment includes a combination of medications and supportive care:

#### Antitoxin

The most important step is prompt administration of diphtheria antitoxin, without waiting for laboratory results. The antitoxin is made from horse serum and works by neutralizing any circulating exotoxin. The doctor must first test the patient for sensitivity to animal serum. Patients who are sensitive (about 10%) must be desensitized with diluted antitoxin, since the antitoxin is the only specific substance that will counteract diphtheria exotoxin. No human antitoxin is available for the treatment of diphtheria.

The dose ranges from 20,000–100,000 units, depending on the severity and length of time of symptoms occurring before treatment. Diphtheria antitoxin is usually given intravenously.

#### Antibiotics

**Antibiotics** are given to wipe out the bacteria, to prevent the spread of the disease, and to protect the patient from developing **pneumonia**. They are not a substitute for treatment with antitoxin. Both adults and children may be given penicillin, ampicillin, or erythromycin. Erythromycin appears to be more effective than penicillin in treating people who are carriers because of better penetration into the infected area.

Cutaneous diphtheria is usually treated by cleansing the wound thoroughly with soap and water, and giving the patient antibiotics for 10 days.

### Supportive care

Diphtheria patients need bed rest with intensive nursing care, including extra fluids, oxygenation, and monitoring for possible heart problems, airway blockage, or involvement of the nervous system. Patients with laryngeal diphtheria are kept in a **croup** tent or high-humidity environment; they may also need throat suctioning or emergency surgery if their airway is blocked.

Patients recovering from diphtheria should rest at home for a minimum of two to three weeks, especially if they have heart complications. In addition, patients should be immunized against diphtheria after recovery, because having the disease does not always induce antitoxin formation and protect them from reinfection.

### Prevention of complications

Diphtheria patients who develop myocarditis may be treated with oxygen and with medications to prevent irregular heart rhythms. An artificial pacemaker may be needed. Patients with difficulty swallowing can be fed through a tube inserted into the stomach through the nose. Patients who cannot breathe are usually put on mechanical respirators.

### Prognosis

The prognosis depends on the size and location of the membrane and on early treatment with antitoxin; the longer the delay, the higher the death rate. The most vulnerable patients are children under age 15 and those who develop pneumonia or myocarditis. Nasal and cutaneous diphtheria are rarely fatal.

### Prevention

Prevention of diphtheria has four aspects:

#### Immunization

Universal immunization is the most effective means of preventing diphtheria. The standard course of immunization for healthy children is three doses of DPT (diphtheria-tetanus-pertussis) preparation given between two months and six months of age, with booster doses given at 18 months and at entry into school. Adults should be immunized at 10 year intervals with Td (tetanus-diphtheria) toxoid. A toxoid is a bacterial toxin that is treated to make it harmless but still can induce immunity to the disease.

## KEY TERMS

- Antitoxin**—An antibody against an exotoxin, usually derived from horse serum.
- Bacillus**—A rod-shaped bacterium, such as the diphtheria bacterium.
- Carrier**—A person who may harbor an organism without symptoms and may transmit it to others.
- Cutaneous**—Located in the skin.
- Diphtheria-tetanus-pertussis (DTP)**—The standard preparation used to immunize children against diphtheria, tetanus, and whooping cough. A so-called “acellular pertussis” vaccine (aP) is usually used since its release in the mid-1990s.
- Exotoxin**—A poisonous secretion produced by bacilli which is carried in the bloodstream to other parts of the body.
- Gram's stain**—A dye staining technique used in laboratory tests to determine the presence and type of bacteria.
- Loeffler's medium**—A special substance used to grow diphtheria bacilli to confirm the diagnosis.
- Myocarditis**—Inflammation of the heart tissue.
- Toxoid**—A preparation made from inactivated exotoxin, used in immunization.

### Isolation of patients

Diphtheria patients must be isolated for one to seven days or until two successive cultures show that they are no longer contagious. Children placed in **isolation** are usually assigned a primary nurse for emotional support.

### Identification and treatment of contacts

Because diphtheria is highly contagious and has a short incubation period, family members and other contacts of diphtheria patients must be watched for symptoms and tested to see if they are carriers. They are usually given antibiotics for seven days and a booster shot of diphtheria/tetanus toxoid.

### Reporting cases to public health authorities

Reporting is necessary to track potential epidemics, to help doctors identify the specific strain of diphtheria, and to see if resistance to penicillin or erythromycin has developed.

## Resources

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Rebecca J. Frey

### Diplegia see Paralysis

### Direct Coombs' test see Coombs' tests

### Direct laryngoscopy see Laryngoscopy

## Discoid lupus erythematosus

### Definition

Discoid lupus erythematosus (DLE) is a disease in which coin-shaped (discoid) red bumps appear on the skin.

### Description

The disease called discoid lupus erythematosus only affects the skin, although similar discoid **skin lesions** can occur in the serious disease called **systemic lupus erythematosus** (SLE). Only about 10% of all patients with DLE will go on to develop the multi-organ disease SLE.

The tendency to develop DLE seems to run in families. Although men or women of any age can develop DLE, it occurs in women three times more frequently than in men. The typical DLE patient is a woman in her 30s.



**Discoloration of the hands is one characteristic of discoid lupus erythematosus.** (Custom Medical Stock Photo. Reproduced by permission.)

### Causes and symptoms

The cause of DLE is unknown. It is thought that DLE (like SLE) may be an autoimmune disorder. **Autoimmune disorders** are those that occur when cells of the immune system are misdirected against the body. Normally, immune cells work to recognize and help destroy foreign invaders like bacteria, viruses, and fungi. In autoimmune disorders, these cells mistakenly recognize various tissues of the body as foreign invaders, and attack and destroy these tissues. In SLE, the misdirected immune cells are antibodies. In DLE, the damaging cells are believed to be a type of white blood cell called a T lymphocyte. The injury to the skin results in inflammation and the characteristic discoid lesions.

In DLE, the characteristic skin lesion is circular and raised. The reddish rash is about 5–10 mm in diameter, with the center often somewhat scaly and lighter in color than the darker outer ring. The surface of these lesions is sometimes described as “warty.” There is rarely any **itching** or **pain** associated with discoid lesions. They tend to appear on the face, ears, neck, scalp, chest, back, and arms. As DLE lesions heal, they leave thickened, scarred areas of skin. When the scalp is severely affected, there may be associated hair loss (**alopecia**).

People with DLE tend to be quite sensitive to the sun. They are more likely to get a **sunburn**, and the sun is likely to worsen their discoid lesions.

### Diagnosis

Diagnosis of DLE usually requires a **skin biopsy**. A small sample of a discoid lesion is removed, specially prepared, and examined under a microscope. Usually, the lesion has certain microscopic characteristics that allow it to be identified as a DLE lesion. Blood tests will not

reveal the type of antibodies present in SLE, and **physical examination** usually does not reveal anything other than the skin lesions. If antibodies exist in the blood, or if other symptoms or physical signs are found, it is possible that the discoid lesions are a sign of SLE rather than DLE.

### Treatment

Treatment of DLE primarily involves the use of a variety of skin creams. **Sunscreens** are used for protection. Steroid creams can be applied to decrease inflammation. Occasionally, small amounts of a steroid preparation will be injected with a needle into a specific lesion. Because of their long list of side effects, steroid preparations taken by mouth are avoided. Sometimes, short-term treatment with oral steroids will be used for particularly severe DLE outbreaks. Medications used to treat the infectious disease **malaria** are often used to treat DLE.

### Alternative treatment

Alternative treatments for DLE include eating a healthy diet, low in red meat and dairy products and high in fish containing omega-3 fatty acids. These types of fish include mackerel, sardines, and salmon. Following a healthy diet is thought to decrease inflammation. Dietary supplements believed to be helpful include **vitamins B, C, E, and selenium**. Vitamin A is also recommended to improve DLE lesions. Constitutional homeopathic treatment can help heal DLE as well as help prevent it developing into SLE.

### Prognosis

For the most part, the prognosis for people with DLE is excellent. While the lesions may be cosmetically unsightly, they are not life threatening and usually do not cause a patient to change his or her lifestyle. Only about 10% of patients with DLE will go on to develop SLE.

### Prevention

DLE cannot be prevented. Recommendations to prevent flares of DLE in patients with the disease include avoiding exposure to sun and consistently using sunscreen.

### Resources

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## KEY TERMS

**Antibody**—Specialized cells of the immune system that can recognize organisms invading the body (like bacteria, viruses, and fungi). The antibodies are then able to start a complex chain of events designed to kill these foreign invaders.

**Autoimmune disorder**—A disorder in which the body's antibodies mistake the body's own tissues for foreign invaders. The immune system then attacks and causes damage to these tissues.

**Immune system**—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

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### PERIODICALS

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 Umansky, Diane. "Living with Lupus." *American Health for Women* 16, no. 5 (June 1997): 92+.

### ORGANIZATIONS

- The American College of Rheumatology. 1800 Century Place, Suite 250, Atlanta, GA 30345. (404) 633-3777. <<http://www.rheumatology.org>>.  
 Lupus Foundation of America. 1300 Piccard Dr., Suite 200, Rockville, MD 20850. (800) 558-0121. <<http://www.lupus.org>>.

Rosalyn Carson-DeWitt, MD

## Disk removal

### Definition

One of the most common types of back surgery is disk removal (diskectomy), the removal of an intervertebral disk, the flexible plate that connects any two adjacent vertebrae in the spine. Intervertebral disks act as shock absorbers, protecting the brain and spinal cord from the impact produced by the body's movements.

### Purpose

About 150,000 Americans undergo disk removal each year in the United States. Removing the invertebral

disk is performed to treat back pain that has lasted at least six weeks as a result of an abnormal disk and that has not responded to conservative treatment. Surgery is also performed if there is pressure on the lumbosacral nerve roots that causes weakness or bowel or bladder dysfunction.

As a person ages, the disks between the vertebrae degenerate and dry out, and the fibers holding them in place tear. Eventually, the disk can form a blister-like bulge, compressing nerves in the spine and causing pain. This is called a "prolapsed" (or herniated) disk. If such a disk causes muscle weakness or interferes with bladder or bowel function because it is pressing on a nerve root, immediate surgery to remove the disk may be needed.

The aim of the surgery is to try to relieve all pressure on nerve roots by removing the pulpy material from the disk, or the disk itself. If it is necessary to remove material from several nearby vertebrae, the spine may become unsteady. In this case, the surgeon will perform a spinal fusion, removing all the disks between two or more vertebrae and roughening the bones so that the vertebrae heal together. Bone strips taken from the patient's leg or hip may be used to help hold the vertebrae together. Spinal fusion decreases pain but it also decreases spinal mobility.

### Precautions

The doctor will obtain x rays, neuroimaging studies, including computed tomography scan (CT scan) myelogram and **magnetic resonance imaging** (MRI), and clinical exams to determine the precise location of the affected disk.

### Description

The surgery is done under general anaesthesia, which puts the patient to sleep and affects the whole body. Operating on the patient's back, the neurosurgeon or orthopedic surgeon makes an opening into the vertebral canal, and then moves the dura and the bundle of nerves called the "cauda equina" (horse's tail) aside, which exposes the disk. If a portion of the disk has moved from between the vertebrae out into the nerve canal, it is simply removed. If the disk itself has become fragmented and partially displaced, or not fragmented but bulging extensively, the surgeon will remove the bulging or displaced part of the disk and the part that lies in the space between the vertebrae.

### Preparation

The patient is given an injection an hour before the surgery to dry up internal fluids and encourage drowsiness.

## KEY TERMS

**Diskectomy**—The surgical removal of a portion of an invertebral disk.

**Dura**—The strongest and outermost of three membranes that protect the brain, spinal cord, and nerves of the cauda equina.

**Herniated disk**—A blisterlike bulging or protrusion of the contents of the disk out through the fibers that normally hold them in place. It is also called a ruptured disk, slipped disk, or displaced disk.

**Intervertebral disk**—Cylindrical elastic-like gel pads that separate and join each pair of vertebrae in the spine.

**Laminectomy**—An operation in which the surgeon cuts through the covering of a vertebra to reach a herniated disk in order to remove it.

**Vertebra**—The bones that make up the back bone (spine).

### Aftercare

After the operation, the patient will awaken lying flat and face down, and must remain this way for several days, changing position only to avoid **bedsores**. There maybe slight pain or stiffness in the back area.

Patients should sleep on a firm mattress and avoid bending at the waist, lifting heavy weights, or sitting in one spot for a long time (such as riding in a car).

After surgery, patients can usually leave the hospital on the fourth or fifth day. They must:

- avoid sitting for more than 15–20 minutes
- use a reclined chair
- avoid bending, twisting, or lifting
- begin gentle walking (indoors or outdoors), gradually increasing
- begin stationary biking or gentle swimming after two weeks
- continue **exercise** for the next four weeks
- slow down if they experience more than minor pain in the back or leg

### Risks

All surgery carries some risk due to heart and lung problems or the anesthesia itself, but this risk is generally

extremely small. (The risk of **death** from general anesthesia for all types of surgery, for example, is only about 1 in 1,600.)

The most common risk of the surgery is infection, which occurs in 1–2% of cases. Rarely, the surgery can damage nerves in the lower back or major blood vessels in front of the disk. Occasionally, there may be some residual **paralysis** of a particular leg or bladder muscle after surgery, but this is the result of the disk problem that necessitated the surgery, not the operation itself.

While disk removals can relieve pain in 90% of cases, there are some people who do not get pain relief, depending on how long they had the condition requiring surgery and other factors.

### Normal results

After about five days, most patients can leave the hospital. They can resume all normal activities, including work, after four to six weeks of recuperation at home.

In properly evaluated patients, there is a very good chance that disk removal will be successful in easing pain. Even in patients over age 60, disk surgery has a “good to excellent” result for 87% of patients. Disk surgery can relieve both back and leg pain, but the greatest pain relief will occur with the leg pain.

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Diskectomy see **Disk removal**

## Dislocations and subluxations

### Definition

In medicine, the terms dislocation and subluxation refer to the displacement of bones that form a joint. These conditions affecting the joint most often result from trauma that causes adjoining bones to no longer align with each other. A partial or incomplete dislocation is called a subluxation.

## Description

In a healthy joint, the bones are normally held together with tough, fibrous bands called ligaments. These ligaments are attached to each bone along with a fibrous sac surrounding the joint called the articular capsule or joint capsule. The ligaments and joint capsule are relatively strong and nonelastic but permit movement within normal limits for each particular joint. In the event of a dislocation, one of the bones making up the joint is forced out of its natural alignment from excessive stretching and tearing of the joint ligaments and capsule. Muscles and tendons surrounding the joint are usually stretched and injured to some degree.

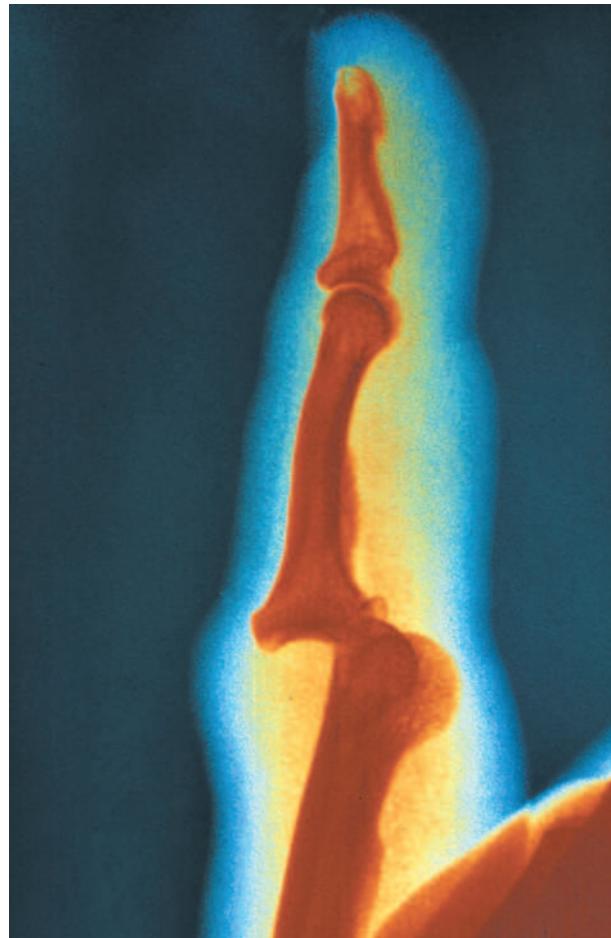
## Causes and symptoms

A violent movement at the joint that exceeds normal limits usually causes a joint dislocation. Although dislocations often result from trauma, they sometimes occur as a result of disease affecting the joint structures. In the process of the dislocation, there is tearing of the ligaments and the articular capsule, which are vital structures for connecting the bone. Following a dislocation, the bones affected are often immobile and the affected limb may be locked in an abnormal position; **fractures** are also a concern with severe dislocations.

Important factors in recognizing a dislocation or subluxation include a history of experiencing a fall or receiving a blow in a particular joint followed by the sudden onset of loss of function to the involved limb. Immediately after the dislocation, the joint almost always swells significantly and feels painful when pressure is applied (point tenderness). If trauma to the joint causing the dislocation or subluxation is violent in nature, small chips of bone can be torn away with the supporting structures. Chronic recurrent dislocations may take place without severe **pain** because of the somewhat slack condition of the surrounding muscles and other supporting tissues. A first-time dislocation is considered and treated as a possible fracture. Risk factors that can increase susceptibility of joint dislocation and subluxation are shallow or abnormally formed joint surfaces present at birth (congenital) and/or other diseases of ligaments and tissue around a joint. Some infants are born with a hip dislocation. Both sexes and all ages are affected.

## Diagnosis

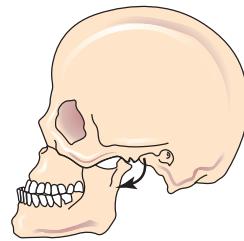
A thorough medical history and physical exam by a physician is the first step in the correct diagnosis of dislocations and subluxations. X rays of the joint and adjacent bones can locate and help determine the extent of dislocated joints.



This x ray shows the dislocation between two bones in a finger. (Photo Researchers, Inc. Reproduced by permission.)

## Treatment

Immediately after the dislocation, the application of ice is helpful to control swelling and decrease pain. If the patient needs to be transported, it is important to prevent the joint from moving (**immobilization**). At times, a cast or splint may be used to immobilize the joint and ensure proper alignment and healing. The treatment of realigning bones following a dislocation is called reduction. This may include simple maneuvers that manipulate the joint to reposition the bones or surgical procedures to restore the joint to its normal position. A general anesthesia or muscle relaxant may be used to help make joint reduction possible by relaxing surrounding muscles in spasm. **Acetaminophen** or **aspirin** are sometimes used to control moderate pain, and narcotics may be prescribed by the physician if the pain is severe. Recurring dislocation may require surgical reconstruction or replacement of the joint. It is not recommended to attempt to reset a dislocated joint outside of a medical



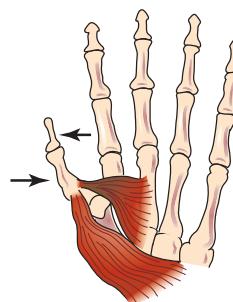
Anterior temporal mandibular dislocation



Subcoracoid dislocation (shoulder)



Posterior dislocation of hip



Dislocation of thumb

**Dislocations and subluxations** refer to the displacement of bones that form a joint. Such conditions most often result from trauma causing adjoining bones to no longer touch each other. A partial or incomplete dislocation is called a subluxation. The illustrations above indicate dislocation of the jaw bone, shoulder blade, hip bone, and the thumb. (Illustration by Electronic Illustrators Group.)

environment with experienced medical personnel, because a fracture may be present.

### Alternative treatment

**Chiropractic** care has been shown to be effective for joint subluxation and dislocation, especially in the spine. Swelling can be addressed using botanical therapies. Bromelain, a pineapple enzyme, and turmeric (*Curcuma longa*) are the most potent botanical remedies for this purpose. Acute homeopathic care with arnica (*Arnica montana*) can reduce the trauma to the body. Ligament and tendon strengthening can be assisted both botanically and homeopathically.

### Prognosis

Joint ligaments have poor blood supply and, therefore, heal slowly. This healing process continues long after the symptoms of the dislocation injury have diminished. Once a joint has been either subluxated or completely dislocated, the connective tissue binding or

holding it in correct alignment is stretched to such an extent that the joint becomes extremely vulnerable to repeated dislocations. However, this chance of recurrent dislocation and subluxation will decrease if a proper **rehabilitation** program is implemented to strengthen surrounding muscles of the joint. Most joint dislocations are curable with prompt treatment. After the dislocation has been corrected, the joint may require immobilization with a cast or sling for two to eight weeks.

### Prevention

When an individual is involved in strenuous sports or heavy work, involved joints may be protected by elastic bandage wraps, tape wraps, knee and shoulder pads, or special support stockings. Keeping the muscles surrounding the joint strong will also help prevent dislocations. Long-term problems may also be prevented by allowing an adequate amount of time for an injured joint to rest and heal prior to resuming full activity.

## KEY TERMS

**Articular capsule**—An envelope of tissue that surrounds a free moving joint, composed of an external layer of white fibrous tissue and an external synovial membrane that secretes a lubricant into the joint.

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Disopyramide see **Antiarrhythmic drugs**

Disproportionate dwarfism see  
**Achondroplasia**

Dissecting aneurysm see **Aortic dissection**

Dissecting hematoma see **Aortic dissection**

Disseminated lupus erythematosus see  
**Systemic lupus erythematosus**

## Description

In order to have a clear picture of these disorders, dissociation should first be understood. Dissociation is a mechanism that allows the mind to separate or compartmentalize certain memories or thoughts from normal consciousness. These split-off mental contents are not erased. They may resurface spontaneously or be triggered by objects or events in the person's environment.

Dissociation is a process that occurs along a spectrum of severity. It does not necessarily mean that a person has a dissociative disorder or other mental illness. A mild degree of dissociation occurs with some physical stressors; people who have gone without sleep for a long period of time, have had "laughing gas" for dental surgery, or have been in a minor accident often have brief dissociative experiences. Another commonplace example of dissociation is a person becoming involved in a book or movie so completely that the surroundings or the passage of time are not noticed. Another example might be driving on the highway and taking several exits without noticing or remembering. Dissociation is related to hypnosis in that hypnotic trance also involves a temporarily altered state of consciousness. Most patients with dissociative disorders are highly hypnotizable.

People in other cultures sometimes have dissociative experiences in the course of religious (in certain trance states) or other group activities. These occurrences should not be judged in terms of what is considered "normal" in the United States.

Moderate or severe forms of dissociation are caused by such traumatic experiences as childhood **abuse**, combat, criminal attacks, brainwashing in hostage situations, or involvement in a natural or transportation disaster. Patients with **acute stress disorder**, **post-traumatic stress disorder** (PTSD), or conversion disorder and somatization disorder may develop dissociative symptoms. Recent studies of trauma indicate that the human brain stores traumatic memories in a different way than normal memories. Traumatic memories are not processed or integrated into a person's ongoing life in the same fashion as normal memories. Instead they are dissociated, or "split off," and may erupt into consciousness from time to time without warning. The affected person cannot control or "edit" these memories. Over a period of time, these two sets of memories, the normal and the traumatic, may coexist as parallel sets without being combined or blended. In extreme cases, different sets of dissociated memories may alter subpersonalities of patients with dissociative identity disorder (**multiple personality disorder**).

The dissociative disorders vary in their severity and the suddenness of onset. It is difficult to give statistics

## Dissociative disorders

### Definition

The dissociative disorders are a group of mental disorders that affect consciousness are defined as causing significant interference with the patient's general functioning, including social relationships and employment.

for their frequency in the United States because they are a relatively new category and are often misdiagnosed. And, criterion for diagnosis require significant impairment in social or vocational functioning.

### **Dissociative amnesia**

Dissociative **amnesia** is a disorder in which the distinctive feature is the patient's inability to remember important personal information to a degree that cannot be explained by normal forgetfulness. In many cases, it is a reaction to a traumatic accident or witnessing a violent crime. Patients with dissociative amnesia may develop depersonalization or trance states as part of the disorder, but they do not experience a change in identity.

### **Dissociative fugue**

Dissociative fugue is a disorder in which a person temporarily loses his or her sense of personal identity and travels to another location where he or she may assume a new identity. Again, this condition usually follows a major stressor or trauma. Apart from inability to recall their past or personal information, patients with dissociative fugue do not behave strangely or appear disturbed to others. Cases of dissociative fugue are more common in wartime or in communities disrupted by a natural disaster.

### **Depersonalization disorder**

Depersonalization disorder is a disturbance in which the patient's primary symptom is a sense of detachment from the self. Depersonalization as a symptom (not as a disorder) is quite common in college-age populations. It is often associated with sleep deprivation or "recreational" drug use. It may be accompanied by "derealization" (where objects in an environment appear altered). Patients sometimes describe depersonalization as feeling like a robot or watching themselves from the outside. Depersonalization disorder may also involve feelings of numbness or loss of emotional "aliveness."

### **Dissociative identity disorder (DID)**

Dissociative identity disorder (DID) is the newer name for multiple personality disorder (MPD). DID is considered the most severe dissociative disorder and involves all of the major dissociative symptoms.

### **Dissociative disorder not otherwise specified (DDNOS)**

DDNOS is a diagnostic category ascribed to patients with dissociative symptoms that do not meet the full criteria for a specific dissociative disorder.

## **Causes and symptoms**

The moderate to severe dissociation that occurs in patients with dissociative disorders is understood to result from a set of causes:

- an innate ability to dissociate easily
- repeated episodes of severe physical or sexual abuse in childhood
- the lack of a supportive or comforting person to counteract abusive relative(s)
- the influence of other relatives with dissociative symptoms or disorders

The relationship of dissociative disorders to childhood abuse has led to intense controversy and lawsuits concerning the accuracy of childhood memories. The brain's storage, retrieval, and interpretation of memories are still not fully understood. Controversy also exists regarding how much individuals presenting dissociative disorders have been influenced by books and movies to describe a certain set of symptoms (scripting).

The major dissociative symptoms are:

### **Amnesia**

Amnesia in a dissociative disorder is marked by gaps in a patient's memory for long periods of time or for traumatic events. Doctors can distinguish this type of amnesia from loss of memory caused by head injuries or drug intoxication, because the amnesia is "spotty" and related to highly charged events and feelings.

### **Depersonalization**

Depersonalization is a dissociative symptom in which the patient feels that his or her body is unreal, is changing, or is dissolving. Some patients experience depersonalization as being outside their bodies or watching a movie of themselves.

### **Derealization**

Derealization is a dissociative symptom in which the external environment is perceived as unreal. The patient may see walls, buildings, or other objects as changing in shape, size, or color. In some cases, the patient may feel that other persons are machines or robots, though the patient is able to acknowledge the unreality of this feeling.

### **Identity disturbances**

Patients with dissociative fugue, DDNOS, or DID often experience confusion about their identities or even assume new identities. Identity disturbances result from the patient having split off entire personality traits

or characteristics as well as memories. When a stressful or traumatic experience triggers the reemergence of these dissociated parts, the patient may act differently, answer to a different name, or appear confused by his or her surroundings.

## Diagnosis

When a doctor is evaluating a patient with dissociative symptoms, he or she will first rule out physical conditions that sometimes produce amnesia, depersonalization, or derealization. These physical conditions include epilepsy, head injuries, brain disease, side effects of medications, substance abuse, intoxication, **AIDS**, **dementia** complex, or recent periods of extreme physical **stress** and sleeplessness. In some cases, the doctor may give the patient an electroencephalogram (EEG) to exclude epilepsy or other seizure disorders.

If the patient appears to be physically normal, the doctor will rule out psychotic disturbances, including **schizophrenia**. In addition, doctors can use some **psychological tests** to narrow the diagnosis. One is a screener, the Dissociative Experiences Scale (DES). If the patient has a high score on this test, he or she can be evaluated further with the Dissociative Disorders Interview Schedule (DDIS) or the Structured Clinical Interview for *DSM-IV* Dissociative Disorders (SCID-D). It is also possible for doctors to measure a patient's hypnotizability as part of a diagnostic evaluation.

## Treatment

Treatment of the dissociative disorders often combines several methods.

### *Psychotherapy*

Patients with dissociative disorders often require treatment by a therapist with some specialized understanding of dissociation. This background is particularly important if the patient's symptoms include identity problems. Many patients with dissociative disorders are helped by group as well as individual treatment.

### *Medications*

Some doctors will prescribe tranquilizers or antidepressants for the **anxiety** and/or depression that often accompany dissociative disorders. Patients with dissociative disorders are, however, at risk for abusing or becoming dependent on medications. As of 2001, there is no drug that can reliably counteract dissociation itself.

### *Hypnosis*

Hypnosis is frequently recommended as a method of treatment for dissociative disorders, partly because hyp-

## KEY TERMS

**Amnesia**—A general medical term for loss of memory that is not due to ordinary forgetfulness. Amnesia can be caused by head injuries, brain disease, or epilepsy, as well as by dissociation.

**Depersonalization**—A dissociative symptom in which the patient feels that his or her body is unreal, is changing, or is dissolving.

**Derealization**—A dissociative symptom in which the external environment is perceived as unreal.

**Dissociation**—A psychological mechanism that allows the mind to split off traumatic memories or disturbing ideas from conscious awareness.

**Fugue**—A dissociative experience during which a person travels away from home, has amnesia for their past, and may be confused about their identity but otherwise appear normal.

**Hypnosis**—The means by which a state of extreme relaxation and suggestibility is induced: used to treat amnesia and identity disturbances that occur in dissociative disorders.

**Multiple personality disorder (MPD)**—An older term for dissociative identity disorder (DID).

**Trauma**—A disastrous or life-threatening event that can cause severe emotional distress, including dissociative symptoms and disorders.

nosis is related to the process of dissociation. Hypnosis may help patients recover repressed ideas and memories. Therapists treating patients with DID sometimes use hypnosis in the process of “fusing” the patient’s alternate personalities.

## Prognosis

Prognoses for dissociative disorders vary. Recovery from dissociative fugue is usually rapid. Dissociative amnesia may resolve quickly, but can become a chronic disorder in some patients. Depersonalization disorder, DDNOS, and DID are usually chronic conditions. DID usually requires five or more years of treatment for recovery.

## Prevention

Since the primary cause of dissociative disorders is thought to involve extended periods of humanly inflicted trauma, prevention depends on the elimination of **child**

**abuse** and psychological abuse of adult prisoners or hostages.

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Dissociative identity disorder see **Multiple personality disorder**

## Diuretics

### Definition

Diuretics are medicines that help reduce the amount of water in the body.

### Purpose

Diuretics are used to treat the buildup of excess fluid in the body that occurs with some medical conditions such as congestive **heart failure**, liver disease, and kidney disease. Some diuretics are also prescribed to treat

high blood pressure. These drugs act on the kidneys to increase urine output. This reduces the amount of fluid in the bloodstream, which in turn lowers blood pressure.

### Description

There are several types of diuretics, also called water pills:

- Loop diuretics, such as bumetanide (Bumex) and furosemide (Lasix), get their name from the loop-shaped part of the kidneys where they have their effect.
- Thiazide diuretics include such commonly used diuretics as hydrochlorothiazide (HydroDIURIL, Esidrix), chlorothiazide (Diuril), and chlorthalidone (Hygroton).
- Potassium-sparing diuretics prevent the loss of potassium, which is a problem with other types of diuretics. Examples of potassium-sparing diuretics are amiloride (Midamor) and triamterene (Dyrenium).

In addition, some medicines contain combinations of two diuretics. The brands Dyazide and Maxzide, for example, contain the thiazide diuretic hydrochlorothiazide with the potassium-sparing diuretic triamterene.

Some nonprescription (over-the-counter) medicines contain diuretics. However, the medicines described here cannot be bought without a physician’s prescription. They are available in tablet, capsule, liquid, and injectable forms.

### Recommended dosage

The recommended dosage depends on the type of diuretic and may be different for different patients. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage, and take the medicine exactly as directed.

### Precautions

Seeing a physician regularly while taking a diuretic is important. The physician will check to make sure the medicine is working as it should and will watch for unwanted side effects.

Some people feel unusually tired when they first start taking diuretics. This effect usually becomes less noticeable over time, as the body adjusts to the medicine.

Because diuretics increase urine output, people who take this medicine may need to urinate more often, even during the night. Health care professionals can help patients schedule their doses to avoid interfering with their sleep or regular activities.

For patients taking the kinds of diuretics that rob potassium from the body, physicians may recommend adding potassium-rich foods or drinks, such as citrus fruits and juices, to the diet. Or they may suggest taking a potassium supplement or taking another medicine that keeps the body from losing too much potassium. If the physician recommends any of these measures, be sure to closely follow his or her directions. Do not make other diet changes without checking with the physician. People who are taking potassium-sparing diuretics should not add potassium to their **diets**, as too much potassium may be harmful.

People who take diuretics may lose too much water or potassium when they get sick, especially if they have severe vomiting and **diarrhea**. They should check with their physicians if they become ill.

These medicines make some people feel lightheaded, dizzy, or faint when they get up after sitting or lying down. Older people are especially likely to have this problem. Drinking alcohol, exercising, standing for long periods, or being in hot weather may make the problem worse. To lessen the problem, get up gradually and hold onto something for support if possible. Avoid drinking too much alcohol and be careful in hot weather or when exercising or standing for a long time.

Anyone who is taking a diuretic should be sure to tell the health care professional in charge before having surgical or dental procedures, medical tests or emergency treatment.

Some diuretics make the skin more sensitive to sunlight. Even brief exposure to sun can cause a severe **sunburn**, **itching**, a rash, redness, or other changes in skin color. While being treated with this medicine, avoid being in direct sunlight, especially between 10 a.m. and 3 p.m.; wear a hat and tightly woven clothing that covers the arms and legs; use a sunscreen with a skin protection factor (SPF) of at least 15; protect the lips with a sun block lipstick; and do not use tanning beds, tanning booths, or sunlamps. People with fair skin may need to use a sunscreen with a higher skin protection factor.

### ***Special conditions***

People who have certain medical conditions or who are taking certain other medicines may have problems if they take diuretics. Before taking these drugs, be sure to let the physician know about any of these conditions:

**ALLERGIES.** Anyone who has had unusual reactions to diuretics or **sulfonamides** (sulfa drugs) in the past should let his or her physician know before using a diuretic. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

**PREGNANCY.** Diuretics will not help the swelling of hands and feet that some women have during **pregnancy**. In general, pregnant women should not use diuretics unless a physician recommends their use. Although studies have not been done on pregnant women, studies of laboratory animals show that some diuretics can cause harmful effects when taken during pregnancy.

**BREASTFEEDING.** Some diuretics pass into breast milk, but no reports exist of problems in nursing babies whose mothers use this medicine. However, thiazide diuretics may decrease the flow of breast milk. Women who are breastfeeding and need to use a diuretic should check with their physicians.

**OTHER MEDICAL CONDITIONS.** Side effects of some diuretics may be more likely in people who have had a recent **heart attack** or who have liver disease or severe kidney disease. Other diuretics may not work properly in people with liver disease or severe kidney disease. Diuretics may worsen certain medical conditions, such as **gout**, **kidney stones**, **pancreatitis**, lupus erythematosus, and hearing problems. In addition, people with diabetes should be aware that diuretics may increase blood sugar levels. People with heart or blood vessel disease should know that some diuretics increase cholesterol or triglyceride levels. The risk of an allergic reaction to certain diuretics is greater in people with bronchial **asthma**. Before using diuretics, people with any of these medical problems should make sure their physicians are aware of their conditions. Also, people who have trouble urinating or who have high potassium levels in their blood may not be able to take diuretics and should check with a physician before using them.

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

### **Side effects**

Some side effects, such as loss of appetite, **nausea** and **vomiting**, stomach cramps, diarrhea and **dizziness**, usually lessen or go away as the body adjusts to the medicine. These problems do not need medical attention unless they continue or interfere with normal activities.

Patients taking potassium-sparing diuretics should know the signs of too much potassium and should check with a physician as soon as possible if any of these symptoms occur:

- irregular heartbeat
- breathing problems
- numbness or tingling in the hands, feet, or lips

## KEY TERMS

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

**Lupus erythematosus**—A chronic disease that affects the skin, joints, and certain internal organs.

**Pancreas**—A gland located beneath the stomach. The pancreas produces juices that help break down food.

**Potassium**—A mineral found in whole grains, meat, legumes, and some fruits and vegetables. Potassium is important for many body processes, including proper functioning of the nerves and muscles.

**Triglyceride**—A substance formed in the body from fat in the diet. Triglycerides are the main fatty materials in the blood. Together with protein, they make up high- and low-density lipoproteins (HDLs and LDLs). Triglyceride levels are important in the diagnosis and treatment of many diseases including high blood pressure, diabetes, and heart disease.

- confusion or nervousness
- unusual tiredness or weakness
- weak or heavy feeling in the legs

Patients taking diuretics that cause potassium loss should know the signs of too little potassium and should check with a physician as soon as possible if they have any of these symptoms:

- fast or irregular heartbeat
- weak pulse
- nausea or vomiting
- dry mouth
- excessive thirst
- muscle cramps or **pain**
- unusual tiredness or weakness
- mental or mood changes

### Interactions

Diuretics 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 a diuretic should let the physician know all other medicines he or she is taking and should ask whether the possible interactions can interfere with drug therapy. Among the drugs that may interact with diuretics are:

- Angiotensin-converting enzyme (ACE) inhibitors, such as benazepril (Lotensin), captopril (Capoten), and enalapril (Vasotec), used to treat high blood pressure. Taking these drugs with potassium-sparing diuretics may cause levels of potassium in the blood to be too high, increasing the chance of side effects.

- Cholesterol-lowering drugs such as cholestyramine (Questran) and colestipol (Colestid). Taking these drugs with combination diuretics such as Dyazide and Maxzide may keep the diuretic from working. Take the diuretic at least one hour before or four hours after the cholesterol-lowering drug.

- Cyclosporine (Sandimmune), a medicine that suppresses the immune system. Taking this medicine with potassium-sparing diuretics may increase the chance of side effects by causing levels of potassium in the blood to be too high.

- Potassium supplements, other medicines containing potassium, or salt substitutes that contain potassium. Taking these with potassium-sparing diuretics may lead to too much potassium in the blood, increasing the chance of side effects.

- Lithium, used to treat **bipolar disorder** (manic-depressive illness). Using this medicine with potassium-sparing diuretics may allow lithium to build up to poisonous levels in the body.

- Digitalis heart drugs, such as digoxin (Lanoxin). Using this medicine with combination diuretics such as triamterene-hydrochlorothiazide (Dyazide, Maxzide) may cause blood levels of the heart medicine to be too high, making side effects such as changes in heartbeat more likely.

The list above does not include every drug that may interact with diuretics. Check with a physician or pharmacist before combining diuretics with any other prescription or nonprescription (over-the-counter) medicine.

Nancy Ross-Flanigan

Diverticulitis see **Diverticulosis and diverticulitis**

## Diverticulosis and diverticulitis

### Definition

Diverticulosis refers to a condition in which the inner, lining layer of the large intestine (colon) bulges

out (herniates) through the outer, muscular layer. These outpouchings are called diverticula. Diverticulitis refers to the development of inflammation and infection in one or more diverticula.

### Description

Diverticula tend to occur most frequently in the last segment of the large intestine, the sigmoid colon. They occur with decreasing frequency as one examines further back toward the beginning of the large intestine. The chance of developing diverticula increases with age, so that by the age of 50, about 20–50% of all people will have some diverticula. By the age of 90, virtually everyone will have developed some diverticula. Most diverticula measure about 3 mm to just over 3 cm in diameter. Larger diverticula, termed giant diverticula, are quite infrequent, but may measure as large as 15 cm in diameter.

### Causes and symptoms

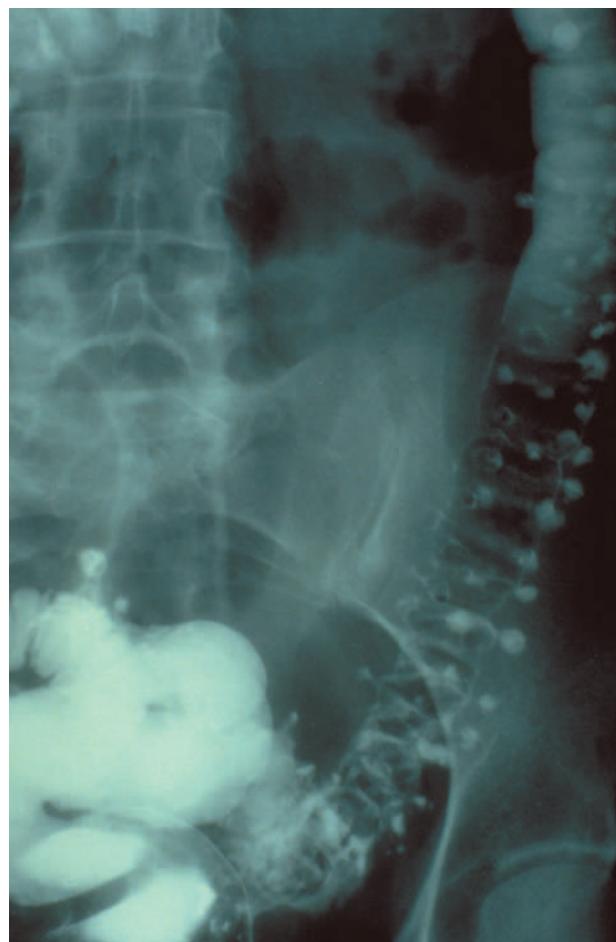
Diverticula are believed to be caused by overly forceful contractions of the muscular wall of the large intestine. As areas of this wall spasm, they become weaker and weaker, allowing the inner lining to bulge through. The anatomically weakest areas of the intestinal wall occur next to blood vessels which course through the wall, so diverticula commonly occur in this location.

Diverticula are most common in the developed countries of the West (North America, Great Britain, northern and western Europe). This is thought to be due to the diet of these countries, which tends to be quite low in fiber. A diet low in fiber results in the production of smaller volumes of stool. In order to move this smaller stool along the colon and out of the rectum, the colon must narrow itself significantly, and does so by contracting down forcefully. This causes an increase in pressure, which, over time, weakens the muscular wall of the intestine and allows diverticular pockets to develop.

The origin of giant diverticula development is not completely understood, although one theory involves gas repeatedly entering and becoming trapped in an already-existing diverticulum, causing stretching and expansion of that diverticulum.

The great majority of people with diverticulosis will remain symptom-free. Many diverticula are quite accidentally discovered during examinations for other conditions of the intestinal tract.

Some people with diverticulosis have symptoms such as **constipation**, cramping, and bloating. It is unclear whether these symptoms are actually caused by the diverticula themselves, or whether some other gastrointestinal



**A barium study x ray showing colonic diverticulosis.** (*Custom Medical Stock Photo. Reproduced by permission.*)

condition (such as **irritable bowel syndrome**) might be responsible. A complication of diverticulosis occurs because many diverticula develop in areas very near blood vessels. Therefore, one serious risk of diverticulosis involves bleeding. Although an infrequent complication, the bleeding can be quite severe. Seventy-five percent of such bleeding episodes occur due to diverticula located on the right side of the colon. About 50% of the time, such bleeding will stop on its own.

One of the most common and potentially serious complications of diverticulosis is inflammation and infection of a particular diverticulum, called diverticulitis.

Diverticulitis is three times more likely to occur in the left side of the large intestine. Since most diverticula are located in the sigmoid colon (the final segment of the large intestine which empties into the rectum), most diverticulitis also takes place in the sigmoid. The elderly have the most serious complications from diverticulitis, although very severe infections can also occur in patients

under the age of 50. Men are three times as likely as women to be stricken with diverticulitis.

Diverticulitis is believed to occur when a hardened piece of stool, undigested food, and bacteria (called a fecalith) becomes lodged in a diverticulum. This blockage interferes with the blood supply to the area, and infection sets in.

An individual with diverticulitis will experience **pain** (especially in the lower left side of the abdomen) and **fever**. In response to the infection and the irritation of nearby tissues within the abdomen, the abdominal muscles may begin to spasm. About 25% of all patients with diverticulitis will have some rectal bleeding, although this rarely becomes severe. Walled-off pockets of infection, called abscesses, may appear within the wall of the intestine, or even on the exterior surface of the intestine. When a diverticulum weakens sufficiently, and is filled to bulging with infected pus, a perforation in the intestinal wall may develop. When the infected contents of the intestine spill out into the abdomen, the severe infection called **peritonitis** may occur. Peritonitis is an infection and inflammation of the lining of the abdominal cavity, the peritoneum. Other complications of diverticulitis include the formation of abnormal connections between two organs that normally do not connect (fistulas; for example, the intestine and the bladder), and scarring outside of the intestine which squeezes off a portion of the intestine, obstructing it.

## Diagnosis

As mentioned, the majority of diverticula do not cause any symptoms, and are often found by coincidence during an examination being performed for some other medical condition.

When diverticula are suspected because a patient begins to have sudden rectal bleeding, the location of the bleeding can be studied by performing an **angiography**. Angiography involves inserting a tiny tube through an artery in the leg, and moving it up into one of the major arteries of the gastrointestinal system. A particular chemical (contrast medium) which will show up on x-ray films is injected, and the area of bleeding is located by looking for an area where the contrast is leaking into the interior (lumen) of the intestine.

A procedure called **endoscopy** provides another method for examining the colon and locating the site of bleeding. In endoscopy, a small, flexible scope (endoscope) is inserted through the rectum and into the intestine. The scope usually bears a fiber-optic camera, which allows the view through this endoscope to be projected onto a television screen. The operator can introduce the endoscope further and further through the intestine to find the location of the bleeding.

Diagnosis of diverticulitis is not difficult in patients with previously diagnosed diverticulosis. The presence of abdominal pain and fever in such an individual would make the suspicion of diverticulitis quite high. Examination of the abdomen will usually reveal tenderness to touch, with the patient's abdominal muscles contracting strongly to protect the tender area. During a rectal exam (performed by inserting a finger into the rectum), a doctor may be able to feel an abnormal mass. Touching this mass may prove painful to the patient.

When a practitioner is suspicious of diverticulitis as the cause for the patient's symptoms, he or she will most likely avoid the types of tests usually used to diagnose gastrointestinal disorders. These include **barium enema** and endoscopy. The concern is that the increased pressure exerted on the intestine during these exams may increase the likelihood of intestinal perforation. After medical treatment for the diverticulitis, these examinations may be performed in order to learn the extent of the patient's disease.

## Treatment

Only about 20% of patients with diverticulosis ever have symptoms which lead them to seek medical help. Most people never know that they have diverticula. For those individuals who have cramping pain and constipation believed to be due to diverticulosis, the usual prescription involves increasing the fiber in the diet. This can be done by adding special diet supplements of bran or psyllium seed, which increase stool volume. Bleeding diverticula can usually be treated by bed rest, with blood **transfusion** needed for more severe bleeding (hemorrhaging). In cases of very heavy hemorrhaging, medications which encourage clotting can be injected during the course of a diagnostic angiography.

While there are almost no situations when uncomplicated diverticulosis requires surgery, giant diverticula always require removal. This is due to the very high chance of infection and perforation of these diverticula. When giant diverticula are diagnosed, the usual treatment involves removing that portion of the intestine.

Treatment for uncomplicated diverticulitis usually requires hospitalization. "Resting the bowel" is a mainstay of treatment, and involves keeping the patient from eating or sometimes even drinking anything by mouth. Therefore, the patient will need to receive fluids through a needle in the vein (intravenous or IV fluids). **Antibiotics** will also be administered through the IV. Some physicians will agree to try treatment at home for very mildly ill patients. These patients will be put on a liquid diet and receive oral antibiotics.

The various complications of diverticulitis need to be treated aggressively, because the **death** rate from such things as perforation and peritonitis is quite high. Abscesses can be drained of their infected contents by inserting a needle through the skin of the abdomen and into the **abscess**. When this is unsuccessful, open abdominal surgery will be required to remove the piece of the intestine containing the abscess. Fistulas require surgical repair, including the removal of the length of intestine containing the origin of the fistula, followed by immediate reconnection of the two free ends of intestine. Peritonitis requires open surgery. The entire abdominal cavity is cleaned by being irrigated (washed) with a warmed sterile saltwater solution, and the damaged piece of intestine is removed. Obstructions require immediate surgery to prevent perforation. Massive, uncontrollable bleeding, while rare, may require removal of part or all of the large intestine.

During any of these types of operations, the surgeon must make an important decision regarding the quantity of intestine which must be removed. When the amount of intestine removed is great, it may be necessary to perform a **colostomy**. A colostomy involves pulling the end of the remaining intestine through the abdominal wall, to the outside. This bit of intestine is then fashioned so that a bag can be fit over it. The patient's waste (feces) collect in the bag, because the intestine no longer connects with the rectum. This colostomy may be temporary, in which case another operation will be required to reconnect the intestine, after some months of substantial healing has occurred. Other times, the colostomy will need to be permanent, and the patient will have to adjust to living permanently with the colostomy bag. Most people with colostomies are able to go on with a very active life.

Occasionally, a patient will have such severe diverticular disease that a surgeon recommends planning ahead, and schedules removal of a portion of the colon. This is done to avoid the high risk of surgery performed after a complication has set in. Certain developments in a patient will identify those patients who are at very high risk of experiencing dangerous complications. Such elective surgery may be recommended:

- when an older individual has had several attacks of diverticulitis
- when someone under the age of 50 has had even one attack
- when treatment does not get rid of a painful mass
- when the intestine appears to be narrowing on x-ray examination (this could suggest the presence of **cancer**)
- when certain patients begin to regularly experience painful urination or urinary infections (this suggests

## KEY TERMS

**Angiography**—An x-ray study of the arteries in a particular part of the body. Angiography is often performed in order to localize internal bleeding.

**Bowel obstruction**—A blockage in the intestine which prevents the normal flow of waste down the length of the intestine.

**Colostomy**—A procedure performed when a large quantity of intestine is removed. The end piece of the intestine leading to the rectum is closed.

**Diverticula**—Outpouchings in the large intestine caused when the inner, lining layer of the large intestine (colon) bulges out (herniates) through the outer, muscular layer.

**Endoscopy**—Examination of an area of the gastrointestinal tract by putting a lighted scope, usually bearing a fiber-optic camera, into the rectum, and passing it through the intestine.

**Fistula**—An abnormal connection formed between two organs that usually have no connection whatsoever.

**Sigmoid colon**—The final portion of the large intestine that empties into the rectum.

that there may be a connection between the intestine and the bladder)

- when there is any question of cancer
- when the diverticular disease appears to be progressing rapidly

## Prognosis

The prognosis for people with diverticula is excellent, with only 20% of such patients ever seeking any medical help for their condition.

While diverticulitis can be a difficult and painful disease, it is usually quite treatable. Prognosis is worse for individuals who have other medical problems, particularly those requiring the use of steroid medications, which increase the chances of developing a serious infection. Prognosis is also worse in the elderly.

## Prevention

While there is no absolutely certain way to prevent the development of diverticula, it is believed that high-fiber **diets** are of help. Foods that are recommended for

their high fiber content include whole grain breads and cereals, and all types of fruits and vegetables. Most experts suggest that individuals take in about 0.71–1.23 oz (20–35 g) of fiber daily. If this is not possible to achieve through a person's diet, there are fiber products which can be mixed into 8 oz (.237 l) of water or juice, and which provide about 0.13–19 oz (4–6 g) of fiber.

## Resources

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### ORGANIZATIONS

National Digestive Diseases Information Clearinghouse. 2 Information Way, Bethesda, MD 20892-3570. (800) 891-5389.  
<http://www.niddk.nih.gov/health/digest/nddic.htm>.

Rosalyn Carson-DeWitt, MD

## Dizziness

### Definition

As a disorder, dizziness is classified into three categories—vertigo, syncope, and nonsyncope nonvertigo. Each category has a characteristic set of symptoms, all related to the sense of balance. In general, syncope is defined by a brief loss of consciousness (**fainting**) or by dimmed vision and feeling uncoordinated, confused, and lightheaded. Many people experience a sensation like syncope when they stand up too fast. Vertigo is the feeling that either the individual or the surroundings are spinning. This sensation is like being on a spinning amusement park ride. Individuals with nonsyncope nonvertigo dizziness feel as though they cannot keep their balance. This feeling may become worse with movement.

### Description

The brain coordinates information from the eyes, the inner ear, and the body's senses to maintain balance. If any of these information sources is disrupted, the brain may not be able to compensate. For example, people sometimes experience **motion sickness** because the information from their body tells the brain that they are sitting still, but information from the eyes indicates that they are moving. The messages don't correspond and dizziness results.

Vision and the body's senses are the most important systems for maintaining balance, but problems in the inner ear are the most frequent cause of dizziness. The inner ear, also called the vestibular system, contains fluid that helps fine tune the information the brain receives from the eyes and the body. When fluid volume or pressure in one inner ear changes, information about balance is altered. The discrepancy gives conflicting messages to the brain about balance and induces dizziness.

Certain medical conditions can cause dizziness, because they affect the systems that maintain balance. For example, the inner ear is very sensitive to changes in blood flow. Because medical conditions such as high blood pressure or low blood sugar can affect blood flow, these conditions are frequently accompanied by dizziness. Circulation disorders are the most common causes of dizziness. Other causes are **head injury**, ear infection, **allergies**, and nervous system disorders.

Dizziness often disappears without treatment or with treatment of the underlying problem, but it can be long term or chronic. According to the National Institutes of Health, 42% of Americans will seek medical help for dizziness at some point in their lives. The costs may exceed a billion dollars and account for five million doctor visits annually. Episodes of dizziness increase with age. Among people aged 75 or older, dizziness is the most frequent reason for seeing a doctor.

### Causes and symptoms

Careful attention to symptoms can help determine the underlying cause of the dizziness. Underlying problems may be benign and easily treated or they may be dangerous and in need of intensive therapy. Not all cases of dizziness can be linked to a specific cause. More than one type of dizziness can be experienced at the same time and symptoms may be mixed. Episodes of dizziness may last for a few seconds or for days. The length of an episode is related to the underlying cause.

The symptoms of syncope include dimmed vision, loss of coordination, confusion, lightheadedness, and sweating. These symptoms can lead to a brief loss of con-

sciousness or fainting. They are related to a reduced flow of blood to the brain; they often occur when a person is standing up and can be relieved by sitting or lying down. Vertigo is characterized by a sensation of spinning or turning, accompanied by nausea, vomiting, ringing in the ears, **headache**, or **fatigue**. An individual may have trouble walking, remaining coordinated, or keeping balance. Non-syncope nonvertigo dizziness is characterized by a feeling of being off balance that becomes worse if the individual tries moving or performing detail-intense tasks.

A person may experience dizziness for many reasons. Syncope is associated with low blood pressure, heart problems, and disorders in the autonomic nervous system, the system of involuntary functions such as breathing. Syncope may also arise from emotional distress, **pain**, and other reactions to outside stressors. Non-syncope nonvertigo dizziness may be caused by rapid breathing, low blood sugar, or **migraine headache**, as well as by more serious medical conditions.

Vertigo is often associated with inner ear problems called vestibular disorders. A particularly intense vestibular disorder, Ménière's disease, interferes with the volume of fluid in the inner ear. This disease, which affects approximately one in every 1,000 people, causes intermittent vertigo over the course of weeks, months, or years. Ménière's disease is often accompanied by ringing or buzzing in the ear, **hearing loss**, and a feeling that the ear is blocked. Damage to the nerve that leads from the ear to the brain can also cause vertigo. Such damage can result from head injury or a tumor. An **acoustic neuroma**, for example, is a benign tumor that wraps around the nerve. Vertigo can also be caused by disorders of the central nervous system and the circulatory system, such as hardening of the arteries (arteriosclerosis), **stroke**, or **multiple sclerosis**.

Some medications cause changes in blood pressure or blood flow. These medications can cause dizziness in some people. Prescription medications carry warnings of such side effects, but common drugs, such as **caffeine** or nicotine can also cause dizziness. Certain **antibiotics** can damage the inner ear and cause hearing loss and dizziness.

Diet may cause dizziness. The role of diet may be direct, as through alcohol intake. It may be also be indirect, as through arteriosclerosis caused by a high-fat diet. Some people experience a slight dip in blood sugar and mild dizziness if they miss a meal, but this condition is rarely dangerous unless the person is diabetic. Food sensitivities or allergies can also be a cause of dizziness. Chronic conditions, such as heart disease, and serious acute problems, such as seizures and strokes, can cause dizziness. However, such conditions usually exhibit other characteristic symptoms.

## Diagnosis

During the initial medical examination, an individual with dizziness should provide a detailed description of the type of dizziness experienced, when it occurs, and how often each episode lasts. A diary of symptoms may help track this information. Report any symptoms that accompany the dizziness, such as a ringing in the ear or nausea, any recent injury or infection, and any medication taken.

Blood pressure, pulse, respiration, and body temperature are checked, and the ear, nose, and throat are scrutinized. The sense of balance is assessed by moving the individual's head to various positions or by tilt-table testing. In tilt-table testing, the person lies on a table that can be shifted into different positions and reports any dizziness that occurs.

Further tests may be indicated by the initial examination. Hearing tests help assess ear damage. x rays, computed tomography scan (CT scan), and **magnetic resonance imaging** (MRI) can pinpoint evidence of nerve damage, tumor, or other structural problems. If a vestibular disorder is suspected, a technique called electromyography (ENG) may be used. ENG measures the electrical impulses generated by eye movements. Blood tests can determine diabetes, **high cholesterol**, and other diseases. In some cases, a heart evaluation may be useful. Despite thorough testing, an underlying cause cannot always be determined.

## Treatment

Treatment is determined by the underlying cause. If an individual has a cold or **influenza**, a few days of bed rest is usually adequate to resolve dizziness. Other causes of dizziness, such as mild vestibular system damage, may resolve without medical treatment.

If dizziness continues, drug therapy may prove helpful. Because circulatory problems often cause dizziness, medication may be prescribed to control blood pressure or to treat arteriosclerosis. Sedatives may be useful to relieve the tension that can trigger or aggravate dizziness. Low blood sugar associated with diabetes sometimes causes dizziness and is treated by controlling blood sugar levels. An individual may be asked to avoid caffeine, nicotine, alcohol, and any substances that cause allergic reactions. A low-salt diet may also help some people.

When other measures have failed, surgery may be suggested to relieve pressure on the inner ear. If the dizziness is not treatable by drugs, surgery, or other means, physical therapy may be used and the patient may be taught coping mechanisms for the problem.

## KEY TERMS

**Acoustic neuroma**—A benign tumor that grows on the nerve leading from the inner ear to the brain. As the tumor grows, it exerts pressure on the inner ear and causes severe vertigo.

**Arteriosclerosis**—Hardening of the arteries caused by high blood cholesterol and high blood pressure.

**Autonomic nervous system**—The part of the nervous system that controls involuntary functions such as breathing and heart beat.

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

**Electronystagmography**—A method for measuring the electricity generated by eye movements. Electrodes are placed on the skin around the eye and the individual is subjected to a variety of stimuli so that the quality of eye movements can be assessed.

**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.

**Vestibular system**—The area of the inner ear that helps maintain balance.

## Alternative treatment

Because dizziness may arise from serious conditions, it is advisable to seek medical treatment. Alternative treatments can often be used alongside conventional medicine without conflict. Relaxation techniques, such as **yoga** and **massage therapy** that focus on relieving tension, are popularly recommended methods for reducing **stress**. Aromatherapists recommend a warm bath scented with essential oils of lavender, geranium, and sandalwood.

Homeopathic therapies can work very effectively for dizziness, and are especially applicable when no organic cause can be identified. An osteopath or chiropractor may suggest adjustments of the head, jaw, neck, and lower back to relieve pressure on the inner ear. Acupuncturists also offer some treatment options for acute and chronic cases of dizziness. Nutritionists may be able to offer advice and guidance in choosing dietary supplements, identifying foods to avoid, and balancing nutritional needs.

## Prognosis

Outcome depends on the cause of dizziness. Controlling or curing the underlying factors usually relieves dizziness. In some cases, dizziness disappears without treatment. In a few cases, dizziness can become a permanent disabling condition and a person's options are limited.

## Prevention

Most people learn through experience that certain activities will make them dizzy and they learn to avoid them. For example, if reading in a car produces motion sickness, an individual leaves reading materials for after the trip. Changes to the diet can also cut down on episodes of dizziness in susceptible people. Relaxation techniques can help ward off tension and **anxiety** that can cause dizziness.

These techniques can help minimize or even prevent dizziness for people with chronic diseases. For example, persons with Ménière's disease may avoid episodes of vertigo by leaving salt, alcohol, and caffeine out of their **diets**. Reducing blood cholesterol can help diminish arteriosclerosis and indirectly treat dizziness.

Some cases of dizziness cannot be prevented. Acoustic neuromas, for example, are not predictable or preventable. When the underlying cause of dizziness cannot be discovered, it may be difficult to recommend preventive measures. Alternative approaches designed to rebalance the body's energy flow, such as **acupuncture** and constitutional **homeopathy**, may be helpful in cases where the cause of dizziness cannot be pinpointed.

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Ruckenstein, Michael J. "A Practical Approach to Dizziness." *Postgraduate Medicine* 97, no. 3 (Mar. 1995): 70.

### ORGANIZATIONS

Ménière's Network. 1817 Patterson St., Nashville, TN 37203. (800) 545-4327. <<http://www.earfoundation.org>>.

The Vestibular Disorders Association. PO Box 4467, Portland, OR 97208-4467. (503) 229-7705. <<http://www.teleport.com/~veda>>.

Julia Barrett

DKA see **Diabetic ketoacidosis**

DLE see **Discoid lupus erythematosus**

Domestic violence see **Abuse**

Donovanosis see **Granuloma inguinale**

Doppler echocardiography see  
**Echocardiography**

## Doppler ultrasonography

### Definition

Doppler ultrasonography is a non-invasive diagnostic procedure that changes sound waves into an image that can be viewed on a monitor.

### Purpose

Doppler ultrasonography can detect the direction, velocity, and turbulence of blood flow. It is frequently used to detect problems with heart valves or to measure blood flow through the arteries. Specifically, it is useful in the work up of **stroke** patients, in assessing blood flow in the abdomen or legs, and in viewing the heart to monitor carotid artery diseases.

### Precautions

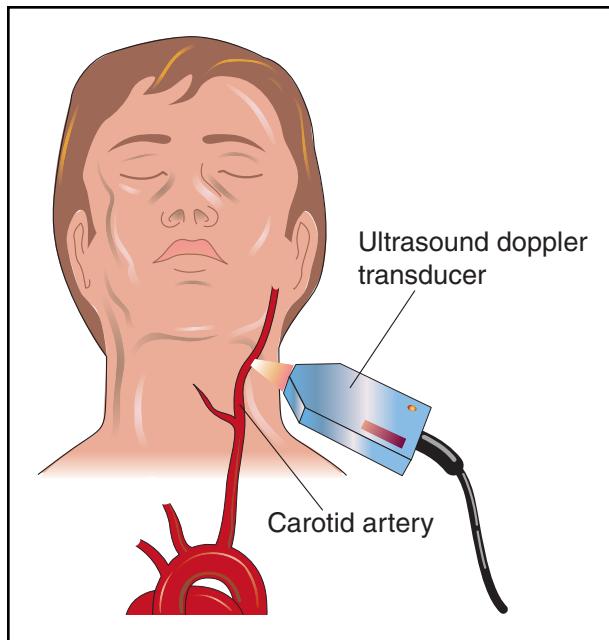
The test is widely used because it is noninvasive, uses no x rays, and gives excellent images. It is harmless, painless, and widely available.

### Description

Doppler ultrasonography makes use of two different principles. The ultrasound principle is this: when a high-frequency sound is produced and aimed at a target, it will be reflected by its target and the reflected sound can be detected back at its origin. In addition, it is known that certain crystals (called piezoelectric crystals) produce an electrical pulse when vibrated by a returning sound.

The Doppler principle is simply that sound pitch increases as the source moves toward the listener and decreases as it moves away.

Medical science utilizes these two principles in the following way. A transducer (sometimes called a probe) containing piezoelectric crystals sends a series of short sound pulses into the body and pauses between each pulse to listen for the returning sounds. The machine then determines the direction and depth of each returning sound and converts this into a point of light on a television monitor. Thousands of these pulses are computed and displayed every second to produce an image of the organ



**Doppler ultrasonography can detect the direction, velocity, and turbulence of blood flow. Because it is non-invasive and uses no x rays, doppler ultrasonography is widely used for numerous diagnostic procedures. (Illustration by Electronic Illustrators Group.)**

being studied. The image allows the doctor to see the organ functioning in real time.

The newest addition to this test is the addition of color. Adding color to the image shows the direction and rate of blood flow more clearly.

During a Doppler ultrasonography procedure the technician will apply a gel to the skin, then place the transducer against the skin at various angles. The transducer sends the information it receives to a television monitor that shows a moving image of the organ being studied. The technician can save these images either on video tape, paper, or x-ray film for further study.

### Preparation

There is no special preparation needed for this test. The ultrasound technician may apply a clear gel to the skin in order to help the transducer move freely over the body.

### Aftercare

No aftercare is necessary.

### Normal results

A Doppler ultrasonography test showing no restricted blood flow, is a normal finding.

## KEY TERMS

**Doppler effect**—The principle that the sound of an object moving toward you has a higher pitch than the sound when it's moving away from you.

**Transducer**—The part of a machine that changes signals in one form into another form.

**Ultrasound**—Sound that is too high for the human ear to hear.

### Abnormal results

Disrupted or obstructed blood flow through the neck arteries may indicate the person is at risk of having a stroke. (Narrowed arterial flow in the legs does not necessarily indicate a risk of stroke.)

### Resources

#### BOOKS

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

*Office Practice of Neurology*. Ed. Martin Samuels and Steven Feske. New York: Churchill Livingstone, 1996.

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Dorothy Elinor Stonely

## Down syndrome

### Definition

Down syndrome is the most common cause of **mental retardation** and malformation in a newborn. It occurs because of the presence of an extra chromosome.

### Description

Chromosomes are the units of genetic information that exist within every cell of the body. Twenty-three distinctive pairs, or 46 total chromosomes, are located within the nucleus (central structure) of each cell. When a baby is conceived by the combining of one sperm cell with one egg cell, the baby receives 23 chromosomes from each parent, for a total of 46 chromosomes. Sometimes, an accident in the production of a sperm or egg cell causes that cell to contain 24 chromosomes. This

event is referred to as nondisjunction. When this defective cell is involved in the conception of a baby, that baby will have a total of 47 chromosomes. The extra chromosome in Down syndrome is labeled number 21. For this reason, the existence of three such chromosomes is sometimes referred to as Trisomy 21.

In a very rare number of Down syndrome cases (about 1–2%), the original egg and sperm cells are completely normal. The problem occurs sometime shortly after fertilization; during the phase where cells are dividing rapidly. One cell divides abnormally, creating a line of cells with an extra chromosome 21. This form of genetic disorder is called a mosaic. The individual with this type of Down syndrome has two types of cells: those with 46 chromosomes (the normal number), and those with 47 chromosomes (as occurs in Down syndrome). Some researchers have suggested that individuals with this type of mosaic form of Down syndrome have less severe signs and symptoms of the disorder.

Another relatively rare genetic accident which can cause Down syndrome is called translocation. During cell division, the number 21 chromosome somehow breaks. A piece of the 21 chromosome then becomes attached to another chromosome. Each cell still has 46 chromosomes, but the extra piece of chromosome 21 results in the signs and symptoms of Down syndrome. Translocations occur in about 3–4% of cases of Down syndrome.

Down syndrome occurs in about one in every 800–1,000 births. It affects an equal number of boys and girls. Less than 25% of Down syndrome cases occur due to an extra chromosome in the sperm cell. The majority of cases of Down syndrome occur due to an extra chromosome 21 within the egg cell supplied by the mother (nondisjunction). As a woman's age (maternal age) increases, the risk of having a Down syndrome baby increases significantly. For example, at younger ages, the risk is about one in 4,000. By the time the woman is age 35, the risk increases to one in 400; by age 40 the risk increases to one in 110; and by age 45 the risk becomes one in 35. There is no increased risk of either mosaicism or translocation with increased maternal age.

### Causes and symptoms

While Down syndrome is a chromosomal disorder, a baby is usually identified at birth through observation of a set of common physical characteristics. Babies with Down syndrome tend to be overly quiet, less responsive, with weak, floppy muscles. Furthermore, a number of physical signs may be present. These include:

- flat appearing face
- small head

- flat bridge of the nose
- smaller than normal, low-set nose
- small mouth, which causes the tongue to stick out and to appear overly large
- upward slanting eyes
- extra folds of skin located at the inside corner of each eye, near the nose (called epicanthal folds)
- rounded cheeks
- small, misshapen ears
- small, wide hands
- an unusual, deep crease across the center of the palm (called a simian crease)
- a malformed fifth finger
- a wide space between the big and the second toes
- unusual creases on the soles of the feet
- overly-flexible joints (sometimes referred to as being double-jointed)
- shorter than normal height

Other types of defects often accompany Down syndrome. About 30–50% of all children with Down syndrome are found to have heart defects. A number of different heart defects are common in Down syndrome, including abnormal openings (holes) in the walls that separate the heart's chambers (**atrial septal defect**, **ventricular septal defect**). These result in abnormal patterns of blood flow within the heart. The abnormal blood flow often means that less oxygen is sent into circulation throughout the body. Another heart defect that occurs in Down syndrome is called **Tetralogy of Fallot**. Tetralogy of Fallot consists of a hole in the heart, along with three other major heart defects.

Malformations of the gastrointestinal tract are present in about 5–7% of children with Down syndrome. The most common malformation is a narrowed, obstructed duodenum (the part of the intestine into which the stomach empties). This disorder, called duodenal atresia, interferes with the baby's milk or formula leaving the stomach and entering the intestine for digestion. The baby often vomits forcibly after feeding, and cannot gain weight appropriately until the defect is repaired.

Other medical conditions that occur in patients with Down syndrome include an increased chance of developing infections, especially ear infections and **pneumonia**; certain kidney disorders; thyroid disease (especially low or hypothyroid); **hearing loss**; vision impairment requiring glasses (corrective lenses); and a 20-times greater chance of developing leukemia (a blood disorder).

Development in a baby and child with Down syndrome occurs at a much slower than normal rate.

Because of weak, floppy muscles (hypotonia), babies learn to sit up, crawl, and walk much later than their normal peers. Talking is also quite delayed. The level of mental retardation is considered to be mild-to-moderate in Down syndrome. The actual IQ range of Down syndrome children is quite varied, but the majority of such children are in what is sometimes known as the trainable range. This means that most people with Down syndrome can be trained to do regular self-care tasks, function in a socially appropriate manner in a normal home environment, and even hold simple jobs.

As people with Down syndrome age, they face an increased chance of developing the brain disease called Alzheimer's (sometimes referred to as **dementia** or senility). Most people have a six in 100 risk of developing Alzheimer's, but people with Down syndrome have a 25 in 100 chance of the disease. **Alzheimer's disease** causes the brain to shrink and to break down. The number of brain cells decreases, and abnormal deposits and structural arrangements occur. This process results in a loss of brain functioning. People with Alzheimer's have strikingly faulty memories. Over time, people with Alzheimer's disease will lapse into an increasingly unresponsive state. Some researchers have shown that even Down syndrome patients who do not appear to have Alzheimer's disease have the same changes occurring to the structures and cells of their brains.

As people with Down syndrome age, they also have an increased chance of developing a number of other illnesses, including **cataracts**, thyroid problems, diabetes, and seizure disorders.

## Diagnosis

Diagnosis is usually suspected at birth, when the characteristic physical signs of Down syndrome are noted. Once this suspicion has been raised, **genetic testing** (chromosome analysis) can be undertaken in order to verify the presence of the disorder. This testing is usually done on a blood sample, although chromosome analysis can also be done on other types of tissue, including skin. The cells to be studied are prepared in a laboratory. Chemical stain is added to make the characteristics of the cells and the chromosomes stand out. Chemicals are added to prompt the cells to go through normal development, up to the point where the chromosomes are most visible, prior to cell division. At this point, they are examined under a microscope and photographed. The photograph is used to sort the different sizes and shapes of chromosomes into pairs. In most cases of Down syndrome, one extra chromosome 21 will be revealed. The final result of such testing, with the photographed chromosomes paired and organized by shape and size, is called the individual's **karyotype**.

## Treatment

No treatment is available to cure Down syndrome. Treatment is directed at addressing the individual concerns of a particular patient. For example, heart defects will many times require surgical repair, as will duodenal atresia. Many Down syndrome patients will need to wear glasses to correct vision. Patients with hearing impairment benefit from **hearing aids**.

A new drug, referred to as a “smart drug,” has been receiving some attention in the treatment of Down syndrome patients. This drug, piracetam, has not been proven to increase intellectual ability, despite testimonials that have been receiving attention on television and the Internet. Piracetam has not been approved for use in the United States, although it is being sold via the Internet. The National Down Syndrome Society and the National Down Syndrome Congress do not recommend the use of this drug as of 2001.

While some decades ago, all Down syndrome children were quickly placed into institutions for lifelong care. Research shows very clearly that the best outlook for children with Down syndrome is a normal family life in their own home. This requires careful support and education of the parents and the siblings. It is a life-changing event to learn that a new baby has a permanent condition that will effect essentially all aspects of his or her development. Some community groups exist to help families deal with the emotional effects of this new information, and to help plan for the baby’s future. Schools are required to provide services for children with Down syndrome, sometimes in separate special education classrooms, and sometimes in regular classrooms (this is called mainstreaming or inclusion).

## Prognosis

The prognosis in Down syndrome is quite variable, depending on the types of complications (heart defects, susceptibility to infections, development of leukemia) of each individual baby. The severity of the retardation can also vary significantly. Without the presence of heart defects, about 90% of children with Down syndrome live into their teens. People with Down syndrome appear to go through the normal physical changes of **aging** more rapidly, however. The average age of **death** for an individual with Down syndrome is about 50–55 years.

Still, the prognosis for a baby born with Down syndrome is better than ever before. Because of modern medical treatments, including **antibiotics** to treat infections and surgery to treat heart defects and duodenal atresia, life expectancy has greatly increased. Community and family support allows people with Down syndrome

to have rich, meaningful relationships. Because of educational programs, some people with Down syndrome are able to hold jobs.

Men with Down syndrome appear to be uniformly sterile (meaning that they are unable to have offspring). Women with Down syndrome, however, are fully capable of having babies. About 50% of these babies, however, will also be born with Down syndrome.

## Prevention

Efforts at prevention of Down syndrome are aimed at **genetic counseling** of couples who are preparing to have babies. A counselor needs to inform a woman that her risk of having a baby with Down syndrome increases with her increasing age. Two types of testing is available during a **pregnancy** to determine if the baby being carried has Down syndrome.

Screening tests are used to estimate the chance that an individual woman will have a baby with Down syndrome. At 14–17 weeks of pregnancy, measurements of a substance called AFP (alpha-fetoprotein) can be performed. AFP is normally found circulating in the blood of a pregnant woman, but may be unusually high or low with certain disorders. Carrying a baby with Down syndrome often causes AFP to be lower than normal. This information alone, or along with measurements of two other hormones, is considered along with the mother’s age to calculate the risk of the baby being born with Down syndrome. These results are only predictions, and are only correct about 60% of the time.

The only way to definitively establish (with about 98–99% accuracy) the presence or absence of Down syndrome in a developing baby, is to test tissue from the pregnancy itself. This is usually done either by **amniocentesis** or **chorionic villus sampling** (CVS). In amniocentesis, a small amount of the fluid in which the baby is floating is withdrawn with a long, thin needle. In chorionic villus sampling, a tiny tube is inserted into the opening of the uterus to retrieve a small sample of the placenta (the organ that attaches the growing baby to the mother via the umbilical cord, and provides oxygen and **nutrition**). Both amniocentesis and CVS allow the baby’s own karyotype to be determined. A couple must then decide whether to use this information in order to begin to prepare for the arrival of a baby with Down syndrome, or to terminate the pregnancy.

Once a couple has had one baby with Down syndrome, they are often concerned about the likelihood of future offspring also being born with the disorder. Most research indicates that this chance remains the same as for any woman at a similar age. However, when the baby

## KEY TERMS

**Chromosome**—The structures that carry genetic information. Chromosomes are located within every cell, and are responsible for directing the development and functioning of all the cells in the body. The normal number is 46 (23 pairs).

**Karyotype**—The specific chromosomal makeup of a particular cell.

**Mental retardation**—A condition where an individual has a lower-than-normal IQ, and thus is developmentally delayed.

**Mosaic**—A term referring to a genetic situation, in which an individual's cells do not have the exact same composition of chromosomes. In Down syndrome, this may mean that some of the individual's cells have a normal 46 chromosomes, while other cells have an abnormal 47 chromosomes.

**Nondisjunction**—A genetic term referring to an event which takes place during cell division, in which a genetic accident causes an egg or sperm cell to have 24 chromosomes, rather than the normal 23.

**Translocation**—A genetic term referring to a situation during cell division in which a piece of one chromosome breaks off and sticks to another chromosome.

**Trisomy**—The condition of having three identical chromosomes, instead of the normal two.

with Down syndrome has the type that results from a translocation, it is possible that one of the two parents is a carrier of that defect. A carrier "carries" the genetic defect, but does not actually have the disorder. When one parent is a carrier of a translocation, the chance of future offspring having Down syndrome is greatly increased. The specific risk will have to be calculated by a genetic counselor.

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- National Down Syndrome Society. 666 Broadway, 8th Floor, New York, NY 10012-2317. (800) 221-4602. <<http://www.ndss.org>>.

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**Down's syndrome** see **Down syndrome**

**Doxazosin** see **Alpha<sub>1</sub>-adrenergic blockers**

**Doxepin** see **Antidepressants, tricyclic**

**Doxycycline** see **Tetracyclines**

**Dracontiasis** see **Guinea worm infection**

**Dracunculiasis** see **Guinea worm infection**

**Drooping eyelid** see **Ptosis**

**Drowning** see **Near-drowning**

**Drug abuse** see **Substance abuse and dependence**

**Drug addiction** see **Substance abuse and dependence**

**Drug dependence** see **Substance abuse and dependence**

## Drug metabolism/interactions

### Definition

Drug metabolism is the process by which the body breaks down and converts medication into active chemical substances.

## Precautions

Drugs can interact with other drugs, foods, and beverages. Interactions can lessen or magnify the desired therapeutic effect of a drug, or may cause unwanted or unexpected side effects. There are thousands of possible drug-to-drug and drug-to-food interactions, and many medications and supplements are contraindicated (not recommended) under certain conditions or in patients with specific diseases and disorders. This is why it is imperative that patients always keep their physician fully informed about all drugs and dietary supplements (including herbal remedies) they are taking.

## Description

The primary site of drug metabolism is the liver, the organ that plays a major role in metabolism, digestion, **detoxification**, and elimination of substances from the body. Enzymes in the liver are responsible for chemically changing drug components into substances known as metabolites. Metabolites are then bound to other substances for excretion through the lungs, or bodily fluids such as saliva, sweat, breast milk, and urine, or through reabsorption by the intestines. The primary mode of excretion is through the kidneys.

The family of liver isoenzymes known as cytochrome P-450 are crucial to drug metabolism. These enzymes (labeled CYP1A2, CYP2C9, CYP2C19, CYP2D6, and CYP3A4) have a catabolic action on substances, breaking them down into metabolites. Consequently, they also act to lower the concentration of medication in the bloodstream.

Drug interactions can occur when one drug inhibits or induces a P-450 that acts on another drug. An example is nicotine, a drug contained in tobacco, and known to induce P-450s. Individuals with liver disease (e.g., **cirrhosis**) may also have insufficient levels of P-450 enzymes. As a result, the concentration of drugs metabolized by these enzymes (e.g., amprenavir and other **protease inhibitors**) remains high and can build up to toxic levels in the bloodstream. In addition, certain medications and foods, such as grapefruit juice, can inactivate or lessen the metabolic activity of P-450s. Changing the drug dosage can alleviate the problem in some cases.

The metabolic rate can vary significantly from person to person, and drug dosages that work quickly and effectively in one individual may not work well for another. Factors such as genetics, environment, **nutrition** and age also influence drug metabolism; infants and elderly patients may have a reduced capacity to metabolize certain drugs, and may require adjustments in dosage.

## Causes and symptoms

Drugs that commonly interact with other medications include:

- **Diuretics.** Diuretics such as hydrochlorothiazide can reduce serum potassium and sodium electrolyte levels when taken with digoxin and lithium, respectively.
- Monoamine oxidase inhibitors (MAOIs). MAOI antidepressants can cause convulsions and other serious side effects when used with tricyclic antidepressants (e.g., Imipramine, Nortriptyline), **selective serotonin reuptake inhibitors** (SSRIs), or sympathomimetic drugs (e.g., amphetamines).
- **Antibiotics.** Antibiotics may reduce the efficiency of oral contraceptives.
- **Metals.** Medications containing metals, such as **antacids** with aluminum additives and iron supplements, can reduce the absorption of **tetracyclines** and fluoroquinolones.
- Drugs that inhibit liver enzyme function. Drugs that slow drug metabolism include ciprofloxacin, erythromycin, fluoxetine, nefazodone, paroxetine, and ritonavir. The therapeutic effect of other medications taken with these drugs may be amplified. Warfarin, a blood thinner, should be used with great caution in individuals taking these drugs.

Foods and beverages that may interact with drugs include:

- Grapefruit juice. Grapefruit juice inhibits the metabolism of many medications, including cyclosporine, felodipine, nifedipine, nitrendipine, nisoldipine, carbamazepine, triazolam, and midazolam.
- Foods and beverages with tyramines. Red wine, malted beers, smoked foods (e.g., fish and meats), dried fruits, and aged cheeses may contain tyramines, and can cause a severe and dangerous elevation in blood pressure when taken with MAOI inhibitors (a class of antidepressants).
- Dairy products. Milk, cream, and other dairy products containing calcium can prevent the absorption of antibiotics such as tetracycline, doxycycline, and ciprofloxacin when they are taken with the drug. In addition, whole milk with vitamin D can cause milk-alkali syndrome in patients taking aluminum hydroxide antacids.
- Caffeinated beverages. The **caffeine** contained in coffee and colas can influence drug metabolism.
- Alcohol. Alcohol is a central nervous system depressant, and should not be taken with other CNS depressants

## KEY TERMS

**Catabolism**—A process of metabolism that breaks down complex substances into simple ones.

**Cirrhosis**—Liver disease characterized by the widespread disruption of the normal liver structure and function.

**CNS depressant**—Anything that depresses, or slows, the sympathetic impulses of the central nervous system (i.e., respiratory rate, heart rate).

**Drug interaction**—A chemical or physiological reaction that can occur when two different drugs are taken together.

**Enzymes**—Organic substances (proteins) composed of amino acids that trigger and regulate chemical reactions in the body. There are over 700 identified human enzymes.

**Liver**—A solid organ located on the right in the upper abdomen. It plays a major role in metabolism, digestion, detoxification, and elimination of substances from the body.

**Metabolism**—The sum of all the physical and chemical processes occurring in the body to organize and maintain life.

**Metabolites**—Substances produced by metabolism or by a metabolic process.

**Milk-alkali syndrome**—Elevated blood calcium levels and alkalosis caused by excessive intake of milk and alkalis. Usually occurs in the treatment of peptic ulcer.

(e.g., antipsychotics, **antihistamines**). In addition, certain fermented beverages may contain tyramines

This list is not all-inclusive and individuals should always let their doctor and pharmacist know when they are taking other medications, herbal remedies, or dietary supplements. Anyone who experiences a serious reaction to a drug that is not consistent with its product labelling should report the event to their doctor and/or the MedWatch adverse event reporting system of the United States Food and Drug Administration (FDA).

### Alternative treatment

The growing use of herbal supplements has also increased the opportunity for adverse drug and herbal interactions. In 2000, the FDA issued a warning on the popular herb **St. John's wort** (*Hypericum perforatum*). The supplement was found to inhibit the effect of indinavir, a protease inhibitor used in the treatment of HIV. It may also affect the action of cyclosporine and other protease inhibitors (e.g., amprenavir, ritonavir). Further clinical studies are still necessary to determine the full metabolic effects of the herb.

Other herbs which may interact with allopathic medications include gingko biloba, ginseng, and garlic, which may all heighten the blood thinning effect of the anticoagulant warfarin. Because herbs are regulated by the FDA as dietary supplements, they do not require the same extensive clinical trials and premarket testing as drugs do before they are cleared for sale in the United States. As such, there is still much to learn about the potential interactions and adverse

effects associated with herbal supplements. Individuals who experience serious side effects from dietary supplements should report them to FDA's MedWatch program.

### Diagnosis

Drug interactions can be difficult to detect. In some cases, adverse reactions may closely resemble the symptoms of the disease or condition the medication was prescribed to treat. Patients who take a number of medications or self-treat with over-the-counter drugs and/or herbal remedies may not be able to determine which drug actually triggered the interaction. A 2001 study by University of Florida researchers found that less than half of the women participating disclosed their use of herbal therapies to their healthcare providers. In cases where a serious drug or herb interaction occurs, withholding this information can delay diagnosis and put the patient at increased risk.

### Treatment

Treatment of a drug interaction is dependant on a number of factors, including the medication(s) or supplements used and the medical history of the patient. A dosage adjustment may reverse the effects of some interactions. Serious or life-threatening interactions will require more aggressive therapies.

### Prevention

Patients with chronic health conditions, particularly those with liver disorders, should always inform their

healthcare professional before taking any over-the-counter (OTC) medications or dietary supplements. Because of the risk for a drug-to-drug interaction, individuals should also let their doctor know if they are taking drugs prescribed by other physicians. Individuals should closely follow instructions for use and package directions on both prescription and over-the-counter drugs. Consulting with a pharmacist and/or physician may be beneficial if package directions are unclear to the patient.

As a rule, grapefruit juice should not be taken with medication unless recommended by a doctor. Patients taking MAOI inhibitors should always check food and beverage labels to ensure tyramines aren't included, and should avoid all fermented drinks.

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## Drug overdose

### Definition

A drug overdose is the accidental or intentional use of a drug or medicine in an amount that is higher than is normally used.

### Description

All drugs have the potential to be misused, whether legally prescribed by a doctor, purchased over-the-counter at the local drug store, or bought illegally on the street. Taken in combination with other drugs or with alcohol, even drugs normally considered safe can cause death or serious long term consequences. Children are particularly at risk for accidental overdose, accounting

for over one million poisonings each year from drugs, alcohol, and other chemicals and toxic substances. People who suffer from depression and who have suicidal thoughts are also at high risk for drug overdose.

### Causes and symptoms

Accidental drug overdose may be the result of misuse of prescription medicines or commonly used medications like **pain** relievers and cold remedies. Symptoms differ depending on the drug taken. Some of the drugs commonly involved in overdoses are listed below along with symptoms and outcomes.

**Acetaminophen** is the generic name for the commonly used pain reliever Tylenol. Overdose of this drug causes liver damage with symptoms that include loss of appetite, tiredness, **nausea and vomiting**, paleness, and sweating. The next stage of symptoms indicates liver failure and includes abdominal pain and tenderness, swelling of the liver, and abnormal blood tests for liver enzymes. In the last stage of this **poisoning**, liver failure advances and the patient becomes jaundiced, with yellowing of the skin and whites of the eyes. They may also experience kidney failure, bleeding disorders, and encephalopathy (swelling of the brain).

Anticholinergic drugs (drugs that block the action of acetylcholine, a neurotransmitter) like atropine, scopolamine, belladonna, **antihistamines**, and antipsychotic agents cause the skin and moist tissues (like in the mouth and nose) to become dry and flushed. Dilated pupils, an inability to urinate, and mental disturbances are also symptoms. Severe toxicity can lead to seizures, abnormal heart rhythms, extremely high blood pressure, and **coma**.

**Antidepressant drugs** like amitriptyline, desipramine, and nortriptyline can cause irregular heart rate, vomiting, low blood pressure, confusion, and seizures. An overdose of antidepressants also causes symptoms similar to those seen with anticholinergic drug overdoses.

**Cholinergic drugs** (drugs that stimulate the parasympathetic nervous system) like carbamate and pilocarpine cause nausea, **diarrhea**, increased secretion of body fluids (sweat, tears, saliva, and urine), **fatigue**, and muscle weakness. Convulsions are possible. Death can occur due to **respiratory failure** and **heart failure**.

**Cocaine** and crack cocaine overdoses cause seizures, high blood pressure, increased heart rate, **paranoia**, and other changes in behavior. **Heart attack or stroke** are serious risks within three days after cocaine overdose.

Depressant drugs (tranquilizers, **antianxiety drugs**, sleeping pills) cause sleepiness, slowed or slurred speech, difficulty walking or standing, blurred vision, impaired ability to think, disorientation, and mood changes. Over-

dose symptoms can include slowed breathing, very low blood pressure, stupor, coma, **shock**, and death.

Digoxin, a drug used to regulate the heart, can cause irregular heart beats, nausea, confusion, loss of appetite, and blurred vision.

Narcotics or opiates are drugs like heroin, morphine, and codeine. Clonidine and diphenoxylate (Lomotil) are also in this category. Overdose with opiate drugs causes **sedation** (sleepiness), low blood pressure, slowed heart rate, and slowed breathing. Pinpoint pupils, where the black centers of the eyes become smaller than normal, are common in opiate overdose. However, if other drugs are taken at the same time as the opiates, they may counteract this effect on the pupils. A serious risk is that the patient will stop breathing.

Salicylates are found in **aspirin** and some creams or ointments used for muscle and joint pain (like Ben-Gay), and creams for **psoriasis**, a skin condition. Initial symptoms are gastrointestinal irritation, **fever**, and vomiting, possibly with blood in the vomit. This overdose will cause **metabolic acidosis** and **respiratory alkalosis**, conditions where the body's acid/base balance is malfunctioning. Symptoms include rapid heart beat and fast breathing. Nervous system symptoms include confusion, **hallucinations**, tiredness, and ringing in the ears. An increased tendency to bleed is also common. Serious complications include acute renal failure, coma, and heart failure. Acute salicylate poisoning can lead to death.

## Diagnosis

Diagnosis of a drug overdose may be based on the symptoms that develop, however, the drug may do extensive damage to the body before significant symptoms develop. If the patient is conscious, he or she may be able to tell what drugs were taken and in what amounts. The patient's recent medical and social history may also help in a diagnosis. For example, a list of medications that the patient takes, whether or not alcohol was consumed recently, even if the patient has eaten in the last few hours before the overdose, can be valuable in determining what was taken and how fast it will be absorbed into the system.

Different drugs have varying effects on the body's acid/base balance and on certain elements in the blood like potassium and calcium. Blood tests can be used to detect changes in body chemistry that may give clues to what drugs were taken. Blood can also be screened for various drugs in the system. Once the overdose drug is identified, blood tests can be used to monitor how fast the drug is being cleared out of the body. Urine tests can also be used to screen for some drugs and to detect changes in the body's chemistry. Blood and urine tests

may show if there is damage to the liver or kidneys as a result of the overdose.

## Treatment

### *Immediate care*

If a drug overdose is discovered or suspected, and the person is unconscious, having convulsions, or is not breathing, call for emergency help immediately. If the person who took the drug is not having symptoms, don't wait to see if symptoms develop; call a poison control center immediately. Providing as much information as possible to the poison control center can help determine what the next course of action should be.

The poison control center, paramedics, and emergency room staff will want to know:

- what drug(s) were taken—try to locate the drug's container.
- how much of the drug was taken
- when was the drug taken
- was the drug taken with alcohol or any other drugs or chemicals
- what is the age of the patient
- what symptoms are the patient experiencing
- is the patient conscious
- is the patient breathing

The poison control center may recommend trying to get the patient to vomit. A liquid called **ipecac** syrup, which is used to induce vomiting, is available from pharmacies without a prescription. Pediatricians may recommend that families keep ipecac syrup on hand in households with children. This medication should be used only on the advice of a medical professional. Vomiting should not be induced if the patient is unconscious.

### *Emergency care*

Emergency medical treatment may include:

- Assessment of the patient's airway and breathing to make sure that the trachea, the passage to the lungs, is not blocked. If needed, a tube may be inserted through the mouth and into the trachea to help the patient breath. This procedure is called intubation.
- Assessment of the patient's heart rate, blood pressure, body temperature, and other physical signs that might indicate the effects of the drug.
- Blood and urine samples may be collected to test for the presence of the suspected overdose drug, and any other drugs or alcohol that might be present.

## KEY TERMS

**Gastric lavage**—Also called a stomach pump. For this procedure, a flexible tube is inserted through the nose, down the throat, and into the stomach and the contents of the stomach are suctioned out. The inside of the stomach is rinsed with a saline (salt water) solution.

**Intubation**—A procedure where a tube is inserted through the mouth and into the trachea keep the airway open and to help the patient breathe.

- Elimination of the drug that has not yet been absorbed is attempted. Vomiting may be induced using ipecac syrup or other drugs that cause vomiting. Ipecac syrup should not be given to patients who overdosed with tricyclic antidepressants, theophylline, or any drug that causes a significant change in mental status. If a patient vomits while unconscious, there is a serious risk of **choking**.
- Gastric lavage, or washing out the stomach, may be attempted. For this procedure a tube flexible tube is inserted through the nose, down the throat, and into the stomach. The contents of the stomach are then suctioned out through the tube. A solution of saline (salt water) is injected into the tube to rinse out the stomach. This solution is then suctioned out. This is the process used when someone has his/her stomach pumped.
- Activated charcoal is sometimes given to absorb the drug.
- Medication to stimulate urination or defecation may be given to try to flush the excess drug out of the body faster.
- Intravenous (IV) fluids may be given. An intravenous line, a needle inserted into a vein, may be put into the arm or back of the hand. Fluids, either sterile saline (salt water solution) or dextrose (sugar water solution), can be administered through this line. Increasing fluids can help to flush the drug out of the system and to reestablish balance of fluids and **minerals** in the body. The pH (acid/base balance) of the body may need to be corrected by administering electrolytes like sodium, potassium, and bicarbonate through this IV line. If drugs need to be administered quickly, they can also be injected directly into the IV line.
- Hemodialysis is a procedure where blood is circulated out of the body, pumped through a dialysis machine, then reintroduced back into the body. This process can be used to filter some drugs out of the blood. It may

also be used temporarily or long term if the kidneys are damaged due to the overdose.

- Antidotes are available for some drug overdoses. An antidote is another drug that counteracts or blocks the overdose drug. For example, acetaminophen overdose can be treated with an oral medication, N-acetylcysteine (Mucomyst), if the level of acetaminophen found in the blood is extremely high. Naloxone is an anti-narcotic drug that is given to counteract narcotic poisoning. Nalmefen or **methadone** may also be used.
- Psychiatric evaluation may be recommended if the drug overdose was taken deliberately.

## Prognosis

While many victims of drug overdose recover without long term effects, there can be serious consequences. Some drug overdoses cause the failure of major organs like the kidneys or liver, or failure of whole systems like the respiratory or circulatory systems. Patients who survive drug overdose may need **kidney dialysis**, kidney or liver transplant, or ongoing care as a result of heart failure, stroke, or coma. Death can occur in almost any drug overdose situation, particularly if treatment is not started immediately.

## Prevention

To protect children from accidental drug overdose, all medications should be stored in containers with child resistant caps. All drugs should be out of sight and out of reach of children, preferably in a locked cabinet. Prescription medications should be used according to directions and only by the person whose name is on the label. Threats of suicide need to be taken seriously and appropriate help sought for people with depression or other mental illness that may lead to suicide.

## Resources

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Altha Roberts Edgren

TDM is important for patients who have other diseases that can affect drug levels, or who take other medicines that may affect drug levels by interacting with the drug being tested. As an example, without drug monitoring, the physician cannot be sure if a patient's lack of response to an antibiotic reflects bacterial resistance, or is the result of failure to reach the proper therapeutic range of antibiotic concentration in the blood. In cases of life-threatening infections, timing of effective antibiotic therapy is critical to success. It is equally crucial to avoid toxicity in a seriously ill patient. Therefore, if toxic symptoms appear with standard dosages, TDM can be used to determine changes in dosing.

Drawn blood, used for TDM, demonstrates a drug action in the body at any specific time, whereas drug levels examined from urine samples reflect the presence of a drug over many days (depending on the rate of excretion). Therefore, blood testing is the procedure of choice when definite data are required. However, for adequate absorption and therapeutic levels to be accurate, it is important to allow for sufficient time to pass between the administration of the medication and the collection of the blood sample.

Blood specimens for drug monitoring can be taken at two different times: during the drug's highest therapeutic concentration ("peak" level), or its lowest ("trough" level). Occasionally called residual levels, trough levels show sufficient therapeutic levels; whereas peak levels show **poisoning** (toxicity). Peak and trough levels should fall within the therapeutic range.

## Drug therapy monitoring

### Definition

Drug therapy monitoring, also known as Therapeutic Drug Monitoring (TDM), is a means of monitoring drug levels in the blood.

### Purpose

TDM is employed to measure blood drug levels so that the most effective dosage can be determined, with toxicity prevented. TDM is also utilized to identify non-compliant patients (those patients who, for whatever reason, either cannot or will not comply with drug dosages as prescribed by the physician).

### Precautions

Because so many different factors influence blood drug levels, the following points should be taken into consideration during TDM: the age and weight of the patient; the route of administration of the drug; the drug's absorption rate, excretion rate, delivery rate, and dosage; other medications the patient is taking; other diseases the patient has; the patient's compliance regarding the drug treatment regimen; and the laboratory methods used to test for the drug.

### Description

TDM is a practical tool that can help the physician provide effective and safe drug therapy in patients who need medication. Monitoring can be used to confirm a blood drug concentration level that is above or below the therapeutic range, or if the desired therapeutic effect of the drug is not as expected. If this is the case, and dosages beyond normal then have to be prescribed, TDM can minimize the time that elapses.

### Preparation

In preparing for this test, the following guidelines should be observed:

- Depending on the drug to be tested, the physician should decide if the patient is to be **fasting** (nothing to eat or drink for a specified period of hours) before the test.
- For patients suspected of symptoms of drug toxicity, the best time to draw the blood specimen is when the symptoms are occurring.
- If there is a question as to whether an adequate dose of the drug is being achieved, it is best to obtain trough (lowest therapeutic concentration) levels.
- Peak (highest concentration) levels are usually obtained one to two hours after oral intake, approximately one hour after intramuscular (IM) administration (a shot in the muscle), and approximately 30 minutes after intravenous (IV) administration. Residual, or trough, levels are usually obtained within 15 minutes of the next scheduled dose.

## Therapeutic Drug Monitoring: Therapeutic And Toxic Range

Drug Level*	Use	Therapeutic Level*	Toxic
Acetaminophen mg/ml	Analgesic, antipyretic	Depends on use	>250
Amikacin mg/ml	Antibiotic	12–25 mg/ml**	>25
Aminophylline ng/ml	Bronchodilator	10–20 mg/ml	>20
Amitriptyline ng/ml	Antidepressant	120–150 ng/ml	>500
Carbamazepine mg/ml	Anticonvulsant	5–12 mg/ml	>12
Chloramphenicol mg/ml	Antibiotic	10–20 mg/ml	>25
Digoxin ng/ml	Cardiotonic	0.8–2.0 ng/ml	>2.4
Gentamicin	Antibiotic	4–12 mg/L	>12 mg/L
Lidocaine	Antiarrhythmic	1.5–5.0 mg/L	>5 mg/ml
Lithium mEq/L	Antimanic	0.7–2.0 mEq/L	>2.0
Nortriptyline ng/ml	Antidepressant	50–150 ng/ml	>500
Phenobarbital mg/ml	Anticonvulsant	10–30 mg/ml	>40
Phenytoin mg/ml	Anticonvulsant	7–20 mg/ml	>30
Procainamide mg/ml	Antiarrhythmic	4–8 mg/ml	>16
Propranolol ng/ml	Antiarrhythmic	50–100 ng/ml	>150
Quinidine mg/ml	Antiarrhythmic	1–4 mg/ml	>10
Theophylline mg/ml	Bronchodilator	10–20 mg/ml	>20
Tobramycin mg/ml	Antibiotic	4–12 mg/ml**	>12
Valproic acid mg/ml	Anticonvulsant	50–100 mg/ml	>100

\* Values are laboratory-specific

\*\*Concentration obtained 30 minutes after the end of a 30-minute infusion.

## Risks

Risks for this test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or feeling lightheaded after blood is drawn, or accumulation of blood under the puncture site (hematoma).

## Resources

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Janis O. Flores

make sure she has discussed the benefits and risks with her physician.

Oxytocin also may be used to control bleeding after delivery or to help make the milk flow in women who are breastfeeding their babies.

## Description

Oxytocin is a hormone and is available only with a physician's prescription. When used to start or continue labor, it is slowly injected into a vein. A nasal spray form is used to increase milk flow in breastfeeding. Some commonly used brand names are Pitocin and Syntocinon.

## Recommended dosage

The dosages given here are average doses. However, doses may be different for different patients. Follow the orders of the physician who prescribed the drug.

### For increasing milk production:

One spray into one or both nostrils, two to three minutes before nursing or using a breast pump.

### For starting or continuing labor:

The physician in charge will determine the appropriate dose.

## Precautions

Oxytocin does not help increase or continue labor in all patients. When it does not help, the physician may deliver the baby by **cesarean section**.

## Drugs used in labor

### Definition

These drugs are used to induce (start) or continue labor.

### Purpose

The drug described here, oxytocin, makes the uterus (womb) contract. Physicians use it to deliberately start labor. Because there are some risks with using oxytocin, this should be done only when there are good medical reasons. Any woman who is being given oxytocin should

## KEY TERMS

**Cesarean section**—The delivery of a baby through a surgical procedure.

**Fetus**—A developing baby inside the womb.

**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.

In women who are especially sensitive to oxytocin, the drug may cause contractions to become too strong. This could tear the uterus or deprive the fetus of blood and oxygen during labor.

Oxytocin does not help improve milk flow in all women who are breastfeeding. Check with a physician if the drug does not seem to be working.

Women with heart disease, high blood pressure, or kidney disease should let their physicians know about these conditions before taking oxytocin. Also, anyone who has had an unusual reaction to oxytocin in the past should inform their physician.

### Side effects

Oxytocin has caused irregular heartbeat and increased bleeding in some women after delivery. It may also cause **jaundice** (yellowing of the eyes and skin) in newborns.

Other side effects are rare, but may include nausea, vomiting, confusion, **dizziness**, convulsions, breathing problems, **headache**, **hives**, skin rash, **itching**, pelvic or abdominal **pain**, and weakness. The nasal spray form may cause watery eyes or irritation of the nose.

### Interactions

Anyone who takes oxytocin should let the physician know all other medicines she is taking.

Nancy Ross-Flanigan

## Dry mouth

### Definition

Dry mouth, known medically as xerostomia, is the abnormal reduction of saliva due to medication, disease, or medical therapy.

### Description

Dry mouth due to the lack of saliva can be a serious medical problem. Decreased salivation can make swallowing difficult, can decrease taste sensation, and can promote **tooth decay**.

### Causes and symptoms

Dry mouth, resulting from thickened or reduced saliva flow, can be caused by a number of factors: medications, both prescription and over-the-counter; systemic diseases, such as anemia or diabetes, manifestations of **Sjögren's syndrome** (as **rheumatoid arthritis**, lupus, chronic hardening and thickening of the skin, or chronic and progressive inflammation of skeletal muscles); infections of the salivary glands; blockage of the salivary ducts caused by stones or tumors forming in the ducts through which the saliva passes; **dehydration**; medical therapies, such as local surgery or radiation; secretion reduction normally involved in the **aging** process; and emotional stress.

### Diagnosis

The diagnosis of dry mouth is not difficult. The patient will state that his or her saliva is very thick or non-existent. Finding the cause of the dry mouth may be more difficult and require some laboratory testing. Salivary gland biopsy for stones or tumors should be performed if indicated.

### Treatment

The treatment of dry mouth involves the management of the condition causing it. If dry mouth is caused by medication, the medication should be changed. If dry mouth is caused by blockage of the salivary ducts, the cause of the blockage should be investigated. When systemic diseases, such as diabetes and anemia, are brought under control dry mouth problems may decrease.

The use of caffeine-containing beverages, alcoholic beverages, and mouthwashes containing alcohol should be minimized. The drinking of water and fruit juices will decrease dry mouth problems. Chewing gum and lemon drops can be used to stimulate saliva flow. Bitters also can initiate salivary flow as long as the salivary glands and ducts are functional. Commercial saliva substitutes are available without prescription and can be used as frequently as needed. Use of a humidifier in the bedroom reduces nighttime oral dryness.

### Prognosis

The prognosis for patients with xerostomia due to medication problems is good, if the offending agent can

## KEY TERMS

**Salivary duct**—Tube through which saliva is carried from the salivary gland to the mouth.

**Salivary gland**—Gland in which saliva forms.

be changed. Dry mouth due to systemic problems may be eliminated or improved once the disease causing the dry mouth is under control. Persistent xerostomia can be managed well with saliva substitutes.

### Prevention

A patient needs to ask his or her health care provider if any medication to be prescribed will cause dry mouth. Patients with persistent xerostomia need to practice good **oral hygiene** and visit a dentist on a regular basis; the lack of adequate saliva can cause severe dental decay. The salivary glands are very sensitive to radiation, so any patient scheduled for **radiation therapy** of the head and neck needs to discuss with the radiation therapist ways to minimize exposure of the salivary glands to radiation.

### Resources

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#### ORGANIZATIONS

American Dental Association. 211 E. Chicago Ave., Chicago, IL 60611. (312) 440-2500. <<http://www.ada.org>>.

American Medical Association. 515 N. State St., Chicago, IL 60612. (312) 464-5000. <<http://www.ama-assn.org>>.

Joseph Knight, PA

Dry skin see **Ichthyosis**

Dual energy x-ray absorptiometry (DXA)  
scan see **Bone density test**

DUB see **Dysfunctional uterine bleeding**

Duchenne muscular dystrophy see  
**Muscular dystrophy**

Duodenal atresia see **Duodenal obstruction**

## Duodenal obstruction

### Definition

Duodenal obstruction is a failure of food to pass out of the stomach either from a complete or partial obstruction.

### Description

The duodenum is the first part of the intestine, into which the stomach, the gall bladder, and the pancreas empty their contents. The pylorus connects the duodenum with the stomach and contains the valve that regulates stomach emptying. Obstruction usually occurs right at this outlet, so that the gall bladder and pancreas are unable to drain their secretions without hindrance.

### Causes and symptoms

Obstruction of the duodenum occurs in adults and infants, each for a different set of reasons. In adults, the usual cause is a peptic ulcer of such antiquity that repeated cycles of injury and scarring have narrowed the pasageway. Medical treatment of ulcers has progressed to the point where such obstinate ulcer disease is rarely seen any more. In infants, the conditions are congenital—either the channel is underdeveloped or the pylorus is overdeveloped. The first type is called duodenal hypoplasia and the second is termed hypertrophic **pyloric stenosis**. In rare cases, the channel may be missing altogether, a condition called duodenal atresia. To say that these anomalies are congenital is not to say their cause is understood. As with most **birth defects**, the specific cause is not known.

Food that cannot exit the stomach in the forward direction will return whence it came. Vomiting is the constant symptom of duodenal obstruction. It may be preceded by **indigestion** and nausea as the stomach attempts to squeeze its contents through an ever narrowing outlet.

Hypertrophic pyloric stenosis appears soon after birth. The infant will vomit feedings, lose weight, and be restless and irritable.

### Diagnosis

X rays taken with contrast material in the stomach readily demonstrate the site of the blockage and often the ulcer that caused it. Gastroscopy is another way to evaluate the problem. In infants, x rays may not be necessary to detect pyloric stenosis. It is often possible to feel the enlarged pylorus, like an olive, deep under the ribs and see the stomach rippling as it labors to force food through.

## Treatment

Bowel obstruction requires a surgeon, sometimes immediately. Newer surgical techniques constantly improve the outcome, but obstruction is a mechanical problem that needs a mechanical solution. Most adults who come to surgery for obstruction have suffered for years from peptic ulcer disease. They will usually benefit from **ulcer surgery** at the same time their obstruction is relieved. The surgeon will therefore select a procedure that combines relief of obstruction with remedy for ulcer disease. There are many choices. In fact, even without obstruction, functional considerations require ulcer surgery to include enhancement of stomach emptying.

To treat an infant with hypertrophic pyloric stenosis, some surgeons have had success with forceful balloon dilation of the pylorus done through a gastroscope, but the standard procedure is to cut across the overdeveloped circular muscle that is constricting the stomach outlet. There are reports of infant hypertrophic pyloric stenosis remitting without surgery following a very careful feeding schedule, but mortality is unacceptably high.

## Prognosis

A functioning and unrestricted intestine is a prerequisite for living independent of the most advanced and continuous medical care available. Achieving this desirable goal is the rule with surgery for duodenal obstructions of all types. The bowel is so malleable that there is a rearrangement to suit every occasion. The variety of possible configurations is limited only by the surgeon's imagination.

## Prevention

Prompt and effective treatment of peptic ulcers will prevent chronic scarring and narrowing. Drugs developed over the past few decades have all but eliminated the need for ulcer surgery.

## Resources

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## KEY TERMS

**Atresia**—Failure to develop; complete absence.

**Contrast agent**—A substance that produces shadows on an x ray so that hollow structures can be more easily seen.

**Gastroscopy**—Looking into the stomach with a flexible viewing instrument called a gastroscope.

**Hypoplasia**—Incomplete development.

**Peptic ulcer**—A wound in the lower stomach and duodenum caused by stomach acid and a newly discovered germ called *Helicobacter pylori*.

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J. Ricker Polsdorfer, MD

Duodenal stenosis see **Duodenal obstruction**

Duodenal ulcers see **Ulcers (digestive)**

Duodenum x rays see **Hypotonic duodenography**

Duplicated ureter see **Congenital ureter anomalies**

Dwarfism see **Achondroplasia; Pituitary dwarfism**

## Dysfunctional uterine bleeding

### Definition

Dysfunctional uterine bleeding is irregular, abnormal uterine bleeding that is not caused by a tumor, infection, or **pregnancy**.

## Description

Dysfunctional uterine bleeding (DUB) is a disorder that occurs most frequently in women at the beginning and end of their reproductive lives. About half the cases occur in women over 45 years of age, and about one fifth occur in women under age 20.

Dysfunctional uterine bleeding is diagnosed when other causes of uterine bleeding have been eliminated. Failure of the ovary to release an egg during the menstrual cycle occurs in about 70% of women with DUB. This is probably related to a hormonal imbalance.

DUB is common in women who have **polycystic ovary syndrome** (cysts on the ovaries). Women who are on dialysis may also have heavy or prolonged periods. So do some women who use an intrauterine device (**IUD**) for birth control.

DUB is similar to several other types of uterine bleeding disorders and sometimes overlaps these conditions.

### *Menorrhagia*

Menorrhagia, sometimes called hypermenorrhea, is another term for abnormally long, heavy periods. This type of period can be a symptom of DUB, or many other diseases or disorders. In menorrhagia, menstrual periods occur regularly, but last more than seven days, and blood loss exceeds 3 oz (88.7 ml). Passing blood clots is common. Between 15–20% of healthy women experience debilitating menorrhagia that interferes with their normal activities. Menorrhagia may or may not signify a serious underlying problem.

### *Metrorrhagia*

Metrorrhagia is bleeding between menstrual periods. Bleeding is heavy and irregular as opposed to ovulatory spotting which is light bleeding, in mid-cycle, at the time of ovulation.

### *Polymenorrhea*

Polymenorrhea describes the condition of having too frequent periods. Periods occur more often than every 21 days, and ovulation usually does not occur during the cycle.

## Causes and symptoms

Dysfunctional uterine bleeding often occurs when the endometrium, or lining of the uterus, is stimulated to grow by the hormone estrogen. When exposure to estrogen is extended, or not balanced by the presence of progesterone, the endometrium continues to grow until it outgrows its blood supply. Then it sloughs off, causing

irregular bleeding. If the bleeding is heavy enough and frequent enough, anemia can result.

Menorrhagia is representative of DUB. It is caused by many conditions including some outside the reproductive system. Causes of menorrhagia include:

- adenomyosis (a benign condition characterized by growths in the area of the uterus)
- imbalance between the hormones estrogen and progesterone
- fibroid tumors
- pelvic infection
- **endometrial cancer** (cancer of the inner mucous membrane of the uterus)
- endometrial polyps
- **endometriosis** (a condition in which endometrial or endometrial-like tissue appears outside of its normal place in the uterus)
- use of an intrauterine device (IUD) for **contraception**
- hypothyroidism
- blood clotting problems (rare)
- lupus erythematosus
- pelvic inflammatory disease
- steroid therapy
- advanced liver disease
- renal (kidney) disease
- chemotherapy (cancer treatment with chemicals)

To diagnose dysfunctional uterine bleeding, many of the potential causes mentioned above must be eliminated. When all potential causes connected with pregnancy, infection, and tumors (benign or malignant) are eliminated, then menorrhagia is presumed to be caused by dysfunctional uterine bleeding.

## Diagnosis

Diagnosis of any menstrual irregularity begins with the patient herself. The doctor will ask for a detailed description of the problem, and take a history of how long it has existed, and any patterns the patient has observed. A woman can assist the doctor in diagnosing the cause of abnormal uterine bleeding by keeping a record of the time, frequency, length, and quantity of bleeding. She should also tell the doctor about any illnesses, including long-standing conditions, like **diabetes mellitus**. The doctor will also inquire about sexual activity, use of contraceptives, current medications, and past surgical procedures.

## Laboratory tests

After taking the woman's history, the gynecologist or family practitioner does a pelvic examination and Pap smear. To rule out specific causes of abnormal bleeding, the doctor may also do a pregnancy test and blood tests to check the level of thyroid hormone. Based on the initial test results, the doctor may want to do tests to determine the level of other hormones that play a role in reproduction. A test of blood clotting time and an adrenal function test are also commonly done.

## Imaging

Imaging tests are important diagnostic tools for evaluating abnormal uterine bleeding. Ultrasound examination of the pelvic and abdominal area is used to help locate **uterine fibroids**, also called uterine leiomyoma, a type of tumor. Visual examination through hysteroscopy—where a camera inside a thin tube is inserted directly into the uterus so that the doctor can see the uterine lining—is also used to assess the condition of the uterus.

Hysterosalpingography can help outline endometrial polyps and fibroids and help detect endometrial cancer. In this procedure an x ray is taken after contrast media has been injected into the cervix. **Magnetic resonance imaging (MRI)** of the pelvic region can also be used to locate fibroids and tumors.

## Invasive procedures

**Endometrial biopsy** (the removal and examination of endometrial tissue) is the most important testing procedure. It allows the doctor to sample small areas of the uterine lining, while cervical biopsy allows the cervix to be sampled. Tissues are then examined for any abnormalities.

Dilation and curettage (D & C), once common is rarely done today for diagnosis of DUB. It is done while the patient is under either general or regional anesthesia. Women over 30 are more likely to need a D & C, as part of the diagnostic procedure, than younger women.

Because DUB is diagnosed by eliminating other possible disorders, diagnosis can take a long time and involve many tests and procedures. Older women are likely to need more extensive tests than adolescents because the likelihood of reproductive cancers is greater in this age group, and therefore must be definitively eliminated before treating bleeding symptoms.

## Treatment

Treatment of DUB depends on the cause of the bleeding and the age of the patient. When the underlying cause of the disorder is known, that disorder is treated.

## KEY TERMS

**Dilation and curettage (D & C)**—A procedure performed under anesthesia during which the cervix is dilated, and tissue lining the uterus is scraped out with a metal spoon-shaped instrument or a suction tube. The procedure can be either diagnostic, or to remove polyps.

**Endometrial biopsy**—The removal of tissue either by suction or scraping of samples of tissue from the uterus. The cervix is not dilated. The procedure has a lower rate of diagnostic accuracy than a D & C, but can be done as an office procedure under local anesthesia.

**Endometrial cancer**—Cancer of the inner mucous membrane of the uterus.

**Fibroids, or fibroid tumors**—Fibroid tumors are non-cancerous (benign) growths in the uterus. They occur in 30–40% of women over age 40, and do not need to be removed unless they are causing symptoms that interfere with a woman's normal activities.

**Hypothyroidism**—A disorder in which the thyroid gland produces too little thyroid hormone causing a decrease in the rate of metabolism with associated effects on the reproductive system.

**Lupus erythematosus**—A chronic inflammatory disease in which inappropriate immune system reactions cause abnormalities in the blood vessels and connective tissue.

**Progesterone**—A hormone naturally secreted by the ovary, or manufactured synthetically, that prepares the uterus for implantation of a fertilized egg.

**Prostaglandins**—A group of chemicals that mediate, or determine the actions of other chemicals in the cell or body.

Otherwise the goal of treatment is to relieve the symptoms to a degree that uterine bleeding does not interfere with a woman's normal activities or cause anemia.

Generally the first approach to controlling DUB is to use **oral contraceptives** that provide a balance between the hormones estrogen and progesterone. Oral contraceptives are often very effective in adolescents and young women in their twenties. NSAIDs (**nonsteroidal anti-inflammatory drugs**), like Naprosyn and Motrin, are also used to treat DUB.

When bleeding cannot be controlled by hormone treatment, surgery may be necessary. Dilation and curettage sometimes relieves the symptoms of DUB. If that fails, endometrial ablation removes the uterine lining, but preserves a woman's uterus. This procedure is sometimes used instead of **hysterectomy**. However, as it affects the uterus, it can only be used when a woman has completed her childbearing years. The prescription of iron is also important to decrease the risk of anemia.

Until the 1980s, hysterectomy often was used to treat heavy uterine bleeding. Today hysterectomy is used less frequently to treat DUB, and then only after other methods of controlling the symptoms have failed. A hysterectomy leaves a woman unable to bear children, and, therefore, is limited largely to women who are unable to, or uninterested in, bearing children. Still, hysterectomy is a common treatment for long-standing DUB in women done with childbearing.

### Alternative treatment

Alternative practitioners concentrate on good **nutrition** as a way to prevent heavy periods that are not caused by uterine fibroids, endometrial polyps, endometriosis, or cancer. Iron supplementation (100 mg per day) not only helps prevent anemia, but also appears to reduce menorrhagia in many women. Other recommended dietary supplements include **vitamins** A and C. Vitamin C improves capillary fragility and enhances iron uptake.

Vitamin E and bioflavonoid supplements are also recommended. Vitamin E can help reduce blood flow, and bioflavonoids help strengthen the capillaries. Vitamin K is known to play a role in clotting and is helpful in situations where heavy bleeding may be due to clotting abnormalities.

Botanical medicines used to assist in treating abnormal bleeding include spotted cranesbill (*Geranium maculatum*), birthroot (*Trillium pendulum*), blue cohosh (*Caulophyllum thalictroides*), witch hazel (*Hamamelis virginiana*), shepherd's purse (*Capsella bursa-pastoris*), and yarrow (*Achillea millefolia*). These are all stiptic herbs that act to tighten blood vessels and tissue. Hormonal balance can also be addressed with herbal formulations containing phytoestrogens and phytoprogestrone.

### Prognosis

Response to treatment for DUB is highly individual and is not easy to predict. The outcome depends largely on the woman's medical condition and her age. Many women, especially adolescents, are successfully treated with hormones (usually oral contraceptives). As a last resort, hysterectomy removes the source of the problem

by removing the uterus, but this operation is not without risk, or the possibility of complications.

### Prevention

Dysfunctional uterine bleeding is not a preventable disorder.

### Resources

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Tish Davidson

## Dyslexia

### Definition

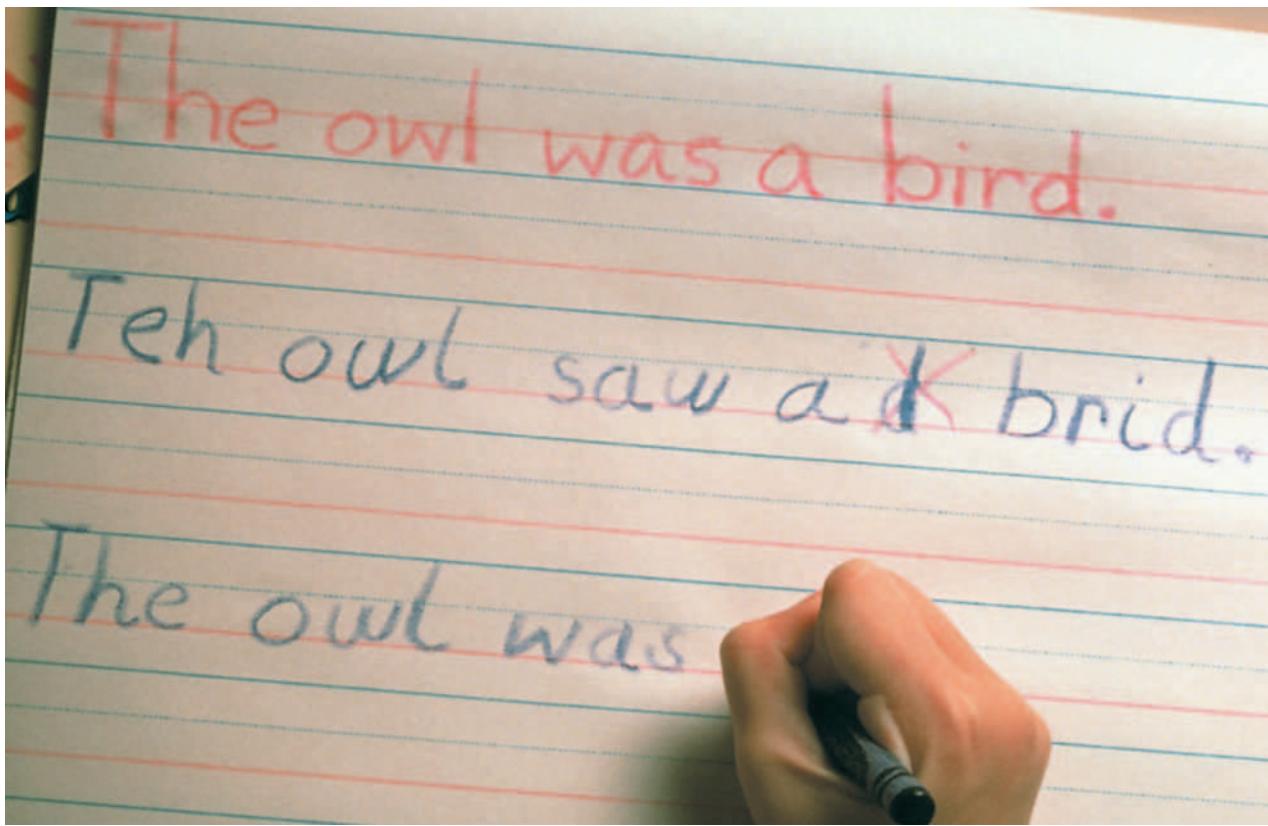
Dyslexia is a learning disability characterized by problems in reading, spelling, writing, speaking, or listening. In many cases, dyslexia appears to be inherited.

### Description

The word dyslexia is derived from the Greek word, *dys* (meaning poor or inadequate) and the word *lexis* (meaning words or language).

The National Institutes of Health estimates that about 15% of the United States population is affected by learning disabilities, mostly with problems in language and reading. The condition appears in all ages, races, and income levels. Dyslexia is not a disease, but describes rather a different kind of mind that learns in a different way from other people. Many people with the condition are gifted and very productive; dyslexia is not at all linked to low intelligence. In fact, intelligence has nothing to do with dyslexia.

Dyslexic children seem to have trouble learning early reading skills, problems hearing individual sounds in words, analyzing whole words in parts, and blending sounds into words. Letters such as "d" and "b" may be confused.



**A student with dyslexia has difficulty copying words.** (Photograph by Will & Deni McIntyre, Photo Researchers, Inc. Reproduced by permission.)

When a person is dyslexic, there is often an unexpected difference between achievement and aptitude. However, each person with dyslexia has different strengths and weaknesses, although many have unusual talents in art, athletics, architecture, graphics, drama, music, or engineering. These special talents are often in areas that require the ability to integrate sight, spatial skills, and coordination.

Often, a person with dyslexia has a problem translating language into thought (such as in listening or reading), or translating thought into language (such as in writing or speaking).

Common characteristics include problems with:

- identifying single words
- understanding sounds in words, sound order, or rhymes
- spelling
- transposing letters in words
- handwriting
- reading comprehension
- delayed spoken language
- confusion with directions, or right/left handedness

- confusion with opposites (up/down, early/late, and so on)
- mathematics

### Causes and symptoms

The underlying cause of dyslexia is not known, although research suggests the condition is often inherited. In 1999, The Centre for Reading Research in Norway presented the first research to study the largest family with reading problems ever known. By studying the reading and writing abilities of close to 80 family members across four generations the researchers reported, for the first time, that chromosome 2 can be involved in the inheritability of dyslexia. When a fault occurs on this gene it leads to difficulties in processing written language. Previous studies have pointed out linkages of other potential dyslexia genes to chromosome 1, chromosome 15 (DYX1 gene), and to chromosome 6 (DYX2 gene). The researchers who pinpointed the newly localized gene on chromosome 2 (DYX3) hope that this finding will lead to earlier and more precise diagnoses of dyslexia.

New research suggests a possible link with a subtle visual problem that affects the speed with which affected

## KEY TERMS

**Spatial skills**—The ability to locate objects in three dimensional world using sight or touch.

people can read. Other experts believe that dyslexia is related to differences in the structure and function of the brain that manifests differently in different people.

### Diagnosis

Anyone who is suspected to have dyslexia should have a comprehensive evaluation, including hearing, vision, and intelligence testing. The test should include all areas of learning and learning processes, not just reading.

As further research pinpoints the genes responsible for some cases of dyslexia, there is a possibility that earlier testing will be established to allow for timely interventions to prevent the onset of the condition and to treat it when it does occur.

Unfortunately, in many schools, a child is not identified as having dyslexia until after repeated failures.

### Treatment

If a child is diagnosed with dyslexia, the parents should find out from the school or the diagnostician exactly what the problem is, and what method of teaching is recommended and why. No single method will work with every child, and experts often disagree as to the best method to use.

The primary focus of treatment is aimed at helping the specific learning problem of each affected person. Most often, this may include modifying teaching methods and the educational environment, since traditional educational methods will not always work with a dyslexic child.

People with dyslexia need a structured language program, with direct instruction in the letter-sound system. Teachers must give the rules governing written language. Most experts agree that the teacher should emphasize the association between simple phonetic units with letters or letter groups, rather than an approach that stresses memorizing whole words.

It is important to teach these students using all the senses: hearing, touching, writing, and speaking, provided by an instructor who is specifically trained in a program that is effective for dyslexic students.

### Prognosis

Many successful and even famous people have dyslexia. How well a person with dyslexia functions in life depends on the way the disability affects that person. There is a great deal of variation among different people with dyslexia, producing different symptoms and different degrees of severity.

Prognosis is usually good if the condition is diagnosed early, and if the person has a strong self image with supportive family, friends, and teachers. It is imperative for a good outcome that the person be involved in a good remedial program.

### Resources

#### BOOKS

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#### ORGANIZATION

International Dyslexia Association (formerly the Orton Dyslexia Society). 8600 LaSalle Rd., Chester Bldg., Ste. 382, Baltimore, MD 21286. (800) ABC-D123.

Learning Disabilities Association. 4156 Library Rd., Pittsburgh, PA 15234. (412) 341-1515.

Beth Kapes

Dyslipidemia see **Hyperlipoproteinemia**

## Dysmenorrhea

### Definition

Dysmenorrhea is the occurrence of painful cramps during menstruation.

### Description

More than half of all girls and women suffer from dysmenorrhea (cramps), a dull or throbbing **pain** that

usually centers in the lower mid-abdomen, radiating toward the lower back or thighs. Menstruating women of any age can experience cramps.

While the pain may be only mild for some women, others experience severe discomfort that can significantly interfere with everyday activities for several days each month.

### Causes and symptoms

Dysmenorrhea is called “primary” when there is no specific abnormality, and “secondary” when the pain is caused by an underlying gynecological problem. It is believed that primary dysmenorrhea occurs when hormone-like substances called “prostaglandins” produced by uterine tissue trigger strong muscle contractions in the uterus during menstruation. However, the level of prostaglandins doesn’t seem to have anything to do with how strong a woman’s cramps are. Some women have high levels of prostaglandins and no cramps, whereas other women with low levels have severe cramps. This is why experts assume that cramps must also be related to other things (such as genetics, **stress**, and different body types) in addition to prostaglandins. The first year or two of a girl’s periods are not usually very painful. However, once ovulation begins, the blood levels of the prostaglandins rise, leading to stronger contractions.

Secondary dysmenorrhea may be caused by **endometriosis**, fibroid tumors, or an infection in the pelvis.

The likelihood that a woman will have cramps increases if she:

- has a family history of painful periods
- leads a stressful life
- doesn’t get enough **exercise**
- uses **caffeine**
- has pelvic inflammatory disease

Symptoms include a dull, throbbing cramping in the lower abdomen that may radiate to the lower back and thighs. In addition, some women may experience **nausea and vomiting**, **diarrhea**, irritability, sweating, or **dizziness**. Cramps usually last for two or three days at the beginning of each menstrual period. Many women often notice their painful periods disappear after they have their first child, probably due to the stretching of the opening of the uterus or because the birth improves the uterine blood supply and muscle activity.

### Diagnosis

A doctor should perform a thorough **pelvic exam** and take a patient history to rule out an underlying condition that could cause cramps.

### KEY TERMS

**Endometriosis**—The growth of uterine tissue outside the uterus.

**Hormone**—A chemical messenger secreted by a gland and released into the blood, which allows it to travel to distant cells where it exerts an effect.

**Ovary**—One of the two almond-shaped glands in the female body that produces the hormones estrogen and progesterone.

**Ovulation**—The monthly release of an egg from an ovary.

**Progesterone**—The hormone produced by the ovary after ovulation that prepares the uterine lining for a fertilized egg.

**Uterus**—The female reproductive organ that contains and nourishes a fetus from implantation until birth.

### Treatment

Secondary dysmenorrhea is controlled by treating the underlying disorder.

Several drugs can lessen or completely eliminate the pain of primary dysmenorrhea. The most popular choice are the **nonsteroidal anti-inflammatory drugs** (NSAIDs), which prevent or decrease the formation of prostaglandins. These include **aspirin**, ibuprofen (Advil), and naproxen (Aleve). For more severe pain, prescription strength ibuprofen (Motrin) is available. These drugs are usually begun at the first sign of the period and taken for a day or two. There are many different types of NSAIDs, and women may find that one works better for them than the others.

If an NSAID is not available, **acetaminophen** (Tylenol) may also help ease the pain. Heat applied to the painful area may bring relief, and a warm bath twice a day also may help. While birth control pills will ease the pain of dysmenorrhea because they lead to lower hormone levels, they are not usually prescribed just for **pain management** unless the woman also wants to use them as a birth control method. This is because these pills may carry other more significant side effects and risks.

New studies of a drug patch containing glyceryl trinitrate to treat dysmenorrhea suggest that it also may help ease pain. This drug has been used in the past to ease preterm contractions in pregnant women.

## Alternative treatment

Simply changing the position of the body can help ease cramps. The simplest technique is assuming the fetal position, with knees pulled up to the chest while hugging a heating pad or pillow to the abdomen. Likewise, several **yoga** positions are popular ways to ease menstrual pain. In the “cat stretch,” position, the woman rests on her hands and knees, slowly arching the back. The pelvic tilt is another popular yoga position, in which the woman lies with knees bent, and then lifts the pelvis and buttocks.

Dietary recommendations to ease cramps include increasing fiber, calcium, and complex carbohydrates, cutting fat, red meat, dairy products, caffeine, salt, and sugar. **Smoking** also has been found to worsen cramps. Recent research suggests that vitamin B supplements, primarily vitamin B<sub>6</sub> in a complex, magnesium, and fish oil supplements (omega-3 fatty acids) also may help relieve cramps.

Other women find relief through visualization, concentrating on the pain as a particular color and gaining control of the sensations. **Aromatherapy** and massage may ease pain for some women. Others find that imagining a white light hovering over the painful area can actually lessen the pain for brief periods.

Exercise may be a way to reduce the pain of menstrual cramps through the brain’s production of endorphins, the body’s own painkillers. And orgasm can make a woman feel more comfortable by releasing tension in the pelvic muscles.

**Acupuncture** and Chinese herbs are another popular alternative treatments for cramps.

## Prognosis

Medication should lessen or eliminate pain.

## Prevention

NSAIDs taken a day before the period begins should eliminate cramps for some women.

## Resources

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Adolescents.” *American Journal of Obstetrics and Gynecology* 174 (Apr. 1996): 335-8.

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## ORGANIZATIONS

American College of Obstetricians and Gynecologists. 409 12th Street, S.W., PO Box 96920

Federation of Feminist Women’s Health Centers. 1469 Humboldt Rd, Suite 200, Chico, CA 96928. (530) 891-1911.

National Women’s Health Network. 514 10th St. NW, Suite 400, Washington, DC 20004. (202) 628-7814. <<http://www.womenshealthnetwork.org>>.

Carol A. Turkington

## Dysmetria see Movement disorders

# Dyspepsia

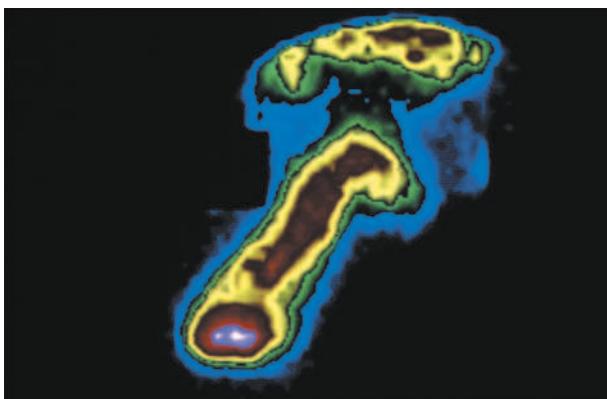
### Definition

Dyspepsia can be defined as painful, difficult, or disturbed digestion, which may be accompanied by symptoms such as **nausea and vomiting**, **heartburn**, bloating, and stomach discomfort.

### Causes and symptoms

The digestive problems may have an identifiable cause, such as bacterial or viral infection, peptic ulcer, gallbladder, or liver disease. The bacteria *Helicobacter pylori* is often found in those individuals suffering from duodenal or gastric ulcers. Investigation of recurrent **indigestion** should rule out these possible causes.

Often, there is no organic cause for the problem, in which case dyspepsia is classified as functional or nonulcer dyspepsia. There is evidence that functional dyspepsia may be related to abnormal motility of the upper gastrointestinal tract (a state known as dysmotility in which the esophagus, stomach, and upper intestine behave abnormally). These patients may respond to a group of drugs called prokinetic agents. A review of eating habits (e.g. chewing with the mouth open, gulping food, or talking while chewing) may reveal a tendency to swallow air. This may contribute to feeling bloated, or to excessive belching. **Smoking**, **caffeine**, alcohol, or carbonated beverages may contribute to the discomfort. When there is sensitivity or allergy to certain food substances, eating those foods may cause gastrointestinal distress. Some medications are associated with indigestion. Stomach problems may also be a response to **stress** or emotional unrest.



**A false-color gamma scan of a human stomach with dyspepsia, or indigestion, during tests to study its rate of emptying.** (Photograph by Jean-Perrin, Custom Medical Stock Photo. Reproduced by permission.)

## Diagnosis

A **physical examination** by a health care professional may reveal mid-abdominal **pain**. A **rectal examination** may be done to rule out bleeding. If blood is found on rectal exam, laboratory studies, including a **blood count** may be ordered. Endoscopy and barium studies may be used to rule out underlying gastrointestinal disease. Upper gastrointestinal x-ray studies using barium may allow for visualization of abnormalities. Endoscopy permits collection of tissue and culture specimens which may be used to further confirm a diagnosis.

## Treatment

The treatment of dyspepsia is based on assessment of symptoms and suspected causative factors. Clinical evaluation is aimed at distinguishing those patients who require immediate diagnostic work-ups from those who can safely benefit from more conservative initial treatment. Some of the latter may require only reassurance, dietary modifications, or antacid use. Medications to block production of stomach acids, prokinetic agents, or antibiotic treatment may be considered. Further diagnostic investigation is indicated if there is severe abdominal pain, pain radiating to the back, unexplained weight loss, difficulty swallowing, a palpable mass, or anemia. Additional work-up is also indicated if a patient does not respond to prescribed medications.

## Prognosis

Statistics show an average of 20% of patients with dyspepsia have duodenal or gastric ulcer disease, 20%

## KEY TERMS

**Anemia**—Diagnosed through laboratory study of the blood, a deficiency in hemoglobin or red blood cells, often associated with paleness or loss of energy.

**Endoscopy**—A diagnostic procedure using a lighted instrument to examine a body cavity or internal organ. Endoscopy permits collection of tissue and culture specimens.

have **irritable bowel syndrome**, fewer than 1% of patients had **cancer**, and the range for functional, or non-ulcer dyspepsia (**gastritis** or superficial erosions), was from 5–40%.

## Resources

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Kathleen D. Wright, RN

Dysphasia see **Aphasia**

Dyspnea see **Shortness of breath**

Dysthymic disorder see **Depressive disorders**

Dystonia see **Movement disorders**

# E

*E. coli* see ***Escherichia coli***

*E. coli* infection see **Enterobacterial infections**

*E. coli* O157:H7 infection see ***Escherichia coli***

Ear canal infection see **Otitis externa**

**examination.** The ears may also be examined if an ear infection is suspected due to **fever**, ear pain, or **hearing loss**. The patient will often be asked to tip the head slightly toward the shoulder so the ear to be examined is pointing up. The doctor or nurse may hold the ear lobe as the speculum is inserted into the ear, and may adjust the position of the otoscope to get a better view of the ear canal and eardrum. Both ears are usually examined, even if there seems to be a problem with just one ear.

## Preparation

No special preparation is required prior to an ear examination with an otoscope. The ear speculum, which is inserted into the ear, is cleaned and sanitized before it is used. The speculums come in various sizes, and the doctor or nurse will select the size that will be most comfortable for the patient's ear.

## Aftercare

If an ear infection is diagnosed, the patient may require treatment with **antibiotics**. If there is a buildup of wax in the ear canal, it might be rinsed or scraped out.

## Risks

This type of ear examination is simple and generally harmless. Caution should always be used any time an object is inserted into the ear. This process could irritate an infected external ear canal and could rupture an eardrum if performed improperly or if the patient moves.

## Normal results

The ear canal is normally skin-colored and is covered with tiny hairs. It is normal for the ear canal to have some yellowish-brown earwax. The eardrum is typically thin, shiny, and pearly-white to light gray in color. The tiny bones in the middle ear can be seen pushing on the

## Purpose

An otoscope is used to look into the ear canal to see the ear drum. Redness or fluid in the eardrum can indicate an ear infection. Some otoscopes can deliver a small puff of air to the eardrum to see if the eardrum will vibrate (which is normal). This type of ear examination with an otoscope can also detect a build up of wax in the ear canal, or a rupture or puncture of the eardrum.

## Precautions

No special precautions are required. However, if an ear infection is present, an ear examination may cause some discomfort or **pain**.

## Description

An ear examination with an otoscope is usually done by a doctor or a nurse as part of a complete **physical**

## KEY TERMS

**Ear speculum**—A cone- or funnel-shaped attachment for an otoscope which is inserted into the ear canal to examine the eardrum.

**Otoscope**—A hand-held instrument with a tiny light and a funnel-shaped attachment called an ear speculum, which is used to examine the ear canal and eardrum.

eardrum membrane like tent poles. The light from the otoscope will reflect off of the surface of the ear drum.

### Abnormal results

An ear infection will cause the eardrum to look red and swollen. In cases where the eardrum has ruptured, there may be fluid draining from the middle ear. A doctor may also see scarring, retraction of the eardrum, or bulging of the eardrum.

### Resources

#### ORGANIZATIONS

American Academy of Otolaryngology-Head and Neck Surgery, Inc. One Prince St., Alexandria VA 22314-3357. (703) 836-4444. <<http://www.entnet.org>>. Ear Foundation. 1817 Patterson St., Nashville, TN 37203. (800) 545-4327. <<http://www.earfoundation.org>>. Hearing Health Information. 2100 W. 3rd St., Los Angeles, CA 90057. (213) 483-4431. National Institute on Deafness and Other Communication Disorders. National Institutes of Health, 31 Center Drive, MSC 2320, Bethesda, MD USA 20892-2320. (800) 241-1044. <<http://www.nidcd.nih.gov>>.

#### OTHER

"Ear Test." *HealthAnswers.com*. <<http://www.healthanswers.com>>.

Altha Roberts Edgren

## Ear surgery

### Definition

Ear surgery is the treatment of diseases, injuries, or deformations of the ear by operation with instruments.

### Purpose

Ear surgery is performed to correct certain types of **hearing loss**, and to treat diseases of, injuries to, or

deformities of the ear's auditory tube, middle ear, inner ear, and auditory and vestibular systems. Ear surgery is commonly performed to treat conductive hearing loss, persistent ear infections, unhealed perforated eardrums, congenital ear defects, and tumors.

Ear surgery is performed on children and adults. In some cases, surgery is the only treatment; in others, it is used only when more conservative medical treatment fails.

### Precautions

The precautions vary, depending on the type of ear surgery under consideration. For example, **stapedectomy** (removal of parts of the middle ear and insertion of prosthesis parts) should not be performed on people with external or middle ear infection or inner ear disease. For people with complete hearing loss in the other ear, it should be performed cautiously. Microsurgery for the removal of a cholesteatoma (a cyst-like mass of cells in the middle ear) should not be performed on patients who are extremely ill or have other medical conditions. Tympanoplasty (any surgical procedure on the eardrum or middle ear) should not be performed on patients with chronic sinus or nasal problems or in some patients with medical problems such as poorly controlled diabetes and heart disease. Surgery for congenital microtia and atresia (absence of normal bodily openings, such as the outer ear canal) should not be performed if the middle ear space is totally or almost totally absent.

### Description

Most ear surgery is microsurgery, performed with an operating microscope to enable the surgeon to view the very small structures of the ear. The use of minimally invasive **laser surgery** for middle ear procedures is growing. Laser surgery reduces the amount of trauma due to vibration, enhances coagulation, and enables surgeons to access hard to reach places in the middle ear. Laser surgery can be performed in an office operating suite. Types of ear surgery include stapedectomy, tympanoplasty, myringotomy and ear tube surgery, ear surgery to repair a **perforated eardrum**, **cochlear implants**, and **tumor removal**.

### Stapedectomy

To restore hearing loss, which is usually due to **otosclerosis**, stapedectomy is performed. Stapedectomy is the removal of all or part of the stapes, one of the bones in the middle ear, and replacement with a tiny prosthesis. An incision is made in the middle ear, the small bones are identified, and the stapes is removed. The stainless steel wire and cellulose sponge prosthesis is inserted, blood

and fluid are drained, and the wound is closed. Performed in a hospital or outpatient surgical facility under local or general anesthetic, full recovery takes about three weeks but hearing should improve immediately.

### **Tympanoplasty**

Tympanoplasty is performed to reconstruct the eardrum after partial or total conductive hearing loss, usually caused by chronic middle ear infections, or perforations that do not heal. This is usually a same day surgery, performed under either local or general anesthesia. After making an incision in the ear to view the perforation, the ear drum is elevated away from the ear canal and lifted forward. If the bones of hearing (ossicular chain) are functioning, tissue is taken from the ear and grafted to the eardrum to close the perforation. A thin sheet of silastic and Gelfoam hold the graft in place. The ear is stitched together, and a sterile patch is placed on the outside of the ear canal. Tympanoplasty is successful in over 90% of all cases. The need for ossicular reconstruction (reconstruction of tiny bones of the middle ear) is sometimes known before surgery and even when identified during surgery, can usually be done while reconstructing the eardrum. If the gap between the anvil bone and the stapes is small, a small piece of bone or cartilage from the patient can be inserted; if it is large, the incus bone is removed, modelled into a prosthesis, and reinserted between the stapes and the malleus. Reconstruction could also be achieved by inserting a strut made from artificial bone. For tympanoplasty with ossicular reconstruction, the patient usually stays in the hospital overnight. The recovery period is about four weeks.

### **Myringotomy and ear tube surgery**

Myringotomy and ear tube surgery is performed to drain ear fluid and prevent ear infections when **antibiotics** don't work or when ear infections are chronic. The process normalizes pressure in the middle ear and decreases fluid accumulation. It is most commonly performed on infants and children, in whom ear infections are most frequent, and may be done on one or both ears. The surgeon makes a small hole in the ear drum, then uses suction to remove fluid. A small ear tube of metal or plastic is inserted into the ear drum to allow continual drainage. The tube prevents infections as long as it stays in place, which varies from six months to three years. When the tube falls out, the hole grows over. As many of 25% of children under the age of two who need ear tubes may need them again. Myringotomy and ear tube surgery is performed in a hospital, using a general anesthetic for most children and a local anesthetic for older children or adults. No anesthetic may be used for infants. The procedure usually takes about two hours. Most patients can go



**Microsurgery being performed in the inner ear.** (Photograph by Hans Halberstadt, Photo Researchers, Inc. Reproduced by permission.)

home the same day; children under three years of age and those with chronic diseases usually stay overnight.

### **Ear surgery for a perforated eardrum**

Ear surgery for a perforated eardrum is only performed in rare cases where it does not heal on its own. In most cases, this is performed in a surgeon's office using a topical anesthetic. The surgeon scratches the undersurface of the eardrum, stimulating the skin to heal and the eardrum to close. A thin patch placed on the eardrum's outer surface allows the skin under the eardrum to heal.

### **Cochlear implants**

Cochlear implants stimulate nerve ends within the inner ear, enabling deaf children to hear. The device has a microphone that remains outside the ear, a processor that selects and codes speech sounds, and a receiver/stimulator to convert the coded sounds to electric signals that stimulate the hearing nerve and are recognized by the brain as sound. During surgery, an incision is made behind and slightly above the ear. A circular hole is drilled in the bone to receive the device's internal coil. The mastoid bone leading to the middle ear is opened to receive the electrodes. The internal coil is inserted and secured, followed by the electrodes. The wound is stitched up and when it heals, an external unit comprised of a stimulator with a microphone is worn behind the ear. Performed in a hospital under general anesthesia, the operation takes about two hours and usually requires a hospital stay overnight. The patient can resume normal activities in two to three weeks.

### **Ear surgery for tumors**

Some ear tumors can be very serious and should be removed surgically. For a tumor on the skin of the ear canal, the skin is removed surgically, the bone beneath it

## KEY TERMS

**Auditory**—Relating to the sense of the organs of hearing.

**Cholesteatoma**—A cystic mass of cells in the middle ear, occurring as a congenital defect or as a serious complication of a disease or traumatic condition of the ear.

**Otologic**—Relating to the study, diagnosis, and treatment of diseases of the ear and related structures.

is drilled away, and a skin graft is placed in the ear canal. If the tumor is near the eardrum, the skin of the ear canal and the eardrum are removed along with the bone surrounding the ear canal. A skin graft is placed on the bare bone. For basal cell cancers and low grade glandular malignancies, surgical resection of the ear canal is adequate. Squamous cell carcinoma, a serious form of **cancer**, of the external ear canal requires radical surgery, followed by **radiation therapy**. Cholesteatoma, a benign tumor caused by an infection in a perforated eardrum that did not heal properly and can destroy the bones of hearing, is removed with microsurgery. **Mastoidectomy** is performed for **mastoiditis**, an inflammation of the middle ear, if medical therapy does not work. Petrous apicectomy is performed to drain the petrous apicitis, the bone between the middle ear and the clivus.

### Ear surgery for congenital ear defects

Congenital atresia, the absence of the external ear canal, and congenital microtia, abnormal growth of the external ear, often occur together, although atresia can occur without microtia. Surgery to reconstruct the ear usually takes place when the child is four or five years old and may require several operations. A facial plastic surgeon and an ear surgeon work together, repairing the microtia first and then the atresia. During surgery, a bony opening is created over the bones of hearing. The surfaces of the bony ear canal are then relined with a skin graft from the thigh or abdomen. Tissue from behind the eardrum is used to create a new eardrum. In many cases, the middle ear will also need to be reconstructed. Surgery is performed in a hospital under general anesthesia.

### Other types of ear surgery

Surgery may also be appropriate to remove multiple bony overgrowths of the ear canal or in rare cases of compromised auditory tube function, to narrow the tube.

## Preparation

The preparation depends upon the type of ear surgery performed. For many procedures, blood and urine studies and hearing tests are conducted.

## Aftercare

The type of aftercare depends upon the type of surgery performed. In most cases, the ear(s) should be kept dry and warm. Non-prescription drugs such as **acetaminophen** can be used for **pain**.

## Risks

The type of risk depends on the type of surgery performed. Total hearing loss is rare.

## Resources

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### ORGANIZATIONS

American Academy of Otolaryngology-Head and Neck Surgery, Inc. One Prince St., Alexandria VA 22314-3357. (703) 836-4444. <<http://www.entnet.org>>.

American Hearing Research Foundation. 55 E. Washington St., Suite 2022, Chicago, IL 60602. (312) 726-9670. <<http://www.american-hearing.org>>.

American Speech-Language-Hearing Association. 10801 Rockville Pike, Rockville, MD 20852. (800) 638-8255. <<http://www.asha.org>>.

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Lori De Milto

Ear tubes see **Myringotomy and ear tubes**

Ear wax impaction see **Cerumen impaction**

Eardrum perforation see **Perforated eardrum**  
 Eastern equine encephalitis see **Arbovirus encephalitis**  
 Eating disorders see **Anorexia nervosa; Bulimia nervosa**  
 Eaton agent pneumonia see **Mycoplasma infections**  
 Ebola virus infection see **Hemorrhagic fevers**  
 Ecchymosis see **Bruises**  
 ECG see **Electrocardiography**

## Echinacea

### Definition

Echinacea, or purple coneflower, is a perennial herb of the Composite family, commonly known as the daisy family. Most often referred to as the purple coneflower, this hardy plant also known as Sampson root, Missouri snakeroot, and rudbeckia. The prominent, bristly seed head inspired the generic name of the plant, taken from the Greek word *echinos* meaning hedgehog.

### Description

Echinacea is a North American prairie native, abundant in the Midwest, and cultivated widely in ornamental and medicinal gardens. The purple-pink rays of the blossom droop downward from a brassy hued center cone composed of many small, tubular florets. The conspicuous flowers bloom singly on stout, prickly stems from mid-summer to autumn. Flower heads may grow to 4 in (10.16 cm) across. The dark green leaves are opposite, entire, lanceolate, toothed, and hairy with three prominent veins. The narrow upper leaves are attached to the stem with stalks. The lower leaves are longer, emerging from the stem without a leaf stalk, and growing to 8 in (20.32 cm) in length. The plant develops deep, slender, black roots. Echinacea propagates easily from seed or by root cuttings. However, due to its increasing popularity as an herbal supplement, echinacea is numbered among the 19 medicinal plants considered at risk by the Vermont nonprofit organization, United Plant Savers.

### Purpose

Three species of echinacea are useful medicinally: *Echinacea augustifolia*, *Echinacea purpurea*, and *Echi-*

*nacea pallida*. The entire plant has numerous medicinal properties that act synergistically to good effect. Echinacea is most often used to boost the immune system and fight infection. Research has shown that echinacea increases production of interferon in the body. It is anti-septic and antimicrobial, with properties that act to increase the number of white blood cells available to destroy bacteria and slow the spread of infection. As a depurative, the herbal extract cleanses and purifies the bloodstream, and has been used effectively to treat **boils**. Echinacea is vulnerary, promoting wound healing through the action of a chemical substance in the root known as caffeic acid glycoside. As an alterative and an immunomodulator, echinacea acts gradually to promote beneficial change in the entire system. It has also been used to treat urinary infection and *Candida albicans* infections. Echinacea is a febrifuge, useful in reducing fevers. It is also useful in the treatment of **hemorrhoids**. A tincture, or a strong decoction of echinacea serves as an effective mouthwash for the treatment of pyorrhea and gingivitis.

Native American plains Indians relied on echinacea as an all-purpose antiseptic. The Sioux tribe valued the root as a remedy for snake bite, the Cheyenne tribe chewed the root to quench thirst, and another tribe washed their hands in a decoction of echinacea to increase their tolerance of heat. European settlers learned of the North American herb's many uses, and soon numerous echinacea-based remedies were commercially available from pharmaceutical companies in the United States. Echinacea was a popular remedy in the United States through the 1930s. It was among many medicinal herbs listed in the *U.S. Pharmacopoeia*, the official United States government listing of pharmaceutical raw materials and recipes. The herb fell out of popular use in the United States with the availability of **antibiotics**. In West Germany, over 200 preparations are made from the species *E. purpurea*. Commercially prepared salves, tinctures, teas, and extracts are marketed using standardized extracts. Echinacea is regaining its status in the United States as a household medicine-chest staple in many homes. It is one of the best-selling herbal supplements in United States health food stores.

Clinical studies have found that the entire plant possesses medicinal properties with varying levels of effectiveness. Echinacea is of particular benefit in the treatment of upper respiratory tract infections. Some research has shown that echinacea activates the macrophages that destroy **cancer** cells and pathogens. When taken after cancer treatments, an extract of the root has been found to increase the body's production of white blood cells. Echinacea has been shown to be most effective when taken at the first sign of illness, rather than when used as a daily preventative. Other research has demonstrated the signifi-

## KEY TERMS

**Alterative**—A medicinal substance that acts gradually to nourish and improve the system.

**Antimicrobial**—A plant substance that acts to inhibit the growth of harmful microorganisms, or acts to destroy them.

**Febrifuge**—A plant substance that acts to prevent or reduce fever.

**Glycoside**—An herbal carbohydrate that exerts powerful effect on hormone-producing tissues. The glycoside breaks down into a sugar and a non-sugar component.

**Lanceolate**—Narrow, leaf shape that is longer than it is wide, and pointed at the end.

**Macrophage**—Specialized cells present throughout the lymphoid tissues of the body that circulate in the bloodstream. Macrophages have a surface marker that stimulates other cells to react to an antigen.

cant effect of *E. purpurea* root on reducing the duration and severity of colds and flu. Some herbal references list only the root as the medicinal part, others include the aerial parts of the plant, particularly the leaf. But research studies in Europe and the United States have concluded that the entire plant is medicinally effective. Most research has been done on the species *E. pallida* and *E. purpurea*. All three species of echinacea are rich in **vitamins** and **minerals**. Echinacea is an herbal source of niacin, chromium, iron, manganese, selenium, silicon, and zinc.

### Preparations

The quality of any herbal supplement depends greatly on the conditions of weather and soil where the herb was grown, the timing and care in harvesting, and the manner of preparation and storage.

Decoction is the best method to extract the mineral salts and other healing components from the coarser herb materials, such as the root, bark, and stems. It is prepared by adding 1 oz (28.4 g) of the dried plant materials, or 2 oz (56.7 g) of fresh plant parts, to 1 pt (0.47 l) of pure, unchlorinated, boiled water in a non-metallic pot. Simmer for about one half hour. Strain and cover. A decoction may be refrigerated for up to two days and retain its healing qualities.

An infusion is the method used to derive benefits from the leaves, flowers, and stems in the form of an herbal tea. Use twice as much fresh, chopped herb as

dried herb. Steep in 1 pt (0.47 l) of boiled, unchlorinated water for 10–15 minutes. Strain and cover. Drink warm, sweetened with honey if desired. A standard dose is three cups per day. An infusion will keep for up to two days in the refrigerator and retain its healing qualities.

A tincture is the usual method to prepare a concentrated form of the herbal remedy. Tinctures, properly prepared and stored, will retain medicinal potency for two years or more. Combine 4 oz (114 g) of finely cut fresh or powdered dry herb with 1 pt (0.47 l) of brandy, gin, or vodka in a glass container. The alcohol should be enough to cover the plant parts and have a 50/50 ratio of alcohol to water. Place the mixture away from light for about two weeks, shaking several times each day. Strain and store in a tightly capped, dark glass bottle. A standard dose is 0.14 oz (4 ml) of the tincture three times a day.

### Precautions

Echinacea is considered safe in recommended doses. Pregnant or lactating women, however, are advised not to take echinacea in injection form. Because the plant has proven immuno-modulating properties, individuals with systemic lupus erythematosus, **rheumatoid arthritis**, **tuberculosis**, leukemia, **multiple sclerosis**, or **AIDS** should consult their physician before using echinacea. Echinacea should not be given to children under two years of age, and it should only be given to children over two in consultation with a physician. Research indicates that echinacea is most effective when taken at first onset of symptoms of cold or flu, and when usage is continued no longer than eight weeks. There is some indication that the herb loses its effectiveness when used over a long period of time. It is necessary to interrupt use for a minimum of several weeks in order to give the body's immune system the opportunity to rest and adjust.

### Side effects

No side effects are reported with oral administration of echinacea, either in tincture, capsule, or as a tea, when taken according to recommended doses. Chills, **fever**, and allergic reactions have been reported in some research studies using an injection of the plant extract.

### Interactions

None reported. When used in combination with other herbs, dosage should be lowered.

### Resources

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Clare Hanrahan

## Echinococcosis

### Definition

Echinococcosis (Hydatid disease) refers to human infection by the immature (larval) form of tapeworm, *Echinococcus*. One of three forms of the *Echinococcus* spp., *E. granulosus*, lives on dogs and livestock, and infects humans through contact with these animals. Allergic reactions and damage to various organs from cyst formation are the most common forms of disease in humans.

### Description

*E. granulosus* is found in many areas of Africa, China, South America, Australia, New Zealand, and Mediterranean and eastern Europe, as well as in parts of the western United States. The parasite lives in regions where dogs and livestock cohabit. Direct exposure to infectious dogs as well as parasitic eggs released into the environment during shedding are both sources of human infection.

In humans, cysts containing the larvae develop after ingestion of eggs. Cysts form primarily in the lungs and liver. Cysts developing in the liver are responsible for about two-thirds of echinococcosis cases. Echinococcosis is a significant public health problem in many areas of

### KEY TERMS

**Allergenic**—A substance capable of causing an allergic reaction.

**Cholangitis**—Infection or inflammation of the bile ducts; often causes abdominal pain, fever, and jaundice.

**Computed tomography (CT) scan**—A specialized x-ray procedure in which cross-sections of the area in question can be examined in detail.

**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.

**Embryo**—The very beginning stages of development of an organism.

**Jaundice**—The yellow-greenish coloring of the skin and eyes due to the presence of bile pigments. The presence of jaundice is usually, but not always, a sign of liver disease.

**Tapeworm**—An intestinal parasite that attaches to the intestine or travels to other organs such as the liver and lungs.

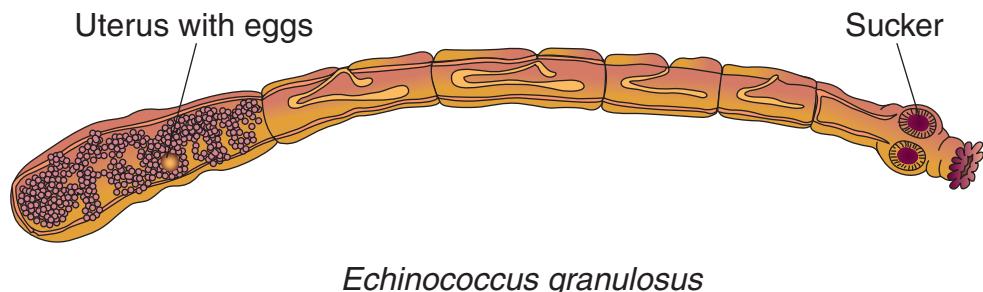
**Ultrasound**—A noninvasive procedure based on changes in sound waves of a frequency that cannot be heard, but respond to changes in tissue composition.

the world, but control programs have decreased the rate of infection in some regions. In Kenya alone, the numbers of persons infected each year is as high as 220 per 100,000 population.

### Causes and symptoms

After ingestion, the eggs develop into embryos within the intestines and then travel to the liver and lungs through major blood vessels. The embryos then begin to form cysts within the liver and lungs, causing damage as they enlarge over a period of five to 20 years. Cysts may become over 8 in (20.3 cm) or more in size and contain a huge amount of highly allergenic fluid. Studies show that while the liver is most often targeted, lungs, brain, heart, and bone can also be affected.

The major symptoms are due to compression damage, blockage of vessels and ducts (such as the bile ducts), and leakage of fluid from cysts. The following symptoms are frequent.



Infection with the larva of *Echinococcus granulosus* (shown above) is responsible for the disease echinococcosis. (Illustration by Electronic Illustrators Group.)

- Liver involvement causes **pain** and eventually **jaundice** or **cholangitis** due to blockage of bile ducts. Infection of cysts leads to abscesses in up to 20%.
- Lung cysts cause **cough** and chest pain.
- Bone cysts cause **fractures** and damage to bone tissue.
- Heart involvement leads to irregularities of heart beat and inflammation of the covering of the heart (pericardium).
- Allergic reactions occur from leakage of cyst fluid that contains antigens. **Itching**, **fever**, and **rashes** are frequent, and fatal allergic reactions (**anaphylaxis**) have been reported. Eosinophils, which are blood cells involved in allergic reactions, are increased in many patients.

## Diagnosis

X rays, **computed tomography scans** (CT scans), and ultrasound are very helpful in detecting cysts. Some cysts will develop characteristic hardening of organ tissues from calcium deposits (calcifications). Blood tests to detect antibodies are useful when positive, but up to 50% of patients have negative results. Examination of aspirated cyst fluid for parasites can be diagnostic, but carries the danger of a fatal allergic reaction. Treatment with anti-parasitic medications before aspiration is reported to decrease allergic complications and decrease the risk of spread during the procedure.

## Treatment

Treatment depends on the size and location of cysts, as well as the symptoms they are producing. Surgical removal of cysts and/or surrounding tissue is the accepted method of treatment, but carries a risk of cyst rupture

with spread or allergic reactions. Recent studies using medication alongside aspiration and drainage of cysts instead of surgery are very encouraging.

The medication albendazole can be taken before or after surgery or alone without surgery. However, its effectiveness as a single treatment is still not known. Multiple courses of medication are often necessary, with cure rates of only about 30%. Response to treatment is best monitored by serial CT scans or similar x-ray studies.

## Prevention

Good hand washing, treating infected dogs, and preventing dogs access to slaughter houses discourage spread of the disease. Limiting the population of stray dogs has also been helpful.

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David Kaminstein, MD

*Echinococcus granulosus* infection see  
**Echinococcosis**



**A patient getting an EKG.** (Photo Researchers. Reproduced by permission.)

## Echocardiography

### Definition

Echocardiography is a diagnostic test that uses ultrasound waves to create an image of the heart muscle. Ultrasound waves that rebound or echo off the heart can show the size, shape, and movement of the heart's valves and chambers as well as the flow of blood through the heart. Echocardiography may show such abnormalities as poorly functioning heart valves or damage to the heart tissue from a past **heart attack**.

### Purpose

Echocardiography is used to diagnose certain cardiovascular diseases. In fact, it is one of the most widely used diagnostic tests for heart disease. It can provide a wealth of helpful information, including the size and shape of the heart, its pumping strength, and the location and extent of any damage to its tissues. It is especially useful for assessing diseases of the heart valves. It not only allows doctors to evaluate the heart valves, but it can detect abnormalities in the pattern of blood flow, such as the backward flow of blood through partly closed heart valves, known as regurgitation. By assessing the motion of the heart wall, echocardiography can help detect the presence and assess the severity of **coronary artery disease**, as well as help determine whether any chest **pain** is related to heart disease. Echocardiography can also help detect **hypertrophic cardiomyopathy**, in which the walls of the heart thicken in an attempt to compensate for heart muscle weakness. The biggest advantage to echocardiography is that it is noninvasive (doesn't involve breaking the skin or entering body cavities) and has no known risks or side effects.

### Precautions

Echocardiography is an extremely safe procedure and no special precautions are required.

### Description

Echocardiography creates an image of the heart using ultra-high-frequency sound waves—sound waves that are too high in frequency to be heard by the human ear. The technique is very similar to ultrasound scanning commonly used to visualize the fetus during **pregnancy**.

An echocardiography examination generally lasts between 15–30 minutes. The patient lies bare-chested on an examination table. A special gel is spread over the chest to help the transducer make good contact and slide smoothly over the skin. The transducer, a small hand-held device at the end of a flexible cable, is placed against the chest. Essentially a modified microphone, the transducer directs ultrasound waves into the chest. Some of the waves get echoed (or reflected) back to the transducer. Since different tissues and blood all reflect ultrasound waves differently, these sound waves can be translated into a meaningful image of the heart, which can be displayed on a monitor or recorded on paper or tape. The patient does not feel the sound waves, and the entire procedure is painless. In fact, there are no known side effects.

Occasionally, variations of the echocardiography test are used. For example, Doppler echocardiography employs a special microphone that allows technicians to measure and analyze the direction and speed of blood flow through blood vessels and heart valves. This makes it especially useful for detecting and evaluating regurgitation through the heart valves. By assessing the speed of blood flow at different locations around an obstruction, it can also help to precisely locate the obstruction.

An **exercise echocardiogram** is an echocardiogram performed during exercise, when the heart muscle must work harder to supply blood to the body. This allows doctors to detect heart problems that might not be evident when the body is at rest and needs less blood. For

## KEY TERMS

**Noninvasive**—Pertaining to a diagnostic procedure or treatment that does not require the skin to be broken or a body cavity to be entered.

**Regurgitation**—Backward flow of blood through a partly closed heart valve.

**Transducer**—A device that converts electrical signals into ultrasound waves and ultrasound waves back into electrical impulses.

**Ultrasound**—Sound waves at a frequency of over 20,000 kHz, often used for diagnostic imaging.

patients who are unable to exercise, certain drugs can be used to mimic the effects of exercise by dilating the blood vessels and making the heart beat faster.

### Preparation

The patient removes any clothing and jewelry above the chest.

### Aftercare

No special measures need to be taken following echocardiography.

### Risks

There are no known risks associated with the use of echocardiography.

### Normal results

A normal echocardiogram shows a normal heart structure and the normal flow of blood through the heart chambers and heart valves. However, a normal echocardiogram does not rule out the possibility of heart disease.

### Abnormal results

An echocardiogram may show a number of abnormalities in the structure and function of the heart, such as:

- thickening of the wall of the heart muscle (especially the left ventricle)
- abnormal motion of the heart muscle
- blood leaking backward through the heart valves (regurgitation)
- decreased blood flow through a heart valve (stenosis)

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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. PO Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

Robert Scott Dinsmoor

Echovirus infections see **Enterovirus infections**

Eclampsia see **Preeclampsia and eclampsia**

ECT see **Electroconvulsive therapy**

Ectopic orifice of the ureter see **Congenital ureter anomalies**

## Ectopic pregnancy

### Definition

In an ectopic **pregnancy**, the fertilized egg implants in a location outside the uterus and tries to develop there. The word ectopic means “in an abnormal place or position.” The most common site is the fallopian tube, the tube that normally carries eggs from the ovary to the uterus. However, ectopic pregnancy can also occur in the ovary, the abdomen, and the cervical canal (the opening from the uterus to the vaginal canal). The phrases tubal pregnancy, ovarian pregnancy, cervical pregnancy, and abdominal pregnancy refer to the specific area of an ectopic pregnancy.

### Description

Once a month, an egg is produced in a woman’s ovary and travels down the fallopian tube where it meets the male’s sperm and is fertilized. In a normal pregnancy the fertilized egg, or zygote, continues on its passage down the fallopian tube and enters the uterus in three to five days. The zygote continues to grow, implanting itself securely in the wall of the uterus. The zygote’s cells develop into the embryo (the organism in its first two months of develop-

ment) and placenta (a spongy structure that lines the uterus and nourishes the developing organism).

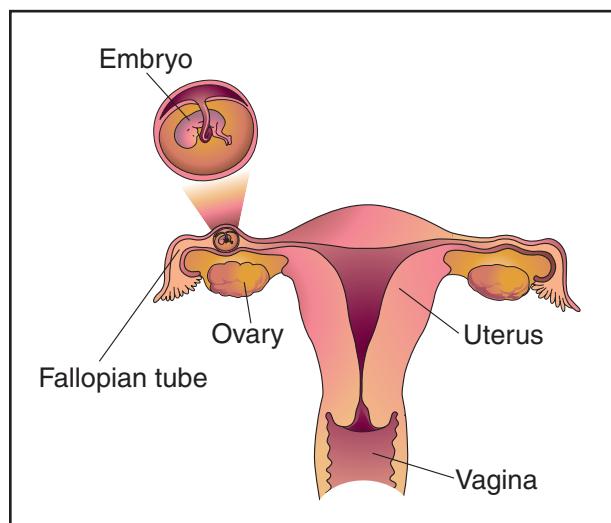
In a tubal ectopic pregnancy, the fertilized egg cannot make it all the way down the tube because of scarring or obstruction. The fallopian tube is too narrow for the growing zygote. Eventually the thin walls of the tube stretch and may burst (rupture), resulting in severe bleeding and possibly the **death** of the mother. More than 95% percent of all ectopic pregnancies occur in the fallopian tube. Only 1.5% develop in the abdomen; less than 1% develop in the ovary or the cervix.

### Causes and symptoms

As many as 50% of women with ectopic pregnancies have a history of **pelvic inflammatory disease** (PID). This is an infection of the fallopian tubes (salpingitis) that can spread to the uterus or ovaries. It is most commonly caused by the organisms *Gonorrhea* and *Chlamydia* and is usually transmitted by sexual intercourse.

Other conditions also increase the risk of ectopic pregnancy. They include:

- **Endometriosis.** A condition in which the tissue that normally lines the uterus is found outside the uterus, and can block a fallopian tube.
- Exposure to diethylstilbestrol (DES) as a fetus. If a woman's mother took DES (a synthetic version of the hormone estrogen) during pregnancy, the woman may have abnormalities in her fallopian tubes that can make ectopic pregnancy more likely.
- Taking hormones. Estrogen and progesterone are hormones that regulate the menstrual cycle and may be in medications prescribed by a doctor for birth control or other reasons. Taking these hormones can affect the interior lining of the fallopian tubes and slow the movement of the fertilized egg down the tube. Women who become pregnant in spite of taking some progesterone-only contraceptives have a greater chance of an ectopic pregnancy. Ectopic pregnancy is also more likely when the ovaries are artificially stimulated with hormones to produce eggs for **in vitro fertilization** (a procedure in which eggs are taken from a woman's body, fertilized, and then placed in the uterus in an attempt to conceive a child).
- Use of an intrauterine device (IUD). These contraceptive devices are designed to prevent fertilized eggs from becoming implanted in the uterus, but they have only a minimal effect on preventing ectopic pregnancies. Therefore, if a woman becomes pregnant while using an IUD for **contraception**, the fertilized egg is more likely to be implanted someplace other than the uterus. For example, among women who become pregnant while using a progesterone-bearing IUD, about 15% have ectopic pregnancies.



**In an ectopic pregnancy, the fertilized egg implants in a location outside the uterus and attempts to develop at that site. The most common site of an ectopic pregnancy is the fallopian tube, but it can occur in the ovary, the abdomen, and the cervical wall. More than 95% of all ectopic pregnancies occur in the fallopian tube.** (Illustration by Electronic Illustrators Group.)

- Surgery on a fallopian tube. The risk of ectopic pregnancy can be as high as 60% after undergoing elective tubal sterilization, a procedure in which the fallopian tubes are severed to prevent pregnancy. Women who have successful surgery to reverse the procedure are also more likely to have an ectopic pregnancy.

### Early symptoms

In an ectopic pregnancy all the hormonal changes associated with a normal pregnancy may occur. The early symptoms include: **fatigue**; nausea; a missed period; breast tenderness; **low back pain**; mild cramping on one side of the pelvis; and abnormal vaginal bleeding, usually spotting.

### Later symptoms

As the embryo grows too large for the confined space in the tube, the first sign that something is wrong may be a stabbing **pain** in the pelvis or abdomen. If the tube has ruptured, blood may irritate the diaphragm and cause shoulder pain. Other warning signs are lightheadedness and **fainting**.

### Diagnosis

To confirm an early diagnosis of ectopic pregnancy, the doctor must determine first that the patient is pregnant and that the location of the embryo is outside the uterus. If an ectopic pregnancy is suspected, the doctor

## KEY TERMS

**Embryo**—In humans, the developing organism from conception until approximately the end of the second month.

**Fallopian tube**—The tube that carries the egg from the ovary to the uterus.

**Human chorionic gonadotropin (hCG)**—A hormone excreted during the development of an embryo or fetus.

**Laparoscopy**—Examination of the contents of the abdominal cavity with a fiberoptic tube inserted through a small incision.

**Laparotomy**—Surgical incision into the abdomen to locate, repair, and/or remove injured or diseased tissues.

**Pelvic inflammatory disease (PID)**—Acute or chronic inflammation in the pelvic cavity, particularly inflammation of the fallopian tubes (salpingitis) and its complications.

**Rupture**—A breaking apart of an organ or tissue.

**Salpingitis**—Inflammation of the fallopian tube.

**Tubal pregnancy**—Pregnancy in one of the fallopian tubes.

**Zygote**—The fertilized egg.

will perform a pelvic examination to locate the source of pain and to detect a mass in the abdomen.

Several laboratory tests of the patient's blood provide information for diagnosis. Measurement of the human chorionic gonadotropin (hCG) level in the patient's blood serum is the most useful laboratory test in the early stages. In a normal pregnancy, the level of this hormone doubles about every two days during the first 10 weeks. In an ectopic pregnancy, the rate of the increase is much slower and the low hCG for the stage of the pregnancy is a strong indication that the pregnancy is abnormal. (It could also represent a **miscarriage** in progress.) The level is usually tested several times over a period of days to determine whether or not it is increasing at a normal rate.

Progesterone levels in the blood are also measured. Lower than expected levels can indicate that the pregnancy is not normal.

An ultrasound examination may provide information about whether or not the pregnancy is ectopic. A device

called a transducer, which emits high frequency sound waves, is moved over the surface of the patient's abdomen or inserted into the vagina. The sound waves bounce off of the internal organs and create an image on a screen. The doctor should be able to see whether or not there is a fetus developing in the uterus after at least five weeks of gestation. Before that point, a normal pregnancy is too small to see.

A culdocentesis may also help confirm a diagnosis. In this procedure a needle is inserted into the space at the top of the vagina, behind the uterus and in front of the rectum. Blood in this area may indicate bleeding from a ruptured fallopian tube.

A **laparoscopy** will enable the doctor to see the patient's reproductive organs and examine an ectopic pregnancy. In this technique, a hollow tube with a light on one end is inserted through a small incision in the abdomen. Through this instrument the internal organs can be observed.

### Treatment

Ectopic pregnancy requires immediate treatment. The earlier the condition is treated, the better the chance to preserve the fallopian tube intact for future normal pregnancies.

### Medical

If the ectopic pregnancy is discovered in a very early stage of development, the drug methotrexate may be given. The best results are obtained when the pregnancy is less than six weeks old and the tubal mass is no more than 1.4 in (3.5 cm) in diameter. Methotrexate, which has been used successfully since 1987, works by inhibiting the growth of rapidly growing cells. (It is also used to treat some cancers.) Most side effects are mild and temporary, but the patient must be monitored after treatment. Usually the medication is injected into the muscle in a single dose, but may also be given intravenously or injected directly into the fallopian tube to dissolve the embryonic tissue. Methotrexate has also been used to treat ovarian, abdominal, and cervical pregnancies that are discovered in the early stages.

### Surgical

When a laparoscopy is done to visualize the ectopic pregnancy, the scope can be fitted with surgical tools and used to remove the ectopic mass immediately after it is identified. The affected fallopian tube can be repaired or removed as necessary. This procedure can be done without requiring the patient to stay in the hospital overnight.

When the pregnancy has ruptured, a surgical incision into the abdomen, or laparotomy, is performed to

stop the immediate loss of blood and to remove the embryo. This usually requires general anesthesia and a hospital stay. Every effort is made to preserve and repair the injured fallopian tube. However, if the fallopian tube has already ruptured, repair is extremely difficult and the tube is usually removed.

### Alternative treatment

Ectopic pregnancy was first described in the eleventh century and was a potentially fatal condition until the advent of surgery and blood transfusions in the early twentieth century. The sophisticated diagnostic tools and surgical procedures developed since the 1970s have equipped modern medicine with the tools to not only save a woman's life, but also to preserve her future fertility.

Although there are herbal remedies for the temporary relief of the common symptoms of **anxiety** and abdominal discomfort, prompt medical treatment is the only sure remedy for ectopic pregnancy.

### Prognosis

Ectopic pregnancies are the leading cause of pregnancy-related deaths in the first trimester and account for 9% of all pregnancy-related deaths in the United States. More than 1% of pregnancies are ectopic, and they are becoming more common. The reason for this increase is not clearly understood, though it is thought that the dramatic increase in **sexually transmitted diseases (STD)** is at least partly responsible.

The earlier an ectopic pregnancy is diagnosed and treated, the better the outcome. The chances of having a successful pregnancy are lower after an ectopic pregnancy, but depend on the extent of permanent fallopian tube damage. If the tube has been spared, chances are as high as 60%. The chances of a successful pregnancy after the removal of one tube are 40%.

### Prevention

Many forms of ectopic pregnancy cannot be prevented. However, tubal pregnancies, which make up the majority of ectopic pregnancies, may be prevented by avoiding conditions that cause damage to the fallopian tubes. Since half of all women who experience ectopic pregnancy have a history of PID, avoiding this infection or getting early diagnosis and treatment for sexually transmitted diseases will decrease the risk of a future problem.

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### ORGANIZATIONS

Resolve. 1310 Broadway, Somerville, MA 02144-1731. (617) 623-0744. <<http://www.resolve.org>>.

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Eczema see **Dermatitis**

ED see **Impotence**

## Edema

### Definition

Edema is a condition of abnormally large fluid volume in the circulatory system or in tissues between the body's cells (interstitial spaces).

### Description

Normally the body maintains a balance of fluid in tissues by ensuring that the same amount of water entering the body also leaves it. The circulatory system transports fluid within the body via its network of blood vessels. The fluid, which contains oxygen and nutrients needed by the cells, moves from the walls of the blood vessels into the body's tissues. After its nutrients are used up, fluid moves back into the blood vessels and returns to the heart. The lymphatic system (a network of channels in the body that carry lymph, a colorless fluid containing white blood cells to fight infection) also absorbs and transports this fluid. In edema, either too much fluid moves from the blood vessels into the tissues, or not enough fluid moves from the tissues back into the blood vessels. This fluid imbalance can cause mild to severe swelling in one or more parts of the body.

### Causes and symptoms

Many ordinary factors can upset the balance of fluid in the body to cause edema, including:

- Immobility. The leg muscles normally contract and compress blood vessels to promote blood flow with walking or running. When these muscles are not used,



**Gross lymphedema in the arm of an elderly woman following radiotherapy treatment for breast cancer.** (Photograph by Dr. P. Marazzi. Photo Researchers, Inc. Reproduced by permission.)

blood can collect in the veins, making it difficult for fluid to move from tissues back into the vessels.

- Heat. Warm temperatures cause the blood vessels to expand, making it easier for fluid to cross into surrounding tissues. High humidity also aggravates this situation.
- Medications. Certain drugs, such as steroids, hormone replacements, **nonsteroidal anti-inflammatory drugs** (NSAIDs), and some blood pressure medications may affect how fast fluid leaves blood vessels.
- Intake of salty foods. The body needs a constant concentration of salt in its tissues. When excess salt is taken in, the body dilutes it by retaining fluid.
- Menstruation and **pregnancy**. The changing levels of hormones affect the rate at which fluid enters and leaves the tissues.

Some medical conditions may also cause edema, including:

- **Heart failure.** When the heart is unable to maintain adequate blood flow throughout the circulatory system, the excess fluid pressure within the blood vessels can

cause shifts into the interstitial spaces. Left-sided heart failure can cause **pulmonary edema**, as fluid shifts into the lungs. The patient may develop rapid, shallow respirations, **shortness of breath**, and a **cough**. Right-sided heart failure can cause pitting edema, a swelling in the tissue under the skin of the lower legs and feet. Pressing this tissue with a finger tip leads to a noticeable momentary indentation.

- Kidney disease. The decrease in sodium and water excretion can result in fluid retention and overload.
- Thyroid or liver disease. These conditions can change the concentration of protein in the blood, affecting fluid movement in and out of the tissues. In advanced liver disease, the liver is enlarged and fluid may build-up in the abdomen.
- Malnutrition. Protein levels are decreased in the blood, and in an effort to maintain a balance of concentrations, fluid shifts out of the vessels and causes edema in tissue spaces.

Some conditions that may cause swelling in just one leg include:

- Blood clots. Clots can cause pooling of fluid and may be accompanied by discoloration and **pain**. In some instances, clots may cause no pain.
- Weakened veins. **Varicose veins**, or veins whose walls or valves are weak, can allow blood to pool in the legs. This is a common condition.
- Infection and inflammation. Infection in leg tissues can cause inflammation and increasing blood flow to the area. Inflammatory diseases, such as **gout** or arthritis, can also result in swelling.
- **Lymphedema**. Blocked lymph channels may be caused by infection, scar tissue, or hereditary conditions. Lymph that can't drain properly results in edema. Lymphedema may also occur after **cancer** treatments, when the lymph system is impaired by surgery, radiation, or **chemotherapy**.
- Tumor. Abnormal masses can compress leg vessels and lymph channels, affecting the rate of fluid movement.

Symptoms vary depending on the cause of edema. In general, weight gain, puffy eyelids, and swelling of the legs may occur as a result of excess fluid volume. Pulse rate and blood pressure may be elevated. Hand and neck veins may be observed as fuller.

## Diagnosis

Edema is a sign of an underlying problem, rather than a disease unto itself. A diagnostic explanation should be sought. Patient history and presenting symptoms, along with laboratory blood studies, if indicated, assist the health professional in determining the cause of the edema.

## Treatment

Treatment of edema is based on the cause. Simple steps to lessen fluid build-up may include:

- Reducing sodium intake. A high sodium level causes or aggravates fluid retention.
- Maintaining proper weight. Being overweight slows body fluid circulation and puts extra pressure on the veins.
- **Exercise**. Regular exercise stimulates circulation.
- Elevation of the legs. Placing the legs at least 12 in (30.5 cm) above the level of the heart for 10–15 minutes, three to four times a day, stimulates excess fluid re-entry into the circulatory system.
- Use of support stocking. Elastic stockings, available at most medical supply or drug stores, will compress the leg vessels, promoting circulation and decreasing pooling of fluid due to gravity.

## KEY TERMS

**Digitalis**—A naturally occurring compound used in the preparation of the medication, digoxin, prescribed to increase the heart rate and strengthen the force of the heart's contractions.

**Diuretics**—Medications used in the treatment of fluid overload, to promote excretion of sodium and water.

**Interstitial spaces**—Areas of the body occurring outside the vessels or organs, between the cells.

**Pitting edema**—A swelling in the tissue under the skin, resulting from fluid accumulation, that is measured by the depth of indentation made by finger pressure over a boney prominence.

• Massage. Massaging the body part can help to stimulate the release of excess fluids, but should be avoided if the patient has blood clots in the veins.

• Travel breaks. Sitting for long periods will increase swelling in the feet and ankles. Standing and/or walking at least every hour or two will help stimulate blood flow.

The three “Ds”—diuretics, digitalis, and diet—are frequently prescribed for medical conditions that result in excess fluid volume. **Diuretics** are medications that promote urination of sodium and water. Digoxin is a digitalis preparation that is sometimes needed to decrease heart rate and increase the strength of the heart's contractions. Dietary recommendations include less sodium in order to decrease fluid retention. Consideration of adequate protein intake is also made.

For patients with lymphedema, a combination of therapies may prove effective. Combined decongestive therapy includes the use of manual lymph drainage (MLD), compression bandaging, garments and pumps, and physical therapy. MLD involves the use of light massage of the subcutaneous tissue where the lymph vessels predominate. Massage begins in an area of the body trunk where there is normal lymph function and proceeds to areas of lymphatic insufficiency, in an effort to stimulate new drainage tract development. (MLD should not be used for patients with active cancer, deep vein clots, congestive heart failure, or cellulitis.) MLD sessions are followed by application of compression garments or pumps. Physical therapy is aimed at strengthening the affected limb and increasing joint mobility.

## Alternative treatment

Dietary changes, in addition to cutting back the amount of sodium eaten, may also help reduce edema. Foods that worsen edema, such as alcohol, **caffeine**, sugar, dairy products, soy sauce, animal protein, chocolate, olives, and pickles, should be avoided. Diuretic herbs can also help relieve edema. One of the best herbs for this purpose is dandelion (*Taraxacum mongolicum*), since, in addition to its diuretic action, it is a rich source of potassium. (Diuretics flush potassium from the body and it must be replaced to avoid potassium deficiency.) **Hydrotherapy** using daily contrast applications of hot and cold (either compresses or immersion) may also be helpful.

## Resources

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### ORGANIZATIONS

Lymphedema and Wound Care Clinic of Austin. 5750 Balcones Dr., Ste. 110, Austin, TX 78731. (512) 453-1930.

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Edrophonium test see **Tensilon test**

## Edwards' syndrome

### Definition

Edwards' syndrome is caused by an extra copy of chromosome 18. For this reason, it is also called trisomy 18 syndrome. The extra chromosome is lethal for most babies born with this condition. It causes major physical abnormalities and severe **mental retardation**, and very few children afflicted with this disease survive beyond a year.

### Description

Humans normally have 23 pairs of chromosomes. Chromosomes are numbered 1–22, and the 23rd pair is composed of the sex chromosomes, X and Y. A person inherits one set of 23 chromosomes from each parent. Occasionally, a genetic error occurs during egg or sperm cell formation. A child conceived with such an egg or sperm cell may inherit an incorrect number of chromosomes.

In the case of Edwards' syndrome, the child inherits three, rather than two, copies of chromosome 18. Trisomy

18 occurs in approximately one in every 3,000 newborns and affects girls more often than boys. Women older than their early thirties have a greater risk of conceiving a child with trisomy 18, but it can occur in younger women.

## Causes and symptoms

A third copy of chromosome 18 causes numerous abnormalities. Most children born with Edwards' syndrome appear weak and fragile, and they are often underweight. The head is unusually small and the back of the head is prominent. The ears are malformed and low-set, and the mouth and jaw are small. The baby may also have a cleft lip or cleft palate. Frequently, the hands are clenched into fists, and the index finger overlaps the other fingers. The child may have clubfeet and toes may be webbed or fused.

Numerous problems involving the internal organs may be present. Abnormalities often occur in the lungs and diaphragm (the muscle that controls breathing), and heart defects and blood vessel malformations are common. The child may also have malformed kidneys and abnormalities of the urogenital system.

## Diagnosis

Physical abnormalities point to Edwards' syndrome, but definitive diagnosis relies on karyotyping. Karyotyping involves drawing the baby's blood or bone marrow for a microscopic examination of the chromosomes. Using special stains and microscopy, individual chromosomes are identified, and the presence of an extra chromosome 18 is revealed.

Trisomy 18 can be detected before birth. If a pregnant woman is older than 35, has a family history of genetic abnormalities, has previously conceived a child with a genetic abnormality, or has suffered earlier miscarriages, she may undergo tests to determine whether her child carries genetic abnormalities. Potential tests include maternal serum analysis or screening, ultrasonography, amniocentesis, and **chorionic villus sampling**.

## Treatment

There is no cure for Edwards' syndrome. Since trisomy 18 babies frequently have major physical abnormalities, doctors and parents face difficult choices regarding treatment. Abnormalities can be treated to a certain degree with surgery, but extreme invasive procedures may not be in the best interests of an infant whose lifespan is measured in days or weeks. Medical therapy often consists of supportive care with the goal of making the infant comfortable, rather than prolonging life.

## KEY TERMS

**Aminocentesis**—A procedure in which a needle is inserted through a pregnant woman's abdomen and into her uterus to withdraw a small sample of amniotic fluid. The amniotic fluid can be examined for signs of disease or other problems afflicting the fetus.

**Chorionic villus sampling**—A medical test that is best done during weeks 10–12 of a pregnancy. The procedure involves inserting a needle into the placenta and withdrawing a small amount of the chorionic membrane for analysis.

**Chromosome**—A structure composed of deoxyribonucleic acid (DNA) contained within a cell's nucleus (center) in where genetic information is stored. Human have 23 pairs of chromosomes, each of which has recognizable characteristics (such as length and staining patterns) that allow individual chromosomes to be identified. Identification is assigned by number (1–22) or letter (X or Y).

**Karyotyping**—A laboratory test used to study an individual's chromosome make-up. Chromosomes are separated from cells, stained, and arranged in

order from largest to smallest so that their number and structure can be studied under a microscope.

**Maternal serum analyte screening**—A medical procedure in which a pregnant woman's blood is drawn and analyzed for the levels of certain hormones and proteins. These levels can indicate whether there may be an abnormality in the unborn child. This test is not a definitive indicator of a problem and is followed by more specific testing such as amniocentesis or chorionic villus sampling.

**Trisomy**—A condition in which a third copy of a chromosome is inherited. Normally only two copies should be inherited.

**Ultrasound**—A medical test that is also called ultrasonography. Sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. An ultrasound of a fetus at weeks 16–20 of a pregnancy can be used to determine structural abnormalities.

## Prognosis

Most children born with trisomy 18 die within their first year of life. The average lifespan is less than two months for 50% of the children, and 90–95% die before their first birthday. The 5–10% of children who survive their first year are severely mentally retarded. They need support to walk, and learning is limited. Verbal communication is also limited, but they can learn to recognize and interact with others.

## Prevention

Edwards' syndrome cannot be prevented.

## Resources

### BOOKS

Gardner, R. J. McKinlay, and Grant R. Sutherland. *Chromosome Abnormalities and Genetic Counseling*. New York: Oxford University Press, 1996.

Jones, Kenneth Lyons. *Smith's Recognizable Patterns of Human Malformation*. 5th ed. W. B. Saunders Co., 1997.

### ORGANIZATIONS

The Chromosome 18 Registry & Research Society. 6302 Fox Head, San Antonio, TX 78247. (210) 657-4968. <<http://www.chromosome18.org>>.

Support Organization for Trisomy 18, 13, and Related Disorders (SOFT). 2982 South Union St., Rochester, NY 14624. (800) 716-7638. <<http://www.trisomy.org>>.

Julia Barrett

EEG see **Electroencephalography**

Egyptian conjunctivitis see **Trachoma**

## Ehlers-Danlos syndrome

### Definition

The Ehlers-Danlos syndromes (EDS) refer to a group of inherited disorders that affect collagen structure and function. Genetic abnormalities in the manufacturing of collagen within the body affect connective tissues, causing them to be abnormally weak.

### Description

Collagen is a strong, fibrous protein that lends strength and elasticity to connective tissues such as the

skin, tendons, organ walls, cartilage, and blood vessels. Each of these connective tissues requires collagen tailored to meet its specific purposes. The many roles of collagen are reflected in the number of genes dedicated to its production. There are at least 28 genes in humans that encode at least 19 different types of collagen. Mutations in these genes can affect basic construction as well as the fine-tuned processing of the collagen.

EDS was originally described by Dr. Van Meekeren in 1682. Dr. Ehlers and Dr. Danlos further characterized the disease in 1901 and 1908, respectively. Today, according to the Ehlers-Danlos National Foundation, one in 5,000 to one in 10,000 people are affected by some form of EDS.

EDS is a group of genetic disorders that usually affects the skin, ligaments, joints, and blood vessels. Classification of EDS types was revised in 1997. The new classification involves categorizing the different forms of EDS into six major sub-types, including classical, hypermobility, vascular, kyphoscoliosis, arthrochalasia, and dermatosparaxis, and a collection of rare or poorly defined varieties. This new classification is simpler and based more on descriptions of the actual symptoms.

#### *Classical type*

Under the old classification system, EDS classical type was divided into two separate types: type I and type II. The major symptoms involved in EDS classical type are the skin and joints. The skin has a smooth, velvety texture and **bruises** easily. Affected individuals typically have extensive scarring, particularly at the knees, elbows, forehead, and chin. The joints are hyperextensible, giving a tendency towards dislocation of the hip, shoulder, elbow, knee, or clavicle. Due to decreased muscle tone, affected infants may experience a delay in reaching motor milestones. Children may have a tendency to develop hernias or other organ shifts within the abdomen. Sprains and partial or complete joint dislocations are also common. Symptoms can range from mild to severe. EDS classical type is inherited in an autosomal dominant manner.

There are three major clinical diagnostic criteria for EDS classical type. These include skin hyperextensibility, unusually wide scars, and joint hypermobility. At this time there is no definitive test for the diagnosis of classical EDS. Both DNA and biochemical studies have been used to help identify affected individuals. In some cases, a **skin biopsy** has been found to be useful in confirming a diagnosis. Unfortunately, these tests are not sensitive enough to identify all individuals with classical EDS. If there are multiple affected individuals in a family, it may be possible to perform prenatal diagnosis using a DNA information technique known as a linkage study.

#### *Hypermobility type*

Excessively loose joints are the hallmark of this EDS type, formerly known as EDS type III. Both large joints, such as the elbows and knees, and small joints, such as toes and fingers, are affected. Partial and total joint dislocations are common, and particularly involve the jaw, knee, and shoulder. Many individuals experience chronic limb and joint **pain**, although x rays of these joints appear normal. The skin may also bruise easily. **Osteoarthritis** is a common occurrence in adults. EDS hypermobility type is inherited in an autosomal dominant manner.

There are two major clinical diagnostic criteria for EDS hypermobility type. These include skin involvement (either hyperextensible skin or smooth and velvety skin) and generalized joint hypermobility. At this time there is no test for this form of EDS.

#### *Vascular type*

Formerly called EDS type IV, EDS vascular type is the most severe form. The connective tissue in the intestines, arteries, uterus, and other hollow organs may be unusually weak, leading to organ or blood vessel rupture. Such ruptures are most likely between ages 20 and 40, although they can occur any time, and may be life-threatening.

There is a classic facial appearance associated with EDS vascular type. Affected individuals tend to have large eyes, a thin pinched nose, thin lips, and a slim body. The skin is thin and translucent, with veins dramatically visible, particularly across the chest.

The large joints have normal stability, but small joints in the hands and feet are loose, showing hyperextensibility. The skin bruises easily. Other complications may include collapsed lungs, premature **aging** of the skin on the hands and feet, and ruptured arteries and veins. After surgery there tends to be poor wound healing, a complication that tends to be frequent and severe. **Pregnancy** also carries the risk complications. During and after pregnancy there is an increased risk of the uterus rupturing and of arterial bleeding. Due to the severe complications associated with EDS type IV, **death** usually occurs before the fifth decade. A study of 419 individuals with EDS vascular type, completed in 2000, found that the median survival rate was 48 years, with a range of six to 73 years. EDS vascular type is inherited in an autosomal dominant manner.

There are four major clinical diagnostic criteria for EDS vascular type. These include thin translucent skin, arterial/intestinal/uterine fragility or rupture, extensive bruising, and characteristic facial appearance. EDS vascular type is caused by a change in the gene COL3A1,

which codes for one of the collagen chains used to build Collage type III. Laboratory testing is available for this form of EDS. A skin biopsy may be used to demonstrate the structurally abnormal collagen. This type of biochemical test identifies more than 95% of individuals with EDS vascular type. Laboratory testing is recommended for individuals with two or more of the major criteria.

DNA analysis may also be used to identify the change within the COL3A1 gene. This information may be helpful for **genetic counseling** purposes. Prenatal testing is available for pregnancies in which an affected parent has been identified and their DNA mutation is known or their biochemical defect has been demonstrated.

#### *Kyphoscoliosis type*

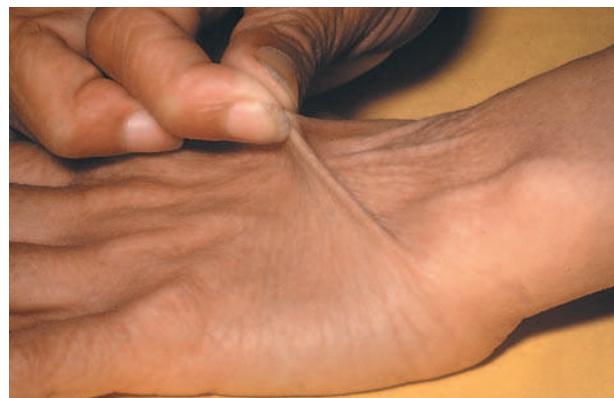
The major symptoms of kyphoscoliosis type, formerly called EDS type VI, are general joint looseness. At birth, the muscle tone is poor, and motor skill development is subsequently delayed. Also, infants with this type of EDS have an abnormal curvature of the spine (**scoliosis**). The scoliosis becomes progressively worse with age, with affected individuals usually unable to walk by age 20. The eyes and skin are fragile and easily damaged, and blood vessel involvement is a possibility. The bones may also be affected as demonstrated by a decrease in bone mass. Kyphoscoliosis type is inherited in an autosomal recessive manner.

There are four major clinical diagnostic criteria for EDS kyphoscoliosis type. These include generally loose joints, low muscle tone at birth, scoliosis at birth (which worsens with age), and a fragility of the eyes, which may give the white area of the eye a blue tint or cause the eye to rupture. This form of EDS is caused by a change in the PLOD gene on chromosome 1, which encodes the enzyme lysyl hydroxylase. A laboratory test is available in which urinary hydroxylysyl pyridinoline is measured. This test, performed on urine is extremely sensitive and specific for EDS kyphoscoliosis type. Laboratory testing is recommended for infants with three or more of the major diagnostic criteria.

Prenatal testing is available if a pregnancy is known to be at risk and an identified affected family member has had positive laboratory testing. An **amniocentesis** may be performed in which fetal cells are removed from the amniotic fluid and enzyme activity is measured.

#### *Arthrochalasia type*

Dislocation of the hip joint typically accompanies arthrochalasia type EDS, formerly called EDS type VIIB. Other joints are also unusually loose, leading to recurrent partial and total dislocations. The skin has a high degree of stretchability and bruises easily. Individuals with this



**Elasticity of the skin is one characteristic of this rare disorder.** (Photograph by Biophoto Associates, Photo Researchers, Inc. Reproduced by permission.)

type of EDS may also experience mildly diminished bone mass, scoliosis, and poor muscle tone. Arthrochalasia type is inherited in an autosomal dominant manner.

There are two major clinical diagnostic criteria for EDS arthrochalasia type. These include severe generalized joint hypermobility and bilateral hip dislocation present at birth. This form of EDS is caused by a change in either of two components of Collage type I, called proa1(I) type A and proa2(I) type B. A skin biopsy may be performed to demonstrate an abnormality in either components. Direct DNA testing is also available.

#### *Dermatosparaxis type*

Individuals with this type of EDS, once called type VIIC, have extremely fragile skin that bruises easily but does not scar excessively. The skin is soft and may sag, leading to an aged appearance even in young adults. Individuals may also experience hernias. Dermatosparaxis type is inherited in an autosomal recessive manner.

There are two major clinical diagnostic criteria for EDS dermatosparaxis type. These include severe skin fragility and sagging or aged appearing skin. This form of EDS is caused by a change in the enzyme called procollagen I N-terminal peptidase. A skin biopsy may be performed for a definitive diagnosis of Dermatosparaxis type.

#### *Other types*

There are several other forms of EDS that have not been as clearly defined as the aforementioned types. Forms of EDS within this category may present with soft, mildly stretchable skin, shortened bones, chronic **diarrhea**, joint hypermobility and dislocation, bladder rupture, or poor wound healing. Inheritance patterns within this group include X-linked recessive, autosomal dominant, and autosomal recessive.

## KEY TERMS

**Arthrochalasia**—Excessive looseness of the joints.

**Blood vessels**—General term for arteries, veins, and capillaries that transport blood throughout the body.

**Cartilage**—Supportive connective tissue that cushions bone at the joints or which connects muscle to bone.

**Collagen**—The main supportive protein of cartilage, connective tissue, tendon, skin, and bone.

**Connective tissue**—A group of tissues responsible for support throughout the body; includes cartilage, bone, fat, tissue underlying skin, and tissues that support organs, blood vessels, and nerves throughout the body.

**Dermatosparaxis**—Skin fragility caused by abnormal collagen.

**Hernia**—A rupture in the wall of a body cavity, through which an organ may protrude.

**Homeopathic**—A holistic and natural approach to healthcare.

**Hyperextensibility**—The ability to extend a joint beyond the normal range.

**Hypermobility**—Unusual flexibility of the joints, allowing them to be bent or moved beyond their normal range of motion.

**Joint dislocation**—The displacement of a bone.

**Kyphoscoliosis**—Abnormal front-to-back and side-to-side curvature of the spine.

**Ligament**—A type of connective tissue that connects bones or cartilage and provides support and strength to joints.

**Osteoarthritis**—A degenerative joint disease that causes pain and stiffness.

**Scoliosis**—An abnormal, side-to-side curvature of the spine.

**Tendon**—A strong connective tissue that connects muscle to bone.

**Uterus**—A muscular, hollow organ of the female reproductive tract. The uterus contains and nourishes the embryo and fetus from the time the fertilized egg is implanted until birth.

**Vascular**—Having to do with blood vessels.

## Causes and symptoms

There are numerous types of EDS, all caused by changes in one of several genes. The manner in which EDS is inherited depends on the specific gene involved. There are three patterns of inheritance for EDS: autosomal dominant, autosomal recessive, and X-linked (extremely rare).

Chromosomes are made up of hundreds of small units known as genes, which contain the genetic material necessary for an individual to develop and function. Humans have 46 chromosomes, which are matched into 23 pairs. Because chromosomes are inherited in pairs, each individual receives two copies of each chromosome and likewise two copies of each gene.

Changes or mutations in genes can cause genetic diseases in several different ways, many of which are represented within the spectrum of EDS. In autosomal dominant EDS, only one copy of a specific gene must be changed for a person to have EDS. In autosomal recessive EDS, both copies of a specific gene must be changed for a person to have EDS. If only one copy of an autosomal recessive EDS gene is changed the person is referred to as a carrier, meaning they do not have any of the signs or symptoms of the disease itself, but carry the possibili-

ty of passing on the disorder to a future child. In X-linked EDS a specific gene on the X chromosome must be changed. However, this affects males and females differently because males and females have a different number of X chromosomes.

As of 2001 the few X-linked forms of EDS fall under the category of X-linked recessive. As with autosomal recessive, this implies that both copies of a specific gene must be changed for a person to be affected. However, because males only have one X-chromosome, they are affected if an X-linked recessive EDS gene is changed on their single X-chromosome. That is, they are affected even though they have only one changed copy. On the other hand, that same gene must be changed on both of the X-chromosomes in a female for her to be affected.

Although there is much information regarding the changes in genes that cause EDS and their various inheritance patterns, the exact gene mutation for all types of EDS is not known.

## Diagnosis

Clinical symptoms such as extreme joint looseness and unusual skin qualities, along with family history, can

lead to a diagnosis of EDS. Specific tests, such as skin biopsies are available for diagnosis of certain types of EDS, including vascular, arthrochalasia, and dermatosparaxis types. A skin biopsy involves removing a small sample of skin and examining its microscopic structure. A urine test is available for the Kyphoscoliosis type.

Management of all types of EDS may include genetic counseling to help the affected individual and their family understand the disorder and its impact on other family members and future children.

If a couple has had a child diagnosed with EDS the chance that they will have another child with the same disorder depends on with what form of EDS the child has been diagnosed and if either parent is affected by the same disease or not.

Individuals diagnosed with an autosomal dominant form of EDS have a 50% chance of passing the same disorder on to a child in each pregnancy. Individuals diagnosed with an autosomal recessive form of EDS have an extremely low risk of having a child with the same disorder.

X-linked recessive EDS is accompanied by a slightly more complicated pattern of inheritance. If a father with an X-linked recessive form of EDS passes a copy of his X chromosome to his children, the sons will be unaffected and the daughters will be carriers. If a mother is a carrier for an X-linked recessive form of EDS, she may have affected or unaffected sons, or carrier or unaffected daughters, depending on the second sex chromosome inherited from the father.

Prenatal diagnosis is available for specific forms of EDS, including kyphoscoliosis type and vascular type. However, prenatal testing is only a possibility in these types if the underlying defect has been found in another family member.

## Treatment

Medical therapy relies on managing symptoms and trying to prevent further complications. There is no cure for EDS.

Braces may be prescribed to stabilize joints, although surgery is sometimes necessary to repair joint damage caused by repeated dislocations. Physical therapy teaches individuals how to strengthen muscles around joints and may help to prevent or limit damage. Elective surgery is discouraged due to the high possibility of complications.

### *Alternative treatment*

There are anecdotal reports that large daily doses 0.04–0.14 oz (1–4 g) of vitamin C may help decrease bruising and aid in wound healing. Constitutional home-

opathic treatment may be helpful in maintaining optimal health in persons with a diagnosis of EDS. An individual with EDS should discuss these types of therapies with their doctor before beginning them on their own. Therapy that does not require medical consultation involves protecting the skin with sunscreen and avoiding activities that place **stress** on the joints.

## Prognosis

The outlook for individuals with EDS depends on the type of EDS with which they have been diagnosed. Symptoms vary in severity, even within one sub-type, and the frequency of complications changes on an individual basis. Some individuals have negligible symptoms while others are severely restricted in their daily life. Extreme joint instability and scoliosis may limit a person's mobility. Most individuals will have a normal lifespan. However, those with blood vessel involvement, particularly those with EDS vascular type, have an increased risk of fatal complications.

EDS is a lifelong condition. Affected individuals may face social obstacles related to their disease on a daily basis. Some people with EDS have reported living with fears of significant and painful skin ruptures, becoming pregnant (especially those with EDS vascular type), their condition worsening, becoming unemployed due to physical and emotional burdens, and social stigmatization in general.

Constant bruises, skin **wounds**, and trips to the hospital take their toll on both affected children and their parents. Prior to diagnosis parents of children with EDS have found themselves under suspicion of **child abuse**.

Some people with EDS are not diagnosed until well into adulthood and, in the case of EDS vascular type, occasionally not until after death due to complications of the disorder. Not only may the diagnosis itself be devastating to the family, but in many cases other family members find out for the first time they are at risk for being affected.

Although individuals with EDS face significant challenges, it is important to remember that each person is unique with their own distinguished qualities and potential. Persons with EDS go on to have families, to have careers, and to be accomplished citizens, surmounting the challenges of their disease.

## Resources

### PERIODICALS

"Clinical and Genetic Features of Ehlers-Danlos Syndrome Type IV, the Vascular Type." *The New England Journal of Medicine* 342, no. 10 (2000).

"Ehlers-Danlos Syndromes: Revised Nosology, Villefranche, 1997." *American Journal of Medical Genetics* 77 (1998): 31–37.

"Living a Restricted Life with Ehlers-Danlos Syndrome." *International Journal of Nursing Studies* 37 (2000): 111–118.

#### ORGANIZATIONS

Ehlers-Danlos National Foundation. 6399 Wilshire Blvd., Ste 203, Los Angeles, CA 90048 (323) 651-3038. Fax: (323) 651-1366. <<http://www.ednf.org>>.

Ehlers-Danlos Support Group- UK. PO Box 335, Farnham, Surrey, GU10 1XJ. UK. <<http://www.atv.ndirect.co.uk>>.

#### OTHER

GeneClinics. <<http://www.geneclinics.org>>.

Java O. Solis, MS

## Ehrlichiosis

### Definition

Ehrlichiosis is a bacterial infection that is spread by ticks. Symptoms include **fever**, chills, **headache**, muscle aches, and tiredness.

### Description

Ehrlichiosis is a tick-borne disease caused by infection with *Ehrlichia* bacteria. Ticks are small, blood-sucking arachnids. Although some ticks carry disease-causing organisms, most do not. When an animal or person is bitten by a tick that carries bacteria, the bacteria are passed to that person or animal during the tick's feeding process. It is believed that the tick must remain attached to the person or animal for at least 24 hours to spread the infection.

There are two forms of ehrlichiosis in the United States; human monocytic ehrlichiosis and human granulocytic ehrlichiosis. Monocytic ehrlichiosis is caused by *Ehrlichia chaffeensis*, which is spread by the Lone Star tick, *Amblyomma americanum*. As of early 1998, about 400 cases of monocytic ehrlichiosis had been reported in 30 states, primarily in the southeastern and south central United States. The bacteria that causes granulocytic ehrlichiosis is not known, but suspected to be either *Ehrlichia equi* or *Ehrlichia phagocytophila*. Granulocytic ehrlichiosis is probably spread by the blacklegged tick *Ixodes scapularis* (which also spreads **Lyme disease**). About 100 cases of granulocytic ehrlichiosis have been reported in Connecticut, Massachusetts,

Rhode Island, Minnesota, New York, and Wisconsin.

### Causes and symptoms

Both forms of ehrlichiosis have similar symptoms, and the illnesses can range from mild to severe and life-threatening. Risk factors include old age and exposure to ticks through work or recreation. Symptoms occur seven to 21 days following a tick bite although patients may not recall being bitten. Fever, tiredness, headache, muscle aches, chills, loss of appetite, confusion, nausea, and vomiting are common to both diseases. A rash may occur.

### Diagnosis

Ehrlichiosis may be diagnosed and treated by doctors who specialize in blood diseases (hematologists) or an infectious disease specialist. Because ehrlichiosis is not very common and the symptoms are not unique, it may be misdiagnosed. A recent history of a tick bite is helpful in the diagnosis. Blood tests will be done to look for antibodies to *Ehrlichia*. Staining and microscopic examination of the blood sample may show *Ehrlichia* bacteria inside white blood cells. Another test, called polymerase chain reaction (PCR), is a very sensitive assay to detect bacteria in the blood sample, but it is not always available.

### Treatment

Antibiotic treatment should begin immediately if ehrlichiosis is suspected, even if laboratory results are not available. Treatment with either tetracycline (Sumycin, Achromycin V) or doxycycline (Monodox, Vibramycin) is recommended. Many patients with ehrlichiosis are admitted to the hospital for treatment.

### Prognosis

For otherwise healthy people, a full recovery is expected following treatment for ehrlichiosis. Elderly patients are at a higher risk for severe disease, which may be fatal. Serious complications include lung or gastrointestinal bleeding. Two to 10 patients out of 100 die from the disease.

### Prevention

The only prevention for ehrlichiosis is to minimize exposure to ticks by staying on the trail when walking through the woods, avoiding tall grasses, wearing long sleeves and tucking pant legs into socks, wearing insect

## KEY TERMS

**Tick-borne disease**—A disease that is spread to animals by the bite of an infected tick.

repellent, and checking for ticks after an outing. Remove a tick as soon as possible by grasping the tick with tweezers and gently pulling.

### Resources

#### BOOKS

McDade, Joseph E., and James G. Olsen. "Ehrlichiosis, Q Fever, Typhus, Rickettsialpox, and Other Rickettsioses." In *Infectious Diseases*. 2nd ed. Philadelphia: W. B. Saunders Co., 1998.

#### OTHER

*Mayo Clinic Online*. 5 Mar. 1998 <<http://www.mayohealth.org>>.

Belinda Rowland, PhD

EKG see **Electrocardiography**

Elder abuse see **Abuse**

ty of injury is determined primarily by the voltage, low voltage can be just as dangerous as high voltage under the right circumstances. People have been killed by shocks of just 50 volts.

How electric shocks affect the skin is determined by the skin's resistance, which in turn is dependent upon the wetness, thickness, and cleanliness of the skin. Thin or wet skin is much less resistant than thick or dry skin. When skin resistance is low, the current may cause little or no skin damage but severely burn internal organs and tissues. Conversely, high skin resistance can produce severe skin **burns** but prevent the current from entering the body.

The nervous system (the brain, spinal cord, and nerves) is particularly vulnerable to injury. In fact, neurological problems are the most common kind of nonlethal harm suffered by electric shock victims. Some neurological damage is minor and clears up on its own or with medical treatment, but some is severe and permanent. Neurological problems may be apparent immediately after the accident, or gradually develop over a period of up to three years.

Damage to the respiratory and cardiovascular systems is most acute at the moment of injury. Electric shocks can paralyze the respiratory system or disrupt heart action, causing instant death. Also at risk are the smaller veins and arteries, which dissipate heat less easily than the larger blood vessels and can develop blood clots. Damage to the smaller vessels is probably one reason why **amputation** is often required following high-voltage injuries.

Many other sorts of injuries are possible after an electric shock, including **cataracts**, kidney failure, and substantial destruction of muscle tissue. The victim may suffer a fall or be hit by debris from exploding equipment. An electric arc may set clothing or nearby flammable substances on fire. Strong shocks are often accompanied by violent muscle spasms that can break and dislocate bones. These spasms can also freeze the victim in place and prevent him or her from breaking away from the source of the current.

### Diagnosis

Diagnosis relies on gathering information about the circumstances of the accident, a thorough **physical examination**, and monitoring of cardiovascular and kidney activity. The victim's neurological condition can fluctuate rapidly and requires close observation. A computed tomography scan (CT scan) or **magnetic resonance imaging** (MRI) may be necessary to check for brain injury.

### Treatment

When an electric shock accident happens at home or in the workplace, the main power should immediately be

## Electric shock injuries

### Definition

Electric shock injuries are caused by lightning or electric current from a mechanical source passing through the body.

### Description

Electric shocks are responsible for about 1,000 deaths in the United States each year, or about 1% of all accidental deaths.

### Causes and symptoms

The severity of injury depends on the current's pressure (voltage), the amount of current (amperage), the type of current (direct vs. alternating), the body's resistance to the current, the current's path through the body, and how long the body remains in contact with the current. The interplay of these factors can produce effects ranging from barely noticeable tingling to instant **death**; every part of the body is vulnerable. Although the severi-

## KEY TERMS

**Antibiotics**—Substances used against microorganisms that cause infection.

**Cataract**—Clouding of the lens of the eye or its capsule (surrounding membrane).

**Computed tomography scan (CT scan)**—A process that uses x rays to create three-dimensional images of structures inside the body.

**Electrolytes**—Substances that conduct electric current within the body and are essential for sustaining life.

**Magnetic resonance imaging (MRI)**—The use of electromagnetic energy to create images of structures inside the body.

**Skin grafting**—A technique in which a piece of healthy skin from the patient's body (or a donor's) is used to cover another part of the patient's body that has lost its skin.

shut off. If that cannot be done, and current is still flowing through the victim, the alternative is to stand on a dry, non-conducting surface such as a folded newspaper, flattened cardboard carton, or plastic or rubber mat and use a non-conducting object such as a wooden broomstick (never a damp or metallic object) to push the victim away from the source of the current. The victim and the source of the current must not be touched while the current is still flowing, for this can electrocute the rescuer. Emergency medical help should be summoned as quickly as possible. People who are trained to perform **cardiopulmonary resuscitation (CPR)** should, if appropriate, begin first aid while waiting for emergency medical help to arrive.

Burn victims usually require treatment at a burn center. Fluid replacement therapy is necessary to restore lost fluids and electrolytes. Severely injured tissue is repaired surgically, which can involve **skin grafting** or amputation. **Antibiotics** and antibacterial creams are used to prevent infection. Victims may also require treatment for kidney failure. Following surgery, physical therapy to facilitate recovery, and psychological counseling to cope with disfigurement, may be necessary.

### Prognosis

Electric shocks cause death in 3–15% of cases. Many survivors require amputation or are disfigured by their burns. Injuries from household appliances and other low-voltage sources are less likely to produce extreme damage.

## Prevention

Parents and other adults need to be alert to possible electric dangers in the home. Damaged electric appliances, wiring, cords, and plugs should be repaired or replaced. Electrical repairs should be attempted only by people with the proper training. Hair dryers, radios, and other electric appliances should never be used in the bathroom or anywhere else they might accidentally come in contact with water. Young children need to be kept away from electric appliances and should be taught about the dangers of electricity as soon as they are old enough. Electric outlets require safety covers in homes with young children.

During thunderstorms, people should go indoors immediately, even if no rain is falling, and boaters should return to shore as rapidly as possible. People who cannot reach indoor shelter should move away from metallic objects such as golf clubs and fishing rods and lie down in low-ground areas. Standing or lying under or next to tall or metallic structures is unsafe. An automobile is appropriate cover, as long as the radio is off. Telephones, computers, hair dryers, and other appliances that can act as conduits for lightning should not be used during thunderstorms.

## Resources

### BOOKS

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Howard Baker

## Electrical nerve stimulation

### Definition

Electrical nerve stimulation, also called transcutaneous electrical nerve stimulation (TENS), is a noninvasive, drug-free **pain management** technique. By sending electrical signals to underlying nerves, the battery-powered TENS device can relieve a wide range of chronic and acute **pain**.

### Purpose

TENS is used to relieve pain caused by a variety of chronic conditions, including:

- neck and lower back pain
- headache/migraine
- arthritis
- post-herpetic **neuralgia** (lingering chronic pain after an attack of **shingles**)

- sciatica (pain radiating from lower back, through the legs, to the foot)
- temporomandibular joint pain
- osteoarthritis
- amputation (phantom limb)
- fibromyalgia (a condition causing aching and stiffness throughout the body)

The device is also effective against short-term pain, such as:

- shingles (painful skin eruptions along the nerves)
- bursitis (inflammation of tissue surrounding a joint)
- childbirth
- post-surgical pain
- fractures
- muscle and joint pain
- sports injuries
- menstrual cramps

### Precautions

Because TENS may interfere with pacemaker function, patients with **pacemakers** should consult a cardiologist before using a TENS unit. Patients should also avoid electrical stimulation in the front of the neck, which can be hazardous. The safety of the device during **pregnancy** has not been established.

TENS doesn't cure any condition; it simply eases pain. Patients who are not sure what is causing their pain should consult a physician before using TENS.

### Description

The TENS device is a small battery-powered stimulator that produces low-intensity electrical signals through electrodes on or near a painful area, producing a tingling sensation that reduces pain. There is no dosage limitation, and the patient controls the amount of pain relief.

Some experts believe TENS works by blocking pain signals in the spinal cord, or by delivering electrical impulses to underlying nerve fibers that lessen the experience of pain. Others suspect that the electrical stimulation triggers the release of natural painkillers in the body.

Patients can rent a TENS unit before buying one, to see if it is effective against their pain.

### Preparation

After TENS has been prescribed, a doctor will refer the patient to a TENS specialist, who will explain how to use the machine. The specialist works with the patient to determine the settings and electrode placements for the best pain relief.

## KEY TERMS

**Fibromyalgia**—A condition characterized by aching and stiffness, fatigue and poor sleep, as well as tenderness at various sites on the body.

**Osteoarthritis**—A painful joint disease aggravated by mechanical stress.

**Phantom limb**—The perception that a limb is present (and throbbing with pain) after it has been amputated.

**Post-herpetic neuralgia**—Lingering pain that can last for years after an attack of shingles.

**Sciatica**—Pain that radiates along the sciatic nerve, extending from the buttock down the leg to the foot.

**Temporomandibular joint pain (TMJ)**—Pain and other symptoms affecting the head, jaw, and face that are caused when the jaw joints and muscles controlling them don't work together correctly.

### Risks

TENS is nonaddictive and completely safe. The only side effect may be a slight skin irritation or redness in some people, which can be prevented by using different gels or electrodes.

### Normal results

The amount of relief a person gets using TENS depends on the underlying cause of the pain, a person's mental state, and whether or not medication is also used. At least one study found that both a real TENS machine and a placebo were equally effective in reducing pain. This suggests that at least part of its effectiveness may be due to the patient's belief in its ability to ease pain.

Carol A. Turkington

## Electrical stimulation of the brain

### Definition

Electrical stimulation of the brain (ESB) is a relatively new technique used to treat chronic **pain** and

## KEY TERMS

**Infarction**—A sudden insufficiency of local blood supply.

**Neuralgia**—Pain extending along one or more nerves.

**Neuropathy**—A functional disturbance or change in the nervous system.

**Parkinson disease**—A chronic neurological illness that causes tremors, stiffness, and difficulty in moving and walking.

**tremors** associated with **Parkinson disease**. ESB is administered by passing an electrical current through an electrode implanted in the brain.

### Purpose

While the implantation of electrodes in the brain is used to treat or diagnose several disorders, the term ESB is limited here to the treatment of tremors, and as a **pain management** tool for patients suffering from back problems and other chronic injuries and illnesses.

### Precautions

An ESB tremor control device, used in treating Parkinson patients, may interfere with or be affected by cardiac **pacemakers** and other medical equipment. As a result, patients with other implanted medical equipment may not be good candidates for the therapy.

### Description

Electrical stimulation of the brain, or deep brain stimulation, is effective in treating tremors in up to 88% of Parkinson disease patients. An electrode is implanted into the thalamus (part of the brain) of the patient, and attached to an electric pulse generator via an extension wire. The pulse generator is implanted into the patient's pectoral, or chest area, and the extension wire is tunneled under the skin. The pulse generator sends out intermittent electrical stimulation to the electrode in the thalamus, which inhibits or partially relieves the tremor. The generator can be turned on and off with a magnet, and needs to be replaced every three to five years.

Similar methods have been used to treat chronic pain that responded unfavorably to conventional therapies. A remote transmitter allows these patients to trigger elec-

tric stimulation to relieve their symptoms on an as-needed basis. Patients with failed back syndrome, trigeminal neuropathy (pertaining to the fifth cranial nerve), and **peripheral neuropathy** fared well for pain control with this treatment, while patients with **spinal cord injury** and postherpetic neuralgia (pain along the nerves following herpes) did poorly.

### Preparation

The patient should be free of any type of infection before undergoing an ESB procedure. He or she may be advised to discontinue any medication for a prescribed period of time before surgery.

### Aftercare

After neurosurgery, patients should undergo regular head dressing changes, minimize exposure to others, and practice good personal hygiene in order to prevent a brain infection. The head may also be kept elevated for a prescribed period of time in order to decrease swelling of the brain.

### Risks

The implantation of electrodes into the brain carries risks of hemorrhage, infarction, infection, and cerebral **edema**. These complications could cause irreversible neurological damage.

Patients with an implanted ESB tremor control device may experience headaches, disequilibrium (a disturbance of the sense of balance), burning or tingling of the skin, or partial **paralysis**.

### Normal results

ESB is effective in pain control for specific conditions. It can provide long-term pain relief with few side effects or complications.

For the control of tremors a deep brain stimulator does provide some relief. It is recommended for patients with tremors severe enough to affect their quality of life.

### Resources

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Oostra, K., M. Van Laere, and B. Scheirlinck. "Use of Electrical Stimulation in Brain-Injured Patients: A Case Report." *Brain Injury* 11, no. 10 (Oct. 1997): 761-4.

#### OTHER

*The Parkinson's Web.* <<http://pdweb.mgh.harvard.edu>>. University of Southern California. *The ANGEL Neurosurgical Information Resource.* <<http://www.usc.edu/hsc/neuro/surgery/angel.html>>.

Paula Anne Ford-Martin



An EKG strip indicating atrial flutter. (Custom Medical Stock Photo. Reproduced by permission.)

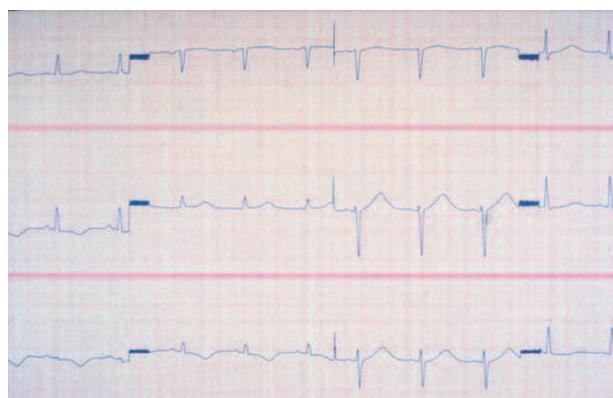
## Electrocardiography

### Definition

Electrocardiography is a commonly used, non-invasive procedure for recording electrical changes in the heart. The record, which is called an electrocardiogram (ECG or EKG), shows the series of waves that relate to the electrical impulses which occur during each beat of the heart. The results are printed on paper or displayed on a monitor. The waves in a normal record are named P, Q, R, S, and T and follow in alphabetical order. The number of waves may vary, and other waves may be present.

### Purpose

Electrocardiography is a starting point for detecting many cardiac problems. It is used routinely in physical examinations and for monitoring the patient's condition during and after surgery, as well as during intensive care. It is the basic measurement used for tests such as exercise tolerance. It is used to evaluate causes of symptoms such as chest pain, shortness of breath, and palpitations.



This EKG strip shows evidence of Wolff-Parkinson-White syndrome. (Custom Medical Stock Photo. Reproduced by permission.)

### Precautions

No special precautions are required.

### Description

The patient disrobes from the waist up, and electrodes (tiny wires in adhesive pads) are applied to specific sites on the arms, legs, and chest. When attached, the electrodes are called leads; three to 12 leads may be employed.

Muscle movement may interfere with the recording, which lasts for several beats of the heart. In cases where rhythm disturbances are suspected to be infrequent, the patient may wear a small Holter monitor in order to record continuously over a 24-hour period; this is known as ambulatory monitoring.

### Preparation

The skin is cleaned to obtain good electrical contact at the electrode positions.

### Aftercare

To avoid skin irritation from the salty gel used to obtain good electrical contact, the skin should be thoroughly cleaned after removal of the electrodes.

### Risks

No complications from this procedure have been observed.

### Normal results

When the heart is operating normally, each part contracts in a specific order. Contraction of the muscle is



**A patient undergoing electrocardiography.** (Russell Curtis, Photo Researchers. Reproduced by permission.)

triggered by an electrical impulse. These electrical impulses travel through specialized cells that form a conduction system. Following this pathway ensures that contractions will occur in a coordinated manner.

When the presence of all waves is observed in the electrocardiogram and these waves follow the order defined alphabetically, the heart is said to show a normal sinus rhythm, and impulses may be assumed to be following the regular conduction pathway.

The heart is described as showing arrhythmia or dysrhythmia when time intervals between waves, the order, or the number of waves do not fit this pattern. Other features that may be altered include the direction of wave deflection and wave widths.

In the normal heart, electrical impulses—at a rate of 60–100 times per minute—originate in the sinus node. The sinus node is located in the first chamber, known as the right atrium, where blood re-enters the heart. After traveling down to the junction between the upper and lower chambers, the signal stimulates the atrioventricular node. From here, after a delay, it passes by specialized routes through the lower chambers or ventricles. In many disease states, the passage of the electrical impulse can be interrupted in a variety of ways, causing the heart to perform less efficiently.

### Abnormal results

Special training is required for interpretation of the electrocardiogram. To summarize the features used in interpretations in the simplest manner, the P wave of the electrocardiogram is associated with the contraction of the atria. The QRS series of waves, or QRS complex, is associated with ventricular contraction, with the T wave coming after the contraction. Finally, the P-Q or P-R interval gives a value for the time taken for the electrical impulse to travel from the atria to the ventricle (normally less than 0.2 sec).

## KEY TERMS

**Ambulatory monitoring**—ECG recording over a prolonged period during which the patient can move around.

**Arrhythmia or dysrhythmia**—Abnormal rhythm in hearts that contract in an irregular way.

**ECG or EKG**—A record of the waves that relate to the electrical impulses produced at each beat of the heart.

**Electrodes**—Tiny wires in adhesive pads that are applied to the body for ECG measurement.

**Fibrillation**—Rapid, uncoordinated contractions of the upper or the lower chambers of the heart.

**Lead**—Name given the electrode when it is attached to the skin.

The cause of dysrhythmia is ectopic beats. Ectopic beats are premature heart beats that arise from a site other than the sinus node—commonly from the atria, atrioventricular node, or the ventricle. When these dysrhythmias are only occasional, they may produce no symptoms, or a feeling of the heart turning over or “flip-flopping” may be experienced. These occasional dysrhythmias are common in healthy people, but they also can be an indication of heart disease.

The varied sources of dysrhythmias provide a wide range of alterations in the form of the electrocardiogram. Ectopic beats that start in the ventricle display an abnormal QRS complex. This can indicate disease associated with insufficient blood supply to the muscle (myocardial **ischemia**). Multiple ectopic sites lead to rapid and uncoordinated contractions of the atria or ventricles. This condition is known as fibrillation. In atrial fibrillation, P waves are absent, and the QRS complex appears at erratic intervals, or “irregularly irregular.”

When the atrial impulse fails to reach the ventricle, a condition known as **heart block** results. If this is partial, the P-R interval (the time for the impulse to reach the ventricle) is prolonged. If complete, the ventricles beat independently of the atria at about 40 beats per minute, and the QRS complex is mostly dissociated from the P wave.

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Van Riper, Sharon, and Ann Luciano. "Basic Cardiac Arrhythmias: A Review for Postanesthesia Care Unit Nurses." *Journal of Postanesthesia Nursing* (Feb. 1994): 2-13.

#### ORGANIZATIONS

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

Alison M. Grant

## Electroconvulsive therapy

### Definition

Electroconvulsive therapy (ECT) is a medical treatment for severe mental illness in which a small, carefully controlled amount of electricity is introduced into the brain. This electrical stimulation, used in conjunction with anesthesia and muscle relaxant medications, produces a mild generalized seizure or convulsion. While used to treat a variety of psychiatric disorders, it is most effective in the treatment of severe depression, and provides the most rapid relief currently available for this illness.

### Purpose

The purpose of electroconvulsive therapy is to provide relief from the signs and symptoms of mental illnesses such as severe depression, **mania**, and **schizophrenia**. ECT is indicated when patients need rapid improvement because they are suicidal, self-injurious, refuse to eat or drink, cannot or will not take medication as prescribed, or present some other danger to themselves. Antidepressant medications, while effective in many cases, may take two to six weeks to produce a therapeutic effect. Antipsychotic medications used to treat mania and schizophrenia have many uncomfortable and sometimes dangerous side effects, limiting their use. In addition, some patients develop **allergies** and therefore are unable to take their medicine.

### Precautions

The most common risks associated with ECT are disturbances in heart rhythm. Broken or dislocated bones occur very rarely.

### Description

The treatment of severe mental illness, such as schizophrenia, using electroconvulsive therapy was introduced in 1938 by two Italian doctors named Cerletti and Bini. In those days many doctors believed that convulsions were incompatible with schizophrenia since, according to their observations, this disease rarely occurred in individuals suffering from epilepsy. They concluded, therefore, that if convulsions could be artificially produced in patients with schizophrenia, the illness could be cured. Some doctors were already using a variety of chemicals to produce seizures, but many of their patients died or suffered severe injuries because the strength of the convulsions could not be well controlled.

Electroconvulsive therapy is among the most controversial of all procedures used to treat mental illness. When it was first introduced, many people were frightened simply because it was called "shock treatment." Many assumed the procedure would be painful, others thought it was a form of electrocution, and still others believed it would cause brain damage. Unfortunately, unfavorable publicity in newspapers, magazines, and movies added to these fears.

Indeed, in those early years, patients and families were rarely educated by doctors and nurses regarding this or other forms of psychiatric treatment. In addition, no anesthesia or **muscle relaxants** were used. As a result, patients had violent seizures, and even though they did not remember them, the procedure itself was frightening.

The way these treatments are given today is very different from the procedures used in the past. Currently, ECT is offered on both an inpatient and outpatient basis. Hospitals have specially equipped rooms with oxygen, suction, and **cardiopulmonary resuscitation (CPR)** in order to deal with the rare emergency.

The treatment is carried out as follows: approximately 30 minutes before the scheduled treatment time, the patient may receive an injection of a medication (such as atropine) that keeps the pulse rate from decreasing too much during the convulsion. Next, the patient is placed on a cot and hooked up to a machine that automatically takes and displays vital signs (temperature, pulse, respiration, and blood pressure) on a television-like monitor. A mild anesthetic is then injected into a vein, followed by a medication (such as Anectine) that relaxes all of the muscles in the body so that the seizure is mild, and the risk of broken bones is virtually eliminated.

When the patient is both relaxed and asleep, an airway is placed in the mouth to aid with breathing. Electrodes are placed on the sides of the head in the temple areas. An electric current is passed through the brain by means of a machine specifically designed for this pur-

pose. The usual dose of electricity is 70–150 volts for 0.1–0.5 seconds. In the first stage of the seizure (tonic phase), the muscles in the body that have not been paralyzed by medication contract for a period of five to 15 seconds. This is followed by the second stage (clonic phase) that is characterized by twitching movements, usually visible only in the toes or in a non-paralyzed arm or leg. These are caused by alternating contraction and relaxation of these same muscles. This stage lasts approximately 10–60 seconds. The entire procedure, from beginning to end, lasts about 30 minutes.

The total number of treatments a patient will receive depends upon many factors such as age, diagnosis, the history of illness, family support, and response to therapy. Patients with depression, for example, usually require six to 12 treatments. Treatments are usually administered every other day, three times a week.

The electrodes may be placed on both sides of the head (bilateral) or one side (unilateral). While bilateral ECT appears to be somewhat more effective, unilateral ECT is preferred for individuals who experience prolonged confusion or forgetfulness following treatment. Many doctors begin treatment with unilateral ECT, then change to bilateral if the patient is not improving.

Post-treatment confusion and forgetfulness are common, though disturbing symptoms associated with ECT. Doctors and nurses must be patient and supportive by providing patients with factual information about recovery. Elderly patients, for example, may become increasingly confused and forgetful as the treatments continue. These symptoms usually subside with time, but a small minority of patients state that they have never fully recovered from these effects.

With the introduction of antipsychotics in the 1950s, the use of ECT became less frequent. These new medications provided relief for untold thousands of patients who suffered greatly from their illness. However, there are a number of side effects associated with these drugs, some of which are irreversible. Another drawback is that some medications do not produce a therapeutic effect for two to six weeks. During this time the patient may present a danger to himself or others. In addition, there are patients who do not respond to medicine or who have severe allergic reactions. For these individuals, ECT may be the only treatment that will help.

## Preparation

Patients and relatives are prepared for ECT by being shown video tapes that explain both the procedure and the risks involved. The physician then answers any questions these individuals may have, and the patient is asked to sign an "Informed Consent Form." This gives the doctor and the hospital permission to administer the treatment.

## KEY TERMS

**Mania**—A mood disorder in which a person experiences prolonged elation or irritability characterized by overactivity that can lead to exhaustion and medical emergencies.

**Relapse**—A return of the signs and symptoms of an illness.

**Schizophrenia**—A severe mental illness in which a person has difficulty distinguishing what is real from what is not real. It is often characterized by hallucinations, delusions, and withdrawal from people and social activities.

Once the form is signed, the doctor performs a complete **physical examination**, and orders a number of tests that can help identify any potential problem. These tests may include a **chest x ray**, an electrocardiogram (ECG), **urinalysis**, spinal x ray, brain wave (EEG), and complete **blood count** (CBC).

Some medications, such as lithium and a type of anti-depressant known as **monoamine oxidase inhibitors**, should be discontinued for some time before treatment. Patients are instructed not to eat or drink for at least eight hours prior to the procedure in order to reduce the possibility of vomiting and **choking**.

## Aftercare

After the treatment, patients are moved to a recovery area. Vital signs are recorded every five minutes until the patient is fully awake, which may take 15–30 minutes. Some initial confusion may be present but usually disappears in a matter of minutes. There may be complaints of **headache**, muscle **pain**, or back pain. Such discomfort is quickly relieved by mild medications such as **aspirin**.

## Risks

Advanced medical technology has substantially reduced the complications associated with ECT. These include slow heart beat (bradycardia), rapid heart beat (tachycardia), memory loss, and confusion. Persons at high risk for ECT include those with recent **heart attack**, uncontrolled blood pressure, brain tumors, and previous spinal injuries.

## Normal results

ECT often produces dramatic improvement in the signs and symptoms of major depression, especially in

elderly individuals, sometimes during the first week of treatment. While it is estimated that 50% of these patients will experience a future return of symptoms, the prognosis for each episode of illness is good. Mania also often responds well to treatment. The picture is not as bright for schizophrenia, which is more difficult to treat and is characterized by frequent relapses.

A few patients are placed on maintenance ECT. This means they return to the hospital every one to two months, as needed, for an additional treatment. These individuals are thus able to keep their illness under control and lead a normal and productive life.

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### ORGANIZATIONS

- National Institutes of Health. 5600 Fishers Lane. Room 7CO2, Rockville, MD 20857. (301) 496-4000. <<http://www.nih.gov>>.

Donald G. Barstow, RN

**Electrocution** see **Electric shock injuries**

## Electroencephalography

### Definition

Electroencephalography, or EEG, is a neurological test that uses an electronic monitoring device to measure and record electrical activity in the brain.

### Purpose

The EEG is a key tool in the diagnosis and management of epilepsy and other seizure disorders. It is also used to assist in the diagnosis of brain damage and disease (e.g., **stroke**, tumors, **encephalitis**), **mental retardation**, **sleep disorders**, degenerative diseases such as **Alzheimer's disease** and **Parkinson's disease**, and certain mental disorders (e.g., **alcoholism**, **schizophrenia**, **autism**).

An EEG may also be used to monitor brain activity during surgery and to determine brain **death**.

### Precautions

Electroencephalography should be administered and interpreted by a trained medical professional only. Data from an EEG is only one element of a complete medical and/or psychological patient assessment, and should never be used alone as the sole basis for a diagnosis.

### Description

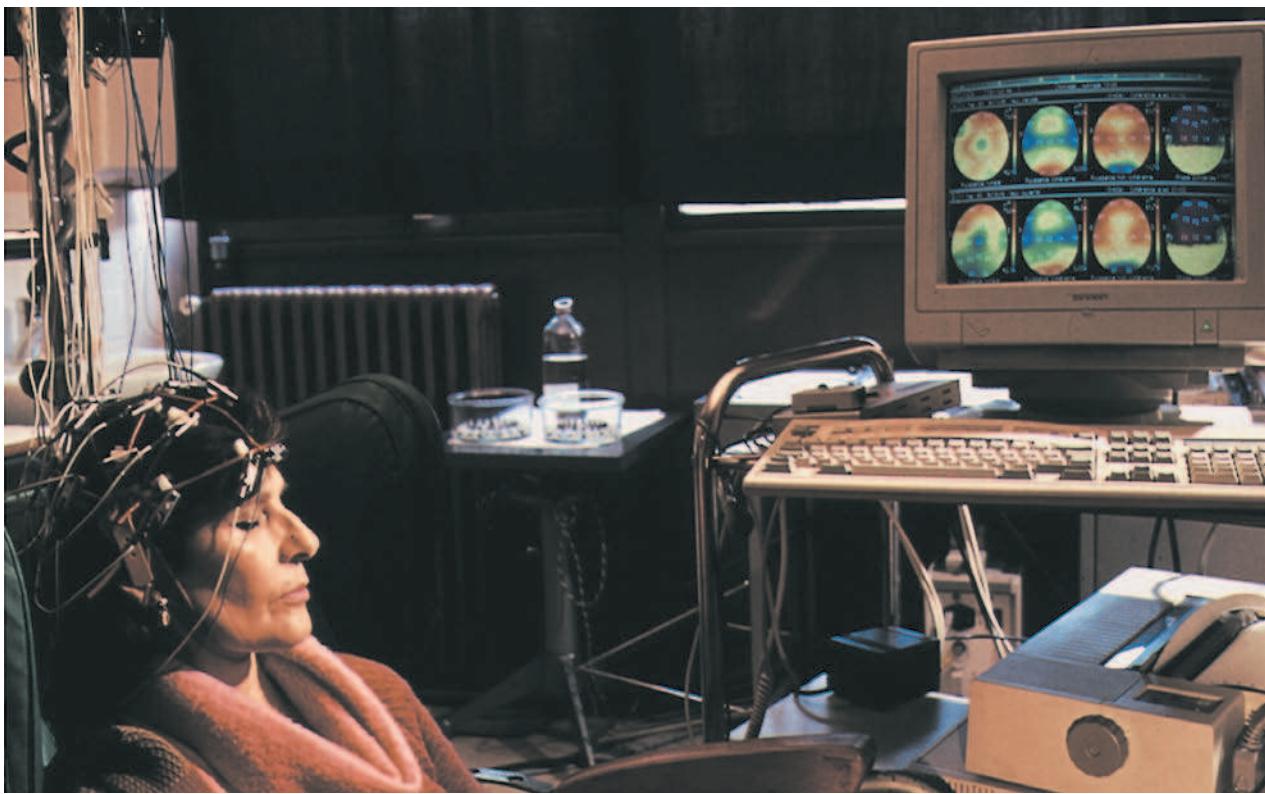
Before the EEG begins, a nurse or technician attaches approximately 16–20 electrodes to the patient's scalp with a conductive, washable paste. Depending on the purpose for the EEG, implantable or invasive electrodes are occasionally used. Implantable electrodes include sphenoidal electrodes, which are fine wires inserted under the zygomatic arch, or cheekbone; and depth electrodes, which are surgically-implanted into the brain. The EEG electrodes are painless, and are used to measure the electrical activity in various regions of the brain.

For the test, the patient lies on a bed, padded table, or comfortable chair and is asked to relax and remain still during the EEG testing period. An EEG usually takes no more than one hour. During the test procedure, the patient may be asked to breathe slowly or quickly; visual stimuli such as flashing lights or a patterned board may be used to stimulate certain types of brain activity. Throughout the procedure, the electroencephalograph machine makes a continuous graphic record of the patient's brain activity, or brainwaves, on a long strip of recording paper or on a computer screen. This graphic record is called an electroencephalogram.

The sleep EEG uses the same equipment and procedures as a regular EEG. Patients undergoing a sleep EEG are encouraged to fall asleep completely rather than just relax. They are typically provided a bed and a quiet room conducive to sleep. A sleep EEG lasts up to three hours.

In an ambulatory EEG, patients are hooked up to a portable cassette recorder. They then go about their normal activities, and take their normal rest and sleep for a period of up to 24 hours. During this period, the patient and patient's family record any symptoms or abnormal behaviors, which can later be correlated with the EEG to see if they represent seizures.

Many insurance plans provide reimbursement for EEG testing. Costs for an EEG range from \$100 to more than \$500, depending on the purpose and type of test (i.e., asleep or awake, and invasive or non-invasive electrodes). Because coverage may be dependent on the dis-



This woman is undergoing an electroencephalogram (EEG) to diagnose Alzheimer's disease. On the computer screen at the right are the colored scans of the electrical activity in her brain. Alzheimer's patients show a specific abnormality in their EEGs. (Photograph by Catherine Pouedras, Photo Researchers, Inc. Reproduced by permission.)

order or illness the EEG is evaluating, patients should check with their individual insurance plan.

### Preparation

Full instructions should be given to EEG patients when they schedule their test. Typically, individuals on medications that affect the central nervous system, such as anticonvulsants, stimulants, or antidepressants, are told to discontinue their prescription for a short time prior to the test (usually one to two days). Patients may be asked to avoid food and beverages that contain **caffeine**, a central nervous system stimulant. However, any such request should be cleared by the treating physician. Patients may also be asked to arrive for the test with clean hair free of spray or other styling products.

Patients undergoing a sleep EEG may be asked to remain awake the night before their test. They may be given a sedative prior to the test to induce sleep.

### Aftercare

If the patient has suspended regular medication for the test, the EEG nurse or technician should advise him when he can begin taking it again.

### Risks

Being off medication for one to two days may trigger seizures. Certain procedures used during EEG may trigger seizures in patients with epilepsy. Those procedures include flashing lights and deep breathing. If the EEG is being used as a diagnostic for epilepsy (i.e., to determine the type of seizures an individual is suffering from), this may be a desired effect, although the patient needs to be monitored closely so that the seizure can be aborted if necessary. This type of test is known as an ictal EEG.

### Normal results

In reading and interpreting brainwave patterns, a neurologist or other physician will evaluate the type of brainwaves and the symmetry, location, and consistency of brainwave patterns. He will also look at the brainwave response to certain stimuli presented during the EEG test (such as flashing lights or noise). There are four basic types of brainwaves: alpha, beta, theta, and delta. "Normal" brainwave patterns vary widely, depending on factors of age and activity. For example, awake and relaxed individuals typically register an alpha wave pattern of eight to 13 cycles per second. Young

## KEY TERMS

**Epilepsy**—A neurological disorder characterized by recurrent seizures with or without a loss of consciousness.

**Ictal EEG**—Used to measure brain activity during a seizure. May be useful in learning more about patients who aren't responding to conventional treatments.

children and sleeping adults may have a delta wave pattern of under four cycles per second.

### Abnormal results

The EEG readings of patients with epilepsy or other seizure disorders display bursts or spikes of electrical activity. In focal epilepsy, spikes are restricted to one hemisphere of the brain. If spikes are generalized to both hemispheres of the brain, multifocal epilepsy may be present.

The diagnostic brainwave patterns of other disorders varies widely. The appearance of excess theta waves (four to eight cycles per second) may indicate brain injury. Brain wave patterns in patients with brain disease, mental retardation, and brain injury show overall slowing. A trained medical specialist should interpret EEG results in the context of the patient's medical history, and other pertinent medical test results.

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Paula Anne Ford-Martin

## Electrolyte disorders

### Definition

An electrolyte disorder is an imbalance of certain ionized salts (i.e., bicarbonate, calcium, chloride, magnesium, phosphate, potassium, and sodium) in the blood.

### Description

Electrolytes are ionized molecules found throughout the blood, tissues, and cells of the body. These molecules, which are either positive (cations) or negative (anions), conduct an electric current and help to balance pH and acid-base levels in the body. Electrolytes also facilitate the passage of fluid between and within cells through a process known as *osmosis* and play a part in regulating the function of the neuromuscular, endocrine, and excretory systems.

The serum electrolytes include:

- Sodium (Na). A positively charged electrolyte that helps to balance fluid levels in the body and facilitates neuromuscular functioning.
- Potassium (K). A main component of cellular fluid, this positive electrolyte helps to regulate neuromuscular function and osmotic pressure.
- Calcium (Ca). A cation, or positive electrolyte, that affects neuromuscular performance and contributes to skeletal growth and blood coagulation.
- Magnesium (Mg). Influences muscle contractions and intracellular activity. A cation.
- Chloride (Cl). An anion, or negative electrolyte, that regulates blood pressure.
- Phosphate (HPO<sub>4</sub>). Negative electrolyte that impacts metabolism and regulates acid-base balance and calcium levels.
- Bicarbonate (HCO<sub>3</sub>). A negatively charged electrolyte that assists in the regulation of blood pH levels. Bicarbonate insufficiencies and elevations cause acid-base disorders (i.e., acidosis, alkalosis).

Medications, chronic diseases, and trauma (i.e., **burns, fractures, etc.**) may cause the concentration of certain electrolytes in the body to become too high (hyper-) or too low (hypo-). When this happens, an electrolyte imbalance, or disorder, results.

### Causes and symptoms

#### Sodium

**HYPERNATREMIA.** Sodium helps the kidneys to regulate the amount of water the body retains or excretes. Consequently, individuals with elevated serum sodium levels also suffer from a loss of fluids, or **dehydration**. **Hypernatremia** can be caused by inadequate water intake, excessive fluid loss (i.e., **diabetes insipidus**, kidney disease, severe burns, and prolonged vomiting or **diarrhea**), or sodium retention (caused by excessive sodium intake or aldosteronism). In addition, certain drugs,

including loop **diuretics**, **corticosteroids**, and antihypertensive medications may cause elevated sodium levels.

Symptoms of hypernatremia include:

- thirst
- orthostatic hypotension
- dry mouth and mucous membranes
- dark, concentrated urine
- loss of elasticity in the skin
- irregular heartbeat (tachycardia)
- irritability
- fatigue
- lethargy
- heavy, labored breathing
- muscle twitching and/or seizures

**HYPONATREMIA.** Up to 1% of all hospitalized patients develop **hyponatremia**, making it one of the most common electrolyte disorders. Diuretics, certain psychoactive drugs (i.e., fluoxetine, sertraline, haloperidol), specific antipsychotics (lithium), vasopressin, chlorpropamide, the illicit drug “ecstasy”, and other pharmaceuticals can cause decreased sodium levels, or hyponatremia. Low sodium levels may also be triggered by inadequate dietary intake of sodium, excessive perspiration, water intoxication, and impairment of adrenal gland or kidney function.

Symptoms of hyponatremia include:

- nausea, abdominal cramping, and/or vomiting
- headache
- edema (swelling)
- muscle weakness and/or tremor
- paralysis
- disorientation
- slowed breathing
- seizures
- **coma**

### Potassium

**HYPERKALEMIA.** **Hyperkalemia** may be caused by ketoacidosis (diabetic coma), myocardial infarction (**heart attack**), severe burns, kidney failure, **fasting**, **bulimia nervosa**, gastrointestinal bleeding, adrenal insufficiency, or **Addison's disease**. Diuretic drugs, cyclosporin, lithium, heparin, ACE inhibitors, **beta blockers**, and trimethoprim can increase serum potassium levels, as can heavy **exercise**. The condition may also be secondary to hypernatremia (low serum concentrations of sodium). Symptoms may include:

- weakness
- nausea and/or abdominal pain
- irregular heartbeat (arrhythmia)
- diarrhea
- muscle pain

**HYPOKALEMIA.** Severe dehydration, aldosteronism, **Cushing's syndrome**, kidney disease, long-term diuretic therapy, certain **penicillins**, laxative abuse, congestive **heart failure**, and adrenal gland impairments can all cause depletion of potassium levels in the bloodstream. A substance known as glycyrrhetic acid, which is found in licorice and chewing tobacco, can also deplete potassium serum levels. Symptoms of **hypokalemia** include:

- weakness
- paralysis
- increased urination
- irregular heartbeat (arrhythmia)
- orthostatic hypotension
- muscle pain
- tetany

### Calcium

**HYPERCALCEMIA.** Blood calcium levels may be elevated in cases of thyroid disorder, **multiple myeloma**, metastatic **cancer**, multiple bone fractures, milk-alkali syndrome, and Paget's disease. Excessive use of calcium-containing supplements and certain over-the-counter medications (i.e., **antacids**) may also cause **hypercalcemia**. Symptoms include:

- fatigue
- constipation
- depression
- confusion
- muscle pain
- nausea and vomiting
- dehydration
- increased urination
- irregular heartbeat (arrhythmia)

**HYPOCALCEMIA.** Thyroid disorders, kidney failure, severe burns, **sepsis**, **vitamin D deficiency**, and medications such as heparin and glucagon can deplete blood calcium levels. Lowered levels cause:

- muscle cramps and spasms
- tetany and/or convulsions
- mood changes (depression, irritability)
- dry skin

- brittle nails
- facial twitching

### Magnesium

**HYPERMAGNESEMIA.** Excessive magnesium levels may occur with end-stage renal disease, Addison's disease, or an overdose of magnesium salts. Hypermagnesemia is characterized by:

- lethargy
- hypotension
- decreased heart and respiratory rate
- muscle weakness
- diminished tendon reflexes

**HYPOMAGNESEMIA.** Inadequate dietary intake of magnesium, often caused by chronic **alcoholism** or **malnutrition**, is a common cause of hypomagnesemia. Other causes include malabsorption syndromes, **pancreatitis**, aldosteronism, burns, **hyperparathyroidism**, digestive system disorders, and diuretic use. Symptoms of low serum magnesium levels include:

- leg and foot cramps
- weight loss
- vomiting
- muscle spasms, twitching, and tremors
- seizures
- muscle weakness
- arrhythmia

### Chloride

**HYPERCHLOREMA.** Severe dehydration, kidney failure, hemodialysis, traumatic brain injury, and aldosteronism can also cause hyperchloremia. Drugs such as boric acid and ammonium chloride and the intravenous (IV) infusion of sodium chloride can also boost chloride levels, resulting in hyperchloremic **metabolic acidosis**. Symptoms include:

- weakness
- headache
- nausea
- cardiac arrest

**HYPOCHLOREMA.** Hypochloremia usually occurs as a result of sodium and potassium depletion (i.e., hyponatremia, hypokalemia). Severe depletion of serum chloride levels causes *metabolic alkalosis*. This alkalinization of the bloodstream is characterized by:

- mental confusion
- slowed breathing

- paralysis
- muscle tension or spasm

### Phosphate

**HYPERPHOSPHATEMIA.** Skeletal fractures or disease, kidney failure, **hypoparathyroidism**, hemodialysis, **diabetic ketoacidosis**, acromegaly, systemic infection, and intestinal obstruction can all cause phosphate retention and build-up in the blood. The disorder occurs concurrently with **hypocalcemia**. Individuals with mild hyperphosphatemia are typically asymptomatic, but signs of severe hyperphosphatemia include:

- tingling in hands and fingers
- muscle spasms and cramps
- convulsions
- cardiac arrest

**HYPOPHOSPHATEMIA.** Serum phosphate levels of 2 mg/dL or below may be caused by hypomagnesemia and hypokalemia. Severe burns, alcoholism, diabetic ketoacidosis, kidney disease, hyperparathyroidism, **hypothyroidism**, Cushing's syndrome, malnutrition, hemodialysis, vitamin D deficiency, and prolonged diuretic therapy can also diminish blood phosphate levels. There are typically few physical signs of mild phosphate depletion. Symptoms of severe hypophosphatemia include:

- muscle weakness
- weight loss
- bone deformities (osteomalacia)

### Diagnosis

Diagnosis is performed by a physician or other qualified healthcare provider who will take a medical history, discuss symptoms, perform a complete **physical examination**, and prescribe appropriate laboratory tests. Because electrolyte disorders commonly affect the neuromuscular system, the provider will test reflexes. If a calcium imbalance is suspected, the physician will also check for Chvostek's sign, a reflex test that triggers an involuntary facial twitch, and Trousseau's sign, a muscle spasm that occurs in response to pressure on the upper arm.

Serum electrolyte imbalances can be detected through blood tests. Blood is drawn from a vein on the back of the hand or inside of the elbow by a medical technician, or phlebotomist, and analyzed at a lab.

Normal levels of electrolytes are:

- Sodium. 135–145 mEq/L (serum)
- Potassium. 3.5–5.5 mEq/L (serum)

## KEY TERMS

**Acid-base balance**—A balance of acidity and alkalinity of fluids in the body that keeps the pH level of blood around 7.35–7.45.

**Aldosteronism**—A condition defined by high serum levels of aldosterone, a hormone secreted by the adrenal gland that is responsible for increasing sodium reabsorption in the kidneys.

**Addison's disease**—A disease characterized by a deficiency in adrenocortical hormones due to destruction of the adrenal gland.

**Bulimia nervosa**—An eating disorder characterized by binging and purging (self-induced vomiting) behaviors.

**Milk-alkali syndrome**—Elevated blood calcium levels and alkalosis caused by excessive intake of milk and alkalis. Usually occurs in the treatment of peptic ulcer.

**Orthostatic hypotension**—A drop in blood pressure that causes faintness or dizziness and occurs when one rises to a standing position. Also known as postural hypotension.

**Osmotic pressure**—Pressure that occurs when two solutions of differing concentrations are separated by a semipermeable membrane, such as a cellular wall, and the lower concentration solute is drawn across the membrane into the higher concentration solute (osmosis).

**Tetany**—A disorder of the nervous system characterized by muscle cramps, spasms of the arms and legs, and numbness of the extremities.

- Calcium. 8.8–10.4 mg/dL (total Ca; serum); 4.7–5.2 mg/dL (unbound Ca; serum)
- Magnesium. 1.4–2.1 mEq/L (plasma)
- Chloride. 100–108 mEq/L (serum)
- Phosphate. 2.5–4.5 mg/dL (plasma; adults)

Standard ranges for test results may vary due to differing laboratory standards and physiological variances (i.e., gender, age, and other factors). Other blood tests that determine pH levels and acid-base balance may also be performed.

### Treatment

Treatment of electrolyte disorders depends on the underlying cause of the problem and the type of elec-

trolyte involved. If the disorder is caused by poor diet or improper fluid intake, nutritional changes may be prescribed. If medications such as diuretics triggered the imbalance, discontinuing or adjusting the drug therapy may effectively treat the condition. Fluid and electrolyte replacement therapy, either intravenously or by mouth, can reverse electrolyte depletion.

Hemodialysis treatment may be required to reduce serum potassium levels in hyperkalemic patients with impaired kidney function. It may also be recommended for renal patients suffering from severe hypermagnesemia.

### Prognosis

A patient's long-term prognosis depends upon the root cause of the electrolyte disorder. However, when treated quickly and appropriately, electrolyte imbalances in and of themselves are usually effectively reversed.

When they are mild, some electrolyte imbalances have few to no symptoms and may pass unnoticed. For example, transient hyperphosphatemia is usually fairly benign. However, long-term elevations of blood phosphate levels can lead to potentially fatal soft tissue and vascular calcifications and bone disease, and severe serum phosphate deficiencies (hypophosphatemia) can cause encephalopathy, coma, and death.

Severe hypernatremia has a mortality rate of 40–60%. Death is commonly due to cerebrovascular damage and hemorrhage resulting from dehydration and shrinkage of the brain cells.

### Prevention

Physicians should use caution when prescribing drugs known to affect electrolyte levels and acid-base balance. Individuals with kidney disease, thyroid problems, and other conditions that may place them at risk for developing an electrolyte disorder should be educated on the signs and symptoms.

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Paula Ford-Martin

## Electrolyte tests

### Definition

Electrolytes are positively and negatively charged molecules, called ions, that are found within cells, between cells, in the bloodstream, and in other fluids throughout the body. Electrolytes with a positive charge include sodium, potassium, calcium, and magnesium; the negative ions are chloride, bicarbonate, and phosphate. The concentrations of these ions in the bloodstream remain fairly constant throughout the day in a healthy person. Changes in the concentration of one or more of these ions can occur during various acute and chronic disease states and can lead to serious consequences.

### Purpose

Tests that measure the concentration of electrolytes are useful in the emergency room and to obtain clues for the diagnosis of specific diseases. Electrolyte tests are used for diagnosing dietary deficiencies, excess loss of nutrients due to urination, vomiting, and **diarrhea**, or abnormal shifts in the location of an electrolyte within the body. When an abnormal electrolyte value is detected, the physician may either act to immediately correct the imbalance directly (in the case of an emergency) or run further tests to determine the underlying cause of the abnormal electrolyte value. Electrolyte disturbances can occur with malfunctioning of the kidney (renal failure), infections that produce severe and continual diarrhea or vomiting, drugs that cause loss of electrolytes in the urine (**diuretics**), **poisoning** due to accidental consumption of electrolytes, or diseases involving hormones that regulate electrolyte concentrations.

### Precautions

Electrolyte tests are performed from routine blood tests. The techniques are simple, automated, and fairly uniform throughout the United States. During the preparation of blood plasma or serum, health workers must take care not to break the red blood cells, especially when testing for serum potassium. Because the concentration of potassium within red blood cells is much higher than in the surrounding plasma or serum, broken cells would cause falsely elevated potassium levels.

### Description

Electrolyte tests are typically conducted on blood plasma or serum, urine, and diarrheal fluids. Electrolytes can be classified in at least five different ways. One way is that some electrolytes tend to exist mostly inside cells,

or are intracellular, while others tend to be outside cells, or are extracellular. Potassium, phosphate, and magnesium occur at much greater levels inside the cell than outside, while sodium and chloride occur at much greater levels extracellularly. A second classification distinguishes those electrolytes that participate directly in the transmission of nerve impulses and those that do not. Sodium, potassium, and calcium are the important electrolytes involved in nerve impulses, and disorders affecting them are most closely associated with neurological disorders. A third classification focuses on electrolytes that are able to form a tight union, or complex, with one another. Calcium and phosphate have the greatest tendency to form complexes with each other. Disorders that cause an increase in either plasma calcium or phosphate can result in the deposit of calcium-phosphate crystals in the soft tissues of the body. A fourth classification concerns those electrolytes that influence the acidity or alkalinity of the bloodstream, also known as the pH. The pH of the bloodstream is normally in the range of 7.35–7.45. A decrease below this range is called acidosis, while a pH above this range is called alkalosis. The electrolytes most closely associated with the pH of the bloodstream are bicarbonate, chloride, and phosphate.

### Preparation

All electrolyte tests can be performed on plasma or serum. Plasma is prepared by withdrawing a blood sample and placing it in a test tube containing a chemical that prevents blood from clotting (an anticoagulant). Serum is prepared by withdrawing a blood sample, placing it in a test tube, and allowing it to clot. The blood spontaneously clots within a minute of withdrawing the blood from a vein. The serum or plasma is then rapidly spun with a centrifuge in order to remove the blood cells or clot.

### Normal results

Electrolyte concentrations are similar whether measured in serum or plasma. Values can be expressed in terms of weight per unit volume (mg/deciliter; mg/dL) or in the number of molecules in a volume, or molarity (moles or millimoles/liter; M or mM). The range of normal values sometimes varies slightly between different age groups, for males and females, and between different analytical laboratories.

The normal level of serum sodium is in the range of 136–145 mM. The normal levels of serum potassium are 3.5–5.0 mM. Note that sodium occurs at a much higher concentration than potassium. The normal concentration of total serum calcium (bound calcium plus free calcium) is in the range of 8.8–10.4 mg/dL. About 40% of the total calcium in the plasma is loosely bound to proteins; this

calcium is referred to as bound calcium. The normal range of free calcium is 4.8–5.2 mg/dL. The normal concentration of serum magnesium is in the range of 2.0–3.0 mg/dL.

The normal concentration range of chloride is 350–375 mg/dL or 98–106 mM. The normal level of phosphate, as expressed as the concentration of phosphorus, is 2.0–4.3 mg/dL. Bicarbonate is an electrolyte that is freely and spontaneously interconvertable with carbonic acid and carbon dioxide. The normal concentration of carbonic acid ( $H_2CO_3$ ) is about 1.35 mM. The normal concentration of bicarbonate ( $HCO_3^-$ ) is about 27 mM. The concentration of total carbon dioxide is the sum of carbonic acid and bicarbonate; this sum is normally in the range of 26–28 mM. The ratio of bicarbonate/carbonic acid is more significant than the actual concentrations of these two forms of carbon dioxide. Its normal value is 27/1.35 (equivalent to 20/1).

## Abnormal results

### *Positively charged electrolytes*

High serum sodium levels (**hypernatremia**) occur at sodium concentrations over 145 mM, with severe hypernatremia over 152 mM. Hypernatremia is usually caused by diseases that cause excessive urination. In these cases, water is lost, but sodium is still retained in the body. The symptoms include confusion and can lead to convulsions and **coma**. Low serum sodium levels (**hyponatremia**) are below 130 mM, with severe hyponatremia at or below 125 mM. Hyponatremia often occurs with severe diarrhea, with losses of both water and sodium, but with sodium loss exceeding water loss. Hyponatremia provokes clinical problems only if serum sodium falls below 125 mM, especially if this has occurred rapidly. The symptoms can be as mild as tiredness but may lead to convulsions and coma.

High serum potassium (**hyperkalemia**) occurs at potassium levels above 5.0 mM; it is considered severe over 8.0 mM. Hyperkalemia is relatively uncommon, but sometimes occurs in patients with kidney failure who take potassium supplements. Hyperkalemia can result in abnormal beating of the heart (cardiac **arrhythmias**). Low serum potassium (**hypokalemia**) occurs when serum potassium falls below 3.0 mM. It can result from low dietary potassium, as during **starvation** or in patients with **anorexia nervosa**; from excessive losses via the kidneys, as caused by diuretic drugs; or by diseases of the adrenal or pituitary glands. Mild hypokalemia causes muscle weakness, while severe hypokalemia can cause **paralysis**, the inability to breathe, and cardiac arrhythmias.

High levels of calcium ions (**hypercalcemia**) occur at free calcium ion concentrations over 5.2 mg/dL or total serum calcium above 10.4 mg/dL. Hypercalcemia

usually occurs when the body dissolves bone at an abnormally fast rate, increasing both serum calcium and serum phosphate. Sudden hypercalcemia can cause vomiting and coma, while prolonged and moderate hypercalcemia results in the deposit of calcium phosphate crystals in the kidneys and eye. **Hypocalcemia** occurs when serum free calcium ions fall below 4.4 mg/dL, or when total serum calcium falls below 8.8 mg/dL. Hypocalcemia can result from **hypoparathyroidism** (low parathyroid hormone), from failure to produce 1,25-dihydroxyvitamin D, from low levels of plasma magnesium, and from phosphate poisoning (the phosphate enters the bloodstream and forms a complex with the free serum calcium). Hypocalcemia can cause depression and muscle spasms.

Hypermagnesemia occurs at serum magnesium levels over 25 mM (60 mg/dL). Hypermagnesemia is rare but can occur with the excessive consumption of magnesium salts. Hypomagnesemia occurs when serum magnesium levels fall below 0.8 mM, and can result from poor **nutrition**. Chronic **alcoholism** is the most common cause of hypomagnesemia, in part because of poor diet. Magnesium levels below 0.5 mM (1.2 mg/dL) cause serum calcium levels to decline. Some of the symptoms of hypomagnesemia, including twitching and convulsions, actually result from the concurrent hypocalcemia. Hypomagnesemia can also result in hypokalemia and thereby cause cardiac arrhythmias.

### *Negatively charged electrolytes*

Serum chloride levels sometimes increase to abnormal levels as an undesirable side effect of medical treatment with sodium chloride or ammonium chloride. The toxicity of chloride results not from the chloride itself, but from the fact that the chloride occurs as the acid, hydrogen chloride (more commonly known as hydrochloric acid, or HCl). An overdose of chloride may cause the accumulation of hydrochloric acid in the bloodstream, with consequent acidosis. **Renal tubular acidosis**, one of many kidney diseases, involves the failure to release acid into the urine. The acidosis produces weakness, **headache**, nausea, and cardiac arrest. Low plasma chloride leads to the opposite situation: a decline in the acid content of the bloodstream. This is known as alkalinization of the bloodstream, or alkalosis. Hydrochloric acid, originally from extracellular fluids, can be lost by vomiting. At its most severe, alkalosis results in paralysis (tetany).

Hyperphosphatemia occurs at serum phosphate levels above 5 mg/dL. It can result from the failure of the kidneys to excrete phosphate into the urine, causing phosphate to accumulate in the bloodstream. Hyperphosphatemia can also be caused by the impaired action of parathyroid hormone and by phosphate poisoning. Severe hyperphos-

phatemia can cause paralysis, convulsions, and cardiac arrest. These symptoms result because the phosphate, occurring in elevated levels, complexes with free serum calcium, resulting in hypocalcemia. Tests for heart function (an electrocardiogram) and parathyroid hormone levels are used in the diagnosis of hyperphosphatemia. Hypophosphatemia occurs if serum phosphorus falls to 2.0 mg/dL or lower. It often results from a shift of inorganic phosphate from the bloodstream to various organs and tissues. This shift can be caused by a rise in pH (alkalization) of the bloodstream, which can occur during hyperventilation, a reaction in various disease states. A shift in phosphate to intracellular tissues may draw calcium away from the bloodstream via the formation of insoluble calcium phosphate crystals within cells, with consequent hypocalcemia. Thus, tests for abnormalities in phosphate metabolism also involve tests for serum calcium.

Bicarbonate metabolism involves several compounds. When dietary starches, sugars, and fats are broken down for energy production, carbon dioxide is created. Much of this carbon dioxide ( $\text{CO}_2$ ) spontaneously converts to carbonic acid ( $\text{H}_2\text{CO}_3$ ), and some of the carbonic acid spontaneously converts to bicarbonate ( $\text{HCO}_3^-$ ) plus a hydrogen ion ( $\text{H}^+$ ). Eventually, almost every molecule of carbon dioxide produced in the body, whether in the form of carbon dioxide, carbonic acid, or bicarbonate, must convert back to carbon dioxide in order to leave via the lungs during normal breathing.

If one holds one's breath, carbon dioxide cannot escape from the lungs, but continues to be generated within the body. This results in an increase in production of carbonic acid. A portion of the carbonic acid breaks apart (dissociates), causing an increase in hydrogen ions in the plasma, with a resulting acidosis. Tests for serum bicarbonate levels are accompanied by tests for acidosis (pH test). Conversely, when one breathes too rapidly (hyperventilation), the carbon dioxide is drawn off from the bloodstream and expelled in the breath at an increased rate. This results in an increase in the rate of combination of bicarbonate with hydrogen ions, resulting in alkalosis. Acidosis and alkalosis can be produced by means other than by altering the rate of breathing. The carbonic acid and bicarbonate in the bloodstream minimize (or buffer) any trend to acidosis or alkalosis. Tests for bicarbonate are generally accompanied by tests for blood pH and possibly tests for kidney malfunction, abnormal hormone function, or gastrointestinal disorders.

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Tom Brody, PhD

## Electromyography

### Definition

Electromyography (EMG) is an electrical recording of muscle activity that aids in the diagnosis of neuromuscular disease.

### Purpose

Muscles are stimulated by signals from nerve cells called motor neurons. This stimulation causes electrical activity in the muscle, which in turn causes contraction. This electrical activity is detected by a needle electrode inserted into the muscle and connected to a recording device. Together, the electrode and recorder are called an electromyography machine. EMG can determine whether a particular muscle is responding appropriately to stimulation, and whether a muscle remains inactive when not stimulated.

EMG is performed most often to help diagnose different diseases causing weakness. Although EMG is a test of the motor system, it may help identify abnormalities of nerves or spinal nerve roots that may be associated with pain or numbness. Other symptoms for which EMG may be useful include numbness, atrophy, stiffness, fasciculation, cramp, deformity, and spasticity. EMG results can help determine whether symptoms are due to a muscle disease or a neurological disorder, and, when combined with clinical findings, usually allow a confident diagnosis.

EMG can help diagnose many muscle and nerve disorders, including:

- muscular dystrophy
- congenital myopathies
- mitochondrial myopathies

## KEY TERMS

**Motor neurons**—Nerve cells that transmit signals from the brain or spinal cord to the muscles.

**Motor unit action potentials**—Spikes of electrical activity recorded during an EMG that reflect the number of motor units (motor neurons and the muscle fibers they transmit signals to) activated when the patient voluntarily contracts a muscle.

- metabolic myopathies
- myotonias
- peripheral neuropathies
- radiculopathies
- nerve lesions
- amyotrophic lateral sclerosis
- polio
- spinal muscular atrophy
- guillain-Barré syndrome
- ataxias
- myasthenias

### Precautions

No special precautions are needed for this test. Patients with a history of bleeding disorder should consult with their treating physician before the test. If a muscle biopsy is planned as part of the diagnostic work-up, EMG should not be performed at the same site, as it may effect the microscopic appearance of the muscle.

### Description

During an EMG test, a fine needle is inserted into the muscle to be tested. This may cause some discomfort, similar to that of an injection. Recordings are made while the muscle is at rest, and then during the contraction. The person performing the test may move the limb being tested, and direct the patient to move it with various levels of force. The needle may be repositioned in the same muscle for further recording. Other muscles may be tested as well. A typical session lasts from 30–60 minutes.

A slightly different test, the *nerve conduction velocity test*, is often performed at the same time with the same equipment. In this test, stimulating and recording electrodes are used, and small electrical shocks are applied to measure the ability of the nerve to conduct electrical signals. This test may cause mild tingling and discomfort

similar to a mild **shock** from static electricity. Evoked potentials may also be performed for additional diagnostic information. Nerve conduction velocity and evoked potential testing are especially helpful when pain or sensory complaints are more prominent than weakness.

### Preparation

No special preparation is needed. The doctor supervising and interpreting the test should be given information about the symptoms, medical conditions, suspected diagnosis, neuroimaging studies, and other test results.

### Aftercare

Minor pain and bleeding may continue for several hours after the test. The muscle may be tender for a day or two.

### Risks

There are no significant risks to this test, other than those associated with any needle insertion (pain, bleeding, bruising, or infection).

### Normal results

There should be some brief EMG activity during needle insertion. This activity may be increased in diseases of the nerve and decreased in long-standing muscle disorders where muscle tissue is replaced by fibrous tissue or fat. Muscle tissue normally shows no EMG activity when at rest or when moved passively by the examiner. When the patient actively contracts the muscle, spikes (motor unit action potentials) should appear on the recording screen, reflecting the electrical activity within. As the muscle is contracted more forcefully, more groups of muscle fibers are recruited or activated, causing more EMG activity.

### Abnormal results

The interpretation of EMG results is not a simple matter, requiring analysis of the onset, duration, amplitude, and other characteristics of the spike patterns.

Electrical activity at rest is abnormal; the particular pattern of firing may indicate denervation (for example, a nerve lesion, radiculopathy, or lower motor neuron degeneration), myotonia, or inflammatory myopathy.

Decreases in the amplitude and duration of spikes are associated with muscle diseases, which also show faster recruitment of other muscle fibers to compensate for weakness. Recruitment is reduced in nerve disorders.

## Resources

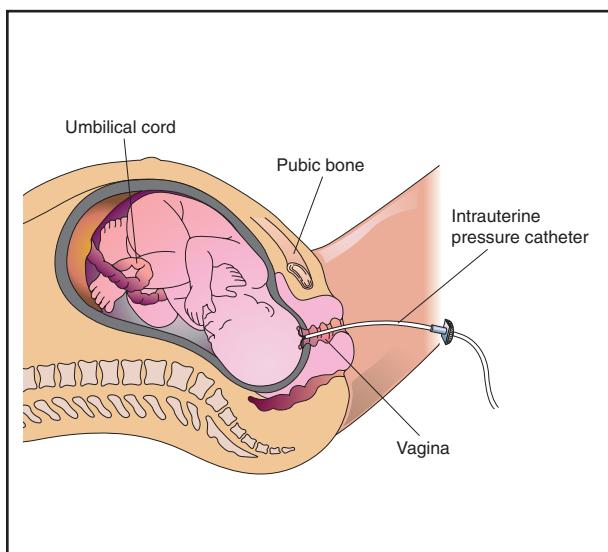
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Richard Robinson



## Electronic fetal monitoring

### Definition

Electronic fetal monitoring (EFM) is a method for examining the condition of a baby in the uterus by noting any unusual changes in its heart rate. Electronic fetal monitoring is performed late in pregnancy or continuously during labor to ensure normal delivery of a healthy baby. EFM can be utilized either externally or internally in the womb.

### Purpose

The heart rate of a fetus undergoes constant adjustment as it responds to its environment and other stimuli. The fetal monitor records an unborn baby's heart rate and graphs it on a piece of paper. Electronic fetal monitoring is usually advised for high-risk pregnancies, when the baby is in danger of distress. Specific reasons for EFM include: babies in a breech position, **premature labor**, and induced labor, among others.

When electronic fetal monitoring was originally introduced in the 1960s and 1970s, the hope was that it would help physicians diagnose fetal hypoxia, or lack of oxygen, in time to prevent damage to the baby. This lack of oxygen, also known as perinatal asphyxia or birth asphyxia, is an important cause of **stillbirth** and newborn deaths. It occurs when there are less than normal amounts of oxygen delivered to the body or an organ and there is build-up of carbon dioxide in the body or tissue. A lack of blood flow to an organ can cause asphyxia. Perinatal asphyxia can occur a long time before birth, shortly before birth, during delivery, or after birth. If the interruption to the supply of oxygen is short, the baby may recover without any damage. If the time is longer, there may be some injury that is reversible. If the time period without oxygen is very long, there may be permanent injury to one or more organs of the body. It is important, to detect any signs of asphyxia as soon as possible. One of the signs is an abnormal heart rate and rhythm in

**Electronic fetal monitoring (EFM)** is performed late in pregnancy or continuously during labor to ensure normal delivery of a healthy baby. EFM can be utilized either externally or internally in the womb. The illustration above shows the internal procedure, in which an electrode is attached directly to the baby's scalp to monitor the heart rate. Uterine contractions are recorded using an intrauterine pressure catheter which is inserted through the cervix into the uterus. (Illustration by the Electronic Illustrators Group.)

the unborn baby, which can be detected by electronic fetal monitoring.

The fetal monitor is a more intricate version of the machine that a health care provider uses to listen to a baby's heartbeat. The monitor that is used during prenatal visits just picks up the sound of the baby's heart beating. The fetal monitor also keeps a continuous paper record of the heart rate. In addition, the fetal monitor can record uterine contractions on the lower part of the paper strip. This helps the doctor or midwife determine how a baby is handling the stress of contractions. The normal pattern is for the baby's heartbeat to drop slightly during a contraction and then go back to normal after the contraction is over. EFM looks for any changes from this normal pattern, particularly if there is a drastic drop in the baby's heart beat or if the heart rate does not recover immediately after a contraction.

Because it is an indirect test, it is not perfect. When an adult complains to a provider about not feeling well, checking the heart rate is only one of many things that the doctor will do. With an unborn baby, however, checking the heart rate is basically the only thing that a doctor or midwife can do.

Fetal monitoring can be helpful in a variety of different situations. During pregnancy, fetal monitoring can be used as a part of **antepartum testing**. If the practitioner

## KEY TERMS

**Breech presentation**—Fetal position in which the buttocks come first.

**A cesarean section**—delivery of a baby through an incision in the mother's abdomen instead of through the vagina.

**Hypoxia**—An oxygen deficiency.

feels that a baby may be at increased risk of problems toward the end of pregnancy, a baby can be checked every week or every other week with a non-stress test. In this test, changes in the baby's heart rate are measured along with the fetus' own movements. The heart rate of a healthy baby should go up whenever she or he moves.

Fetal monitoring is used on and off during early labor. As labor progresses, more monitoring is often needed. Usually, as the time for delivery nears, the monitor is left on continuously since the end of labor tends to be the most stressful time for the baby.

A baby who is having trouble in labor will show characteristic changes in heart rate after a contraction (late decelerations). If a baby is not receiving enough oxygen to withstand the stress of labor and delivery is many hours away, a **cesarean section** (C-section) may be necessary.

### Description

Using the external fetal monitor is simple and painless. Two elastic belts are placed around the mother's abdomen. One belt holds a listening device in place while the other belt holds the contraction monitor. The nurse or midwife adjusts the belts to get the best readings from each device.

Sometimes, it is difficult to hear the baby's heartbeat with the external monitoring device. Other times, the monitor may show subtle signs of a developing problem. In either case, the doctor or midwife may recommend that the external belt be replaced with an internal monitor.

The internal monitor is an electronic wire that rests directly on the baby's head. The provider can place it on the baby's head during an internal exam. The internal monitor can only be used when the cervix is already open. This device provides a more accurate record of the baby's heart rate.

### Preparation

There are no special preparations needed for fetal monitoring.

### Risks

External EFM poses no direct risks to the baby. However, because of being connected to the machine, the mother cannot walk around. This inactivity may prolong labor and reduce oxygen levels in the mother's blood, both of which can be detrimental to the unborn baby. Another problem is that electronic fetal monitoring seems to be associated with an increase in caesarian deliveries. There is a concern that EFM can give false alarms of distress in the baby, and that this can lead to unneeded caesarians. With internal monitoring, there is a higher risk for infection. For these and other reasons, the United States Preventive Services Task Force states that there is some evidence that using electronic fetal monitoring on low-risk women in labor might not be indicated. Many physicians, however, continue to use EFM routinely, and believe it to be of value in both low-risk and high-risk labors.

### Normal results

An unborn baby's heart rate normally ranges from 120–160 beats per minute (bpm). A baby who is receiving enough oxygen through the placenta will move around. The monitor strip will show the baby's heart rate rising briefly as he/she moves (just as an adult's heart rate rises when he/she moves).

The baby's monitor strip is considered to be reactive when the baby's heart rate rises at least 20 bpm above the baseline heart rate for at least 20 seconds. This must occur at least twice in a 20-minute period. A reactive heart rate tracing (also known as a reactive non-stress test) is considered a sign of the baby's well being.

### Abnormal results

If the baby's heart rate drops very low or rises very high, this signals a serious problem. In either of these cases it is obvious that the baby is in distress and must be delivered soon. However, many babies who are having problems do not give such clear signs.

During a contraction, the flow of oxygen (from the mother) through the placenta (to the baby) is temporarily stopped. It is as if the baby has to hold its breath during each contraction. Both the placenta and the baby are designed to withstand this condition. Between contractions, the baby should be receiving more than enough oxygen to do well during the contraction.

The first sign that a baby is not getting enough oxygen between contractions is often a drop in the baby's heart rate after the contraction (late deceleration). The baby's heart rate recovers to a normal level between contractions, only to drop again after the next contraction. This is also a more subtle sign of distress.

These babies will do fine if they are delivered in a short period of time. Sometimes, these signs develop long before delivery is expected. In that case, a C-section may be necessary.

## Resources

### PERIODICALS

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noninvasive tests because electrodes are placed directly on heart tissue. This allows the electrophysiologist to determine the specific location of an arrhythmia and, oftentimes, correct it during the same procedure. This corrective treatment is permanent and considered a cure, and, in many cases, the patient may not need to take heart medications.

EP studies may be helpful in assessing:

- certain tachycardias or bradycardias of unknown cause.
- patients who have been resuscitated after experiencing sudden cardiac death.
- various symptoms of unknown cause, such as chest pain, shortness of breath, fatigue, or syncope (dizziness/fainting).
- response to anti-arrhythmic therapy.

### Precautions

Pregnant patients should not undergo an EP study because of exposure to radiation during the study, which may be harmful to the growing baby.

Patients who have coronary artery disease may need to have that treated before having an EP study.

### Description

The rhythmic pumping action of the heart, which is essentially a muscle, is the result of electrical impulses traveling throughout the walls of the four heart chambers. These impulses originate in the sinoatrial (SA) node, which are specialized cells situated in the top right chamber of the heart: the right atrium. Normally, the SA node, acting like a spark plug, spontaneously generates the impulses, which travel through specific pathways throughout the atria to the atrioventricular (AV) node. The AV node is a relay station, sending the impulses to more specialized muscle fibers throughout the bottom chambers of the heart: the ventricles. If these pathways become damaged or blocked or if extra (abnormal) pathways exist, the heart's rhythm may be altered (perhaps too slow, too fast, or irregular), which can seriously affect the heart's pumping ability.

The patient is transported to the x-ray table in the EP lab and connected to various monitors. Sterile sheets are placed over him or her. A minimum of two catheters are inserted into the right femoral (thigh) vein in the groin area. Depending on the type of arrhythmia, the number of catheters used in an EP test and their route to the heart may vary. For certain tachycardias, two more catheters may be inserted in the left groin and one in the internal jugular (neck) vein or in the subclavian (below the clavi-

## Electrophysiology study of the heart

### Definition

An electrophysiology (EP) study of the heart is a nonsurgical analysis of the electrical conduction system (normal or abnormal) of the heart. The test employs cardiac catheters and sophisticated computers to generate electrocardiogram (EKG) tracings and electrical measurements with exquisite precision from within the heart chambers.

The EP study can be performed solely for diagnostic purposes. It also is performed to pinpoint the exact location of electrical signals (cardiac mapping) in conjunction with a therapeutic procedure called **catheter ablation**.

The test is simple, not painful, and performed in a special laboratory under controlled clinical circumstances by cardiologists and nurses who subspecialize in electrophysiology.

### Purpose

A cardiologist may recommend an EP study when the standard EKG, Holter monitor, event recorder, **stress test**, echocardiogram, or angiogram cannot provide enough information to evaluate an abnormal heart rhythm, called an arrhythmia.

An EP study also may be beneficial in diagnosing a suspected arrhythmia in a patient who shows symptoms of an arrhythmia but in whom it could not be detected from other tests.

The purpose and great value of an EP study is that it offers more detailed information to the doctor about the electrical activity in the heart than the aforementioned



An electrophysiologist nurse monitors a patient's heart rhythm during an electrophysiology study for tachycardia. (Photograph by Collette Placek. Reproduced by permission.)

cle) vein. The catheters are about 0.08 in (2 mm) in diameter, about the size of a spaghetti noodle. The catheters used in catheter ablation are slightly larger.

With the help of fluoroscopy (x rays on a television screen), all the catheters are guided to several specific locations in the heart. Typically, four to 10 electrodes are located on the end of the catheters, which have the ability to send electrical signals to stimulate the heart (called pacing) and to receive electrical signals from the heart—but not at the same time (just as a walkie-talkie cannot send and receive messages at the same time).

First, the electrodes are positioned to receive signals from inside the heart chambers. This allows the doctor to measure how fast the electrical impulses travel currently in the patient's heart. These measurements are called the patient's baseline measurements. Next, the electrodes are positioned to pace: The EP team actually tries to induce

(sometimes in combination with various heart drugs) the arrhythmia that the patient has previously experienced so the team can observe it in a controlled environment, compare it to the patient's clinical or spontaneous arrhythmia, and decide how to treat it.

Once the arrhythmia is induced and the team determines it can be treated with catheter ablation, cardiac mapping is performed to locate precisely the origin and route of the abnormal pathway. When this is accomplished, the ablating electrode catheter is positioned directly against the abnormal pathway, and high radio-frequency energy is delivered through the electrode to destroy (burn) the tissue in this area.

### Preparation

The following preparations are made for an EP study:

- the patient may be advised to stop taking certain medications, especially heart drugs, that may interfere with the test results.
- blood tests usually are ordered the week before the test.
- the patient undergoes conscious **sedation** (awake but relaxed) during the test. This is accomplished quite often with the anesthetic drugs VersedR (Roche laboratories) and fentanyl.
- a local anesthetic is injected at the site of catheter insertion.

### Aftercare

The patient needs to rest flat in bed for several hours after the procedure to allow healing at the catheter insertion sites.

The patient often returns home either the same day of the test or the next day. Someone should drive him or her home.

The doctor may prescribe drugs and/or insert an AFCD to treat the arrhythmia and may do a possible follow-up EP study.

### Risks

The EP diagnostic study and catheter ablation are low-risk procedures. There is a small risk of bleeding and/or infection at the site of catheter insertion, but this occurs less than 1% of the time. Blood clot formation occurs only two in 1,000 instances and is minimized with blood thinner medications administered during the procedure. Vascular injuries causing hemorrhage or **thrombophlebitis** are possible but occur less than 0.7% of the time. Cardiac perforations occur only in one or two per 1,000 instances. If the right internal jugular vein is accessed, the small possibility of puncturing the lung with the catheter exists, which, at worst, could cause a collapsed lung.

Because **ventricular tachycardia** or fibrillation (lethal **arrhythmias**) may be induced in the patient, the EP lab personnel must be prepared to defibrillate the patient as necessary.

### Normal results

The heart initiates and conducts electrical impulses normally.

### Abnormal results

Confirmation of arrhythmias, such as:

- supraventricular tachycardias
- ventricular arrhythmias

## KEY TERMS

**Ablation**—Remove or destroy, such as by burning or cutting.

**Angiogram**—X ray of a blood vessel after special x-ray dye has been injected into it.

**Bradycardia**—Slow heartbeat.

**Cardiac catheter**—Long, thin, flexible tube, that is threaded into the heart through a blood vessel.

**Cardiologist**—Doctor who specializes in diagnosing and treating heart diseases.

**Echocardiogram**—Ultrasound image of the heart.

**Electrocardiogram**—Tracing of the electrical activity of the heart.

**Electrode**—Medium for conducting an electrical current—in this case, platinum wires.

**Electrophysiology**—Study of how electrical signals in the body relate to physiologic function.

**Event recorder**—A small machine, worn by a patient usually for several days or weeks, that is activated by the patient to record his or her EKG when a symptom is detected.

**Fibrillation**—Rapid, random contraction (quivering).

**Holter monitor**—A small machine, worn by a patient usually for 24 hours, that continuously records the patient's EKG during usual daily activity.

**Stress test**—Recording a patient's EKG during exercise.

**Supraventricular tachycardia**—A fast heart beat that originates above the ventricles.

**Tachycardia**—Fast heartbeat.

**Vascular**—Pertaining to blood vessels.

- accessory (extra) pathways
- bradycardias

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## ORGANIZATIONS

Cardiac Arrhythmia Research and Education Foundation  
 (C.A.R.E.) 2082 Michelson Dr. #301 Irvine, CA 92612  
 (800)-404-9500. <<http://www.longqt.com>>.

Medtronics Manufacturer of Therapeutic Devices. 710  
 Medtronic Parkway NE, Minneapolis, MN 55432-5604.  
 (800) 328-2518. <<http://www.medtronic.com>>.

Midwest Heart Specialists. Physician Office Building, 3825  
 Highland Ave., Tower 2, Ste. 400, Downers Grove, IL  
 60515. (630) 719-4799. <<http://www.midwestheart.com>>.

United States Catheter Instruments (USCI). 129 Concord Road  
 Billerica, MA 01821. (800) 826-2273.

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Electroshock therapy see **Electroconvulsive therapy**

## Elephantiasis

### Definition

The word elephantiasis is a vivid and accurate term for the syndrome it describes: the gross (visible) enlargement of the arms, legs, or genitals to elephantoid size.

### Description

True elephantiasis is the result of a parasitic infection caused by three specific kinds of round worms. The long, threadlike worms block the body's lymphatic system—a network of channels, lymph nodes, and organs that helps maintain proper fluid levels in the body by draining lymph from tissues into the bloodstream. This blockage causes fluids to collect in the tissues, which can lead to great swelling, called "lymphedema." Limbs can swell so enormously that they resemble an elephant's foreleg in size, texture, and color. This is the severely disfiguring and disabling condition of elephantiasis.

There are a few different causes of elephantiasis, but the agents responsible for most of the elephantiasis in the world are filarial worms: white, slender round worms found in most tropical and subtropical places. They are transmitted by particular kinds (species) of mosquitoes, that is, bloodsucking insects. Infection with these worms is called "lymphatic filariasis" and over a long period of time can cause elephantiasis.

Lymphatic filariasis is a disease of underdeveloped regions found in South America, Central Africa, Asia, the Pacific Islands, and the Caribbean. It is a disease that has been present for centuries, as ancient Persian and Indian writings clearly described elephant-like swellings of the

arms, legs, and genitals. It is estimated that 120 million people in the world have lymphatic filariasis, as of 1997. The disease appears to be spreading, in spite of decades of research in this area.

Other terms for elephantiasis are Barbados leg, elephant leg, morbus herculeus, mal de Cayenne, and myelolymphangioma.

Other situations that can lead to elephantiasis are:

- a protozoan disease called **leishmaniasis**
- a repeated streptococcal infection
- the surgical removal of lymph nodes (usually to prevent the spread of **cancer**)
- a hereditary birth defect

### Causes and symptoms

Three kinds of round worms cause elephantiasis filariasis: *Wuchereria bancrofti*, *Brugia malayi*, and *Brugia timori*. Of these three, *W. bancrofti* makes up about 90% of the cases. Man is the only known host of *W. bancrofti*.

*Culex, Aedes*, and *Anopheles* mosquitoes are the carriers of *W. bancrofti*. *Anopheles* and *Mansonia* mosquitoes are the carriers of *B. malayi*. In addition, *Anopheles* mosquitoes are the carriers of *B. timori*.

Infected female mosquitoes take a blood meal from a human, and, in doing so, introduce larval forms of the particular parasite they carry to the person. These larvae migrate toward a lymphatic channel, then travel to various places within the lymphatic system, usually positioning themselves in or near lymph nodes throughout the body. During this time, they mature into more developed larvae and eventually into adult worms. Depending upon the species of round worm, this development can take a few months or more than a year. The adult worms grow to about 1 in (2.5 cm) to 4 in (10 cm) long.

The adult worms can live from about three to eight years. Some have been known to live to 20 years, and in one case 40 years. The adult worms begin reproducing numerous live embryos, called microfilariae. The microfilariae travel to the bloodstream, where they can be ingested by a mosquito when it takes a blood meal from the infected person. If they are not ingested by a mosquito, the microfilariae die within about 12 months. If they are ingested by a mosquito, they continue to mature. They are totally dependent on their specific species of mosquito to develop further. The cycle continues when the mosquito takes another blood meal.

Most of the symptoms an infected person experiences are due to the blockage of the lymphatic system by the adult worms and due to the substances (excretions and secretions) produced by the worms.

The body's allergic reactions may include repeated episodes of **fever**, shaking chills, sweating, headaches, vomiting, and **pain**. Enlarged lymph nodes, swelling of the affected area, skin ulcers, bone and joint pain, tiredness, and red streaks along the arm or leg also may occur. Abscesses can form in lymph nodes or in the lymphatic vessels. They may appear at the surface of the skin as well.

Long-term infection with lymphatic filariasis can lead to **lymphedema**, hydrocele (a buildup of fluid in any saclike cavity or duct) in the scrotum, and elephantiasis of the legs, scrotum, arms, penis, breasts, and vulvae. The most common site of elephantiasis is the leg. It typically begins in the ankle and progresses to the foot and leg. At first the swollen leg may feel soft to the touch but eventually becomes hard and thick. The skin may appear darkened or warty and may even crack, allowing bacteria to infect the leg and complicate the disease. The microfilariae usually don't cause injury. In some instances, they cause "eosinophilia," an increased number of eosinophils (a type of white blood cells) in the blood.

This disease is more intense in people who never have been exposed to lymphatic filariasis than it is in the native people of tropical areas where the disease occurs. This is because many of the native people often are immunologically tolerant.

## Diagnosis

The only sure way to diagnose lymphatic filariasis is by detecting the parasite itself, either the adult worms or the microfilariae.

Microscopic examination of the person's blood may reveal microfilariae. But many times, people who have been infected for a long time do not have microfilariae in their bloodstream. The absence of them, therefore, does not mean necessarily that the person is not infected. In these cases, examining the urine or hydrocele fluid or performing other clinical tests is necessary.

Collecting blood from the individual for microscopic examination should be done during the night when the microfilariae are more numerous in the bloodstream. (Interestingly, this is when mosquitoes bite most frequently.) During the day microfilariae migrate to deeper blood vessels in the body, especially in the lung. If it is decided to perform the blood test during the day, the infected individual may be given a "provocative" dose of medication to provoke the microfilariae to enter the bloodstream. Blood then can be collected an hour later for examination.

Detecting the adult worms can be difficult because they are deep within the lymphatic system and difficult to get to. Biopsies usually are not performed because they usually don't reveal much information.



**Man suffering from elephantiasis.** (Photograph by C. James Webb, Phototake NYC. Reproduced by permission.)

## Treatment

The drug of choice in treating lymphatic filariasis is diethylcarbamazine (DEC). The trade name in the United States is Hetrazen.

The treatment schedule is typically 2 mg/kg per day, three times a day, for three weeks. The drug is taken in tablet form.

DEC kills the microfilariae quickly and injures or kills the adult worms slowly, if at all. If all the adult worms are not killed, remaining paired males and females may continue to produce more larvae. Therefore, several courses of DEC treatment over a long time period may be necessary to rid the individual of the parasites.

DEC has been shown to reduce the size of enlarged lymph nodes and, when taken long-term, to reduce elephantiasis. In India, DEC has been given in the form of a medicated salt, which helps prevent spread of the disease.

## KEY TERMS

**Antigen**—Any substance (usually a protein) that causes an immune response by the body to produce antibodies.

**Filarial**—Threadlike. The word “filament” is formed from the same root word.

**Host**—A person or animal in which a parasite lives, is nourished, grows, and reproduces.

**Lymph**—A watery substance that collects in the tissues and organs of the body and eventually drains into the bloodstream.

**Lymphatic system**—A network composed of vessels, lymph nodes, the tonsils, the thymus gland, and the spleen. It is responsible for transporting fluid and nutrients to the bloodstream and for maturing certain blood cells that are part of the body’s immune system.

**Lymphedema**—The unnatural accumulation of lymph in the tissues of the body, which results in swelling in that area.

**Protozoa**—(Plural form of protozoan) Single-celled organisms (not bacteria) of which about 30 kinds cause disease in humans.

**Streptococcal**—Pertaining to any of the *Streptococcus* bacteria. These organisms can cause pneumonia, skin infections, and many other diseases.

The side effects of DEC almost all are due to the body’s natural allergic reactions to the dying parasites rather than to the DEC itself. For this reason, DEC must be given carefully to reduce the danger to the individual. Side effects may include fever, chills, **headache**, **dizziness**, **nausea and vomiting**, **itching**, and joint pain. These side effects usually occur within the first few days of treatment. These side effects usually subside as the individual continues taking the drug.

There is an alternate treatment plan for the use of DEC. This plan is designed to kill the parasites slowly (to reduce allergic reactions to the dead microfilariae and dying adult worms within the body). Lower doses of DEC are taken for the first few days, followed by the higher dose of 2 mg/kg per day for the remaining three weeks. In addition, steroids may be prescribed to prevent the individual’s body from reacting severely to the dead worms.

Another drug used is Ivermectin. Early research studies of Ivermectin show that it is excellent in killing

microfilariae, but the effects of this drug on the adult worms are still being investigated. It is probable that patients will need to continue using DEC to kill the adult worms. Mild side effects of Ivermectin include headache, fever, and myalgia.

Other means of managing lymphatic filariasis are pressure bandages to wrap the swollen limb and elastic stockings to help reduce the pressure. Exercising and elevating a bandaged limb also can help reduce its size.

Surgery can be performed to reduce elephantiasis by removing excess fatty and fibrous tissue, draining the swelled area, and removing the dead worms.

## Prognosis

With DEC treatment, the prognosis is good for early and mild cases of lymphatic filariasis. The prognosis is poor, however, for heavy parasitic infestations.

## Prevention

The two main ways to control this disease are to take DEC preventively, which has shown to be effective, and to reduce the number of carrier insects in a particular area.

Avoiding mosquito bites with insecticides and insect repellents is helpful, as is wearing protective clothing and using bed netting.

Much effort has been made in cleaning the breeding sites (stagnant water) of mosquitoes near people’s homes in areas where filariasis is found.

Before visiting countries where lymphatic filariasis is found, it would be wise to consult a travel physician to learn about current preventative measures.

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## ORGANIZATIONS

National Institute of Allergies and Infectious Diseases, Division of Microbiology and Infectious Diseases. Building 31, Room. 7A-50, 31 Center Drive MSC 2520, Bethesda, MD 20892. <<http://www.niaid.nih.gov>>.

National Lymphedema Network (NLN). 2211 Post St., Suite 404, San Francisco, CA 94115. (800) 541-3259. <<http://www.hooked.net>>.

National Organization for Rare Disorders. PO Box 8923, New Fairfield, CT 06812-8923. (800) 999-6673. <<http://www.rarediseases.org>>.

Collette L. Placek

Enzyme-linked Immunosorbant (ELISA) see  
**AIDS tests**

# Embolism

## Definition

An embolism is an obstruction in a blood vessel due to a blood clot or other foreign matter that gets stuck while traveling through the bloodstream. The plural of embolism is emboli.

## Description

Emboi have moved from the place where they were formed through the bloodstream to another part of the body, where they obstruct an artery and block the flow of blood. The emboli are usually formed from blood clots but are occasionally comprised of air, fat, or tumor tissue. Embolic events can be multiple and small, or single and massive. They can be life-threatening and require immediate emergency medical care. There are three general categories of emboli: arterial, gas, and pulmonary. Pulmonary emboli are the most common.

## Arterial embolism

In arterial emboli, blood flow is blocked at the junction of major arteries, most often at the groin, knee, or thigh. Arterial emboli are generally a complication of heart disease. An **arterial embolism** in the brain (cere-

bral embolism) causes **stroke**, which can be fatal. An estimated 5–14% of all strokes are caused by cerebral emboli. Arterial emboli to the extremities can lead to tissue **death** and **amputation** of the affected limb if not treated effectively within hours. Intestines and kidneys can also suffer damage from emboli.

## Gas embolism

Gas emboli result from the compression of respiratory gases into the blood and other tissues due to rapid changes in environmental pressure, for example, while flying or scuba diving. As external pressure decreases, gases (like nitrogen) that are dissolved in the blood and other tissues become small bubbles that can block blood flow and cause organ damage.

## Pulmonary embolism

In a **pulmonary embolism**, a common illness, blood flow is blocked at a pulmonary artery. When emboli block the main pulmonary artery, and in cases where there are no initial symptoms, a pulmonary embolism can quickly become fatal. According to the American Heart Association, an estimated 600,000 Americans develop pulmonary emboli annually and 60,000 die from it.

A pulmonary embolism is difficult to diagnose. Less than 10% of patients who die from a pulmonary embolism were diagnosed with the condition. More than 90% of cases of pulmonary emboli are complications of **deep vein thrombosis**, blood clots in the deep vein of the leg or pelvis.

## Causes and symptoms

Arterial emboli are usually a complication of heart disease where blood clots form in the heart's chambers. Gas emboli are caused by rapid changes in environmental pressure that could happen when flying or scuba diving. A pulmonary embolism is caused by blood clots that travel through the blood stream to the lungs and block a pulmonary artery. More than 90% of the cases of pulmonary embolism are a complication of deep vein thrombosis, which typically occurs in patients who have had **orthopedic surgery** and patients with **cancer** or other chronic illnesses like congestive **heart failure**.

Risk factors for arterial and pulmonary emboli include: prolonged bed rest, surgery, **childbirth**, **heart attack**, stroke, congestive heart failure, cancer, **obesity**, a broken hip or leg, **oral contraceptives**, sickle cell anemia, chest trauma, certain congenital heart defects, and old age. Risk factors for gas emboli include: scuba diving, amateur plane flight, **exercise**, injury, obesity, **dehy-**



**A close up view of a pulmonary embolism.** (Custom Medical Stock Photo. Reproduced by permission.)

**dration**, excessive alcohol, colds, and medications such as narcotics and **antihistamines**.

Common symptoms of a pulmonary embolism include:

- labored breathing, sometimes accompanied by chest pain
- a rapid pulse
- a **cough** that may produce sputum
- a low-grade **fever**
- fluid build-up in the lungs

Less common symptoms include:

- coughing up blood
- pain caused by movement or breathing
- leg swelling
- bluish skin
- fainting
- swollen neck veins

Symptoms of an arterial embolism include:

- severe pain in the area of the embolism
- pale, bluish cool skin
- numbness
- tingling
- muscular weakness or **paralysis**

## Diagnosis

An embolism can be diagnosed through the patient's history, a physical exam, and diagnostic tests. For arterial emboli, cardiac ultrasound and/or arteriography are ordered. For a pulmonary embolism, a **chest x ray**, lung scan, pulmonary **angiography**, **electrocardiography**,

arterial blood gas measurements, and **venography** or venous ultrasound could be ordered.

### Diagnosing an arterial embolism

Ultrasound uses sound waves to create an image of the heart, organs, or arteries. The technician applies gel to a hand-held transducer then presses it against the patient's body. The ultrasound's sound waves arteries are converted into an image that can be displayed on a monitor. Performed in an outpatient diagnostic laboratory, the test takes 30–60 minutes.

An arteriogram is an x ray in which a contrast medium is injected to make the arteries visible on the x ray. It can be performed in a radiology unit, outpatient clinic, or diagnostic center of a hospital.

### Diagnosing a pulmonary embolism

A chest x ray can show fluid build-up and detect other respiratory diseases. The perfusion lung scan shows poor flow of blood in areas beyond blocked arteries. The patient inhales a small amount of radiopharmaceutical and pictures of airflow into the lungs are taken with a gamma camera. Then a different radiopharmaceutical is injected into an arm vein and lung blood flow is scanned. A normal result essentially rules out a pulmonary embolism. A lung scan can be performed in a hospital or an outpatient facility and takes about 45 minutes.

Pulmonary angiography is the most reliable test for diagnosing a pulmonary embolism but it is not used often because it is expensive, invasive, and not readily available in most hospitals. Pulmonary angiography is a radiographic test which involves injection of a radio contrast agent to show the pulmonary arteries. A cinematic camera records the blood flow through the patient, who lies on a table. Pulmonary angiography is usually performed in a hospital's radiology medicine department and takes 30–60 minutes.

An electrocardiograph shows the heart's electrical activity and helps distinguish a pulmonary embolism from a heart attack. Electrodes covered with conducting jelly are placed on the patient's chest, arms, and legs. Impulses of the heart's activity are traced on paper. The test takes about 10 minutes.

Arterial blood gas measurements are sometimes helpful but, alone, they are not diagnostic for pulmonary embolism. Blood is taken from an artery instead of a vein, usually in the wrist.

Venography is used to look for the most likely source of a pulmonary embolism, deep vein thrombosis. It is very accurate, but it is not used often, because it is painful, expensive, exposes the patient to a fairly high dose of radiation, and can cause complications. Venogra-

phy identifies the location, extent, and degree of attachment of the blood clots and enables the condition of the deep leg veins to be assessed. A contrast solution is injected into a foot vein through a catheter. The physician observes the movement of the solution through the vein with a fluoroscope while a series of x rays are taken. Venography takes between 30–45 minutes and can be done in a physician's office, a laboratory, or a hospital. Radionuclide venography, in which a radioactive isotope is injected, is occasionally used, especially if a patient has had reactions to contrast solutions. Venous ultrasound is the preferred evaluation of leg veins.

## Treatment

Patients with emboli require immediate hospitalization. They are generally treated with clot-dissolving and/or clot-preventing drugs. **Thrombolytic therapy** to dissolve blood clots is the definitive treatment for a very severe pulmonary embolism. Streptokinase, urokinase, and recombinant tissue plasminogen activator (TPA) are used. Heparin is the anticoagulant drug of choice for preventing formation of blood clots. Warfarin, an oral anti-coagulant, is sometimes used concurrently and is usually continued after the hospitalization.

In the case of an arterial embolism, the affected limb is placed in a dependent position and kept warm. Embolectomy is the treatment of choice in the majority of early cases of arterial emboli in the extremities. In this procedure, a balloon-tipped catheter is inserted into the artery to remove thromboembolic matter.

With a pulmonary embolism, oxygen therapy is often used to maintain normal oxygen concentrations. For people who can't take anticoagulants and in some other cases, surgery may be needed to insert a device that filters blood returning to the heart and lungs.

## Prognosis

Of patients hospitalized with an arterial embolism, 25–30% die, and 5–25% require amputation of a limb. About 10% of patients with a pulmonary embolism die suddenly within the first hour of onset of the condition. The outcome for all other patients is generally good; only 3% of patients die who are properly diagnosed early and treated. In cases of an undiagnosed pulmonary embolism, about 30% of patients die.

## Prevention

Embolism can be prevented in high risk patients through antithrombotic drugs such as heparin, venous interruption, gradient elastic stockings, and intermittent pneumatic compression of the legs. The combination of

## KEY TERMS

**Anticoagulants**—Drugs that suppress, delay, or prevent blood clots. Anticoagulants are used to treat embolisms.

**Artery**—A blood vessel that carries blood from the heart to other body tissues. Embolisms obstruct arteries.

**Deep vein thrombosis**—A blood clot in the calf's deep vein. This frequently leads to pulmonary embolism if untreated.

**Embo**—Clots or other substances that travel through the blood stream and get stuck in an artery, blocking circulation.

**Thrombolytics**—Drugs that dissolve blood clots. Thrombolytics are used to treat embolisms.

graduated compression stockings and low-dose heparin is significantly more effective than low-dose heparin alone.

Gradient elastic stockings, also called anti-embolism stockings, decrease the risk of blood clots by compressing superficial leg veins and forcing blood into the deep veins. They can be knee-, thigh-, or waist-length. Many physicians order the use of stockings before surgery and until there is no longer an elevated risk of developing blood clots. The risk of deep vein thrombosis after surgery is reduced 50% with the use of these stockings. The American Heart Association recommends that the use of graduated compression stockings be considered for all high-risk surgical patients.

Intermittent pneumatic compression involves wrapping knee- or thigh-high cuffs around the legs to prevent blood clots. The cuffs are connected to a pump which inflates and deflates, mimicking the heart's normal pumping action and reducing the pooling of blood. Intermittent pneumatic compression can be used during surgery and recovery and continues until there is no longer an elevated risk of developing blood clots. The American Heart Association recommends the use of intermittent pneumatic compression for patients who cannot take anticoagulants, for example, spinal cord and brain trauma patients.

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Lori De Milto

**EMG** see **Electromyography**

**Emollient bath** see **Therapeutic baths**

## Emphysema

### Definition

Emphysema is a chronic respiratory disease where there is over-inflation of the air sacs (alveoli) in the lungs, causing a decrease in lung function, and often, breathlessness.

### Description

Emphysema is the most common cause of **death** from respiratory disease in the United States, and is the fourth most common cause of death overall. There are

1.8 million Americans with the disease, which ranks fifteenth among chronic conditions that cause limitations of activity. The disease is usually caused by **smoking**, but a small number of cases are caused by an inherited defect.

Normally functioning lungs are elastic, efficiently expanding and recoiling as air passes freely through the bronchus to the alveoli, where oxygen is moved into the blood and carbon dioxide is filtered out. When a person inhales cigarette smoke or certain other irritants, his or her immune system responds by releasing substances that are meant to defend the lungs against the smoke. These substances can also attack the cells of the lungs, but the body normally inhibits such action with the release of other substances. In smokers and those with the inherited defect, however, no such prevention occurs and the lung tissue is damaged in such a way that it loses its elasticity. The small passageways (bronchioles) leading to the alveoli collapse, trapping air within the alveoli. The alveoli, unable to recoil efficiently and move the air out, over expand and rupture. As the disease progresses, coughing and **shortness of breath** occur. In the later stages, the lungs cannot supply enough oxygen to the blood. Emphysema often occurs with other respiratory diseases, particularly chronic **bronchitis**. These two diseases are often referred to as one disorder called chronic obstructive pulmonary disease (COPD).

Emphysema is most common among people aged 50 and older. Those with inherited emphysema may experience the onset as early as their thirties or forties. Men are more likely than women to develop emphysema, but female cases are increasing as the number of female smokers rises.

### Causes and symptoms

Heavy cigarette smoking causes about 80–90% of all emphysema cases. However a few cases are the result of an inherited deficiency of a substance called alpha-1-antitrypsin (AAT). The number of Americans with this deficiency is relatively small, probably no greater than 70,000. Pipe, cigar, and marijuana smoking can also damage the lungs. While a person may be less likely to inhale cigar and pipe smoke, these types of smoke can also impair lung function. Marijuana smoke may be even more damaging because it is inhaled deeply and held in by the smoker.

The symptoms of emphysema develop gradually over many years. It is a common occurrence for many emphysema patients to have lost over half of their functioning lung tissue before they become aware that something is wrong. Shortness of breath, a chronic mild **cough** (which may be productive of large amounts of dark, thick sputum, and often dismissed as "smoker's cough"), and sometimes weight loss are associated with emphysema.

Initially, a patient may only notice shortness of breath when he or she is exercising. However, as the disease progresses, it will occur with less exertion or no exertion at all. Emphysema patients may also develop an enlarged, or “barrel,” chest. Other symptoms may be skipped breaths, difficulty sleeping, morning headaches, increased difficulty breathing while lying down, chronic **fatigue**, and swelling of the feet, ankles, or legs. Those with emphysema are at risk for a variety of other complications resulting from weakened lung function, including **pneumonia**.

### Diagnosis

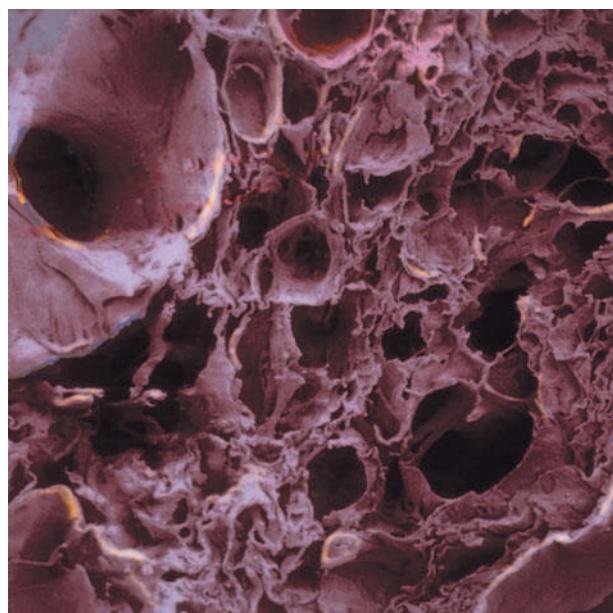
A variety of pulmonary function tests may be ordered. In the early stages of emphysema, the only result may be dysfunction of the small airways. Patients with emphysema may show an increase in the total amount of air that is in the lungs (total lung capacity), but a decrease in the amount of air that can be breathed out after taking a deep breath (vital capacity). With severe emphysema, vital capacity is substantially below normal. Spirometry, a procedure that measures air flow and lung volume, helps in the diagnosis of emphysema.

A **chest x ray** is often ordered to aid in the diagnosis of emphysema, though patients in the early stages of the disease may have normal findings. Abnormal findings on the chest x ray include over-inflation of the lungs and an abnormally increased chest diameter. The diaphragm may appear depressed or flattened. In addition, patients with advanced emphysema may show a smaller or vertical heart. The physician may observe blisters in the lungs and bulging of the accessory muscles of the respiratory system. Late in the disease, an EKG will show signs of right ventricular failure in the heart and increased hemoglobin due to lower levels of oxygen in the patient’s blood.

### Treatment

Treatment methods for emphysema do not cure or reverse the damage to the lungs. However, they may slow the progression of the disease, relieve symptoms, and help control possibly fatal complications. The first step in treatment for smokers is to quit, so as to prevent any further deterioration of breathing ability. Smoking cessation programs may be effective. Consistent encouragement along with the help of health care professionals as well as family and friends can help increase the success rate of someone attempting to quit.

If the patient and the health care team develop and maintain a complete program of respiratory care, disability can be decreased, acute episodes of illness may be prevented, and the number of hospitalizations reduced. However, only quitting smoking has been shown to slow down the progression of the disease, and among all other



**A scanning electron microscopy (SEM) of lung tissue indicating emphysema.** (Photograph by Hossler, Ph.D., Custom Medical Stock Photo. Reproduced by permission.)

treatments, only oxygen therapy has shown an increase in the survival rate.

Home oxygen therapy may improve the survival times in those patients with advanced emphysema who also have low blood oxygen levels. It may improve the patient’s tolerance of **exercise**, as well as improve their performance in certain aspects of brain function and muscle coordination. The functioning of the heart may also improve with an increased concentration of oxygen in the blood. Oxygen may also decrease **insomnia** and headaches. Some patients may only receive oxygen at night, but studies have illustrated that it is most effective when administered at least 18, but preferably 24 hours per day. Portable oxygen tanks prescribed to patients carry a limited supply and must be refilled on a regular basis by a home health provider. Medicare and most insurance companies cover a large proportion of the cost of home oxygen therapy. Patients should be instructed regarding special safety issues involving the transport and presence of oxygen in the home.

A variety of medications may be used in the treatment of emphysema. Usually the patient responds best to a combination of medicines, rather than one single drug.

**Bronchodilators** are sometimes used to help alleviate the patient’s symptoms by relaxing and opening the airways. They can be inhaled, taken by mouth, or injected. Another category of medication often used is **corticosteroids** or steroids. These help to decrease the



**X ray showing emphysema in the lungs.** (Photo Researchers. Reproduced by permission.)

inflammation of the airway walls. They are occasionally used if bronchodilators are ineffective in preventing airway obstruction. Some patients' lung function improves with corticosteroids, and inhaled steroids may be beneficial to patients with few side effects. A variety of **antibiotics** are frequently given at the first sign of a respiratory infection, such as increased amounts of sputum, or if there has been a change in the color of the sputum. **Expectorants** can help to loosen respiratory secretions, enabling the patient to more easily expel them from the airways.

Many of the medications prescribed involve the use of a metered dose inhaler (MDI) that may require special instruction to be used correctly. MDIs are a convenient and safe method of delivering medication to the lungs. However, if they are used incorrectly the medication will not get to the right place. Proper technique is essential for the medication to be effective.

For some patients, surgical treatment may be the best option. Lung volume reduction surgery is a surgical procedure in which the most diseased parts of the lung are removed to enable the remaining lung and breathing muscles to work more efficiently. Preliminary studies suggest improved survival rates and better functioning with the surgery. Another surgical procedure used for emphysema patients is **lung transplantation**. Transplantation may involve one or both lungs. However, it is a risky and expensive procedure, and donor organs may not be available.

For those patients with advanced emphysema, keeping the air passages reasonably clear of secretions can prove difficult. Some common methods for mobilizing and removing secretions include:

- Postural drainage. This helps to remove secretions from the airways. The patient lies in a position that allows gravity to aid in draining different parts of the lung. This is often done after the patient inhales an aerosol medication. The basic position involves the patient lying on the bed with his chest and head over the side and the forearms resting on the floor.
- Chest percussion. This technique involves lightly clapping the back and chest, and may help to loosen thick secretions.
- Coughing and deep breathing. These techniques may aid the patient in bringing up secretions.
- Aerosol treatments. These treatments may involve solutions of saline, often mixed with a bronchodilator, which are then inhaled as an aerosol. The aerosols thin and loosen secretions. A treatment normally takes 10 to 15 minutes, and is given three or four times a day.

Patients with COPD can learn to perform a variety of self-help measures that may help improve their symptoms and their ability to participate in everyday activities. These measures include:

- Avoiding any exposure to dusts and fumes.
- Avoiding air pollution, including the cigarette smoke of others.
- Avoiding other people who have infections like the cold or flu. Get a pneumonia **vaccination** and a yearly flu shot.
- Drinking plenty of fluids. This helps to loosen respiratory secretions so they can be brought up more easily through coughing.
- Avoiding extreme temperatures of heat or cold. Also avoiding high altitudes. (Special precautions can be taken that may enable the emphysema patient to fly on a plane.)
- Maintaining adequate nutritional intake. Normally a high protein diet taken in many small feedings is recommended.

### Alternative treatment

Many patients are interested in whether any alternative treatments for emphysema are available. Some practitioners recommend supplements of antioxidant nutrients. There have also been some studies indicating a correlation between a low Vitamin A levels and COPD, with suggestions that supplements of vitamin A might be ben-

## KEY TERMS

**Alveoli**—Small cells or cavities. In the lungs, these are air sacs where oxygen enters the blood and carbon dioxide is filtered out.

**Pulmonary**—Related to or associated with the lungs.

eficial. Aromatherapists have used essential oils like eucalyptus, lavender, pine, and rosemary to help relieve nasal congestion and make breathing easier. The herb elecampane may act as an expectorant to help patients clear mucus from the lungs. The patient should discuss these remedies with their health care practitioner prior to trying them, as some may interact with the more traditional treatments that are already being used.

### Prognosis

Emphysema is a serious and chronic disease that cannot be reversed. If detected early, the effects and progression can be slowed, particularly if the patient stops smoking immediately. Complications of emphysema include higher risks for pneumonia and acute bronchitis. Overall, the prognosis for patients with emphysema is poor, with a survival rate for all those with COPD of four years, and even less for emphysema. However, individual cases vary and many patients can live much longer with supplemental oxygen and other treatment measures.

### Prevention

The best way to prevent emphysema is to avoid smoking. Even patients with inherited emphysema should avoid smoking, as it especially worsens the onset and severity. If patients quit smoking as soon as evidence of small airway obstruction begins, they can significantly improve their prognosis.

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

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## Empyema

### Definition

Empyema is a condition in which pus and fluid from infected tissue collects in a body cavity. The name comes from the Greek word *empyein* meaning pus-producing (suppurate). Empyema is most often used to refer to collections of pus in the space around the lungs (pleural cavity), but sometimes refers to similar collections in the gall bladder or the pelvic cavity. Empyema in the pleural cavity is sometimes called empyema thoracis, or empyema of the chest, to distinguish it from empyema elsewhere in the body.

### Description

Empyema may have a number of causes but is most frequently a complication of **pneumonia**. Its development can be divided into three phases: an acute phase in which the body cavity fills with a thin fluid containing some pus; a second stage in which the fluid thickens and a fibrous, coagulation protein (fibrin) begins to accumulate within the cavity; and a third or chronic stage in which the lung or other organ is encased within a thick covering of fibrous material.

### Causes and symptoms

Empyema thoracis can be caused by a number of different organisms, including bacteria, fungi, and amebas, in connection with pneumonia, chest **wounds**, chest surgery, lung abscesses, or a ruptured esophagus. The infective organism can get into the pleural cavity either through the bloodstream or other circulatory system, in secretions from lung tissue, or on the surfaces of surgical instruments or objects that cause open chest wounds. The most common organisms that cause empyema are the following bacteria: *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Staphylococcus aureus*. *S. aureus* is the most common cause in all age groups, accounting for 90% of cases of empyema in infants and children. Pelvic empyema in

## KEY TERMS

**Abscess**—An area of inflamed and injured body tissue that fills with pus.

**Decortication**—Surgical removal of the fibrous peel that covers the lungs in third-stage empyema.

**Empyema**—The collection of pus in a body cavity, particularly the lung or pleural cavity.

**Fibrin**—A fibrous blood protein vital to coagulation and blood clot formation.

**Percussion**—A diagnostic technique in which the back, chest, or abdomen is tapped to determine whether body cavities contain abnormal fluid.

**Pleural cavity**—The space surrounding the lungs, including the membranes covering the lungs and lining the inside of the chest wall.

**Pneumonia**—Inflammation of the lungs usually caused by a virus, bacteria, or other organism.

**Resection**—The surgical removal of part of an organ or body structure, as in rib resection.

**Suppurate**—To produce or discharge pus.

**Thoracentesis**—A procedure in which fluid is withdrawn from the pleural cavity through a needle inserted between the ribs. The fluid may be withdrawn either for diagnostic tests or to drain the cavity.

**Video-assisted thoracic surgery (VATS)**—A technique used to aid in the placement of chest tubes or when performing decortications when treating advanced empyema.

women is most often caused by *Bacteroides* strains or *Pseudomonas aeruginosa*. In elderly, chronically ill, or alcoholic patients, empyema is often caused by *Klebsiella pneumoniae* species of bacteria.

When the disease organisms arrive in the cavity surrounding the lungs, they infect the tissues that cover the lungs and line the chest wall. As the body attempts to fight off the infection, the cavity fills up with tissue fluid, pus, and dead tissue cells. Empyema of the gall bladder or pelvis results from similar reactions to infection in those parts of the body.

The signs and symptoms of empyema vary somewhat according to the location of the infection and its severity. In empyema thoracis, patients usually exhibit symptoms of pneumonia, including **fever**, **cough**, **fatigue**, **shortness of breath**, and chest **pain**. They may

prefer to lie on the side of the body affected by the empyema. Family members may notice **bad breath**. In severe cases, the patient may become dehydrated, cough up blood or greenish-brown sputum, run a fever as high as 105°F (40.6°C), or fall into a **coma**.

Patients with thoracic empyema may develop potentially life-threatening complications if the condition is not treated. The infected tissues may develop large collections of pus (abscesses) that can rupture into the patient's airway, or the infection may spread to the tissues surrounding the heart. In extreme cases the empyema may spread to the brain by means of bacteria carried in the bloodstream.

In pelvic empyema, the infection produces large amounts of thick, foul-smelling pus that is rapidly replaced even after drainage. Empyema of the gallbladder is marked by intense pain on the upper right side of the abdomen, high fever, and rigidity of the muscles over the infected area.

## Diagnosis

A physician may consider the possibility of empyema thoracis in patients with pneumonia or other symptoms of lung infection. When listening to sounds within the patient's chest with a stethoscope, the sounds of breathing will be partly muffled and harder to hear in the patients with empyema. The area of the chest over the infection will sound dull when tapped or thumped (percussed). On an x ray, empyema thoracis will appear as a cloudy or opaque area. The amount of fluid present in the pleural cavity can be estimated using an ultrasound imaging procedure. The diagnosis of empyema, however, has to be confirmed with laboratory tests because its symptoms can be caused by other disease conditions.

The diagnosis of empyema is usually confirmed by analyzing a sample of fluid taken from the pleural cavity. The sample is obtained by a procedure called **thoracentesis**. In this procedure, the patient is given a local anesthetic, a needle is inserted into the pleural cavity through the back between the ribs on the infected side, and a sample of fluid is withdrawn. If the patient has empyema, there will be a very high level of one particular kind of immune cell (white blood cells), a high level of protein, and a very low level of blood sugar. The fluid can also be tested for the specific disease organism by staining or tissue cultures. In some cases, the color, smell, or consistency of the tissue fluid also helps to confirm the diagnosis.

## Treatment

Empyema is treated using a combination of medications and surgical techniques. Treatment with medication involves intravenously administering a two-week course

of **antibiotics**. It is important to give antibiotics as soon as possible to prevent first-stage empyema from progressing to its later stages. The antibiotics most commonly used are penicillin and vancomycin. Patients experiencing difficulty breathing are also given oxygen therapy.

Surgical treatment of empyema has two goals: drainage of the infected fluid and closing up of the space left in the pleural cavity. If the infection is still in its early stages, the fluid can be drained by thoracentesis. In second-stage empyema, the surgeon will insert a chest tube in the patient's rib cage or remove part of a rib (rib resection) in order to drain the fluid. In third-stage empyema, the surgeon may cut or peel away the thick fibrous layer coating the lung. This procedure is called decortication. When the fibrous covering is removed, the lung will expand to fill the space in the chest cavity. The doctor can use video-assisted **thoracic surgery** (VATS) techniques to position the chest tube or to perform a limited decortication. The VATS technique allows a physician to see within the body during certain surgical procedures. Empyema of the gallbladder is a serious condition that is treated with intravenous antibiotics and surgical removal of the gallbladder.

## Prognosis

The prognosis for recovery is generally good, except in those cases with complications, such as a **brain abscess** or blood **poisoning**, or cases caused by certain types of streptococci.

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Rebecca J. Frey

Enalapril see **Angiotensin-converting enzyme inhibitors**

## Encephalitis

### Definition

Encephalitis is an inflammation of the brain, usually caused by a direct viral infection or a hypersensitivity reaction to a virus or foreign protein. Brain inflammation caused by a bacterial infection is sometimes called cerebritis. When both the brain and spinal cord are involved, the disorder is called encephalomyelitis. An inflammation of the brain's covering, or meninges, is called **meningitis**.

### Description

Encephalitis is an inflammation of the brain. The inflammation is a reaction of the body's immune system to infection or invasion. During the inflammation, the brain's tissues become swollen. The combination of the infection and the immune reaction to it can cause **headache** and a **fever**, as well as more severe symptoms in some cases.

Approximately 2,000 cases of encephalitis are reported to the Centers for Disease Control in Atlanta, GA each year. The viruses causing primary encephalitis can be epidemic or sporadic. The **polio** virus is an epidemic cause. Arthropod-borne viral encephalitis is responsible for most epidemic viral encephalitis. The viruses live in animal hosts and mosquitos that transmit the disease. The most common form of non-epidemic or sporadic encephalitis is caused by the herpes simplex virus, type 1 (HSV-1) and has a high rate of **death**. **Mumps** is another example of a sporadic cause.

### Causes and symptoms

#### Causes

There are more than a dozen viruses that can cause encephalitis, spread by either human-to human contact or by animal bites. Encephalitis may occur with several common viral infections of childhood. Viruses and viral diseases that may cause encephalitis include:

- **chickenpox**
- **measles**
- **mumps**
- Epstein-Barr virus (EBV)

- cytomegalovirus infection
- HIV
- herpes simplex
- herpes zoster (**shingles**)
- herpes B
- polio
- **rabies**
- mosquito-borne viruses (arboviruses)

Primary encephalitis is caused by direct infection by the virus, while secondary encephalitis is due to a post-infectious immune reaction to viral infection elsewhere in the body. Secondary encephalitis may occur with measles, chickenpox, mumps, **rubella**, and EBV. In secondary encephalitis, symptoms usually begin five to 10 days after the onset of the disease itself and are related to the breakdown of the myelin sheath that covers nerve fibers.

In rare cases, encephalitis may follow **vaccination** against some of the viral diseases listed above. **Creutzfeldt-Jakob disease**, a very rare brain disorder caused by an infectious particle called a prion, may also cause encephalitis.

Mosquitoes spread viruses responsible for equine encephalitis (eastern and western types), St. Louis encephalitis, California encephalitis, and **Japanese encephalitis**. **Lyme disease**, spread by ticks, can cause encephalitis, as can Colorado tick fever. Rabies is most often spread by animal bites from dogs, cats, mice, raccoons, squirrels, and bats and may cause encephalitis.

Equine encephalitis is carried by mosquitoes that do not normally bite humans but do bite horses and birds. It is occasionally picked up from these animals by mosquitoes that do bite humans. Japanese encephalitis and St. Louis encephalitis are also carried by mosquitoes. The risk of contracting a mosquito-borne virus is greatest in mid- to late summer, when mosquitoes are most active, in those rural areas where these viruses are known to exist. Eastern equine encephalitis occurs in eastern and southeastern United States; western equine and California encephalitis occur throughout the West; and St. Louis encephalitis occurs throughout the country. Japanese encephalitis does not occur in the United States, but is found throughout much of Asia. The viruses responsible for these diseases are classified as arbovirus and these diseases are collectively called **arbovirus encephalitis**.

Herpes simplex encephalitis, the most common form of sporadic encephalitis in western countries, is a disease with significantly high mortality. It occurs in children and adults and both sides of the brain are affected. It is theorized that brain infection is caused by the virus moving

from a peripheral location to the brain via two nerves, the olfactory and the trigeminal (largest nerves in the skull).

Herpes simplex encephalitis is responsible for 10% of all encephalitis cases and is the main cause of sporadic, fatal encephalitis. In untreated patients, the rate of death is 70% while the mortality is 15–20% in patients who have been treated with acyclovir. The symptoms of herpes simplex encephalitis are fever, rapidly disintegrating mental state, headache, and behavioral changes.

### Symptoms

The symptoms of encephalitis range from very mild to very severe and may include:

- headache
- fever
- lethargy (sleepiness, decreased alertness, and **fatigue**)
- malaise
- nausea and vomiting
- visual disturbances
- tremor
- decreased consciousness (drowsiness, confusion, **delirium**, and unconsciousness)
- stiff neck
- seizures

Symptoms may progress rapidly, changing from mild to severe within several days or even several hours.

### Diagnosis

Diagnosis of encephalitis includes careful questioning to determine possible exposure to viral sources. Tests that can help confirm the diagnosis and rule out other disorders include:

- Blood tests. These are to detect antibodies to viral antigens, and foreign proteins.
- Cerebrospinal fluid analysis (spinal tap). This detects viral antigens, and provides culture specimens for the virus or bacteria that may be present in the cerebrospinal fluid.
- Electroencephalogram (EEG).
- CT and MRI scans.

A **brain biopsy** (surgical gathering of a small tissue sample) may be recommended in some cases where treatment to date has been ineffective and the cause of the encephalitis is unclear. Definite diagnosis by biopsy may allow specific treatment that would otherwise be too risky.

## Treatment

Choice of treatment for encephalitis will depend on the cause. Bacterial encephalitis is treated with **antibiotics**. Viral encephalitis is usually treated with **antiviral drugs** including acyclovir, ganciclovir, foscarnet, ribavarin, and AZT. Viruses that respond to acyclovir include herpes simplex, the most common cause of sporadic (non-epidemic) encephalitis in the United States.

The symptoms of encephalitis may be treated with a number of different drugs. **Corticosteroids**, including prednisone and dexamethasone, are sometimes prescribed to reduce inflammation and brain swelling. **Anticonvulsant drugs**, including dilantin and phenytoin, are used to control seizures. Fever may be reduced with **acetaminophen** or other fever-reducing drugs.

A person with encephalitis must be monitored carefully, since symptoms may change rapidly. Blood tests may be required regularly to track levels of fluids and salts in the blood.

## Prognosis

Encephalitis symptoms may last several weeks. Most cases of encephalitis are mild, and recovery is usually quick. Mild encephalitis usually leaves no residual neurological problems. Overall, approximately 10% of those with encephalitis die from their infections or complications such as secondary infection. Some forms of encephalitis have more severe courses, including herpes encephalitis, in which mortality is 15–20% with treatment, and 70–80% without. Antiviral treatment is ineffective for eastern equine encephalitis, and mortality is approximately 30%.

Permanent neurological consequences may follow recovery in some cases. Consequences may include personality changes, memory loss, language difficulties, seizures, and partial **paralysis**.

## Prevention

Because encephalitis is due to infection, it may be prevented by avoiding the infection. Minimizing contact with others who have any of the viral illness listed above may reduce the chances of becoming infected. Most infections are spread by hand-to-hand or hand-to-mouth contact; frequent hand washing may reduce the likelihood of infection if contact cannot be avoided.

Mosquito-borne viruses may be avoided by preventing mosquito bites. Mosquitoes are most active at dawn and dusk, and are most common in moist areas with standing water. Minimizing exposed skin and use of mosquito repellents on other areas can reduce the chances of being bitten.

## KEY TERMS

**Cerebrospinal fluid analysis**—A analysis that is important in diagnosing diseases of the central nervous system. The fluid within the spine will indicate the presence of viruses, bacteria, and blood. Infections such as encephalitis will be indicated by an increase of cell count and total protein in the fluid.

**Computerized tomography (CT) Scan**—A test to examine organs within the body and detect evidence of tumors, blood clots, and accumulation of fluids.

**Electroencephalogram (EEG)**—A chart of the brain waves picked up by the electrodes placed on the scalp. Changes in brain wave activity can be an indication of nervous system disorders.

**Inflammation**—A response from the immune system to an injury. The signs are redness, heat, swelling, and pain.

**Magnetic Resonance Imaging (MRI)**—MRI is diagnostic radiography using electromagnetic energy to create an image of the central nervous system (CNS), blood system, and musculoskeletal system.

**Vaccine**—A preparation containing killed or weakened microorganisms used to build immunity against infection from that microorganism.

**Virus**—A very small organism that can only live within a cell. They are unable to reproduce outside that cell.

Vaccines are available against some viruses, including polio, herpes B, Japanese encephalitis, and equine encephalitis. Rabies vaccine is available for animals; it is also given to people after exposure. Japanese encephalitis vaccine is recommended for those traveling to Asia and staying in affected rural areas during transmission season.

## Resources

### BOOKS

*Harrison's Principles of Internal Medicine*. Ed. Anthony S.

Fauci, et al. New York: McGraw-Hill, 1997.

*Hurst, J. Willis. Medicine for the Practicing Physician*. Stamford: Appleton & Lange, 1988.

*Office Practice of Neurology*. Ed. Martin Samuels and Steven Feske. New York: Churchill Livingstone, 1996.

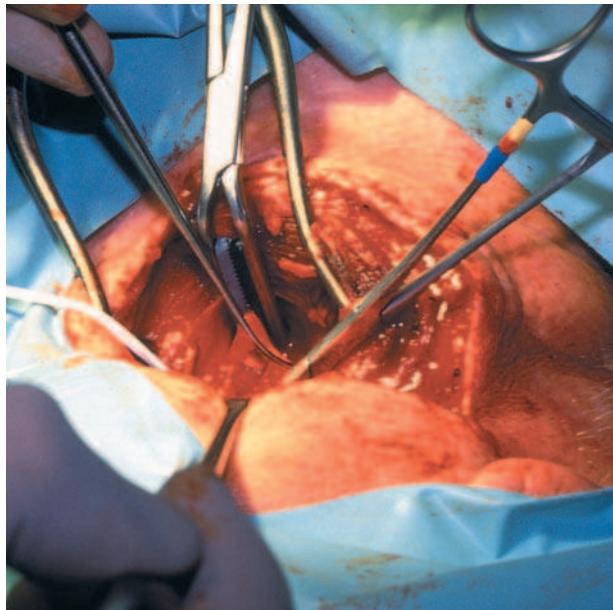
*The Merck Manual*. Whitehouse Station, NJ: Merck and Co. Inc., 1996.

**ORGANIZATIONS**

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

Richard Robinson

Encephalocele see **Congenital brain defects**



## Endarterectomy

### Definition

Endarterectomy is an operation to remove or bypass the fatty deposits, or blockage, in an artery narrowed by the buildup of fatty tissue (**atherosclerosis**).

### Purpose

Removing the fatty deposits restores normal blood flow to the part of the body supplied by the artery. An endarterectomy is performed to treat cerebrovascular disease in which there is a serious reduction of blood supply to the brain (carotid endarterectomy), or to treat **peripheral vascular disease** (impaired blood supply to the legs).

Endarterectomy is most often performed on one of the two main arteries in the neck (the carotids) opening the narrowed arteries leading to the brain. When performed by an experienced surgeon, the practice is extremely effective, reducing the risk of **stroke** by up to 70%. Recent studies indicate it is effective in preventing stroke, even among those patients who had no warning signs except narrowed arteries detected by their doctors on a routine exam.

### Precautions

Before the surgery, a full medical exam is usually done to assess any specific health problems, such as diabetes, high blood pressure, heart disease, or stroke. If possible, reversible health problems, such as cigarette **smoking** or being overweight, should be corrected.

### Description

#### *Carotid artery disease*

Every person has four carotid arteries (the internal and external carotids on each side of the neck) through which blood from the heart moves into the brain. If one of these arteries becomes blocked by fat and cholesterol, the patient may have a range of symptoms, including:

In this procedure, surgeons are removing plaque from the carotid artery. (Custom Medical Stock Photo. Reproduced by permission.)

- weakness in one arm, leg, half of the face, or one entire side of the body
- numbness tingling
- **paralysis** of an arm, leg, or face
- slurred speech
- dizziness
- confusion, **fainting**, or **coma**
- stroke

Removing this fatty buildup, or bypassing a blocked segment, may restore blood flow to the brain, eliminate or decrease the symptoms, and lessen the risk of a stroke.

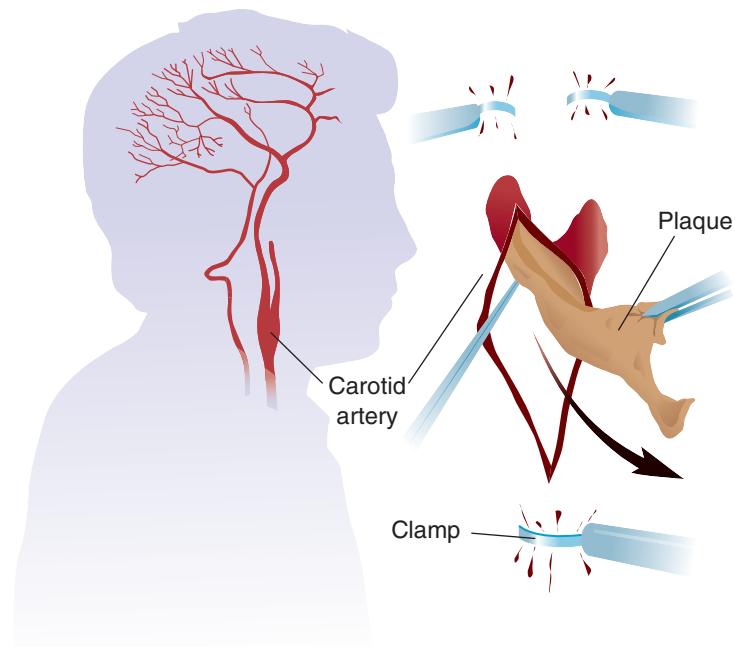
#### *Peripheral vascular disease*

When the blood vessels in the legs (and sometimes the arms) become narrowed, this can restrict blood flow and cause **pain** in the affected area. In severe cases, the tissue may die, requiring **amputation**.

The narrowing is usually caused by buildup of fatty plaques in the vessels, often as the result of smoking, high blood pressure, or poorly-controlled **diabetes mellitus**. The vessels usually narrow slowly, but it's possible for a blood clot to form quickly, causing sudden severe pain in the affected leg or arm.

#### *Procedure*

Endarterectomy is a delicate operation that may require several hours. The surgeon begins by making an



**Plaque is removed from the carotid artery by clamping the artery, cutting the plaque out, and closing the opening back up.**  
*(Illustration by Argosy Inc.)*

incision over the blocked artery and inserting a tube above and below the blockage to redirect the blood flow while the artery is opened.

Next, the surgeon removes the fat and cholesterol buildup, along with any blood clots that have formed, with a blunt dissecting instrument. Then the surgeon bathes the clean wall in salt solution combined with heparin, an anticoagulant. Then the surgeon stitches the artery just enough so that the bypass shunt tube can be removed, and then he/she stitches the artery completely closed. After checking to make sure no blood is leaking, the surgeon next closes the skin incision with stitches.

The operation should improve symptoms, although its long-term effects may be more limited, since arterial narrowing is rarely confined to one area of one artery. If narrowing is a problem throughout the body, arterial reconstructive surgery may be required.

The total cost of an endarterectomy, including diagnostic tests, surgery, hospitalization, and follow-up care, will vary according to hospital, doctor, and area of the country where the operation is performed, but a patient

can expect to pay in the range of \$15,000. Patients who are very young, very old, or very ill, or who need more extensive surgery, may require more expensive treatment.

### Preparation

Before surgery, the doctor pinpoints the location of the narrowed artery with an x-ray procedure called **angiography**. For surgery to be effective, the degree of narrowing should be at least 70%, but it should not be total. Patients undergoing angiography are given a local anesthetic, but the endarterectomy itself requires the use of a general anesthesia.

### Aftercare

After the surgery, the patient spends the first two days lying flat in bed. Patients who have had carotid endarterectomy should not bend the neck sharply during this time. Because the blood flow to the brain is now greatly increased, patients may experience a brief but severe **headache**, or lightheadedness. There may be a slight loss of sensation in the skin, or maybe a droop in

## KEY TERMS

**Carotid arteries**—The four principal arteries of the neck and head. There are two common carotid arteries, each of which divides into the two main branches (internal and external).

**Diabetes mellitus**—A disorder in which the pancreas doesn't produce enough (or any) insulin. As a result, the blood levels of sugar become very high. Among other things, diabetes can lead to the breakdown of small blood vessels and a high risk of atherosclerosis and high blood pressure.

**Stroke**—Damage to the part of the brain caused by an interruption of the blood supply. In some cases, small pieces of plaque in the carotid artery may break loose and block an artery in the brain. A narrowed carotid artery also can be the source of blood clots travelling to the brain, or the artery can become completely clogged, blocking all blood flow to the brain.

the mouth, if any of the nerves in the neck were lightly bruised during surgery. In time, this should correct itself.

### Risks

The amount of risk depends on the hospital, the skill of the surgeon, and the severity of underlying disease. Patients who have just had an acute stroke are at greatest risk. During carotid artery surgery, blood flow is interrupted through the artery, so that paralysis and other stroke symptoms may occur. These may resolve after surgery, or may result in permanent stroke. Paralysis is usually one-sided; other stroke symptoms may include loss of half the field of vision, loss of sensation, double vision, speech problems, and personality changes. Risks of endarterectomy to treat either carotid artery or peripheral vascular disease include:

- reactions to anesthesia
- bleeding
- infection
- blood clots

### Normal results

The results after successful surgery are usually striking. The newly opened artery should help to restore normal blood flow. In carotid endarterectomy, surgery should prevent the risk of brain damage and stroke. However, the

buildup of fat and cholesterol usually affects all arteries, not just the one that was operated on. Affected arteries in other parts of the body may be equally clogged and potentially dangerous. Even arteries that were operated electively will likely begin to clog up again after the surgery.

For this reason, lifestyle changes (no smoking, low fat, low cholesterol diet) are important, especially if diet and lifestyle contributed to the development of the problem in the first place.

## Resources

### BOOKS

"Carotid Endarterectomy." In *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.

### PERIODICALS

"Better Blood Flow: Surgery May Strike Down Stroke Risk." *Prevention* 47 (1 Feb. 1995): 50-52.

### ORGANIZATIONS

National Institute of Neurological Disorders and Stroke. PO Box 5801, Bethesda, MD 20824. (800) 352-9424. <<http://www.ninds.nih.gov/index.htm>>.

National Institute of Neurological Disorders at the Neurology Institute. PO Box 5801, Bethesda, MD 20824.

Carol A. Turkington

Endemic syphilis see **Bejel**

Endocardial resection see **Myocardial resection**

## Endocarditis

### Definition

The endocardium is the inner lining of the heart muscle, which also covers the heart valves. When the endocardium becomes damaged, bacteria from the blood stream can become lodged on the heart valves or heart lining. The resulting infection is known as endocarditis.

### Description

The endocardium lines all four chambers of the heart—two at the top (the right and left atria) and two at the bottom (the right and left ventricles)—through which blood passes as the heart beats. It also covers the four valves (the tricuspid valve, the pulmonary valve, the mitral valve, and the aortic valve), which normally open

and close to allow the blood to flow in only one direction through the heart during each contraction.

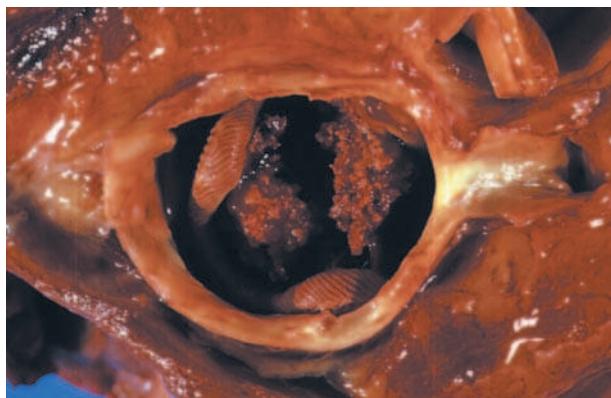
For the heart to pump blood efficiently, the four chambers must contract and relax, and the four valves must open and close, in a well coordinated fashion. By damaging the valves or the walls of the heart chambers, endocarditis can interfere with the ability of the heart to do its job.

Endocarditis rarely occurs in people with healthy, normal hearts. Rather, it most commonly occurs when there is damage to the endocardium. The endocardium may be affected by a congenital heart defect, such as **mitral valve prolapse**, in which blood leaks through a poorly functioning mitral valve back into the heart. It may also be damaged by a prior scarring of the heart muscle, such as **rheumatic fever**, or replacement of a heart valve. Any of these conditions can damage the endocardium and make it more susceptible to infection.

Bacteria can get into the blood stream (a condition known as **bacteremia**) in a number of different ways: It may spread from a localized infection such as a urinary tract infection, **pneumonia**, or skin infection or get into the blood stream as a result of certain medical conditions, such as severe **periodontal disease**, **colon cancer**, or inflammatory bowel disease. It can enter the blood stream during minor procedures, such as periodontal surgery, tooth extractions, teeth cleaning, tonsil removal, prostate removal, or endoscopic examination. It can also be introduced through in-dwelling catheters, which are used for intravenous medications, intravenous feeding, or dialysis. In people who use intravenous drugs, the bacteria can enter the blood stream through unsterilized, contaminated needles and syringes. (People who are prone to endocarditis generally need to take prescribed **antibiotics** before certain surgical or dental procedures to help prevent this infection.)

If not discovered and treated, infective endocarditis can permanently damage the heart muscle, especially the valves. For the heart to work properly, all four valves must be functioning well, opening at the right time to let blood flow in the right direction and closing at the right time to keep the blood from flowing in the wrong direction. If the valve is damaged, this may allow blood to flow backward—a condition known as regurgitation. As a result of a poorly functioning valve, the heart muscle has to work harder to pump blood and may become weakened, leading to **heart failure**. Heart failure is a chronic condition in which the heart is unable to pump blood well enough to supply blood adequately to the body.

Another danger associated with endocarditis is that the vegetation formed by bacteria colonizing on heart valves may break off, forming emboli. These emboli



**A close-up view of an infected artificial heart valve showing bacterial endocarditis (the granulated tissue at center of image). When infection occurs early after surgery, it is likely that organisms have gained entry during the operative period. This type of infection is usually caused by *Staphylococcus epidermidis* and *S. aureus* and is treated with antibiotic drugs. (Photograph by Dr. E. Walker, Photo Researchers, Inc. Reproduced by permission.)**

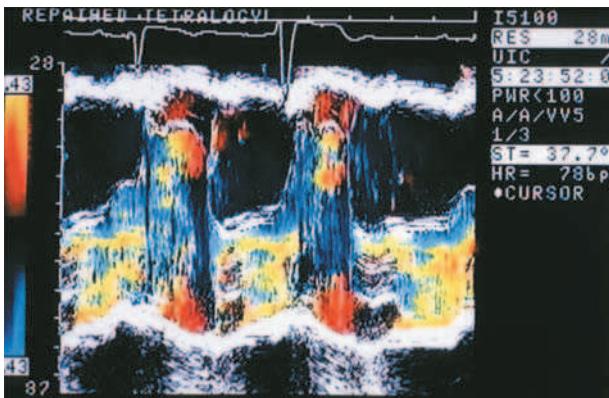
may travel through the circulation and become lodged in blood vessels. By blocking the flow of blood, emboli can starve various tissues of nutrients and oxygen, damaging them. For instance, an embolus lodged in the blood vessels of the lungs may cause pneumonia-like symptoms. An embolus may also affect the brain, damaging nerve tissue, or the kidneys, causing kidney disease. Emboli may also weaken the tiny blood vessels called capillaries, causing hemorrhages (leaking blood vessels) throughout the body.

### Causes and symptoms

Most cases of infective endocarditis occur in people between the ages of 15 and 60, with a median age at onset of about 50 years. Men are affected about twice as often as women are. Other factors that put people at increased risk for endocarditis are congenital heart problems, heart surgery, previous episodes of endocarditis, and intravenous drug use.

While there is no single specific symptom of endocarditis, a number of symptoms may be present. The most common symptom is a mild **fever**, which rarely goes above 102°F (38.9°C). Other symptoms include chills, weakness, **cough**, trouble breathing, headaches, aching joints, and loss of appetite.

Emboli may also cause a variety of symptoms, depending on their location. Emboli throughout the body may cause Osler's nodes, small, reddish, painful bumps most commonly found on the inside of fingers and toes. Emboli may also cause petechiae, tiny purple



**This echocardiogram shows an aortic regurgitation due to endocarditis, an infection of the lining membrane of the cardiac chambers.** (Custom Medical Stock Photo. Reproduced by permission.)

or red spots on the skin, resulting from hemorrhages under the skin's surface. Tiny hemorrhages resembling splinters may also appear under the fingernails or toenails. If emboli become lodged in the blood vessels of the lungs, they may cause coughing or **shortness of breath**. Emboli lodged in the brain may cause symptoms of a mini-stroke, such as numbness, weakness, or **paralysis** on one side of the body or sudden vision loss or double vision. Emboli may also damage the kidneys, causing blood to appear in the urine. Sometimes the capillaries on the surface of the spleen rupture, causing the spleen to become enlarged and tender to the touch. Any-one experiencing any of these symptoms should seek medical help immediately.

## Diagnosis

Doctors begin the diagnosis by taking a history, asking the patient about the symptoms mentioned above. During a **physical examination**, the doctor may also uncover signs such as fever, an enlarged spleen, signs of kidney disease, or hemorrhaging. Listening to the patient's chest with a stethoscope, the doctor may also hear a heart murmur. A heart murmur may indicate abnormal flow of blood through one of the heart chambers or valves.

Doctors take a sample of the patient's blood to test it for bacteria and other microorganisms that may be causing the infection. They usually also use a test called **echocardiography**, which uses ultrasound waves to make images of the heart, to check for abnormalities in the structure of the heart wall or valves. One of the tell-tale signs they look for in echocardiography is vegetation, the abnormal growth of tissue around a valve composed of blood platelets, bacteria, and a clotting protein

called fibrin. Another tell-tale sign is regurgitation, or the backward flow of blood, through one of the heart valves. A normal echocardiogram does not exclude the possibility of endocarditis, but an abnormal echocardiogram can confirm its presence. If an echocardiogram cannot be done or its results are inconclusive, a modified technique called **transesophageal echocardiography** is sometimes performed. Transesophageal echocardiography involves passing an ultrasound device into the esophagus to get a clearer image of the heart.

## Treatment

When doctors suspect infective endocarditis, they will admit the patient to a hospital and begin treating the infection before they even have the results of the **blood culture**. Their choice of antibiotics depends on what the most likely infecting microorganism is. Once the results of the blood culture become available, the doctor can adjust the medications, using specific antibiotics known to be effective against the specific microorganism involved.

Unfortunately, in recent years, the treatment of endocarditis has become more complicated as a result of antibiotic resistance. Over the past few years, especially as antibiotics have been overprescribed, more and more strains of bacteria have become increasingly resistant to a wider range of antibiotics. For this reason, doctors may need to try a few different types of antibiotics—or even a combination of antibiotics—to successfully treat the infection. Antibiotics are usually given for about one month, but may need to be given for an even longer period of time if the infection is resistant to treatment.

Once the fever and the worst of the symptoms have gone away, the patient may be able to continue antibiotic therapy at home. During this time, the patient should make regular visits to the health care team for further testing and physical examination to make sure that the antibiotic therapy is working, that it is not causing adverse side effects, and that there are no complications such as emboli or heart failure. The patient should alert the health-care team to any symptoms that could indicate serious complications: For instance, trouble breathing or swelling in the legs could indicate congestive heart failure. **Headache**, joint **pain**, blood in the urine, or **stroke** symptoms could indicate an embolus, and fever and chills could indicate that the treatment is not working and the infection is worsening. Finally, **diarrhea**, rash, **itching**, or joint pain may suggest a bad reaction to the antibiotics. Anyone experiencing any of these symptoms should alert the health care team immediately.

In some cases, surgery may be needed. These include cases of congestive heart failure, recurring

## KEY TERMS

**Aortic valve**—The valve between the left ventricle of the heart and the aorta.

**Bacteremia**—An infection caused by bacteria in the blood.

**Congestive heart failure**—A condition in which the heart muscle cannot pump blood as efficiently as it should.

**Echocardiography**—A diagnostic test using reflected sound waves to study the structure and motion of the heart muscle.

**Embolus**—A bit of foreign material, such as gas, a piece of tissue, or tiny clot, that travels in the circulation until it becomes lodged in a blood vessel.

**Endocardium**—The inner wall of the heart muscle, which also covers the heart valves.

**Mitral valve**—The valve between the left atrium and the left ventricle of the heart.

**Osler's nodes**—Small, raised, reddish, tender areas

associated with endocarditis, commonly found inside the fingers or toes.

**Petechiae**—Tiny purple or red spots on the skin associated with endocarditis, resulting from hemorrhages under the skin's surface.

**Pulmonary valve**—The valve between the right ventricle of the heart and the pulmonary artery.

**Transducer**—A device that converts electrical signals into ultrasound waves and ultrasound waves back into electrical impulses.

**Transesophageal echocardiography**—A diagnostic test using an ultrasound device, passed into the esophagus of the patient, to create a clear image of the heart muscle.

**Tricuspid valve**—The valve between the right atrium and the right ventricle of the heart.

**Vegetation**—An abnormal growth of tissue around a valve, composed of blood platelets, bacteria, and a protein involved in clotting.

emboli, infection that doesn't respond to treatment, poorly functioning heart valves, and endocarditis involving prosthetic (artificial) valves. The most common surgical treatment involves cutting away (debriding) damaged tissue and replacing the damaged valve.

### Prognosis

If left untreated, infective endocarditis continues to progress and is always fatal. However, if it is diagnosed and properly treated within the first six weeks of infection, the infection can be completely cured in about 90% of the cases. The prognosis depends on a number of factors, such as the patient's age and overall physical condition, the severity of the diseases involved, the exact site of the infection, how vulnerable the microorganisms are to antibiotics, and what kind of complications the endocarditis may be causing.

### Prevention

Some people are especially prone to endocarditis. These include people with past episodes of endocarditis, those with congenital heart problems or heart damage from rheumatic fever, and those with artificial heart valves. Intravenous drug users are also at increased risk.

Anyone who falls into a high-risk category should alert his or her health-care professionals before undergoing any surgical or dental procedures. High-risk patients must be treated in advance with antibiotics before these procedures to minimize the risk of infection.

### Resources

#### BOOKS

*The Patient's Guide to Medical Tests*. Ed. Barry L. Zaret, et al. Boston: Houghton Mifflin, 1997.

#### PERIODICALS

Auten, Gramce M., and Victor Del Bene. "Endocarditis: Current Guidelines on Prophylaxis, Diagnosis, and Treatment." *Consultant* 36 (May 1996): 973-78.

#### ORGANIZATIONS

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

National Heart, Lung and Blood Institute. PO Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. <<http://www.nhlbi.nih.gov>>.

Robert Scott Dinsmoor

Endocrine pancreatic cancer see **Pancreatic cancer, endocrine**

## Endometrial biopsy

### Definition

Endometrial biopsy is a procedure in which a sample of the endometrium (tissue lining the inside of the uterus) is removed for microscopic examination.

### Purpose

The test is most often performed to find out the cause of abnormal uterine bleeding. Abnormal bleeding includes bleeding between menstrual periods, excessive bleeding during a menstrual period, or bleeding after **menopause**. Since abnormal uterine bleeding can indicate **cancer**, an endometrial biopsy is done to rule out **endometrial cancer** or hyperplasia (a potentially pre-cancerous condition).

Endometrial biopsies are also done as a screening test for endometrial cancer in postmenopausal women on **hormone replacement therapy**. Hormone replacement therapy usually requires a woman to take estrogen and progesterone. An endometrial biopsy is particularly useful in cases where postmenopausal women take estrogen, but cannot take progesterone. Estrogen in the system without the balancing effect of progesterone has been linked to an increased risk of endometrial cancer.

An endometrial biopsy can also be used as part of an **infertility** exam to rule out problems with the development of the endometrium. This condition is called luteal phase defect and can cause the endometrium to not support a **pregnancy**. An endometrial biopsy can also be used to evaluate the problem of repeated early miscarriages.

### Precautions

If the endometrial biopsy is being done to investigate why a woman is unable to get pregnant, the test must be performed at a specific time during the menstrual cycle. Since the test evaluates whether the endometrium is developed adequately to support implantation and growth of a fertilized egg, it is critical to perform the test approximately three days before the expected menstrual period.

### Description

The test is performed by a doctor who specializes in women's reproductive health (an obstetrician/gynecologist). The test is performed either in the doctor's office or in a local hospital. The patient may be asked to take **pain** medication (like Motrin or Aleve) an hour or so before the procedure. A local anesthetic may be injected into the cervix in order to decrease pain and discomfort during the procedure.

### KEY TERMS

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

**Cervix**—The opening of the uterus extending into the vagina.

**Endometrium**—The layer lining the inner cavity of the uterus; this layer changes daily throughout the menstrual cycle.

**Uterus**—The hollow, muscular female organ that supports the development and nourishment of the unborn baby during pregnancy.

The woman will be asked to lie on her back with knees apart and feet in stirrups. The doctor will first conduct a thorough exam of the pelvic region, including the vulva (the external genitals), vagina, and uterus. A speculum (an instrument that is used to hold the walls of the vagina open) will be inserted into the vagina. A small, hollow plastic tube is then passed into the uterine cavity. A small piece of the uterine lining is sucked out with a plunger that is attached to the tube. Once the sample is obtained, the instruments are removed. The sample is sent to the laboratory for microscopic examination.

The patient may experience some pain when the cervix is grasped. The patient may also feel some cramping, pressure, and discomfort when the instruments are inserted into the uterus and the tissue sample is collected.

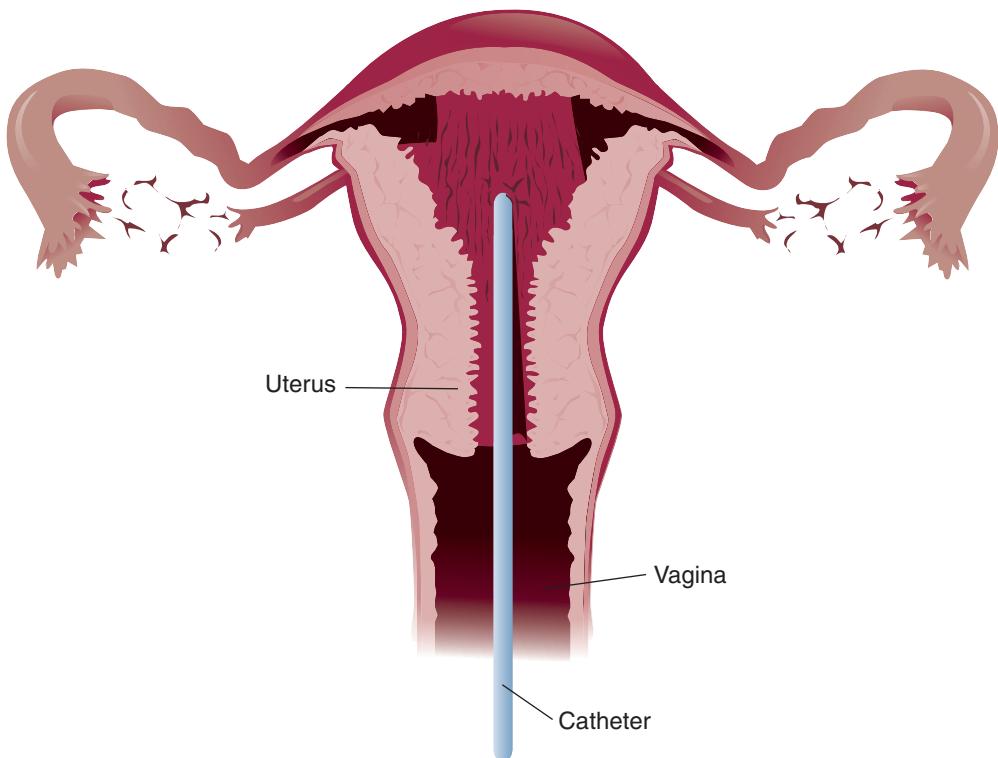
### Preparation

For the small number of endometrial biopsies that are done as part of infertility testing, a pregnancy test is also often performed before the procedure. Since the biopsy is performed late in the menstrual cycle, it is possible that the woman may be pregnant.

### Aftercare

The biopsy may cause a small amount of bleeding (spotting). The woman can resume normal activities right away. If cramping becomes severe, heavy bleeding occurs, or the woman develops a high temperature, the doctor should be notified immediately.

If the test is being done to determine the cause of infertility, the onset of the menstrual period following the biopsy should be reported to the doctor. This will allow the doctor to correctly predict if the endometrium has been developing at the expected rate.



A catheter is inserted into the uterus to remove uterine cells for further examination. (Illustration by Argosy Inc.)

## Risks

The risks of an endometrial biopsy are very small. There is a possibility that prolonged bleeding may occur after the procedure. There is also a slight chance of an infection. Very rarely, there are instances when the uterus is pierced (perforated) or the cervix is torn because of the biopsy.

## Normal results

Most biopsies are done to rule out endometrial cancer or endometrial hyperplasia. A normal result shows no cancerous or precancerous cells. Normal results also show that the uterine lining is changing at the proper rate. If it is, then the results of the biopsy are said to be "in-phase" because the tissue looks appropriate and has developed normally for the late phase of the menstrual cycle.

## Abnormal results

If the endometrium is not developing at the appropriate rate, the results are said to be "out-of-phase" or abnormal. The endometrium has not developed appropriately and cannot support a pregnancy. This condition is

called luteal phase defect and may need to be treated with progesterone.

Abnormal appearance of the cells forming the uterine tissue could also indicate uterine cancer, or the presence of fibroids or polyps in the uterus.

## Resources

### BOOKS

*The Merck Manual of Diagnosis and Therapy*. 16th ed. Ed. Robert Berkow. Rahway, NJ: Merck Research Laboratories, 1992.  
Piotrowski, Nancy A., ed. "Endometrial Biopsy." In *Magill's Medical Guide Health and Illness Supplement*. Vol. 4. Pasadena: Salem Press, 1996.

### 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, NY 10022. (800) 992-2623. <<http://www.cancerresearch.org>>. Gynecologic Cancer Foundation. 401 North Michigan Ave., Chicago, IL 60611. (800) 444-4441. National Cancer Institute. Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD 20892-2580. (800) 422-6237. <<http://www.nci.nih.gov>>.

Lata Cherath, PhD

## Endometrial cancer

### Definition

Endometrial **cancer** develops when the cells that make up the inner lining of the uterus (the endometrium) become abnormal and grow uncontrollably.

### Description

Endometrial cancer (also called uterine cancer) is the fourth most common type of cancer among women and the most common gynecologic cancer. Approximately 34,000 women are diagnosed with endometrial cancer each year. In 1998, approximately 6,300 women died from this cancer. Although endometrial cancer generally occurs in women who have gone through **menopause** and are 45 years of age or older, 30% of the women with endometrial cancer are younger than 40 years of age. The average age at diagnosis is 60 years old.

The uterus, or womb, is the hollow female organ that supports the development of the unborn baby during **pregnancy**. The uterus has a thick muscular wall and an inner lining called the endometrium. The endometrium is very sensitive to hormones and it changes daily during the menstrual cycle. The endometrium is designed to provide an ideal environment for the fertilized egg to implant and begin to grow. If pregnancy does not occur, the endometrium is shed causing the menstrual period.

More than 95% of uterine cancers arise in the endometrium. The most common type of uterine cancer is adenocarcinoma. It arises from an abnormal multiplication of endometrial cells (atypical adenomatous hyperplasia) and is made up of mature, specialized cells (well-differentiated). Less commonly, endometrial cancer arises without a preceding hyperplasia and is made up of poorly differentiated cells. The more common of these types are the papillary serous and clear cell carcinomas. Poorly differentiated endometrial cancers are often associated with a less promising prognosis.

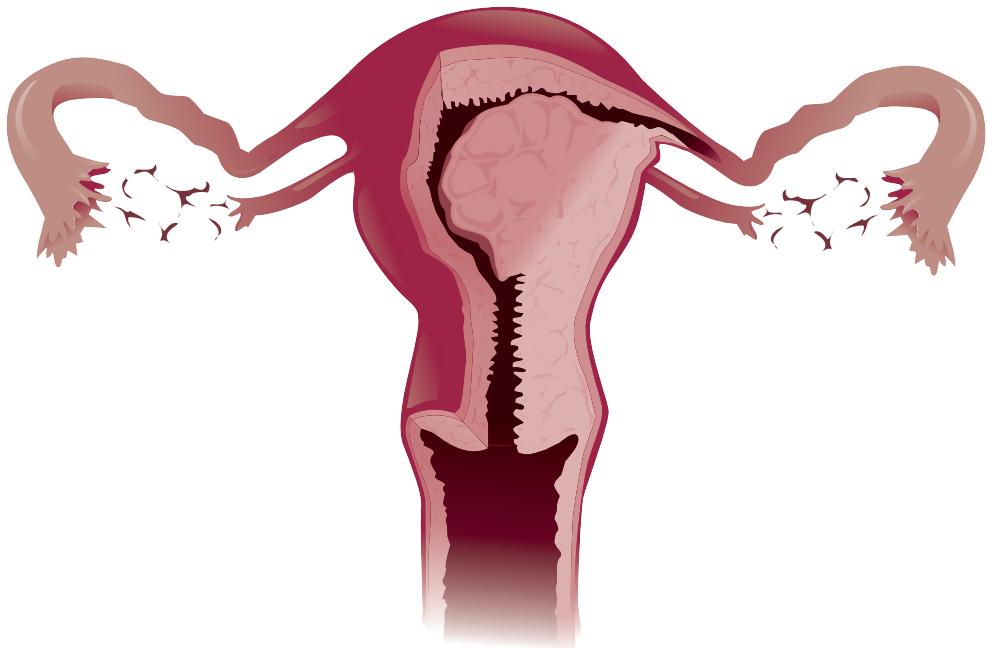
The highest incidence of endometrial cancer in the United States is in Caucasians, Hawaiians, Japanese, and African Americans. American Indians, Koreans, and Vietnamese have the lowest incidence. African American and Hawaiian women are more likely to be diagnosed with advanced cancer and, therefore, have a higher risk of dying from the disease.

### Causes and symptoms

Although the exact cause of endometrial cancer is unknown, it is clear that high levels of estrogen, when not balanced by progesterone, can lead to abnormal

growth of the endometrium. Factors that increase a woman's risk of developing endometrial cancer are:

- **Age.** The risk is considerably higher in women who are over the age of 50 and have gone through menopause.
- **Obesity.** Being overweight is a very strong risk factor for this cancer. Fatty tissue can change other normal body chemicals into estrogen, which can promote endometrial cancer.
- **Estrogen replacement therapy.** Women receiving estrogen supplements after menopause have a 12 times higher risk of getting endometrial cancer if progesterone is not taken simultaneously.
- **Diabetes.** Diabetics have twice the risk of getting this cancer as nondiabetic women. It is not clear if this risk is due to the fact that many diabetics are also obese and hypertensive. One 1998 study found that women who were obese and diabetic were three times more likely to develop endometrial cancer than women who were obese but nondiabetic. This study also found that nonobese diabetics were not at risk of developing endometrial cancer.
- **Hypertension.** High blood pressure (or hypertension) is also considered a risk factor for uterine cancer.
- **Irregular menstrual periods.** During the menstrual cycle, there is interaction between the hormones estrogen and progesterone. Women who do not ovulate regularly are exposed to high estrogen levels for longer periods of time. If a woman does not ovulate regularly, this delicate balance is upset and may increase her chances of getting uterine cancer.
- **Early first menstruation or late menopause.** Having the first period at a young age (the mean age of menses is 12.16 years in African American girls and 12.88 years in caucasian girls) or going through menopause at a late age (over age 51) seem to put women at a slightly higher risk for developing endometrial cancer.
- **Tamoxifen.** This drug, which is used to treat or prevent **breast cancer**, increases a woman's chance of developing endometrial cancer. Tamoxifen users tend to have more advanced endometrial cancer with an associated poorer survival rate than those who do not take the drug. In many cases, however, the value of tamoxifen for treating breast cancer and for preventing the cancer from spreading far outweighs the small risk of getting endometrial cancer.
- **Family history.** Some studies suggest that endometrial cancer runs in certain families. Women with inherited mutations in the BRCA1 and BRCA2 genes are at a higher risk of developing breast, ovarian, and other gynecologic cancers. Those with the hereditary non-



Cancer located in the uterus. (Illustration by Argosy Inc.)

polyposis colorectal cancer gene have a higher risk of developing endometrial cancer.

- Breast, ovarian, or **colon cancer**. Women who have a history of these other types of cancer are at an increased risk of developing endometrial cancer.
- Low parity or nulliparity. Endometrial cancer is more common in women who have born few (low parity) or no (nulliparity) children. The high levels of progesterone produced during pregnancy has a protective effect against endometrial cancer. The results of one study suggest that nulliparity is associated with a lower survival rate.
- Infertility. Risk is increased due to nulliparity or the use of fertility drugs.
- Polycystic ovary syndrome. The increased level of estrogen associated with this abnormality raises the risk of cancers of the breast and endometrium.

The most common symptom of endometrial cancer is unusual vaginal spotting, bleeding, or discharge. In women who are near menopause (perimenopausal), symptoms of endometrial cancer could include bleeding between periods (intermenstrual bleeding), heavy bleeding that lasts for more than seven days, or short menstrual cycles (fewer than 21 days). For women who have

gone through menopause, any vaginal bleeding or abnormal discharge is suspect. **Pain** in the pelvic region and the presence of a lump (mass) are symptoms that occur late in the disease.

### Diagnosis

If endometrial cancer is suspected, a series of tests will be conducted to confirm the diagnosis. The first step will involve taking a complete personal and family medical history. A **physical examination**, which will include a thorough pelvic examination, will also be done.

The doctor may order an **endometrial biopsy**. This is generally performed in the doctor's office and does not require anesthesia. A thin, flexible tube is inserted through the cervix and into the uterus. A small piece of endometrial tissue is removed. The patient may experience some discomfort, which can be minimized by taking an anti-inflammatory medication (like Advil or Motrin) an hour before the procedure.

If an adequate amount of tissue was not obtained by the endometrial biopsy, or if the biopsy tissue looks abnormal but confirmation is needed, the doctor may perform a **dilatation and curettage** (D & C). This procedure is done in the outpatient surgery department of a hospital

## KEY TERMS

**Adjuvant therapy**—A treatment done when there is no evidence of residual cancer in order to aid the primary treatment. Adjuvant treatments for endometrial cancer are radiation therapy, chemotherapy, and hormone therapy.

**Atypical adenomatous hyperplasia**—The overgrowth of the endometrium. This precancerous condition is estimated to progress to cancer in one third of the cases.

**Dilation and curettage (D & C)**—A procedure in which the doctor opens the cervix and uses a special instrument to scrape tissue from the inside of the uterus.

**Endometrial biopsy**—A procedure in which a sample of the endometrium is removed and examined under a microscope.

**Endometrium**—The mucosal layer lining the inner cavity of the uterus. The endometrium's structure changes with age and with the menstrual cycle.

**Estrogen**—A female hormone responsible for stimulating the development and maintenance of female secondary sexual characteristics.

**Estrogen replacement therapy (ERT)**—A treatment in which estrogen is used therapeutically during menopause to alleviate certain symptoms such as hot flashes. ERT has also been shown to reduce the risk of osteoporosis and heart disease in women.

**Progesterone**—A female hormone that acts on the inner lining of the uterus and prepares it for implantation of the fertilized egg.

**Progestins**—A female hormone, like progesterone, that acts on the inner lining of the uterus.

and takes about an hour. The patient may be given general anesthesia. The doctor dilates the cervix and uses a special instrument to scrape tissue from inside the uterus.

The tissue that is obtained from the biopsy or the D & C is sent to a laboratory for examination. If cancer is found, then the type of cancer will be determined. The treatment and prognosis depends on the type and stage of the cancer.

Trans-vaginal ultrasound may be used to measure the thickness of the endometrium. For this painless procedure, a wand-like ultrasound transducer is inserted into the vagina to enable visualization and measurement of the uterus, the thickness of the uterine lining, and other pelvic organs.

Other possible diagnostic procedures include sonohysterography and **hysteroscopy**. For sonohysterography, a small tube is passed through the cervix and into the uterus. A small amount of a salt water (saline) solution is injected through the tube to open the space within the uterus and allow ultrasound visualization of the endometrium. For hysteroscopy, a wand-like camera is passed through the cervix to allow direct visualization of the endometrium. Both of these procedures cause discomfort, which may be reduced by taking an anti-inflammatory medication prior to the procedure.

### Treatment

#### Clinical staging

The International Federation of Gynecology and Obstetrics (FIGO) has adopted a staging system for

endometrial cancer. The stage of cancer is determined after surgery. Endometrial cancer is categorized into four stages (I, II, III, and IV) that are subdivided (A, B, and possibly C) based on the depth or spread of cancerous tissue. Seventy percent of all uterine cancers are stage I, 10–15% are stage II, and the remainder are stages III and IV. The cancer is also graded (G1, G2, and G3) based upon microscopic analysis of the aggressiveness of the cancer cells.

The FIGO stages for endometrial cancer are:

- Stage I. Cancer is limited to the uterus.
- Stage II. Cancer involves the uterus and cervix.
- Stage III. Cancer has spread out of the uterus but is restricted to the pelvic region.
- Stage IV. Cancer has spread to the bladder, bowel, or other distant locations.

The mainstay of treatment for most stages of endometrial cancer is surgery. **Radiation therapy**, hormonal therapy, and **chemotherapy** are additional treatments (called adjuvant therapy). The necessity of adjuvant therapy is a controversial topic which should be discussed with the patient's treatment team.

#### Surgery

Most women with endometrial cancer, except those with stage IV disease, are treated with a **hysterectomy**. A simple hysterectomy involves the removal of the uterus. In a bilateral **salpingo-oophorectomy** with total hysterectomy, the ovaries, fallopian tubes, and uterus are

removed. This may be necessary because endometrial cancer often spreads to the ovaries first. The lymph nodes in the pelvic region may also be biopsied or removed to check for metastasis. Hysterectomy is traditionally performed through an incision in the abdomen (laparotomy), however, endoscopic surgery (**laparoscopy**) with vaginal hysterectomy is also being used. Women with stage I disease may require no further treatment. However, those with higher grade disease will receive adjuvant therapy.

### Radiation therapy

The decision to use radiation therapy depends on the stage of the disease. Radiation therapy may be used before surgery (preoperatively) and/or after surgery (postoperatively). Radiation given from a machine that is outside the body is called external radiation therapy. Sometimes applicators containing radioactive compounds are placed inside the vagina or uterus. This is called internal radiation therapy or brachytherapy and requires hospitalization.

Side effects are common with radiation therapy. The skin in the treated area may become red and dry. **Fatigue**, upset stomach, **diarrhea**, and nausea are also common complaints. Radiation therapy in the pelvic area may cause the vagina to become narrow (vaginal stenosis), making intercourse painful. **Premature menopause** and some problems with urination may also occur.

### Chemotherapy

Chemotherapy is usually reserved for women with stage IV or recurrent disease because this therapy is not a very effective treatment for endometrial cancer. The **anticancer drugs** are given by mouth or intravenously. Side effects include stomach upset, vomiting, appetite loss, hair loss, mouth or vaginal sores, fatigue, menstrual cycle changes, and premature menopause. There is also an increased chance of infections.

### Hormonal therapy

Hormonal therapy uses drugs like progesterone to slow the growth of endometrial cells. These drugs are usually available as pills. This therapy is usually reserved for women with advanced or recurrent disease. Side effects include fatigue, fluid retention, and appetite and weight changes.

### Alternative treatment

Although alternative and complementary therapies are used by many cancer patients, very few controlled studies on the effectiveness of such therapies exist. Mind-body techniques, such as prayer, **biofeedback**, visualization, **meditation**, and **yoga**, have not shown any effect in reduc-

ing cancer, but they can reduce **stress** and lessen some of the side effects of cancer treatments. Clinical studies of hydrazine sulfate found that it had no effect on cancer and even worsened the health and well-being of the study subjects. One clinical study of the drug amygdalin (Laetrile) found that it had no effect on cancer. Laetrile can be toxic and has caused deaths. Shark cartilage, although highly touted as an effective cancer treatment, is an improbable therapy that has not been the subject of clinical study.

The American Cancer Society has found that the “metabolic diets” pose serious risk to the patient. The effectiveness of the macrobiotic, Gerson, and Kelley **diets** and the Manner metabolic therapy has not been scientifically proven. The FDA was unable to substantiate the anti-cancer claims made about the popular Cancell treatment.

There is no evidence for the effectiveness of most over-the-counter herbal cancer remedies. Some herbals have shown an anticancer effect. As shown in clinical studies, Polysaccharide krestin, from the mushroom *Coriolus versicolor*, has significant effectiveness against cancer. In a small study, the green alga *Chlorella pyrenoidosa* has been shown to have anticancer activity. In a few small studies, evening primrose oil has shown some benefit in the treatment of cancer.

### Prognosis

Because it is possible to detect endometrial cancer early, the chances of curing it are excellent. The five year survival rates for endometrial cancer by stage are: 90%, stage I; 60%, stage II; 40%, stage III; and 5%, stage IV. Endometrial cancer most often spreads to the lungs, liver, bones, brain, vagina, and certain lymph nodes.

### Prevention

Women (especially postmenopausal women) should report any abnormal vaginal bleeding or discharge to the doctor. Controlling obesity, blood pressure, and diabetes can help to reduce the risk of this disease. Women on estrogen replacement therapy have a substantially reduced risk of endometrial cancer if progestins are taken simultaneously. Long term use of birth control pills has been shown to reduce the risk of this cancer. Women who have irregular periods may be prescribed birth control pills to help prevent endometrial cancer. Women who are taking tamoxifen and those who carry the hereditary non-polyposis colorectal cancer gene should be screened regularly, receiving annual pelvic examinations.

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- Cancer Research Institute, National Headquarters. 681 Fifth Ave., New York, NY 10022. (800) 992-2623. <<http://www.cancerresearch.org>>.
- Gynecologic Cancer Foundation. 401 North Michigan Ave., Chicago, IL 60611. (800) 444-4441. <<http://www.wcn.org>>.
- National Cancer Institute, National Institutes of Health. 9000 Rockville Pike, Bethesda, MD 20892. (800) 422-6237. <<http://cancernet.nci.nih.gov>>.

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Lata Cherath  
Belinda Rowland, Ph.D.

## Endometriosis

### Definition

Endometriosis is a condition in which bits of the tissue similar to the lining of the uterus (endometrium) grow

in other parts of the body. Like the uterine lining, this tissue builds up and sheds in response to monthly hormonal cycles. However, there is no natural outlet for the blood discarded from these implants. Instead, it falls onto surrounding organs, causing swelling and inflammation. This repeated irritation leads to the development of scar tissue and adhesions in the area of the endometrial implants.

### Description

Endometriosis is estimated to affect 7% of women of childbearing age in the United States. It most commonly strikes between the ages of 25 and 40. Endometriosis can also appear in the teen years, but never before the start of menstruation. It is seldom seen in postmenopausal women.

Endometriosis was once called the "career woman's disease" because it was thought to be a product of delayed childbearing. The statistics defy such a narrow generalization; however, pregnancy may slow the progress of the condition. A more important predictor of a woman's risk is if her female relatives have endometriosis. Another influencing factor is the length of a woman's menstrual cycle. Women whose periods last longer than a week with an interval of less than 27 days between them seem to be more prone to the condition.

Endometrial implants are most often found on the pelvic organs—the ovaries, uterus, fallopian tubes, and in the cavity behind the uterus. Occasionally, this tissue grows in such distant parts of the body as the lungs, arms, and kidneys. Newly formed implants appear as small bumps on the surfaces of the organs and supporting ligaments and are sometimes said to look like "powder burns." **Ovarian cysts** may form around endometrial tissue (endometriomas) and may range from pea to grapefruit size. Endometriosis is a progressive condition that usually advances slowly, over the course of many years. Doctors rank cases from minimal to severe based on factors such as the number and size of the endometrial implants, their appearance and location, and the extent of the scar tissue and adhesions in the vicinity of the growths.

### Causes and symptoms

Although the exact cause of endometriosis is unknown, a number of theories have been put forward. Some of the more popular ones are:

- **Implantation theory.** Originally proposed in the 1920s, this theory states that a reversal in the direction of menstrual flow sends discarded endometrial cells into the body cavity where they attach to internal organs and seed endometrial implants. There is considerable evidence to support this explanation. Reversed menstrual

flow occurs in 70–90% of women and is thought to be more common in women with endometriosis. However, many women with reversed menstrual flow do not develop endometriosis.

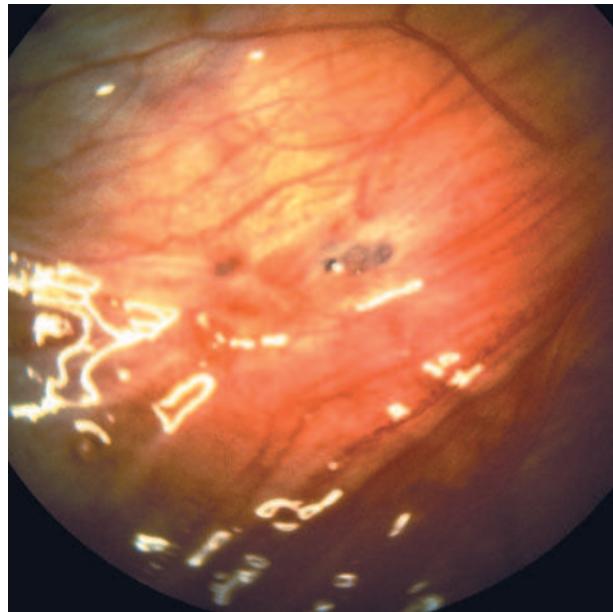
- **Vascular-lymphatic theory.** This theory suggests that the lymph system or blood vessels (vascular system) is the vehicle for the distribution of endometrial cells out of the uterus.
- **Coelomic metaplasia theory.** According to this hypothesis, remnants of tissue left over from prenatal development of the woman's reproductive tract transforms into endometrial cells throughout the body.
- **Induction theory.** This explanation postulates that an unidentified substance found in the body forces cells from the lining of the body cavity to change into endometrial cells.

In addition to these theories, the following factors are thought to influence the development of endometriosis:

- **Heredity.** A woman's chance of developing endometriosis is seven times greater if her mother or sisters have the disease.
- **Immune system function.** Women with endometriosis may have lower functioning immune systems that have trouble eliminating stray endometrial cells. This would explain why a high percentage of women experience reversed menstrual flow while relatively few develop endometriosis.
- **Dioxin exposure.** Some research suggests a link between the exposure to dioxin (TCDD), a toxic chemical found in weed killers, and the development of endometriosis.

While many women with endometriosis suffer debilitating symptoms, others have the disease without knowing it. Paradoxically, there does not seem to be any relation between the severity of the symptoms and the extent of the disease. The most common symptoms are:

- **Menstrual pain.** Pain in the lower abdomen that begins a day or two before the menstrual period starts and continues through to the end is typical of endometriosis. Some women also report lower back aches and pain during urination and bowel movement, especially during their periods.
- **Painful sexual intercourse.** Pressure on the vagina and cervix causes severe pain for some women.
- **Abnormal bleeding.** Heavy menstrual periods, irregular bleeding, and spotting are common features of endometriosis.
- **Infertility.** There is a strong association between endometriosis and infertility, although the reasons for this have not been fully explained. It is thought that



An endoscopic view of endometriosis on pelvic wall. (Custom Medical Stock Photo. Reproduced by permission.)

the build up of scar tissue and adhesions blocks the fallopian tubes and prevents the ovaries from releasing eggs. Endometriosis may also affect fertility by causing hormonal irregularities and a higher rate of early miscarriage.

## Diagnosis

If a doctor suspects endometriosis, the first step will be to perform a **pelvic exam** to try to feel if implants are present. Very often there is no strong evidence of endometriosis from a physical exam. The only way to make a definitive diagnosis is through minor surgery called a **laparoscopy**. A laparoscope, a slender scope with a light on the end, is inserted into the woman's abdomen through a small incision near her belly button. This allows the doctor to examine the internal organs for endometriotic growths. Often, a sample of tissue is taken for later examination in the laboratory. Endometriosis is sometimes discovered when a woman has abdominal surgery for another reason such as **tubal ligation** or **hysterectomy**.

Various imaging techniques such as ultrasound, computed tomography scan (CT scan), or **magnetic resonance imaging** (MRI) can offer additional information but aren't useful in making the initial diagnosis. A blood test may also be ordered because women with endometriosis have higher levels of the blood protein CA125. Testing for this substance before and after treatment can predict a recurrence of the disease, but the test is not reliable as a diagnostic tool.

## Treatment

How endometriosis is treated depends on the woman's symptoms, her age, the extent of the disease, and her personal preferences. The condition cannot be fully eradicated without surgery. Conservative treatment focuses on managing the pain, preserving fertility, and delaying the progress of the condition.

### Pain relief

Over-the-counter pain relievers such as **aspirin** and **acetaminophen** (Tylenol) are useful for mild cramping and menstrual pain. Prescription-strength and over-the-counter **nonsteroidal anti-inflammatory drugs** (NSAIDs), such as ibuprofen (Motrin, Advil) and naproxen (Naprosyn), are also effective. If pain is severe, a doctor may prescribe narcotic medications, although these can be addicting and are rarely used.

### Hormonal treatments

Hormonal therapies effectively tame endometriosis but also act as contraceptives. A woman who is hoping to become pregnant would take these medications for a period of time, then try to conceive within several months of discontinuing treatment.

- Oral contraceptives. Continuously taking estrogen-progestin pills tricks the body into thinking it is pregnant. This state of pseudopregnancy means reduced pelvic pain and a temporary withering of endometrial implants.
- Danazol (Danocrine) and gestrinone are synthetic male hormones that lower estrogen levels, prevent menstruation, and shrink endometrial tissues. On the downside, they lead to weight gain and menopause-like symptoms, and cause some women to develop masculine characteristics.
- Progestins. Medroxyprogesterone (Depo-Provera) and related drugs may also be used in treating endometriosis. They have been proven effective in minimizing pain and halting the progress of the condition, but are rarely used because of the high rate of side effects.
- Gonadotropin-releasing hormone (GnRH) agonists. These estrogen-inhibiting drugs successfully limit pain and prevent the growth of endometrial implants. They can cause **menopause** symptoms, however, and doses have to be regulated to prevent bone loss associated with low estrogen levels.

### Surgery

Removing the uterus, ovaries, and fallopian tubes is the only permanent method of eliminating endometriosis. This is an extreme measure that deprives a woman of

her ability to bear children and forces her body into menopause. Endometrial implants and ovarian cysts can be removed with **laser surgery** performed through a laparoscope. For women with minimal endometriosis, this technique is usually successful in reducing pain and slowing the condition's progress. It may also help infertile women increase their chances of becoming pregnant.

## Alternative treatment

Although severe endometriosis should not be self-treated, many women find they can help their condition through alternative therapies. Taking vitamin B complex combined with **vitamins C, E**, and the **minerals calcium, magnesium, and selenium** can help the depression and lack of energy that may accompany endometriosis. B vitamins also counteract the side effects of hormonal drugs. Other women have found relief when they turned to a macrobiotic diet. Less extreme **diets** that cut out sugar, salt, and processed foods are sometimes helpful as well. Mind-body therapies such as relaxation and visualization help women cope with pain. Other avenues to combat pain include **acupuncture** and **biofeedback** techniques. Still other women report positive results after being treated by chiropractors or homeopathic doctors.

## Prognosis

Most women who have endometriosis have minimal symptoms and do well. Overall, endometriosis symptoms come back in an average of 40% of women over the five years following treatment. With hormonal therapy, pain returned after five years in 37% of patients with minimal symptoms and 74% of those with severe cases. The highest success rate from conservative treatment followed complete removal of implants using laser surgery. Eighty percent of these women were still pain-free five years later. In cases that don't respond to these treatments, a woman and her doctor may consider surgery to remove her reproductive organs.

## Prevention

There is no proven way to prevent endometriosis. One study, however, indicated that girls who begin participating in aerobic **exercise** at a young age are less likely to develop the condition.

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## KEY TERMS

**Adhesions**—Web-like scar tissue that may develop as a result of endometriosis and bind organs to one another.

**Dioxin**—A toxic chemical found in weed killers that has been linked to the development of endometriosis.

**Endometrial implants**—Growth of endometrial tissue that attach to organs, primarily in the pelvic cavity.

**Endometrium**—The tissue lining the uterus that grows and sheds each month during a woman's menstrual cycle.

**Estrogen**—A female hormone that promotes the growth of endometrial tissue.

**Hormonal therapy**—Use of hormone medications to inhibit menstruation and relieve the symptoms of endometriosis.

**Laparoscopy**—A diagnostic procedure for endometriosis performed by inserting a slender, wand-like instrument through a small incision in the woman's abdomen.

**Menopause**—The end of a woman's menstrual periods when the body stops making estrogen.

**Retrograde menstruation**—Menstrual flow that travels into the body cavity rather than being expelled through the uterus.

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Endometriosis Association International Headquarters. 8585 North 76th Place, Milwaukee, WI 53223. (800) 992-3636.  
<http://EndometriosisAssn.org>.

Stephanie Slon

Endometritis see **Pelvic inflammatory disease**

## Endorectal ultrasound

### Definition

Endorectal ultrasound (ERUS) is a procedure where a probe is inserted into the rectum and high frequency

sound waves (ultrasound waves) are generated. The pattern of echoes as they bounce off tissues is converted into a picture (sonogram) on a television screen.

### Purpose

ERUS is used as a diagnostic procedure in **rectal cancer** to determine stage of the tumor and as a postradiation, presurgical examination to assess extent of tumor shrinkage. ERUS can also be used in cases of anal fistula (an abnormal passage) and problems with the anal sphincter muscles (muscles that control the opening and closing of the anus).

### Precautions

Normal precautions should be taken with any diagnostic procedure. Since the population in which this procedure is normally done is elderly, the imaging staff should be extra cautious about stressing the patient. The procedure is invasive and may be embarrassing to some. Other patients may be anxious about their medical condition since endorectal ultrasounds are not routine. This places an added burden on already stressed hearts and nervous systems. Physicians, nurses, and technicians may need to be prepared for **stress** reactions that could include the heart, **asthma**, or anxious behaviors.

### Description

ERUS has been used as a means to determine the depth of rectal cancers and to assess whether the tumor has affected surrounding tissues. This pre-treatment procedure has proven to be an accurate tool for tailoring surgery for patients.

Problems with interpretation of the sonograms after radiation and before surgery have resulted in tumors being identified that were merely the formation of fibrous tissues that remained after the tumors had been eliminated by the radiation. Yet, some of the fibrous areas actually hid residual tumors. Rectal anatomy itself can affect the accuracy of ultrasound reading. This makes ERUS problematic in determining the amount of tumor reduction a patient has after **radiation therapy**.

### Preparation

The patient must evacuate the bowels completely before the procedure is done. This usually is assisted though the use of several **enemas**. The patient may be told to adhere to a liquid diet the day prior to doing this procedure. The probe is inserted, usually with little discomfort for the patient since it will only be examining the first few inches of the colon.

## KEY TERMS

- Anal sphincter muscles**—Muscles that control the opening and closing of the anus.
- Fistula**—An abnormal passage.
- Sonogram**—The picture formed by the pattern of echoes from an ultra sound.
- Ultrasound waves**—High frequency sound waves.

### Aftercare

Since ERUS is a minor invasive procedure, there is no aftercare.

### Risks

There are no risks to having an ultrasound.

### Normal results

Normal results after an endorectal ultrasound are normal, healthy tissues.

### Abnormal results

Abnormal results range from any number of congenital deformities in the lining of the rectum to serious rectal cancers.

### Resources

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Janie F. Franz

## Endoscopic retrograde cholangiopancreatography

### Definition

Endoscopic retrograde cholangiopancreatography (ERCP) is a technique in which a hollow tube called an endoscope is passed through the mouth and stomach to the duodenum (the first part of the small intestine). This procedure was developed to examine abnormalities of the bile ducts, pancreas, and gallbladder. It was developed during the late 1960s and is used today to diagnose and treat blockages of the bile and pancreatic ducts.

The term has three parts to its definition:

- endoscopic refers to the use of an endoscope
- retrograde refers to the injection of dye up into the bile ducts in a direction opposing, or against, the normal flow of bile down the ducts
- cholangiopancreatography means visualization of the bile ducts (cholangio) and pancreas (pancreato)

### Purpose

Until the 1970s, methods to visualize the bile ducts produced images that were of relatively poor quality and often misleading; in addition, the pancreatic duct could not be examined at all. Patients with symptoms related to the bile ducts or pancreatic ducts frequently needed surgery to diagnose and treat their conditions.

Using ERCP, physicians can obtain high-quality x rays of these structures and identify areas of narrowing (strictures), cancers, and gallstones. This procedure can help determine whether bile or pancreatic ducts are blocked; it also identifies where they are blocked along with the cause of the blockage. ERCP may then be used to relieve the blockage. For patients requiring surgery or additional procedures for treatment, ERCP outlines the anatomical changes for the surgeon.

### Precautions

The most important precaution is that the examination should be performed by an experienced physician. The procedure is much more technically difficult than many other gastrointestinal endoscopic studies. Patients

should seek physicians with experience performing ERCP. Patients should inform the physician about any **allergies** (including allergies to contrast dyes, iodine, or shellfish), medication use, and medical problems. Occasionally, patients may need to be admitted to the hospital after the procedure.

## Description

After **sedation**, a specially adapted endoscope is passed through the mouth, through the stomach, then into the duodenum. The opening to ducts that empty from the liver and pancreas is identified, and a plastic tube or catheter is placed into the orifice (opening). Contrast dye is then injected into the ducts, and with the assistance of a radiologist, pictures are taken.

## Preparation

The upper intestinal tract must be empty for the procedure, so patients should not eat or drink for at least six to 12 hours before the exam. Patients should ask the physician about taking their medications before the procedure.

## Aftercare

Someone should be available to take the person home after the procedure and stay with them for a while; patients will not be able to drive themselves because they undergo sedation during this test. **Pain** or any other unusual symptoms should be reported to the physician.

## Risks

ERCP-related complications can be broken down into those related to medications used during the procedure, the diagnostic part of the procedure, and those related to endoscopic therapy. The overall complication rate is 5–10%; most of those occur when diagnostic ERCP is combined with a therapeutic procedure. During the exam, the endoscopist can cut or stretch structures (such as the muscle leading to the bile duct) to treat the cause of the patient's symptoms. Although the use of sedatives carries a risk of decreasing cardiac and respiratory function, it is very difficult to perform these procedures without these drugs.

The major complications related to diagnostic ERCP are **pancreatitis** (inflammation of the pancreas) and **cholangitis** (inflammation of the bile ducts). **Bacteremia** (the passage of bacteria into the blood stream) and perforation (hole in the intestinal tract) are additional risks.

## Normal results

Because certain standards have been set for the normal diameter or width of the pancreatic duct and bile

## KEY TERMS

**Endoscope, endoscopy**—An endoscope used in the field of gastroenterology is a hollow, thin, flexible tube that uses a lens or miniature camera to view various areas of the gastrointestinal tract. When the procedure is performed to examine the bile ducts or pancreas, the organs are not viewed directly, but rather indirectly through the injection of contrast. The performance of an exam using an endoscope is referred to as endoscopy. Diagnosis through biopsies or other means and therapeutic procedures can also be done using these instruments.

**Visualization**—The process of making an internal organ visible. A radiopaque substance is introduced into the body, then an x-ray picture of the desired area is taken.

ducts, measurements using x rays are taken to determine if the ducts are too large (dilated) or too narrow (stricture). The ducts and gallbladder should be free of stones or tumors.

## Abnormal results

When areas in the pancreatic or bile ducts (including those in the liver) are too wide or too narrow compared with the standard, the test is considered abnormal. Once these findings are demonstrated using ERCP, symptoms are usually present; they generally do not change without treatment. Stones, identified as opaque or solid structures within the ducts, are also considered abnormal. Masses or tumors may also be seen, but sometimes the diagnosis is made not by direct visualization of the tumor, but by indirect signs, such as a single narrowing of one of the ducts. Overall, ERCP has an excellent record in diagnosing these abnormalities.

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David S. Kaminstein

Endoscopic sclerotherapy see **Sclerotherapy for esophageal varices**

## Endoscopic sphincterotomy

### Definition

Endoscopic sphincterotomy or endoscopic retrograde sphincterotomy (ERS) is a relatively new endoscopic technique developed to examine and treat abnormalities of the bile ducts, pancreas and gallbladder. The procedure was developed as an extension to the diagnostic examination, ERCP (**endoscopic retrograde cholangiopancreatography**); with the addition of "sphincterotomy," abnormalities found during the study could be treated at the same time without the need for invasive surgery.

The term ERS has three parts to its definition:

- endoscopic refers to the use of an endoscope
- retrograde refers to the insertion of the endoscope *up* into the ducts in a direction opposite to or against the normal flow of bile *down* the ducts
- sphincterotomy, which means cutting of the sphincter or muscle that lies at the juncture of the intestine with both the bile and pancreatic ducts

### Purpose

Until the 1970s, patients with symptoms related to disease of the bile ducts or pancreas frequently needed surgery to diagnose the cause and treat any abnormalities. ERCP allowed physicians for the first time to obtain high quality x rays of the common bile and pancreatic ducts, and detect areas of narrowing (strictures), stones, and tumors. ERCP was not initially designed for treatment. ERS was developed shortly after and enabled physicians to treat the abnormalities identified by the injection of dye and x rays.

The revolutionary technique made possible the endoscopic removal of stones and stretching of areas of narrowing (strictures). It has since been expanded to include drainage of bile from blocked ducts and treatment of various abnormalities of the pancreas.

### Precautions

The most important precaution related to both ERCP and ERS is to have the procedure performed by an experienced physician. ERS is technically more difficult than many other gastrointestinal endoscopic studies, including ERCP. Patients should inquire as to the physician's experience with the procedure. The physician should also be informed of any **allergies**, medication use, and medical problems.

### Description

ERS is generally performed only after ERCP has been successfully accomplished and detail of the anatomy and abnormalities is known. During ERS, a number of various instruments are inserted through the endoscope in order to "cut" or stretch the sphincter. Once this is done, additional instruments are passed that enable the removal of stones and the stretching of narrowed regions of the ducts. Drains (stents) can also be used to prevent a narrowed area from rapidly returning to its previously narrowed state.

### Preparation

The upper intestinal tract must be empty for the procedure, so patients must not eat or drink for at least six to 12 hours before the exam. Patients need to inquire about taking their medications before the procedure. Some patients may require **antibiotics** before and/or after the procedure. When possible, **aspirin** or NSAIDS should not be taken within several days before the procedure, because they interfere with blood clotting.

### Aftercare

When ERS is performed, physicians will usually want to observe the patient closely for several hours to ensure that there are no signs of complications. **Pain** or any other unusual symptoms should be reported. Admission to the hospital may be advised.

### Risks

ERS complications are related either to the drugs used during the procedure, or the results of dye injection or cutting of tissue. The overall complication rate is 5–10%. During the exam, the endoscopist can cut or

## KEY TERMS

**Endoscope, Endoscopy**—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. 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.

**NSAIDS**—This abbreviation stands for non-steroidal anti-inflammatory drugs, which are medications such as Ibuprofen that are used to control pain and inflammation. Most may be purchased over the counter. One of their major side effects is that they decrease the effect of the normal blood clotting factors in blood. In patients undergoing surgical or endoscopic procedures, this can lead to an increased risk of bleeding.

stretch structures (such as the muscle leading to the bile duct) to treat the cause of the patient's symptoms. Cutting or stretching of these structures can sometimes cause a hole or perforation. The use of sedatives also carries a risk of decreasing cardiac and respiratory function, however, it is very difficult to perform these procedures without these drugs.

Other major complications related to ERCP or ERS are **pancreatitis** (inflammation of the pancreas) and **cholangitis** (inflammation of the bile ducts). **Bacteremia** (the passage of bacteria into the blood stream) and bleeding are also risks.

### Normal results

Certain standards have been set for the diameter or width of the pancreatic and bile ducts. Measurements by x ray are used to determine if the ducts are too large (dilated) or too narrow (strictured). Lastly, the ducts and gallbladder should be free of any solid particles, such as stones, and free of areas of narrowing.

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## Enemas

### Definition

An enema is the insertion of a solution into the rectum and lower intestine.

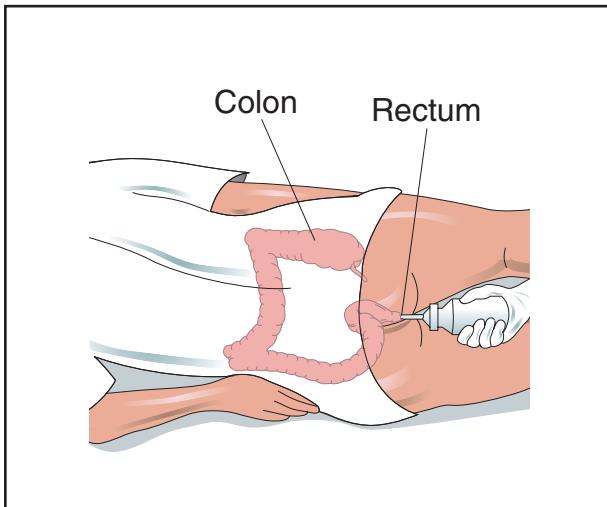
### Purpose

Enemas may be given for the following purposes:

- to remove feces when an individual is constipated or impacted,
- to remove feces and cleanse the rectum in preparation for an examination,
- to remove feces prior to a surgical procedure to prevent contamination of the surgical area,
- to administer drugs or anesthetic agents.

### Precautions

The rectal tube used for infusion of the enema solution should be smooth and flexible to decrease the possibility of damage to the mucous membrane that lines the rectum. Tap water is commonly used for adults but should not be used for infants because of the danger of electrolyte (substance that conducts electric current within the body and is essential for sustaining life) imbalance. The colon absorbs water, and repeated tap



**Enemas may be given for the following purposes:** to remove feces when an individual is constipated, or to remove feces and cleanse the rectum in preparation for an examination, or prior to surgery to prevent contamination. There are two types of enemas: the high enema, given to cleanse the large bowel, and the low enema, to cleanse only the lower bowel.

(Illustration by Electronic Illustrators Group.)

water enemas can cause cardiovascular overload and electrolyte imbalance. Similarly, repeated saline enemas can cause increased absorption of fluid and electrolytes into the bloodstream, resulting in overload. Individuals receiving frequent enemas should be observed for overload symptoms that include **dizziness**, sweating, or vomiting.

Soap suds and saline used for cleansing enemas can cause irritation of the lining of the bowel, with repeated use or a solution that is too strong. Only white soap should be used; the bar should not have been previously used, to prevent infusing undesirable organisms into the individual receiving the enema. Common household detergents are considered too strong for the rectum and bowel. The commercially prepared castile soap is preferred, and should be used in concentration no greater than 5 cc soap to 1,000 cc of water.

## Description

Cleansing enemas act by stimulation of bowel activity through irritation of the lower bowel, and by distension with the volume of fluid instilled. When the enema is administered, the individual is usually lying on the left side, which places the sigmoid colon (lower portion of bowel) below the rectum and facilitates infusion of fluid. The length of time it takes to administer an enema depends on the amount of fluid to be infused. The amount of fluid administered will vary depending on the

age and size of the person receiving the enema, however general guidelines would be:

- Infant: 250 cc or less
- Toddler and preschooler: 500 cc or less
- School-aged child: 500–1,000 cc
- Adult: 750–1,000 cc

Some may differentiate between high and low enemas. A high enema, given to cleanse as much of the large bowel as possible, is usually administered at higher pressure and with larger volume (1,000 cc), and the individual changes position several times in order for the fluid to flow up into the bowel. A low enema, intended to cleanse only the lower bowel, is administered at lower pressure, using about 500 cc of fluid.

Oil retention enemas serve to lubricate the rectum and lower bowel, and soften the stool. For adults, about 150–200 cc of oil is instilled, while in small children, 75–150 cc of oil is considered adequate. Salad oil or liquid petrolatum are commonly used at a temperature of 91°F (32.8°C). There are also commercially prepared oil retention enemas. The oil is usually retained for one to three hours before it is expelled.

The rectal tube used for infusion of the solution, usually made of rubber or plastic, has two or more openings at the end through which the solution can flow into the bowel. The distance to which the tube must be inserted is dependent upon the age and size of the patient. For adult, insertion is usually 3–4 in (7.5–10 cm); for children, approximately 2–3 in (5–7.5 cm); and for infants, only 1–1.5 in (2.5–3.75 cm). The rectal tube is lubricated before insertion with a water soluble lubricant to ease insertion and decrease irritation to the rectal tissues.

The higher the container of solution is placed, the greater the force in which the fluid flows into the patient. Routinely, the container should be no higher than 12 in (30 cm) above the level of the bed; for a high cleansing enema, the container may be 12–18 in (30–45 cm) above the bed level, because the fluid is to be instilled higher into the bowel.

## Preparation

The solution used in the procedure is measured, mixed, and warmed before administration of the enema.

## Aftercare

If necessary, a specimen will be collected for diagnostic evaluation. If the enema was given to alleviate **constipation**, the better approach to combatting consti-

## KEY TERMS

**Electrolyte**—A substance that conducts electric current within the body and is essential for sustaining life.

**Intestine**—Also called the bowels and divided into large and small intestine, they extend from the stomach to the anus, where waste products exit the body. The small intestine is about 20 ft (6.1 m) long and the large intestine, about 5 ft (1.5 m) long.

**Rectum**—The portion of bowel just before the anus. The prefix *recto* is used with a variety of words in relation to conditions that affect the rectum.

pation in the future is with a high fiber diet (five to six servings of whole grain foods) and adequate fluid intake (seven to eight glasses of water per day). Regular **exercise** and going to the bathroom when necessary will also help. If constipation is a chronic problem, medical help should be consulted to determine if there is underlying disorder.

### Risks

Habitual use of enemas as a means to combat constipation can make the problem even more severe when their use is discontinued. Enemas should be used only as a last resort for treatment of constipation and with a doctor's recommendation. Enemas should not be administered to individuals who have recently had colon or rectal surgery, a **heart attack**, or who suffer from an unknown abdominal condition or an irregular heartbeat.

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Kathleen D. Wright, RN

## Enlarged prostate

### Definition

A non-cancerous condition that affects many men past 50 years of age, enlarged prostate makes urinating

more difficult by narrowing the urethra, a tube running from the bladder through the prostate gland. It can be effectively treated by surgery and, today, by certain drugs.

### Description

The common term for enlarged prostate is BPH, which stands for benign (non-cancerous) prostatic hyperplasia or hypertrophy. Hyperplasia means that the prostate cells are dividing too rapidly, increasing the total number of cells, and, therefore, the size of the organ itself. Hypertrophy simply means "enlargement." BPH is part of the **aging** process. The actual changes in the prostate may start as early as the 30s but take place very gradually, so that significant enlargement and symptoms usually do not appear until after age 50. Past this age the chances of the prostate enlarging and causing urinary symptoms become progressively greater. More than 40% of men in their 70s have an enlarged prostate. Symptoms generally appear between ages 55–75. About 10% of all men eventually will require treatment for BPH.

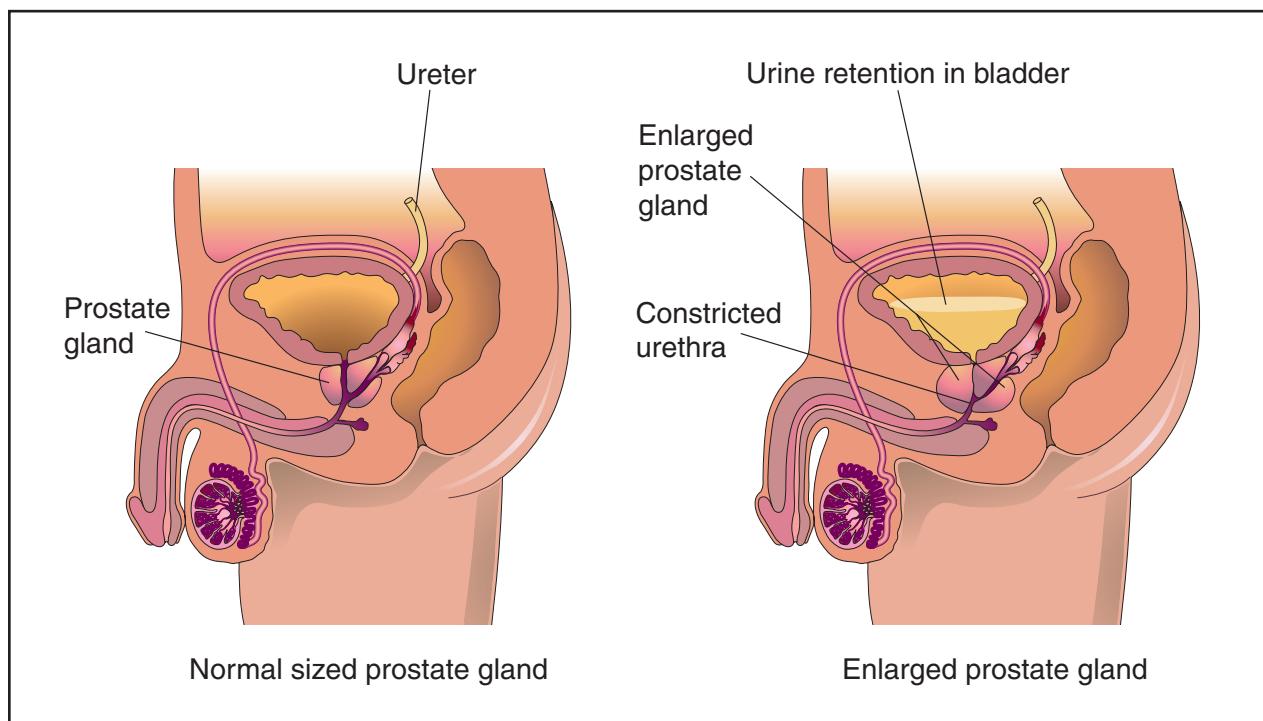
BPH has been viewed as a rare condition in African, Chinese and other Asian peoples for reasons that are not clear.

### Causes and symptoms

The cause of BPH is a mystery, but age-related changes in the levels of hormones circulating in the blood may be a factor. Whatever the cause, an enlarging prostate gradually narrows the urethra and obstructs the flow of urine. Even though the muscle in the bladder wall becomes stronger in an attempt to push urine through the smaller urethra, in time, the bladder fails to empty completely at each urination. The urine that collects in the bladder can become infected and lead to stone formation. The kidneys themselves may be damaged by infection or by urine constantly "backing up."

When the enlarging prostate gland narrows the urethra, a man will have increasing trouble starting the urine stream. Because some urine remains behind in the bladder, he will have to urinate more often, perhaps two or three times at night (nocturia). The need to urinate can become very urgent and, in time, urine may dribble out to stain a man's clothing. Other symptoms of BPH are a weak and sometimes a split stream and general aching or **pain** in the perineum (the area between the scrotum and anus). Some men may have considerable enlargement of the prostate before even mild symptoms develop.

If a man must strain hard to force out the urine, small veins in the bladder wall and urethra may rupture,



An enlarged prostate is a non-cancerous condition in which the narrowing of the urethra makes the elimination of urine more difficult. It most often occurs in men over age 50. (Illustration by Electronic Illustrators Group.)

causing blood to appear in the urine. If the urinary stream becomes totally blocked, the urine collecting in the bladder may cause severe discomfort, a condition called acute urinary retention. Urine that stagnates in the bladder can easily become infected. A burning feeling during urination and **fever** are clues that infection may have developed. Finally, if urine backs up long enough it may increase pressure in the kidneys, though this rarely causes permanent kidney damage.

## Diagnosis

When a man's symptoms point to BPH, the first thing the physician will want to do is a digital **rectal examination**, inserting a finger into the anus to feel whether—and how much—the prostate is enlarged. A smooth prostate surface suggests BPH, whereas a distinct lump in the gland might mean **prostate cancer**. The next step is a blood test for a substance called prostate-specific antigen or PSA. Between 30–50% of men with BPH have an elevated PSA level. This does not mean **cancer** by any means, but other measures are needed to make sure that the prostate enlargement is in fact benign. An ultrasound exam of the prostate, which is entirely safe and delivers no radiation, can show whether it is enlarged and may show that cancer is present.

If digital or ultrasound examination of the prostate raises the suspicion of cancer, most urologists will recommend that a prostatic tissue biopsy be performed. This is usually done using a lance-like instrument that is inserted into the rectum. It pierces the rectal wall and, guided by the physician's finger, obtains six to eight pieces of prostatic tissue that are sent to the laboratory for microscopic examination. If cancer is present, the prognosis and treatment are changed accordingly.

A catheter placed through the urethra and into the bladder can show how much urine remains in the bladder after the patient urinates—a measure of how severe the obstruction is. Another and very simple test for obstruction is to have the man urinate into a uroflowmeter, which measures the rate of urine flow. A very certain—though invasive—way of confirming obstruction from an enlarged prostate is to pass a special viewing instrument called a cystoscope into the bladder, but this is not often necessary.

It is routine to check a urine sample for an increased number of white blood cells, which may mean there is infection of the bladder or kidneys. The same sample may be cultured to show what type of bacterium is causing the infection, and which **antibiotics** will work best. The state of the kidneys may be checked in two ways: imaging by either ultrasound or injecting a dye (the intra-

venous urogram, or pyelogram); or a blood test for creatinine, which collects in the blood when the kidneys cannot eliminate it.

## Treatment

### *Drugs*

A class of drugs called alpha-adrenergic blockers, which includes phenoxybenzamine and doxazosin, relax the muscle tissue surrounding the bladder outlet and lining the wall of the urethra to permit urine to flow more freely. These drugs improve obstructive symptoms, but do not keep the prostate from enlarging. Other drugs (finasteride is a good example) do shrink the prostate and may delay the need for surgery. Symptoms may not, however, improve until the drug has been used for three months or longer. Antibiotic drugs are given promptly whenever infection is diagnosed. Some medications, including **antihistamines** and some **decongestants**, can make the symptoms of BPH suddenly worse and even cause acute urinary retention, and therefore should be avoided.

### *Intermediate treatments*

When drugs have failed to control symptoms of BPH but the physician does not believe that conventional surgery is yet needed, a procedure called transurethral needle ablation may be tried. In the office and using local anesthesia, a needle is inserted into the prostate and radiofrequency energy is applied to destroy the tissue that is obstructing urine flow. Another new approach is microwave hyperthermia, using a device called the Prostatron to deliver microwave energy to the prostate through a catheter. This procedure is done at an outpatient surgery center.

### *Surgery*

For many years the standard operation for BPH has been transurethral resection (TUR) of the prostate. Under general or spinal anesthesia, a cystoscope is passed through the urethra and prostate tissue surrounding the urethra is removed using either a cutting instrument or a heated wire loop. The small pieces of prostate tissue are washed out through the scope. No incision is needed for TUR. There normally is some blood in the urine for a few days following the procedure. In a few men—less than 5% of all those having TUR—urine will continue to escape unintentionally. Other uncommon complications include a temporary rise in blood pressure with mental confusion, which is treated by giving salt solution. Impotence—the inability to achieve lasting penile erections—does occur, but probably in fewer than 10% of patients. A narrowing or stricture rarely develops in the urethra, but this can be treated fairly easily.

## KEY TERMS

**Catheter**—A rubber or plastic tube placed through the urethra into the bladder to remove excess urine when the flow of urine is cut off, or to prevent urinary infection.

**Creatinine**—One of the “waste” substances normally excreted by the kidneys into the urine. When urine flow is slowed, creatinine may collect in the blood and cause toxic effects.

**Hyperplasia**—A condition where cells, such as those making up the prostate gland, rapidly divide abnormally and cause the organ to become enlarged.

**Hypertrophy**—A technical term for enlargement, as in BPH (benign prostatic hypertrophy).

**Urethra**—In males, the tube that conducts urine from the bladder through the penis to the outside of the body. When narrowed by an enlarging prostate, symptoms of BPH develop.

**Urinary retention**—The result of progressive obstruction of the urethra by an enlarging prostate, causing urine to remain in the bladder even after urination.

Alternatives to TUR, some only recently introduced, include:

- Laser ablation of the prostate. Laser energy is applied to the prostate through a special fiber passed through a cystoscope. The procedure is done in an operating room, and several patients have retained urine postoperatively.
- Transurethral incision of the prostate. Less invasive than standard TUR, an incision is made through the prostate to open up the part of the urethra passing through it. This may work well in men whose prostate is not grossly enlarged.
- Transurethral vaporization. A small roller ball is used to break up and vaporize the obstructing prostatic tissue, rather than cutting it away as in standard TUR. This is equally successful but patients usually can leave the hospital within 24 hours, and there is less blood loss.
- If the prostate is greatly enlarged—as is the case in about 5–10% of those diagnosed, an incision is made to perform an open **prostatectomy**, removing the entire gland under direct vision.

## Alternative treatment

An extract of the **saw palmetto** (*Serenoa repens* or *S. serrulata*) has been shown to stop or decrease the hyperplasia of the prostate. Symptoms of BPH will improve after taking the herb for one to two months, but continued use is recommended.

## Prognosis

In a man without symptoms whose prostate is enlarged, it is hard to predict when urinary symptoms will develop and how rapidly they will progress. For this reason some specialists (urologists) advise a period of "watchful waiting." When BPH is treated by conventional TUR, there is a small risk of complications but, in the great majority of men, urinary symptoms will be relieved and their quality of life will be much enhanced. In the future, it is possible that the less invasive forms of surgical treatment will be increasingly used to achieve results as good as those of the standard operation. It also is possible that new medications will be developed that shrink the prostate and eliminate obstructive symptoms so that surgery can be avoided altogether.

## Prevention

Whether or not BPH is caused by hormonal changes in aging men, there is no known way of preventing it. Once it does develop and symptoms are present that interfere seriously with the patient's life, timely medical or surgical treatment will reliably prevent symptoms from getting worse. Also, if the condition is treated before the prostate has become grossly enlarged, the risk of complications is minimal. One of the potentially most serious complications of BPH, urinary infection (and possible infection of the kidneys), can be prevented by using a catheter to drain excess urine out of the bladder so that it does not collect, stagnate, and become infected.

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### ORGANIZATIONS

- Prostate Health Council. American Foundation for Urologic Disease. 1128 N. Charles St., Baltimore, MD 21201. (800) 242-AFUD.

David A. Cramer, MD

*Entamoeba histolytica* infection see  
**Amebiasis**

Enteric fever see **Typhoid fever**

Enterically transmitted non-A non-B see  
**Hepatitis E**

## Enterobacterial infections

### Definition

Enterobacterial infections are disorders of the digestive tract and other organ systems produced by a group of gram-negative, rod-shaped bacteria called Enterobacteriaceae. Gram-negative means that the organisms do not retain the violet color of the dye used to make Gram stains. The most troublesome organism in this group is *Escherichia coli*. Other enterobacteria are species of *Salmonella*, *Shigella*, *Klebsiella*, *Enterobacter*, *Serratia*, *Proteus*, and *Yersinia*.

### Description

Enterobacterial infections can be produced by bacteria that normally live in the human digestive tract without causing serious disease, or by bacteria that enter from the outside. In many cases these infections are nosocomial, which means that they can be acquired in the hospital. *Klebsiella* and *Proteus* sometimes cause **pneumonia**, ear and sinus infections, and urinary tract infections. *Enterobacter* and *Serratia* often cause bacterial infection of the blood (**bacteremia**), particularly in patients with weakened immune systems.

**Diarrhea** caused by enterobacteria is a common problem in the United States. It is estimated that each person in the general population has an average of 1.5 episodes of diarrhea each year, with higher rates in children, institutionalized people, and Native Americans. This type of enterobacterial infection can range from a minor nuisance to a life-threatening disorder, especially in infants, elderly persons, **AIDS** patients, and malnourished people. Enterobacterial infections are one of the two leading killers of children in developing countries.

### Causes and symptoms

#### Causes

Enterobacterial infections in the digestive tract typically start when the organisms invade the mucous tissues that line the digestive tract. They may be bacteria that are

already present in the stomach and intestines, or they may be transmitted by contaminated food and water. It is also possible for enterobacterial infections to spread by person-to-person contact. The usual incubation period is 12–72 hours.

**ESCHERICHIA COLI INFECTIONS.** *E. coli* infections cause most of the enterobacterial infections in the United States. The organisms are categorized according to whether they are invasive or noninvasive. Noninvasive types of *E. coli* include what are called enteropathogenic *E. coli*, or EPEC, and enterotoxigenic *E. coli*, or ETEC. EPEC and ETEC types produce a bacterial poison (toxin) in the stomach that interacts with the digestive juices and causes the patient to lose large amounts of water through the intestines.

The invasive types of *E. coli* are called enterohemorrhagic *E. coli*, or EHEC, and enteroinvasive *E. coli*, or EIEC. These subtypes invade the stomach tissues directly, causing tissue destruction and bloody stools. EHEC can produce complications leading to **hemolytic-uremic syndrome** (HUS), a potentially fatal disorder marked by the destruction of red blood cells and kidney failure. EHEC has become a growing problem in the United States because of outbreaks caused by contaminated food. A particular type of EHEC known as O157:H7 has been identified since 1982 in undercooked hamburgers, unpasteurized milk, and apple juice. Between 2–7% of infections caused by O157:H7 develop into HUS.

### Symptoms

The symptoms of enterobacterial infections are sometimes classified according to the type of diarrhea they produce.

**WATERY DIARRHEA.** Patients infected with ETEC, EPEC, some types of *Salmonella*, and some types of *Shigella* develop a watery diarrhea. These infections are located in the small intestine, result from bacterial toxins interacting with digestive juices, do not produce inflammation, and do not usually need treatment with **antibiotics**.

**BLOODY DIARRHEA (DYSENTERY).** Bloody diarrhea is sometimes called dysentery. It is produced by EHEC, EIEC, some types of *Salmonella*, some types of *Shigella*, and *Yersinia*. In dysentery, the infection is located in the colon, cells and tissues are destroyed, inflammation is present, and antibiotic therapy is usually required.

**NECROTIZING ENTEROCOLITIS (NEC).** **Necrotizing enterocolitis** (NEC) is a disorder that begins in newborn infants shortly after birth. Although NEC is not yet fully understood, it is thought that it results from a bacterial or viral invasion of damaged intestinal tissues. The disease organisms then cause the **death** (necrosis) of bowel tis-

sue or **gangrene** of the bowel. NEC is primarily a disease of **prematurity**; 60–80% of cases occur in high-risk preterm infants. NEC is responsible for 2–5% of cases in newborn intensive care units (NICU). Enterobacteriaceae that have been identified in infants with NEC include *Salmonella*, *E. coli*, *Klebsiella*, and *Enterobacter*.

## Diagnosis

### Patient history

The diagnosis of enterobacterial infections is complicated by the fact that viruses, protozoa, and other types of bacteria can also cause diarrhea. In most cases of mild diarrhea, it is not critical to identify the organism because the disorder is self-limiting. Some groups of patients, however, should have stool tests. They include:

- patients with bloody diarrhea,
- patients with watery diarrhea who have become dehydrated,
- patients with watery diarrhea that has lasted longer than three days without decreasing in amount,
- patients with disorders of the immune system.

The patient history is useful for public health reasons as well as helping the doctor determine what type of enterobacterium may be causing the infection. The doctor will ask about the frequency and appearance of the diarrhea as well as other digestive symptoms. If the patient is nauseated and vomiting, the infection is more likely to be located in the small intestine. If the patient is running a **fever**, a diagnosis of dysentery is more likely. The doctor will also ask if anyone else in the patient's family or workplace is sick. Some types of enterobacteriaceae are more likely to cause group outbreaks than others. Other questions include the patient's food intake over the last few days and whether he or she has recently traveled to countries with **typhoid fever** or **cholera** outbreaks.

### Physical examination

The most important parts of the **physical examination** are checking for signs of severe fluid loss and examining the abdomen to rule out typhoid fever. The doctor will look at the inside of the patient's mouth and evaluate the skin for signs of **dehydration**. The presence of a skin rash and an enlarged spleen suggests typhoid rather than a bacterial infection. If the patient's abdomen hurts when the doctor examines it, a diagnosis of dysentery is more likely.

### Laboratory tests

The most common test that is used to identify the cause of diarrhea is the stool test. Examining a stool sample under a microscope can help to rule out parasitic and

protozoal infections. Routine stool cultures, however, cannot be used to identify any of the four types of *E. coli* that cause intestinal infections. ETEC, EPEC, and EIEC are unusual in the United States and can usually be identified only by specialists in research laboratories. Because of concern about EHEC outbreaks, however, most laboratories in the United States can now screen for O157:H7 with a test that identifies its characteristic toxin. All patients with bloody diarrhea should have a stool sample tested for *E. coli* O157:H7.

### Treatment

The initial treatment of enterobacterial diarrhea is usually empiric. Empiric means that the doctor treats the patient on the basis of the visible symptoms and professional experience in treating infections, without waiting for laboratory test results. Since the results of stool cultures can take as long as two days, it is important to prevent dehydration. The patient will be given fluids to restore the electrolyte balance and paregoric to relieve abdominal cramping.

Newborn infants and patients with immune system disorders will be given antibiotics intravenously once the organism has been identified. Gentamicin, tobramycin, and amikacin are being used more frequently to treat enterobacterial infections because many of the organisms are becoming resistant to ampicillin and cephalosporin antibiotics.

### Alternative treatment

Alternative treatments for diarrhea are intended to relieve the discomfort of abdominal cramping. Most alternative practitioners advise consulting a medical doctor if the patient has sunken eyes, dry eyes or mouth, or other signs of dehydration.

#### *Herbal medicine*

Herbalists may recommend cloves taken as an infusion or ginger given in drop doses to control intestinal cramps, eliminate gas, and prevent vomiting. Peppermint (*Mentha piperita*) or chamomile (*Matricaria recutita*) tea may also ease cramps and intestinal spasms.

#### *Homeopathy*

Homeopathic practitioners frequently recommend *Arsenicum album* for diarrhea caused by contaminated food, and *Belladonna* for diarrhea that comes on suddenly with mucus in the stools. *Veratrum album* would be given for watery diarrhea, and *Podophyllum* for diarrhea with few other symptoms.

## KEY TERMS

**Dysentery**—A type of diarrhea caused by infection and characterized by mucus and blood in the stools.

**Empirical treatment**—Medical treatment that is given on the basis of the doctor's observations and experience.

***Escherichia coli***—A type of enterobacterium that is responsible for most cases of severe bacterial diarrhea in the United States.

**Hemolytic-uremic syndrome (HUS)**—A potentially fatal complication of *E. coli* infections characterized by kidney failure and destruction of red blood cells.

**Necrotizing enterocolitis (NEC)**—A disorder in newborns caused by bacterial or viral invasion of vulnerable intestinal tissues.

**Nosocomial infections**—Infections acquired in hospitals.

**Toxin**—A poison produced by certain types of bacteria.

### Prognosis

The prognosis for most enterobacterial infections is good; most patients recover in about a week or 10 days without needing antibiotics. HUS, on the other hand, has a mortality rate of 3–5% even with intensive care. About a third of the survivors have long-term problems with kidney function, and another 8% develop high blood pressure, seizure disorders, and blindness.

### Prevention

The World Health Organization (WHO) offers the following suggestions for preventing enterobacterial infections, including *E. coli* O157:H7 dysentery:

- Cook ground beef or hamburgers until the meat is thoroughly done. Juices from the meat should be completely clear, not pink or red. All parts of the meat should reach a temperature of 70°C (158°F) or higher.
- Do not drink unpasteurized milk or use products made from raw milk.
- Wash hands thoroughly and frequently, especially after using the toilet.
- Wash fruits and vegetables carefully, or peel them. Keep all kitchen surfaces and serving utensils clean.

- If drinking water is not known to be safe, boil it or drink bottled water.
- Keep cooked foods separate from raw foods, and avoid touching cooked foods with knives or other utensils that have been used with raw meat.

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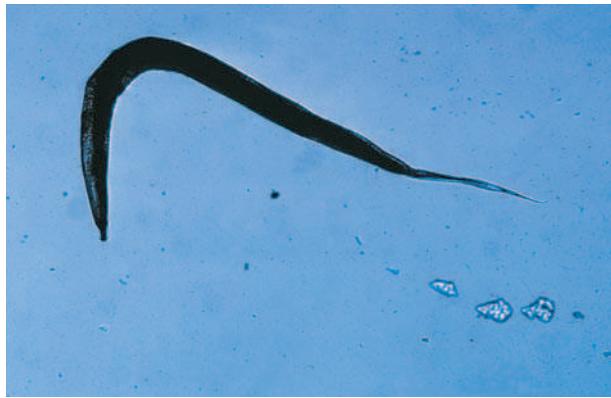
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The pinworm of the genus *Enterobius* pictured above is the source of this infestation occurring in children. (Photo Researchers, Inc. Reproduced by permission.)

## Description

Enterobiasis is also called seatworm infection or oxyuriasis. In the United States, enterobiasis is the most common worm infection, and some estimate that approximately 10% of the United States population is infected. Worldwide, approximately 200 million people are infected. Enterobiasis can affect people of any age, but is most common among children ages 5–14 and particularly affects those in the daycare setting.

## Causes and symptoms

The disease is highly contagious and is caused by a parasitic worm called *Enterobius vermicularis*. The adult female worm is about the size of a staple (approximately 0.4 in [1 cm] long and 0.02 in [0.5 mm] wide) and has a pointed tip. The disease is transmitted by ingesting the eggs of the pinworm. These eggs travel to the small intestine where, after approximately one month, they hatch and mature into adult worms. During the night, the female adult worms travel to the area around the anus and deposit eggs in the folds of the anal area. A single female pinworm can lay 10,000 eggs and, after laying eggs, dies. The eggs are capable of causing infection after six hours at body temperature.

Significant itching in the anal region is caused by the movement of the adult worm as the eggs are deposited. When an individual scratches the anal region, the tiny eggs get under the finger nails and in the underwear and night clothes. Anything the individual touches with the contaminated fingers, for example, toys, bedding, blankets, bathroom door knobs, or sinks, becomes contaminated. The eggs are very hardy and can live on surfaces for two to three weeks. Anyone touching these contaminated surfaces can ingest the eggs and become infected. An individ-

## Enterobiasis

### Definition

Enterobiasis, or pinworm infection as it is commonly called, is an intestinal infection caused by the parasitic roundworm called *Enterobius vermicularis*. The most common symptom of this irritating, but not particularly dangerous, disease is **itching** around the anal area.

## KEY TERMS

**Anus**—The opening through which feces are eliminated.

**Hemorrhoid**—An area around the anus where veins become dilated and the tissue swells, causing itching and pain.

**Rectum**—The end of the large intestine in which feces collects for elimination through the anus.

**Vaginitis**—Inflammation of the vagina.

ual can also become infected by inhaling and swallowing the eggs, for example, when the bedcovers are shaken.

Many individuals with enterobiasis exhibit no symptoms. When present, however, symptoms of the infection begin approximately two weeks after ingesting the pinworm eggs. The main symptom is itching around the anus. Because the itching intensifies at night, when the female worms come to the anus to lay eggs, it often leads to disrupted sleep and irritability. Poor sleeping at night in small children can be related to pinworms. Occasionally, the itching causes some bleeding and bruising in the region, and secondary bacterial infections can occur. In females, the itching may spread to the vagina and sometimes causes an infection of the vaginal region (vaginitis). Enterobiasis usually lasts one to two months.

## Diagnosis

First, a physician will rule out other potential causes of the itching, such as **hemorrhoids**, lice, or fungal or bacterial infection. Once these have been ruled out, an accurate diagnosis of enterobiasis will require that either the eggs or the adult worms are detected. Rarely, the adult worms are seen as thin, yellowish-white threads, about 0.4 in (1 cm) long, in the stools of the infected person. Usually, an hour or so after the individual goes to sleep, the adult female worms may be seen moving around laying eggs if a flashlight is shone at the rectal area.

An easier method is to observe the eggs under the microscope. In order to collect a specimen for laboratory diagnosis, the physician may provide a paddle with a sticky adhesive on one side, or an individual may be instructed to place a piece of shiny cellophane tape sticky side down against the anal opening. The best time to perform this test is at night or as soon as the individual wakes up in the morning, before having a bowel movement or taking a bath or shower. The pinworm eggs will stick to the tape, which can then be placed on a specimen

slide. When under a microscope in the laboratory, the eggs will be clearly visible.

## Treatment

In order to treat the disease, either mebendazole (Vermox) or pyrantel pamoate (Pin-X) will be given in two oral doses spaced two weeks apart. These medications eradicate the infection in approximately 90% of cases. Re-infection is common and several treatments may be required. Because the infection is easily spread through contact with contaminated clothing or surfaces, it is recommended that all family members receive the therapeutic dose. Sometimes a series of six treatments are given, each spaced two weeks apart. If family members continue to be infected, a source outside the house may be responsible.

To relieve the rectal itching, a shallow warm bath with either half a cup of table salt, or Epsom salts is recommended. Also, application of an ointment containing zinc oxide or regular petroleum jelly can be used to relieve rectal itching.

## Prognosis

Pinworms cause little damage and can be easily eradicated with proper treatment. Full recovery is expected.

## Prevention

The disease can be prevented by treating all the infected cases and thus eliminating the source of infection. Some ways to keep from catching or spreading the disease include the following recommendations:

- wash hands thoroughly before handling food and eating
- keep finger nails short and clean
- avoiding scratching the anal area
- take early morning showers to wash away eggs deposited overnight
- once the infection has been identified, and treatment is started, change the bed linen, night clothes, and underwear daily
- machine wash linens in hot water and dry with heat to kill any eggs
- open the blinds or curtains since eggs are sensitive to sunlight

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Lata Cherath, PhD

Enterohemorrhagic *E. coli* see *Escherichia coli*

## Enterostomy

### Definition

An enterostomy is an operation in which the surgeon makes a passage into the patient's small intestine through the abdomen with an opening to allow for drainage or to insert a tube for feeding. The opening is called a stoma, from the Greek word for mouth. Enterostomies may be either temporary or permanent. They are classified according to the part of the intestine that is used to create the stoma. If the ileum, which is the lowest of the three sections of the small intestine, is used to make the stoma, the operation is called an ileostomy. If the jejunum, which is the middle section of the small intestine, is used, the operation is called a jejunostomy. Some people use the word *ostomy* as a word that covers all types of enterostomies.

### Purpose

Enterostomies are performed in order to create a new opening for the passage of fecal matter when normal intestinal functioning is interrupted or when diseases of the intestines cannot be treated by medications or less radical surgery. Some situations that may require enterostomies include:

- Healing of inflamed bowel segments. Enterostomies performed for this reason are usually temporary.
- Emergency treatment of gunshot or other penetrating **wounds** of the abdomen. An enterostomy is needed to prevent the contents of the intestine from causing a serious inflammation of the inside of the abdominal cavity (**peritonitis**). These enterostomies are also often temporary.
- Placement of a tube for enteral feeding. Enteral feeding is a method for conveying nutritional solutions directly into the stomach or jejunum through a tube. Tube enterostomies may be long-term but are not permanent.
- Removal of diseased sections of the intestines. Ileostomies performed for this reason are permanent.

The most common disorders requiring permanent ileostomy are **Crohn's disease**, **familial polyposis**, and **ulcerative colitis**. Familial polyposis and ulcerative colitis are serious health risks because they can develop into **cancer**.

- Treatment of advanced cancer or other causes of intestinal obstruction.

### Precautions

Enterostomies are usually performed only as emergency treatments for traumatic injuries in the abdomen or as final measures for serious disorders of the intestines. Most patients do not refuse to have the operation performed when the need for it is explained to them. A small minority, however, refuse enterostomies because of strong psychological reactions to personal disfigurement and the need to relearn bowel habits.

### Description

#### Ileostomy

Ileostomies represent about 25% of enterostomies. They are performed after the surgeon removes a diseased colon and sometimes the rectum as well. The most common ileostomy is called a Brooke ileostomy after the English surgeon who developed it. In a Brooke ileostomy, the surgeon makes the stoma in the lower right section of the abdomen. The ileum is pulled through an opening (incision) in the muscle layer. The surgeon then turns the cut end of the intestine inside out and sews it to the edges of the hole. He or she then positions an appliance for collecting the fecal material. The appliance consists of a plastic bag that fits over the stoma and lies flat against the abdomen. The patient is taught to drain the bag from time to time during the day. Ileostomies need to be emptied frequently because the digested food contains large amounts of water. Shortly after the operation, the ileostomy produces 1–2 qt (0.9–1.91) of fluid per day; after a month or two of adjustment, the volume decreases to 1–2 pt (0.5–0.9) per day.

**KOCK POUCH (CONTINENT ILEOSTOMY).** The Kock pouch is a variation of the basic ileostomy and is named for its Swedish inventor. In the Kock technique, the surgeon forms a pouch inside the abdominal cavity behind the stoma that collects the fecal material. The stoma is shaped into a valve to prevent fluid from leaking onto the patient's abdomen. The patient then empties the pouch several times daily by inserting a tube (catheter) through the valve. The Kock technique is sometimes called a continent ileostomy because the fluid is contained inside the abdomen. It is successful in 70–90% of patients who have it done.

## KEY TERMS

**Crohn's disease**—A disease of the intestines that causes inflammation leading to scarring, thickening of the walls of the intestine, and eventual obstruction.

**Duodenum**—The first of the three segments of the small intestine. The duodenum connects the stomach and the jejunum.

**Enteral nutrition**—A technique for feeding patients with liquid formulas conveyed directly into the stomach or jejunum through tubes.

**Enterostomal therapist (ET)**—A specialized counselor, usually a registered nurse, who provides ostomy patients with education and counseling before the operation. After surgery, the ET helps the patient learn to take care of the stoma and appliance, and offers long-term emotional support.

**Familial polyposis**—A disease that runs in families in which lumps of tissue (polyps) form inside the colon. Familial polyposis may develop into cancer.

**Ileum**—The third segment of the small intestine, connecting the jejunum and the large intestine.

**Jejunum**—The second of the three segments of the small intestine, connecting the duodenum and the ileum.

**Kock pouch**—A type of ileostomy in which the surgeon forms an artificial rectum from a section of the ileum. A Kock pouch is sometimes called a continent ileostomy because it is drained with a tube.

**Ostomy**—A common term for all types of enterostomies.

**Stoma**—The surgically constructed mouth or passage between the intestine and the outside of the patient's body.

**Tube enterostomy**—An enterostomy performed to allow the insertion of a feeding tube into the jejunum or stomach.

**Ulcerative colitis**—A disease of the colon characterized by inflammation of the mucous lining, ulcerated areas of tissue, and bloody diarrhea.

### *Jejunostomy*

A jejunostomy is similar to an ileostomy except that the stoma is placed in the second section of the small intestine rather than the third. Jejunostomies are performed less frequently than ileostomies. They are almost always temporary procedures.

### *Tube enterostomies*

Tube enterostomies are operations in which the surgeon makes a stoma into the stomach itself or the jejunum in order to insert a tube for liquid nutrients. Tube enterostomies are performed in patients who need tube feeding for longer than six weeks, or who have had recent mouth or nose surgery. As long as the patient's intestinal tract can function, **tube feedings** are considered preferable to intravenous feeding. Enteral **nutrition** is safer than intravenous fluids and helps to keep the patient's digestive tract functioning.

### **Preparation**

Preoperative preparation includes both patient education and physical preparation.

### *Patient education*

If the patient is going to have a permanent ileostomy, the doctor will explain what will happen during the operation and why it is necessary. Most patients are willing to accept an **ostomy** as an alternative to the chronic **pain** and **diarrhea** of ulcerative colitis or the risk of cancer from other intestinal disorders. The patient can also meet with an enterostomal therapist (ET) or a member of the United Ostomy Association, which is a support group for people with ostomies.

### *Medical preparation*

The patient is prepared for surgery with an evaluation of his or her nutritional status, possible need for blood transfusions, and **antibiotics** if necessary. If the patient does not have an intestinal obstruction or severe inflammation, he or she may be given a large quantity of a polyethylene glycol (PEG) solution to cleanse the intestines before surgery.

### **Aftercare**

Aftercare of an enterostomy is both psychological and medical.

### Medical aftercare

If the enterostomy is temporary, aftercare consists of the usual monitoring of surgical wounds for infection or bleeding. If the patient has had a permanent ileostomy, aftercare includes learning to use the appliance or empty the Kock pouch; learning to keep the stoma clean; and readjusting bathroom habits. Recovery takes a long time because major surgery is a **shock** to the system and the intestines take several days to resume normal functioning. The patient's fluid intake and output will be checked frequently to minimize the risk of **dehydration**.

### Patient education

Ileostomy patients must learn to watch their fluid and salt intake. They are at greater risk of becoming dehydrated in hot weather, from **exercise**, or from diarrhea. In some cases they may need extra bananas or orange juice in the diet to keep up the level of potassium in the blood.

Patient education includes social concerns as well as physical self-care. Many ileostomy patients are worried about the effects of the operation on their close relationships and employment. If the patient has not seen an ET before the operation, the aftercare period is a good time to find out about self-help and support groups. The ET can also evaluate the patient's emotional reactions to the ostomy.

### Risks

Enterostomies are not considered high-risk operations by themselves. About 40% of ileostomy patients have complications afterward, however; about 15% require minor surgical corrections. Possible complications include:

- skin irritation caused by leakage of digestive fluids onto the skin around the stoma, irritation is the most common complication of ileostomies
- diarrhea
- the development of abscesses
- gallstones or stones in the urinary tract
- inflammation of the ileum
- odors (can often be prevented by a change in diet)
- intestinal obstruction
- a section of the bowel pushing out of the body (pro-lapse)

### Normal results

Normal results include recovery from the surgery with few or no complications. About 95% of people with ostomies recover completely, are able to return to work,

and consider themselves to be in good health. Many ileostomy patients enjoy being able to eat a full range of foods rather than living on a restricted diet. Some patients, however, need to be referred to psychotherapists to deal with depression or other emotional problems after the operation.

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- United Ostomy Association, Inc. (UOA). 19772 MacArthur Blvd., Suite 200, Irvine, CA 92612-2405. (800) 826-0826. <<http://www.uoa.org>>

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## Enterovirus infections

### Definition

Enteroviruses are so named because they reproduce initially in the gastrointestinal tract after infection occurs. Despite this, they usually do not lead to intestinal symptoms; rather it is their spread to organs, such as the nervous system, heart, skin, and others that causes disease. Enteroviruses are part of a larger group of viruses known as Picornaviruses. The word comes from the combination of "pico" (Spanish, meaning "a little bit"), and RNA (ribonucleic acid, an important component of genetic material).

### Description

There are four groups of enteroviruses: Coxsackievirus, Echovirus, ungrouped Enterovirus, and Y Poliovirus.

Viruses are generally divided into those that use DNA (desoxyribonucleic acid) or RNA as their genetic material; all enteroviruses are RNA viruses. They are found worldwide, but infection is more common in areas of poor hygiene and overcrowding.

Although most cases of enterovirus do not produce symptoms, some five to 10 million individuals in the United States each year suffer from one of the enteroviral diseases. Illness is more common in the very young. While there are close to seventy different strains of enteroviruses, over 70% of infections are caused by only 10 types.

The virus is most commonly transmitted by the fecal-oral route (contamination of fingers or objects by human waste material); in some instances transmission is through contaminated food or water. Passage of some strains of virus by way of air droplets can lead to respiratory illness. Infection of fetuses by way of the placenta has also been documented. Breast milk contains antibodies which can protect newborns.

The incubation period for most enteroviruses ranges from two to 14 days. In areas of temperate climate, infections occur mainly in the summer and fall.

### Causes and symptoms

Enteroviruses are believed to be the cause of at least 10 distinct illnesses. Once they enter the body, they multiply in the cells that line the gastrointestinal tract, and eventually reach sites of lymphatic tissue (such as the tonsils). While most of these diseases are of short duration and do not cause significant injury, some can produce severe illness.

The main syndromes caused by the various enteroviruses are the following:

- Summer gripe (nonspecific febrile illness). This is the most common syndrome, and is characterized by flu-like symptoms of **fever**, **headache**, and weakness, that typically last three to four days. Many patients also develop upper respiratory symptoms and some **nausea and vomiting**. One of the major ways to distinguish this disease from **influenza**, is the fact that gripe most often occurs in the summer.
- Generalized disease of the newborn is a potentially serious infection in which infants from one week to three months of age develop a syndrome that can be difficult to distinguish from a severe bacterial infection. Fever, irritability, and decreased responsiveness or excessive sleepiness are the major symptoms. Inflammation of heart muscle (**myocarditis**), low blood pressure, hepatitis, and **meningitis** sometimes complicate the illness.

- Aseptic meningitis **encephalitis** is a well known syndrome caused by this group of viruses. In fact, enteroviruses are responsible for over 90% of cases of aseptic meningitis, and most often hits children and young adults. Headache, fever, avoidance of light, and **eye pain** are characteristic. Drowsiness may be prominent, and other symptoms include **sore throat**, **cough**, muscle pain, and rash. Occasionally, not only the meninges—the covering around the brain and spinal cord—is infected, but also brain tissue itself, producing encephalitis. The illness resolves after about a week or so, and permanent damage is unusual. Enteroviruses can also produce the Guillain-Barré syndrome, which involves weakness and **paralysis** of the extremities and even the muscles of respiration.

- Pleurodynia (Bornholm's disease) is due to viral infection and inflammation of the chest and abdominal muscles used for breathing. Pain occurs as acute episodes, lasting 30 minutes or so. Coxsackie B virus is the usual cause of the illness.
- Myocarditis and/or **pericarditis** involves infection of the heart muscle (myocardium) and the covering around the heart (pericardium). Infants and young adults are the most susceptible, and for some reason, over two-thirds of cases occur in males. The disease usually begins as an upper respiratory tract infection with cough, **shortness of breath**, and fever. Chest pain, increasing shortness of breath, irregularities of cardiac rhythm, and **heart failure** sometimes develop. Some patients wind up with long term heart failure if the heart muscle is significantly affected.
- Exanthems is the medical term for **rashes**, and enterovirus is the number one cause of summer and fall rashes in children. They occur anywhere on the body, and often resemble diseases such as **measles**.
- Hand-foot-and-mouth disease occurs initially as a sore throat (often involving the tongue as well), and is followed by a rash on the hands, and sometimes the feet. The rash often forms small blisters, which lead to ulcers. Symptoms generally resolve within a week. A specific Coxsackievirus (A16) is the most frequent cause of this highly infectious disease.
- Herpangina is most often caused by one of the Coxsackie A viruses, and appears as the acute onset of fever and sore throat. This last symptom is particularly severe, as the virus produces multiple ulcers in the throat. Swallowing becomes very painful; symptoms can persist for several weeks.
- Acute hemorrhagic **conjunctivitis** involves viral infection of the conjunctiva, which is a covering around the eye. Pain, blurred vision, aversion to light, and a discharge from the eye are the main symptoms. Headache

and fever occur in about one in five patients. The disease runs its course in about 10 days.

A number of other illnesses have been attributed to enteroviruses, including **pneumonia** and other respiratory infections, myositis or muscle inflammation, arthritis, and acute inflammation of the kidneys. It is clear then that these viruses produce a number of various illnesses, most often in younger age groups.

## Diagnosis

In the majority of cases, diagnosis is based on the characteristic symptoms that the virus produces (such as the chest pain in pleurodynia). Rarely is it necessary to identify a specific strain of virus causing the illness. It is more important to be certain that the infection is due to a virus which does not require treatment with **antibiotics**.

Culture, or growing the organism outside of the body, is helpful only when obtained from areas that tend to indicate recent infection, such as from swollen joints, cerebrospinal fluid, or blood. Cultures from other areas, such as the throat, can be misleading. This is because the virus may remain for long periods of time in places with a large amount of lymphatic tissue. As a rule, cultures done early in the illness are more likely to identify the virus.

New techniques that involve identification of viral genetic material (PCR) are useful in certain cases, but are not indicated for routine testing.

## Treatment

As noted above, enterovirus is capable of attacking many different organs and producing a variety of symptoms. Most infections are mild and improve without complications, and require no specific therapy. When the virus attacks critical organs however, such as the heart, respiratory muscles, nervous system, etc., then specialized care is often needed.

As of 2001, no effective antiviral medication for enterovirus has undergone investigation in patients, though some drugs appear promising for the future. In some patients who are unable to produce antibodies (hypogammaglobulinemia), administrating antibodies themselves is helpful.

## Prognosis

The overall outlook for enterovirus infection depends on the organs involved, and the immune condition of the individual patient. Unless vital organs are involved or immunity is abnormal, infection causes few problems. On the other hand, patients who have diseases

## KEY TERMS

**Antibodies**—Proteins that are formed by the body and play a role in defense against infection.

**Antibiotic**—A medication that is designed to kill or weaken bacteria.

**Meninges**—Outer covering of the spinal cord and brain. Infection is called meningitis, which can lead to damage to the brain or spinal cord and lead to death.

that affect antibody production can develop chronic infection of the brain or meninges.

## Prevention

In the hospital setting, the best means of avoiding transmission of infection is the use of good hand-washing practices and other appropriate precautions (gowns and gloves for hospital staff). The virus is found in feces for up to one week after infection; therefore precautions that isolate waste material (enteric precautions) will help decrease the chance of spreading the illness.

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David Kaminstein, MD

Entropy see **Eyelid disorders**

Enuresis see **Bed-wetting**

Environmental medicine see **Wilderness medicine**

## Enzyme therapy

### Definition

Enzyme therapy is a plan of dietary supplements of plant and animal enzymes used to facilitate the digestive process and improve the body's ability to maintain balanced metabolism.

### Purpose

In traditional medicine, enzyme supplements are often prescribed for patients suffering from disorders that affect the digestive process, such as **cystic fibrosis**, Gaucher's disease, and **celiac disease**. A program of enzyme supplementation is rarely recommended for healthy patients. However, proponents of enzyme therapy believe that such a program is beneficial for everyone. They point to enzymes' ability to purify the blood, strengthen the immune system, enhance mental capacity, cleanse the colon, and maintain proper pH balance in urine. They feel that by improving the digestive process, the body is better able to combat infection and disease.

Some evidence exists that pancreatic enzymes derived from animal sources are helpful in **cancer** treatment. The enzymes may be able to dissolve the coating on cancer cells and may make it easier for the immune system to attack the cancer.

A partial list of the wide variety of complaints and illnesses that can be treated by enzyme therapy includes:

- AIDS
- anemia
- alcohol consumption
- anxiety
- acute inflammation
- back pain
- cancer
- colds
- chronic **fatigue** syndrome
- colitis

- constipation
- **diarrhea**
- food **allergies**
- gastritis
- gastric duodenal ulcer
- gout
- headaches
- hepatitis
- hypoglycemia
- infections
- mucous congestion
- multiple sclerosis
- nervous disorders
- nutritional disorders
- obesity
- premenstrual syndrome (PMS)
- stress

### Description

#### Origins

Enzymes are protein molecules used by the body to perform all of its chemical actions and reactions. The body manufactures several thousands of enzymes. Among them are the digestive enzymes produced by the stomach, pancreas, small intestine, and the salivary glands of the mouth. Their energy-producing properties are responsible for not only the digestion of nutrients, but their absorption, transportation, metabolization, and elimination as well.

Enzyme therapy is based on the work of Dr. Edward Howell in the 1920s and 1930s. Howell proposed that enzymes from foods work in the stomach to pre-digest food. He advocated the consumption of large amounts of plant enzymes, theorizing that if the body had to use less of its own enzymes for digestion, it could store them for maintaining metabolic harmony. Four categories of plant enzymes are helpful in pre-digestion: protease, amylase, lipase, and cellulase. Cellulase is particularly helpful because the body is unable to produce it.

Animal enzymes, such as pepsin extracted from the stomach of pigs, work more effectively in the duodenum. They are typically used for the treatment of nondigestive ailments.

The seven categories of food enzymes and their activities

- amylase breaks down starches
- cellulase breaks down fibers
- lactase breaks down dairy products
- lipase breaks down fats
- maltase breaks down grains
- protease breaks down proteins
- sucrase breaks down sugars

Enzyme theory generated further interest as the human diet became more dependent on processed and cooked foods. Enzymes are extremely sensitive to heat, and temperatures above 118°F (48°C) destroy them. Modern processes of pasteurization, canning, and microwaving are particularly harmful to the enzymes in food.

Enzyme supplements are extracted from plants like pineapple and papaya and from the organs of cows and pigs. The supplements are typically given in tablet or capsule form. Pancreatic enzymes may also be given by injection. The dosage varies with the condition being treated. For nondigestive ailments, the supplements are taken in the hour before meals so that they can be quickly absorbed into the blood. For digestive ailments, the supplements are taken immediately before meals accompanied by a large glass of fluids. Pancreatic enzymes may be accompanied by doses of vitamin A.

## 7Preparations

No special preparations are necessary before beginning enzyme therapy. However, it is always advisable to talk to a doctor or pharmacist before purchasing enzymes and beginning therapy.

## Precautions

People with allergies to beef, pork, pineapples, and papaya may suffer allergic reactions to enzyme supplements. Tablets are often coated to prevent them from breaking down in the stomach, and usually shouldn't be chewed or crushed. People who have difficulty swallowing pills can request enzyme supplements in capsule form. The capsules can then be opened and the contents sprinkled onto soft foods like applesauce.

## Side effects

Side effects associated with enzyme therapy include **heartburn, nausea and vomiting**, diarrhea, bloating, gas, and **acne**. According to the principles of therapy, these are temporary cleansing symptoms. Drinking eight to ten glasses of water daily and getting regular **exercise** can reduce the discomfort of these side effects. Individuals may also experience an increase in bowel move-

## KEY TERMS

**Celiac disease**—A chronic disease characterized by defective digestion and use of fats.

**Cystic fibrosis**—A genetic disease that causes multiple digestive, excretion, and respiratory complications. Among the effects, the pancreas fails to provide secretions needed for the digestion of food.

**Duodenum**—The first part of the small intestine.

**Gaucher's disease**—A rare genetic disease caused by a deficiency of enzymes needed for the processing of fatty acids.

**Metabolism**—The system of chemical processes necessary for living cells to remain healthy.

ments, perhaps one or two per day. This is also considered a positive effect.

Plant enzymes are safe for pregnant women, although they should always check with a doctor before using enzymes. Pregnant women should avoid animal enzymes. In rare cases, extremely high doses of enzymes can result in a build up of uric acid in the blood or urine and can cause a break down of proteins.

## Research and general acceptance

In the United States, the Food and Drug Administration (FDA) has classified enzymes as a food. Therefore, they can be purchased without a prescription. However, insurance coverage is usually dependent upon the therapy resulting from a doctor's orders.

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Mary McNulty

Eosinophilic granuloma see **Histiocytosis X**

## Eosinophilic pneumonia

### Definition

Eosinophilic **pneumonia** is a group of diseases in which there is an above normal number of eosinophils in the lungs and blood.

### Description

Eosinophilia is an increase in the number of eosinophils. Eosinophilic pneumonia is characterized by a large number of eosinophils in the lungs, usually in the absence of an infectious disease. Eosinophils are one of the white blood cells and are classified as a granulocyte. They are part of the non-specific immune system and participate in inflammatory reactions. Eosinophils contain cationic molecules that are useful for destroying infectious agents, especially helminthic parasites (worms). There are several types of eosinophilic pneumonia. Loffler's pneumonia is a temporary infiltration of eosinophils into the lungs. The patient will feel tired, have a **cough**, spasms of the bronchial airway, and difficulty breathing. Loffler's pneumonia will clear spontaneously, but slowly over the course of about a month. Another form of eosinophilic pneumonia, pulmonary infiltrates with eosinophilia (PIE), is a more serious and potentially fatal disease. In PIE, the patient experiences **asthma**, pulmonary infiltrates, disorders of the peripheral nervous system, central nervous systems symptoms, and periarthritis nodosa.

### Causes and symptoms

Pneumonia with eosinophils occurs as part of a hypersensitivity reaction. A hypersensitivity reaction is an over-reaction of the immune system to a particular stimulus. As part of the hypersensitive reaction, cells of the immune system are produced in increased numbers and migrate into areas targeted by the hypersensitivity reaction. In the case of eosinophilic pneumonia, the lungs are the target. Generally, eosinophilia pneumonia is not a reaction to an infection. There is a correlation between asthma and eosinophilic pneumonia. Eosinophilic pneumonia can also be caused by drugs and, in some people, by polluted air. The symptoms range from mild (coughing, **wheezing**, and **shortness of breath**) to severe and life threatening (severe shortness of breath and difficulty getting enough oxygen). The symptoms may resolve spontaneously or can persist for long periods of time. In a few cases, the disease may rapidly produce life-threatening pneumonia.

### KEY TERMS

**Infiltrates**—Cells or body fluids that have passed into a tissue or body cavity.

**Sputum**—Material coughed up from the throat or lungs.

### Diagnosis

Since eosinophilia is common to a number of conditions, the physician must rule out asthma and infection by helminths when diagnosing eosinophilic pneumonia. A whole **blood count** will reveal an increased number of eosinophils in the blood. An x ray of the lungs may show the presence of infiltrates (the eosinophils and fluid). If sputum is produced in coughing, eosinophils will be seen instead of the more normal profile of granulocytes seen when an infectious agent is present.

### Treatment

Eosinophilic pneumonia may not respond to drugs used to treat asthma. Eosinophilic pneumonia is usually treated with steroids, particularly glucocorticosteroids. Steroids are not effective against infectious agents, but the main disease process in eosinophilic pneumonia is an inflammatory reaction, not a response to infection. When eosinophilia is produced as a consequence of asthma or an infection by helminths, treatment of the asthma or helminths will reduce the eosinophilia.

### Resources

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Ephedrine see **Bronchodilators**

Epicondylitis see **Tennis elbow**

Epidemic icterus see **Hepatitis A**

Epidemic typhus see **Typhus**

Epidemic viral gastroenteritis see **Rotavirus infections**

# Epidermolysis bullosa

## Definition

Epidermolysis bullosa (EB) is a group of rare inherited skin diseases that are characterized by the development of blisters following minimal pressure to the skin. Blistering often appears in infancy in response to simply being held or handled. In rarer forms of the disorder, EB can be life-threatening. There is no cure for the disorder. Treatment focuses on preventing and treating **wounds** and infection.

## Description

Epidermolysis bullosa has three major forms and at least 16 subtypes. The three major forms are EB simplex, junctional EB, and dystrophic EB. These can range in severity from mild blistering to more disfiguring and life-threatening disease. Physicians diagnose the form of the disease based on where the blister forms in relation to the epidermis (the skin's outermost layer) and the deeper dermis layer.

The prevalence of epidermolysis varies among different populations. A study in Scotland estimated the prevalence to be one in 20,400. Researchers in other parts of the world estimate the prevalence to be one in 100,000. This variance is due to the variability of expression. Many cases of epidermolysis bullosa are often not accurately diagnosed and thus, are not reported.

## Causes and symptoms

EB can be inherited as the result of a dominant genetic abnormality (only one parent carries the abnormal gene) or a recessive genetic abnormality (both parents carry the abnormal gene).

EB simplex results from mutations in genes responsible for keratin 5 and 14, which are proteins that give cells of the epidermis its structure. EB simplex is transmitted in an autosomal dominant fashion.

Dystrophic EB is caused by mutations in genes for type VII collagen, the protein contained in the fibers anchoring the epidermis to the deeper layers of the skin. The genetic mutations for junctional EB are found in the genes responsible for producing the protein Laminin-5. Dystrophic EB is an autosomal disorder and will only result if both parents transmit an abnormal gene during conception.

EB simplex, the most common form of EB, is the least serious form of the disease. In most affected individuals, the blisters are mild and do not scar after they heal. Some forms of EB simplex affect just the hands and

## KEY TERMS

**Collagen**—The main supportive protein of cartilage, connective tissue, tendon, skin, and bone.

**Dermis**—The layer of skin beneath the epidermis.

**Epidermis**—The outermost layer of the skin.

**Keratin**—A tough, nonwater-soluble protein found in the nails, hair, and the outermost layer of skin. Human hair is made up largely of keratin.

feet. Other forms of EB simplex can lead to more widespread blistering, as well as hair loss and missing teeth. Recurrent blistering is annoying but not life threatening.

The second, or junctional, form of EB does not lead to scarring. However, skin on the areas prone to blistering, such as elbows and knees, often shrinks. In one variation of junctional EB, called gravis junctional EB of Herlitz, the blistering can be so severe that affected infants may not survive due to massive infection and **dehydration**.

The third form of EB, dystrophic EB, varies greatly in terms of severity, but more typically affects the arms and legs. In one variation, called Hallopeau-Siemens EB, repeated blistering and scarring of the hands and feet causes the fingers and toes to fuse, leaving them dysfunctional and with a mitten-like appearance.

## Diagnosis

Physicians and researchers distinguish between the three major subtypes of EB based on which layer of the epidermis separates from the deeper dermis layer of the skin below. Patients suspected of having EB should have a fresh blister biopsied for review. This sample of tissue is examined under an electron microscope or under a conventional microscope using a technique called immunofluorescence, which helps to map the underlying structure.

Knowing that a family member has EB can help establish the diagnosis, but it is possible that parents or siblings will show no sign of the disease, either because it is caused by a new genetic mutation, or because the parents are carriers of the recessive trait and do not display the disease.

## Treatment

The most important treatment for EB is daily wound care. Because the skin is very fragile, care must be taken to be certain that dressing changes do not cause further damage. Tape should not be applied directly to skin and

bandages should be soaked off. Infection is a major concern, so a topical antibiotic, such as bacitracin, mupirocin, or sulfadiazine, should be routinely applied. Among persons with recessive dystrophic EB, the anticonvulsant phenytoin is sometimes effective because it decreases production of an enzyme that breaks down collagen.

## Prognosis

The prognosis of EB varies depending on the subtype of the disease. Individuals with EB simplex can live long, fulfilling lives. The severity of the junctional and dystrophic forms of EB can vary greatly. Infants affected with some forms of the disease often do not survive infancy; other forms can lead to severe scarring and disfigurement.

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- American Academy of Dermatology. PO Box 4014, 930 N. Meacham Rd., Schaumburg, IL 60168-4014. (847) 330-0230. Fax: (847) 330-0050. <<http://www.aad.org>>.
- Dystrophic Epidermolysis Bullosa Research Association of America (DebRA). 40 Rector St., Suite 1403, New York,

NY 10006. (212) 513-4090. Fax: (212) 513-4099. <[staff.debra@exario.net](mailto:staff.debra@exario.net)>. <<http://www.debra.org>>.

Dystrophic Epidermolysis Bullosa Research Association of United Kingdom, (DebRA). 13 Wellington Bus. Park, Dukes Ride, Crowthorne, Berkshire, RG45 6LS. UK 011-01344 771961. <[admin@debra.org.uk](mailto:admin@debra.org.uk)> <<http://www.debra.org.uk>>.

National Epidermolysis Bullosa Registry. University of North Carolina at Chapel Hill, Bolin Heights Bldg. #1, CB# 3369, Chapel Hill, NC 27514-3369. (919) 966-2007. Fax: (919) 966-7080. <[eb\\_registry@med.unc.edu](mailto:eb_registry@med.unc.edu)> <[http://www.med.unc.edu/derm/nebr\\_site](http://www.med.unc.edu/derm/nebr_site)>.

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## Epididymitis

### Definition

Epididymitis is inflammation or infection of the epididymis. In this long coiled tube attached to the upper part of each testicle, sperm mature and are stored before ejaculation.

### Description

Epididymitis is the most common cause of pain in the scrotum. The acute form is usually associated with the most severe pain and swelling. If symptoms last for more than six weeks after treatment begins, the condition is considered chronic.

Epididymitis can occur any time after the onset of puberty but is most common between the ages of 18 and 40. It is especially common among members of the military who exercise for extended periods without emptying their bladders.

Factors that increase the risk of developing epididymitis include:

- infection of the bladder, kidney, prostate, or urinary tract
- other recent illness

- narrowing of the urethra (the tube that drains urine from the bladder)
- use of a urethral catheter.

### Causes and symptoms

Although epididymitis can be caused by the same organisms that cause some **sexually transmitted diseases** (STDs) or occur after prostate surgery, the condition is generally due to pus-generating bacteria associated with infections in other parts of the body.

Epididymitis can also be caused by injury or infection of the scrotum or by irritation from urine that has accumulated in the vas deferens (the duct through which sperm travels after leaving the epididymis).

Epididymitis is characterized by sudden redness and swelling of the scrotum. The affected testicle is hard and sore, and the other testicle may feel tender. The patient has chills and **fever** and usually has acute **urethritis** (inflammation of the urethra).

Enlarged lymph nodes in the groin cause scrotal pain that intensifies throughout the day and may become so severe that walking normally becomes impossible.

### Diagnosis

Laboratory tests used to diagnose epididymitis include:

- urinalysis and urine culture
- examination of discharges from the urethra and prostate gland
- blood tests to measure white-cell counts

### Treatment

Because epididymitis that affects both testicles can make a man sterile, antibiotic therapy must be initiated as soon as symptoms appear. To prevent reinfection, medication must be taken exactly as prescribed, even if the patient's symptoms disappear or he begins to feel better. Over-the-counter anti-inflammatories can relieve pain but should not be used without the approval of a family physician or urologist.

Bed rest is recommended until symptoms subside, and patients are advised to wear athletic supporters when they resume normal activities. If pain is severe, a local anesthetic like lidocaine (Xylocaine) may be injected directly into the spermatic cord.

### Self-care

A patient who has epididymitis should not drink beverages that contain **caffeine**. To prevent **constipation**,

he should use stool softeners or eat plenty of fruit, nuts, whole grain cereals, and other foods with laxative properties.

An ice bag wrapped in a towel can reduce pain and swelling but should be removed from the inflamed area for a few minutes every hour to prevent **burns**.

Strenuous activity should be avoided until symptoms disappear. Sexual activity should not be resumed until a month after symptoms disappear.

If a second course of treatment doesn't eradicate stubborn symptoms, longterm anti-inflammatory therapy may be recommended. In rare instances, chronic symptoms require surgery.

### Surgery

Each of the surgical procedures used to treat epididymitis is performed under local anesthesia on an outpatient basis. Both of them cause sterility.

Epididymectomy involves removing the inflamed section of the epididymis through a small incision in the scrotum.

Bilateral **vasectomy** prevents fluid and sperm from passing through the epididymis. This procedure is usually performed on men who have chronic epididymitis or on elderly patients undergoing prostate surgery.

### Prognosis

Pain generally subsides 24–72 hours after treatment begins. Complete healing may take weeks or months.

### Prevention

Using condoms and not having sex with anyone who has an STD can prevent some cases of epididymitis.

### Resources

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Epidural abscess see **Central nervous system infections**

Epidural anesthetic see **Anesthesia, local**

# Epiglottitis

## Definition

Epiglottitis is an infection of the epiglottis, which can lead to severe airway obstruction.

## Description

When air is inhaled (inspired), it passes through the nose and the nasopharynx or through the mouth and the oropharynx. These are both connected to the larynx, a tube made of cartilage. The air continues down the larynx to the trachea. The trachea then splits into two branches, the left and right bronchi (bronchial tubes). These bronchi branch into smaller air tubes that run within the lungs, leading to the small air sacs of the lungs (alveoli).

Either food, liquid, or air may be taken in through the mouth. While air goes into the larynx and the respiratory system, food and liquid are directed into the tube leading to the stomach, the esophagus. Because food or liquid in the bronchial tubes or lungs could cause a blockage or lead to an infection, the airway is protected. The epiglottis is a leaf-like piece of cartilage extending upwards from the larynx. The epiglottis can close down over the larynx when someone is eating or drinking, preventing these food and liquids from entering the airway.

Epiglottitis is an infection and inflammation of the epiglottis. Because the epiglottis may swell considerably, there is a danger that the airway will be blocked off by the very structure designed to protect it. Air is then unable to reach the lungs. Without intervention, epiglottitis has the potential to be fatal.

Epiglottitis is primarily a disease of two to seven-year-old children, although older children and adults can also contract it. Boys are twice as likely as girls to develop this infection. Because epiglottitis involves swelling and infection of tissues, which are all located at or above the level of the epiglottis, it is sometimes referred to as supraglottitis (*supra*, meaning above). About 25% of all children with this infection also have **pneumonia**.

## Causes and symptoms

The most common cause of epiglottitis is infection with the bacteria called *Haemophilus influenzae type b*. Other types of bacteria are also occasionally responsible for this infection, including some types of *Streptococcus* bacteria and the bacteria responsible for causing **diphtheria**.

A patient with epiglottitis typically experiences a sudden **fever**, and begins having severe throat and neck

**pain**. Because the swollen epiglottis interferes significantly with air movement, every breath creates a loud, harsh, high-pitched sound referred to as **stridor**. Because the vocal cords are located in the larynx just below the area of the epiglottis, the swollen epiglottis makes the patient's voice sound muffled and strained. Swallowing becomes difficult, and the patient may drool. The patient often leans forward and juts out his or her jaw, while struggling for breath.

Epiglottitis strikes suddenly and progresses quickly. A child may begin complaining of a **sore throat**, and within a few hours be suffering from extremely severe airway obstruction.

## Diagnosis

Diagnosis begins with a high level of suspicion that a quickly progressing illness with fever, sore throat, and airway obstruction is very likely to be epiglottitis. If epiglottitis is suspected, no efforts should be made to look at the throat, or to swab the throat in order to obtain a culture for identification of the causative organism. These maneuvers may cause the larynx to go into spasm (laryngospasm), completely closing the airway. These procedures should only be performed in a fully-equipped operating room, so that if laryngospasm occurs, a breathing tube can be immediately placed in order to keep the airway open.

An instrument called a laryngoscope is often used in the operating room to view the epiglottis, which will appear cherry-red and quite swollen. An x-ray picture taken from the side of the neck should also be obtained. The swollen epiglottis has a characteristic appearance, called the "thumb sign."

## Treatment

Treatment almost always involves the immediate establishment of an artificial airway: inserting a breathing tube into the throat (intubation); or making a tiny opening toward the base of the neck and putting a breathing tube into the trachea (tracheostomy). Because the patient's apparent level of distress may not match the actual severity of the situation, and because the disease's progression can be quite surprisingly rapid, it is preferable to go ahead and place the artificial airway, rather than adopting a wait-and-see approach.

Because epiglottitis is caused by a bacteria, **antibiotics** such as cefotaxime, ceftriaxone, or ampicillin with sulbactam should be given through a needle placed in a vein (intravenously). This prevents the bacteria that are circulating throughout the bloodstream from causing infection elsewhere in the body.

## KEY TERMS

**Epiglottis**—A leaf-like piece of cartilage extending upwards from the larynx, which can close like a lid over the trachea to prevent the airway from receiving any food or liquid being swallowed.

**Extubation**—Removal of a breathing tube.

**Intubation**—Putting a breathing tube into the airway.

**Laryngospasm**—Spasm of the larynx.

**Larynx**—The part of the airway lying between the pharynx and the trachea.

**Nasopharynx**—The part of the airway into which the nose leads.

**Oropharynx**—The part of the airway into which the mouth leads.

**Supraglottitis**—Another term for epiglottitis.

**Trachea**—The part of the airway that leads into the bronchial tubes.

**Tracheostomy**—A procedure in which a small opening is made in the neck and into the trachea. A breathing tube is then placed through this opening.

## Prognosis

With treatment (including the establishment of an artificial airway), only about 1% of children with epiglottitis die. Without the artificial airway, this figure jumps to 6%. Most patients recover from the infection, and can have the breathing tube removed (extubation) within a few days.

## Prevention

Prevention involves the use of a vaccine against *H. influenzae type b* (called the Hib vaccine). It is given to babies at two, four, six, and 15 months. Use of this vaccine has made epiglottitis a very rare occurrence.

## Resources

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## ORGANIZATIONS

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Epilepsy see **Seizure disorder**

Epinephrine see **Bronchodilators**

## Episiotomy

### Definition

An episiotomy is a surgical incision made in the area between the vagina and anus (perineum). This is done during the last stages of labor and delivery to expand the opening of the vagina to prevent tearing during the delivery of the baby.

### Purpose

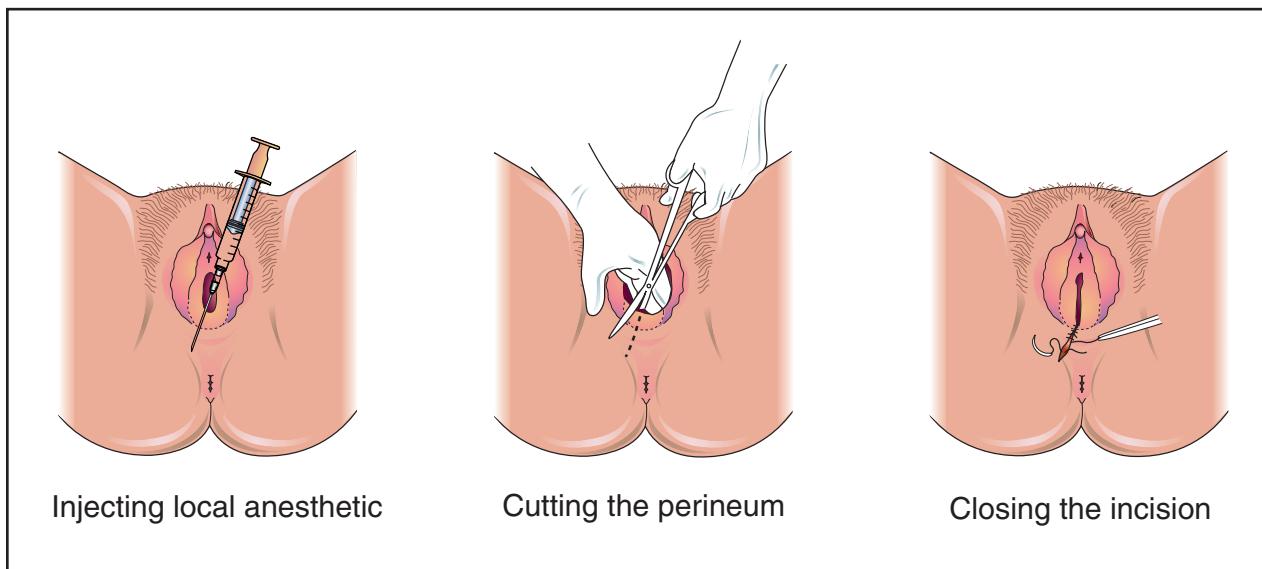
This procedure is usually done during the delivery or birthing process when the vaginal opening does not stretch enough to allow the baby to be delivered without tearing the surrounding tissue.

### Precautions

Prior to the onset of labor, pregnant women may want to discuss the use of episiotomy with their care providers. It is possible that, with adequate preparation and if the stages of labor and delivery are managed with adequate coaching and support, the need for an episiotomy may be reduced.

### Description

An episiotomy is a surgical incision, usually made with sterile scissors, in the perineum as the baby's head is being delivered. This procedure may be used if the tissue around the vaginal opening begins tearing or does not seem to be stretching enough to allow the baby to be delivered.



An episiotomy is a surgical incision made in the perineum, the area of tissue between the vaginal opening and the anus, during the birthing process. This procedure may be used if the tissue around the vaginal opening begins to tear or is not stretching enough to allow the baby to be delivered vaginally. In the United States, the rate of episiotomies being performed is estimated at 65–95%. (Illustration by Electronic Illustrators Group.)

In most cases, the physician makes a midline incision along a straight line from the lowest edge of the vaginal opening toward the anus. In other cases, the episiotomy is performed by making a diagonal incision across the midline between the vagina and anus. This method is used much less often, may be more painful, and may require more healing time than the midline incision. After the baby is delivered through the extended vaginal opening, the incision is closed with stitches. A local anesthetic agent may be applied or injected to numb the area before it is sewn up (sutured).

Several reasons are cited for performing episiotomies. Some experts believe that an episiotomy speeds up the birthing process, making it easier for the baby to be delivered. This can be important if there is any sign of distress that may harm the mother or baby. Because tissues in this area may tear during the delivery, another reason for performing an episiotomy is that a clean incision is easier to repair than a jagged tear and may heal faster. Although the use of episiotomy is sometimes described as protecting the pelvic muscles and possibly preventing future problems with **urinary incontinence**, it is not clear that the procedure actually helps.

The use of episiotomy during the birthing process is fairly widespread in the United States. Estimates of episiotomy use in hospitals range from 65–95% of deliveries, depending on how many times the mother has given birth previously. This routine use of episiotomy is being reexamined in many hospitals and health care settings.

However, an episiotomy is always necessary during a forceps delivery because of the size of the forceps.

### Preparation

It may be possible to avoid the need for an episiotomy. Pregnant women may want to talk with their care providers about the use of episiotomy during the delivery. Kegel exercises are often recommended during the **pregnancy** to help strengthen the pelvic floor muscles. Prenatal perineal massage may help to stretch and relax the tissue around the vaginal opening. During the delivery process, warm compresses can be applied to the area along with the use of perineal massage. Coaching and support are also important during the delivery process. A slowed, controlled pushing during the second stage of labor (when the mother gets the urge to push) may allow the tissues to stretch rather than tear. Also, an upright birthing position (rather than one where the mother is lying down) may decrease the need for an episiotomy.

### Aftercare

The area of the episiotomy may be uncomfortable or even painful for several days. Several practices can relieve some of the **pain**. Cold packs can be applied to the perineal area to reduce swelling and discomfort. Use of the **Sitz bath** available at the hospital or birth center can ease the discomfort, too. This unit circulates warm water over the area. A squirt bottle with water can be used to clean the area after urination or defecation rather

## KEY TERMS

**Kegel exercises**—A series of contractions and relaxations of the muscles in the perineal area. These exercises are thought to strengthen the pelvic floor and may help prevent urinary incontinence in women.

**Perineum**—The area between the opening of the vagina and the anus in a woman, or the area between the scrotum and the anus in a man.

**Sitz bath**—A shallow tub or bowl, sometimes mounted above a toilet, that allows the perineum and buttocks to be immersed in circulating water.

**Urinary incontinence**—The inability to prevent the leakage or discharge of urine. This situation becomes more common as people age, and is more common in women who have given birth to more than one child.

than wiping with tissue. Also, the area should be patted dry rather than wiped. Cleansing pads soaked in witch hazel (such as Tucks) are very effective for cleaning the area and also feel soothing.

### Risks

Several side effects of episiotomy have been reported, including infection, increased pain, prolonged healing time, and increased discomfort once sexual intercourse is resumed. There is also the risk that the episiotomy incision will be deeper or longer than is necessary to permit the birth of the infant. There is a risk of increased bleeding.

### Normal results

In a normal and well managed delivery, an episiotomy may be avoided altogether. If an episiotomy is deemed to be necessary, a simple midline incision will be made to extend the vaginal opening without additional tearing or extensive trauma to the perineal area. Although there may be some pain associated with the healing of the episiotomy incision, relief can usually be provided with mild pain relievers and supportive measures, such as the application of cold packs.

### Abnormal results

An episiotomy incision that is too long or deep may extend into the rectum, causing more bleeding and an

increased risk of infection. Additional tearing or tissue damage may occur beyond the episiotomy incision, leaving a cut and a tear to be repaired.

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Altha Roberts Edgren

Epispadias see **Hypospadias and epispadias**

Epistaxis see **Nosebleed**

EPS see **Electrophysiology study of the heart**

## Epstein-Barr virus test

### Definition

The Epstein-Barr virus test is a blood test, or group of tests, to determine the presence or absence of antibodies in the blood stream directed against proteins of the Epstein-Barr virus, the cause of **infectious mononucleosis**.

### Purpose

The test is primarily used to detect whether first time infection (called primary infection) with the Epstein-Barr virus is currently occurring, or has occurred within a short period of time. The pattern of the antibodies detected can, however, tell if the person has never been infected with the Epstein-Barr virus, or if the infection occurred in the more distant past. These tests are mostly utilized in the diagnosis of Epstein-Barr virus-associated infectious mononucleosis when the more common diagnostic test, the heterophile antibody, is negative, or in situations where the infection is manifesting unusual symptoms. Therefore, the tests are often not needed in a situa-

tion where a doctor believes that a person has mononucleosis and the heterophile test (also called the monospot test) is positive.

In addition, Epstein-Barr virus testing is usually not needed in the evaluation of a patient who has long-lasting **fatigue**, and may have the **chronic fatigue syndrome**. Initially, it was thought that discovering a particular pattern of antibodies to this virus was helpful in the diagnosis of chronic fatigue syndrome, but this no longer appears to be the case.

### Precautions

As in any blood test, standard precautions should be performed to prevent infection at the site where the blood is obtained, and to prevent excess bleeding. Normally, the site is cleaned with an antiseptic liquid prior to the blood being obtained; a sterile non-reusable needle and syringe are used; and, once the needle is removed, pressure is placed at the site until bleeding has stopped.

### Description

These tests are more often performed in a consulting laboratory than at a physician's office or in a hospital laboratory. Like most antibody tests, they are performed on serum, the liquid part of the blood obtained after the whole blood is allowed to clot in a tube. Antibodies can be detected against several components of the Epstein-Barr virus (EBV). These components are the EBV early antigen (EA), the viral capsid antigen (VCA), and the nuclear antigen (EBNA). These several antigens are different proteins that are produced in the process (stages) of the virus' growth.

At the time of infection with Epstein-Barr virus, antibodies to EA are found and usually last for four to six months only. This antibody, however, persists substantially longer in about 10% of persons who have had EBV infection in the more remote past. The absence of antibody to EA when other EBV antibodies are present strongly suggests that first time infection with EBV occurred in the past.

Antibody to VCA is found both early and late in EBV infection. At the time of infection, antibody of both the IgM and IgG types are detectable. After four to six months, usually, only the IgG antibody against VCA can be found.

Unlike antibodies to EA and VCA, antibody to EBNA does not usually develop until recovery from first time infection of this virus. Therefore, finding detectable amounts of antibody to EBNA during an illness which might be caused by EBV makes the causal relationship very unlikely.

### Preparation

The skin area from which the blood sample will be obtained is wiped with an antiseptic such as alcohol or iodine.

### Aftercare

The aftercare is similar to that for any blood test. Usually, pressure is applied to the area for several moments until bleeding stops. If the results are difficult to interpret, it may be necessary to re-test later, after waiting one to three weeks. The change in the amounts of antibody detected between the two tests can be particularly useful, at times, in helping to make a diagnosis.

### Risks

There are no risks over and above those of having blood drawn for any other purpose. These tests are more expensive than many other blood tests but are usually covered by medical insurance.

### Normal results

The pattern of the three antibodies can be used to determine whether the person has not had infection with EBV to this point (is susceptible to infection); is currently, or recently, infected with EBV for the first time; or has had first time infection with EBV sometime in the past (more than six months ago).

If one defines "normal" results as either not having EBV in the past, and call that category one; or having had it in the past, and call that category two. Most young children below the age of five will fall into category one, while most adults over the age of 20 years will fall into category two.

The results for susceptibility are:

- antibody to EA = negative
- antibody to VCA (either IgM or IgG) = negative
- antibody to EBNA = Negative

The results for past infection are:

- antibody to EA = negative (90% of time)
- antibody to VCA IgM = negative
- antibody to VCA IgG = positive
- antibody to EBNA = positive

It is important to realize that the Epstein-Barr virus, like all the human herpes viruses, does not totally leave the body after the patient recovers from illness. With EBV, the virus will intermittently recur in the saliva of people without any symptoms. Such people will have a

test pattern of previous infection. It is this group of people who can transmit EBV to others without themselves being ill.

### Abnormal results

The results for current or recent infection are:

- antibody to EA = positive
- antibody to VCA IgM = positive
- antibody to VCA IgG = positive
- antibody to EBNA = negative

Without the pattern of the three antibodies, it can be difficult to be accurate in interpretation. The presence of antibody to VCA IgM is the best single test for current or recent first time infection.

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**ERCP** see **Endoscopic retrograde cholangiopancreatography**

Erectile disorder see **Impotence**

Ergotamine see **Antimigraine drugs**

Erosive gastritis see **Gastritis**

## Erysipelas

### Definition

Erysipelas is a skin infection that often follows **strep throat**.

### Description

Erysipelas, also called St. Anthony's fire, is caused by infection by Group A *Streptococci*. This same type of

### KEY TERMS

**Bacteremia**—The presence of bacteria in the blood.

**Streptococcus**—A bacteria that causes erysipelas and strep throat, as well as other infections.

bacteria is responsible for such infections as strep throat, and infections of both surgical and other kinds of **wounds** in the skin. The infection occurs most often in young infants and the elderly.

### Causes and symptoms

Erysipelas usually occurs rather abruptly. When the preceding infection was strep throat, the rash begins on the face. Occasionally, when the preceding infection was of a wound from an injury or operation, the rash will appear on an arm or leg.

Classically, the usual presentation is a bright-red, butterfly-shaped rash appearing across the bridge of the nose and the cheeks. It is hot to the touch, painful, shiny, and swollen, with clearly defined margins. The edges of the rash are a raised ridge, hard to the touch. There may be fluid-filled bumps scattered along the area. The rash spreads rapidly. Some patients have swelling of the eyelids, sometimes so severe that their eyes swell shut. The patient may have **fever**, chills, loss of energy, **nausea and vomiting**, and swollen, tender lymph nodes. In severe cases, walled-off areas of pus (abscesses) may develop beneath the skin. If left untreated, the streptococcal bacteria may begin circulating in the bloodstream (a condition called **bacteremia**). A patient may then develop an overwhelming, systemic infection called **sepsis**, with a high risk of **death**.

### Diagnosis

The rash of erysipelas is very characteristic, raising the practitioner's suspicion towards that diagnosis, especially when coupled with a history of recent strep infection. Attempts to culture (grow) the bacteria from a sample of the rash usually fail. When the bacteria are present in the blood, they may be grown in a laboratory, and identified under a microscope. Other laboratory tests involve reacting fluorescently-tagged antibodies with a sample of the patient's infected tissue. This type of test may be successful in positively identifying the streptococcal bacteria.

## Treatment

Penicillin is the drug of choice for treating erysipelas. It can usually be given by mouth, although in severe cases (or in cases of diagnosed bacteremia) it may be given through a needle placed in a vein (intravenously).

Even with antibiotic treatment, swelling may continue to spread. Other symptoms, such as fever, **pain**, and redness, usually decrease rapidly after penicillin is started. Cold packs and pain relievers may help decrease discomfort. Within about five to 10 days, the affected skin may begin drying up and flaking off.

## Prognosis

With prompt treatment, the prognosis from erysipelas is excellent. Delay of treatment, however, increases the chance for bacteremia and the potential for death from overwhelming sepsis. This is particularly true of people with weakened immune systems (babies, the elderly, and people ill with other diseases, especially Acquired Immunodeficiency Syndrome, or **AIDS**). Frequently, an individual who has had erysipelas will have it occur again in the same location.

## Prevention

Prevention involves appropriate and complete treatment of **streptococcal infections**, including strep throat and wound infections.

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Erythema infectiosum see **Fifth disease**

## Erythema multiforme

### Definition

Erythema multiforme is a skin disease that causes lesions and redness around the lesions.

### Description

Erythema multiforme appears on the skin and the mucous membranes (the lining of the mouth, digestive tract, vagina, and other organs). Large, symmetrical red blotches appear all over the skin in a circular pattern. On mucous membranes, it begins as blisters and progresses to ulcers. A more advanced form, called Stevens-Johnson syndrome, can be severe and even fatal.

### Causes and symptoms

Erythema multiforme has many causes, most commonly are drugs. Penicillin, **sulfonamides**, certain epilepsy drugs, **aspirin**, and **acetaminophen** are the most likely medication-induced causes. Erythema multiforme can also be caused by certain diseases. Herpes virus and mycoplasma **pneumonia** are likely infectious causes.

### Diagnosis

The appearance of the rash is sufficiently unique to identify it on sight. Having identified it, the physician will determine the underlying cause.

### Treatment

Erythema multiforme is inadvertently treated when the causative agent, whether it be a drug or a disease, is treated. In severe cases, cortisone-like medication is often used along with general supportive measures and prevention of infection.

### Prognosis

As a rule, the rash abates by itself without damaging the skin. Only in the case of infection, severe blistering, or continued use of an offending drug does complications occur.

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## KEY TERMS

**Herpes virus**—Viruses that can infect the skin, mucous membranes, and brain, and they are responsible for such diseases as herpes simplex, chicken pox, and shingles.

**Mycoplasma pneumonia**—An incomplete bacterium that infects the lung.

## KEY TERMS

**Biopsy**—Surgical removal of tissue for diagnostic purposes.

**Panniculitis**—Inflammation of fatty tissue.

Once the skin problem has been diagnosed, its underlying cause must then be identified. A lengthy evaluation may ensue, and often times the cause remains unknown.

## Treatment

Painful nodules can be treated with mild **pain killers** and local application of ice packs. Medical attention will be directed toward the underlying disease.

The nodules will eventually disappear, leaving no trace behind.

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Erythremia see **Polycythemia vera**

## Erythroblastosis fetalis

### Definition

Erythroblastosis fetalis refers to two potentially disabling or fatal blood disorders in infants: Rh incompatibility disease and ABO incompatibility disease. Either disease may be apparent before birth and can cause fetal **death** in some cases. The disorder is caused by incompatibility between a mother's blood and her unborn baby's blood. Because of the incompatibility, the mother's immune system may launch an immune response against the baby's red blood cells. As a result, the baby's blood

## Erythema nodosum

### Definition

Erythema nodosum is a skin disorder characterized by painful red nodules appearing mostly on the shins.

### Description

Erythema nodosum is an eruption of tender red lumps on both shins and occasionally the arms and face. Bruising often accompanies the nodule formation. Erythema nodosum is most prevalent in young adults.

### Causes and symptoms

Erythema nodosum can be caused by many important and treatable diseases. Among them are **tuberculosis**, several fungal lung infections, **leprosy**, inflammatory bowel disease, and some potentially dangerous bacterial infections. Drugs can also induce erythema nodosum. The most common are penicillin, **sulfonamides**, and birth control pills.

### Diagnosis

There are a few other skin eruptions that mimic erythema nodosum, so the physician may have to perform a biopsy to sort them out. There are a few types of *panniculitis*, fat inflammation, that may signal a **cancer** somewhere in the body, and there are other kinds of inflammation that may confuse the diagnosis.

cells are destroyed, and the baby may suffer severe anemia (deficiency in red blood cells), brain damage, or death.

## Description

Red blood cells carry several types of proteins, called antigens, on their surfaces. The A, B, and O antigens are used to classify a person's blood as type A, B, AB, or O. Each parent passes one A, B, or O antigen gene to their child. How the genes are paired determines the person's blood type.

A person who inherits an A antigen gene from each parent has type A blood; receiving two B antigen genes corresponds with type B blood; and inheriting A and B antigen genes means a person has type AB blood. If the O antigen gene is inherited from both parents, the child has type O blood; however, the pairing of A and O antigen genes corresponds with type A blood; and if the B antigen gene is matched with the O antigen gene, the person has type B blood.

Another red blood cell antigen, called the Rh factor, also plays a role in describing a person's blood type. A person with at least one copy of the gene for the Rh factor has Rh-positive blood; if no copies are inherited, the person's blood type is Rh-negative. In blood typing, the presence of A, B, and O antigens, plus the presence or absence of the Rh-factor, determine a person's specific blood type, such as A-positive, B-negative, and so on.

A person's blood type has no effect on health. However, an individual's immune system considers only that person's specific blood type, or a close match, acceptable. If a radically different blood type is introduced into the bloodstream, the immune system produces antibodies, proteins that specifically attack and destroy any cell carrying the foreign antigen.

Determining a person's blood type is very important if she becomes pregnant. Blood cells from the unborn baby (fetal red blood cells) can cross over into the mother's bloodstream, especially at delivery. If the mother and her baby have compatible blood types, the crossover does not present any danger. However, if the blood types are incompatible, the mother's immune system manufactures antibodies against the baby's blood.

Usually, this incompatibility is not a factor in a first pregnancy, because few fetal blood cells reach the mother's bloodstream until delivery. The antibodies that form after delivery cannot affect the first child. In later pregnancies, fetuses and babies may be in grave danger. The danger arises from the possibility that the mother's antibodies will attack the fetal red blood cells. If this happens, the fetus or baby can suffer severe health effects and may die.

There are two types of incompatibility diseases: Rh incompatibility disease and ABO incompatibility disease. Both diseases have similar symptoms, but Rh disease is much more severe, because anti-Rh antibodies cross over the placenta more readily than anti-A or anti-B antibodies. (The immune system does not form antibodies against the O antigen.) Therefore, a greater percentage of the baby's blood cells are destroyed by Rh disease.

Both incompatibility diseases are uncommon in the United States due to medical advances over the last 50 years. For example, prior to 1946 (when newborn blood transfusions were introduced) 20,000 babies were affected by Rh disease yearly. Further advances, such as suppressing the mother's antibody response, have reduced the incidence of Rh disease to approximately 4,000 cases per year.

Rh disease only occurs if a mother is Rh-negative and her baby is Rh-positive. For this situation to occur, the baby must inherit the Rh factor gene from the father. Most people are Rh-positive. Only 15% of the caucasian population is Rh-negative, compared to 5–7% of the african american population and virtually none of Asian populations.

ABO incompatibility disease is almost always limited to babies with A or B antigens whose mothers have type O blood. Approximately one third of these babies show evidence of the mother's antibodies in their bloodstream, but only a small percentage develop symptoms of ABO incompatibility disease.

## Cause and symptoms

Rh disease and ABO incompatibility disease are caused when a mother's immune system produces antibodies against the red blood cells of her unborn child. The antibodies cause the baby's red blood cells to be destroyed and the baby develops anemia. The baby's body tries to compensate for the anemia by releasing immature red blood cells, called erythroblasts, from the bone marrow.

The overproduction of erythroblasts can cause the liver and spleen to become enlarged, potentially causing liver damage or a ruptured spleen. The emphasis on erythroblast production is at the cost of producing other types of blood cells, such as platelets and other factors important for blood clotting. Since the blood lacks clotting factors, excessive bleeding can be a complication.

The destroyed red blood cells release the blood's red pigment (hemoglobin) which degrades into a yellow substance called bilirubin. Bilirubin is normally produced as red blood cells die, but the body is only equipped to handle a certain low level of bilirubin in the bloodstream at one time. Erythroblastosis fetalis overwhelms the removal system, and high levels of bilirubin

## KEY TERMS

**Amniocentesis**—A procedure in which a needle is inserted through a pregnant woman's abdomen and into her uterus to withdraw a small sample of amniotic fluid. The amniotic fluid can be examined for sign of disease or other problems afflicting the fetus.

**Amniotic fluid**—The fluid that surrounds a fetus in the uterus.

**Anemia**—A condition in which there is an abnormally low number of red blood cells in the bloodstream. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

**Antibody**—A protein molecule produced by the immune system in response to a protein that is not recognized as belonging in the body.

**Antigen**—A protein that can elicit an immune response in the form of antibody formation. With regard to red blood cells, the major antigens are A, B, O, and the Rh factor.

**Bilirubin**—A yellow-colored end-product of hemoglobin degradation. It is normally present at very low levels in the bloodstream; at high levels, it produces jaundice.

**Cordocentesis**—A procedure for delivering a blood transfusion to a fetus. It involves a fine needle being threaded through a pregnant woman's abdomen and into the umbilical cord with the aid of ultrasound imaging.

**Hemoglobin**—A molecule in red blood cells that transports oxygen and gives the cells their characteristic color.

**Hydrops fetalis**—A condition in which a fetus or newborn baby accumulates fluids, causing swollen arms and legs and impaired breathing.

**Hyperbilirubinemia**—A condition in which bilirubin accumulates to abnormally high levels in the bloodstream

**Placenta**—A protective membrane that surrounds and protects the fetus during pregnancy.

**Platelet**—A blood factor that is important in forming blood clots.

**Rh factor**—An antigen that is found on the red blood cells of most people. If it is present, the blood type is referred to as Rh-positive; if absent, the blood type is Rh-negative.

accumulate, causing hyperbilirubinemia, a condition in which the baby becomes jaundiced. The **jaundice** is apparent from the yellowish tone of the baby's eyes and skin. If hyperbilirubinemia cannot be controlled, the baby develops kernicterus. The term kernicterus means that bilirubin is being deposited in the brain, possibly causing permanent damage.

Other symptoms that may be present include high levels of insulin and low blood sugar, as well as a condition called hydrops fetalis. Hydrops fetalis is characterized by an accumulation of fluids within the baby's body, giving it a swollen appearance. This fluid accumulation inhibits normal breathing, because the lungs cannot expand fully and may contain fluid. If this condition continues for an extended period, it can interfere with lung growth. Hydrops fetalis and anemia can also contribute to heart problems.

### Diagnosis

Erythroblastosis fetalis can be predicted before birth by determining the mother's blood type. If she is Rh-negative, the father's blood is tested to determine whether he

is Rh-positive. If the father is Rh-positive, the mother's blood will be checked for antibodies against the Rh factor. A test that demonstrates no antibodies is repeated at week 26 or 27 of the pregnancy. If antibodies are present, treatment is begun.

In cases in which incompatibility is not identified before birth, the baby suffers recognizable characteristic symptoms such as anemia, hyperbilirubinemia, and hydrops fetalis. The blood incompatibility is uncovered through blood tests such as the Coombs test, which measures the level of maternal antibodies attached to the baby's red blood cells. Other blood tests reveal anemia, abnormal blood counts, and high levels of bilirubin.

### Treatment

When a mother has antibodies against her unborn infant's blood, the pregnancy is watched very carefully. The antibodies are monitored and if levels increase, **amniocentesis**, fetal umbilical cord blood sampling, and ultrasound are used to assess any effects on the baby. Trouble is indicated by high levels of bilirubin in the amniotic fluid or baby's blood, or if the ultrasound

reveals hydrops fetalis. If the baby is in danger, and the pregnancy is at least 32–34 weeks along, labor is induced. Under 32 weeks, the baby is given blood transfusions while still in the mother's uterus.

There are two techniques that are used to deliver a blood **transfusion** to a baby before birth. In the first, a needle is inserted through the mother's abdomen and uterus, and into the baby's abdomen. Red blood cells injected into the baby's abdominal cavity are absorbed into its bloodstream. In early pregnancy or if the baby's bilirubin levels are gravely high, cordocentesis is performed. This procedure involves sliding a very fine needle through the mother's abdomen and, guided by ultrasound, into a vein in the umbilical cord to inject red blood cells directly into the baby's bloodstream.

After birth, the severity of the baby's symptoms are assessed. One or more transfusions may be necessary to treat anemia, hyperbilirubinemia, and bleeding. Hyperbilirubinemia is also treated with **phototherapy**, a treatment in which the baby is placed under a special light. This light causes changes in how the bilirubin molecule is shaped, which makes it easier to excrete. The baby may also receive oxygen and intravenous fluids containing electrolytes or drugs to treat other symptoms.

## Prognosis

In many cases of blood type incompatibility, the symptoms of erythroblastosis fetalis are prevented with careful monitoring and blood type screening. Treatment of minor symptoms is typically successful and the baby will not suffer long-term problems.

Nevertheless, erythroblastosis is a very serious condition for approximately 4,000 babies annually. In about 15% of cases, the baby is severely affected and dies before birth. Babies who survive pregnancy may develop kernicterus, which can lead to deafness, speech problems, **cerebral palsy**, or **mental retardation**. Extended hydrops fetalis can inhibit lung growth and contribute to **heart failure**. These serious complications are life threatening, but with good medical treatment, the fatality rate is very low. According to the U.S. Centers for Disease Control and Prevention, there were 21 infant deaths in the United States during 1996 that were attributable to hemolytic disease (erythroblastosis fetalis) and jaundice.

## Prevention

With any pregnancy, whether it results in a live birth, **miscarriage**, **stillbirth**, or abortion, blood typing is a universal precaution against blood compatibility disease. Blood types cannot be changed, but adequate forewarning allows precautions and treatments that limit the danger to unborn babies.

If an Rh-negative woman gives birth to an Rh-positive baby, she is given an injection of immunoglobulin G, a type of antibody protein, within 72 hours of the birth. The immunoglobulin destroys any fetal blood cells in her bloodstream before her immune system can react to them. In cases where this precaution is not taken, antibodies are created and future pregnancies may be complicated.

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Julia Barrett

# Erythrocyte sedimentation rate

## Definition

The erythrocyte sedimentation rate (ESR), or sedimentation rate (sed rate), is a measure of the settling of red blood cells in a tube of blood during one hour. The rate is an indication of inflammation and increases in many diseases.

## Purpose

ESR is increased in rheumatoid diseases, most infections, and in **cancer**. An advanced rate doesn't diagnose a specific disease, but it does indicate that an underlying disease may be present.

A physician can use ESR to monitor a person with an associated disease. When the disease worsens, the ESR increases; when the disease improves, the ESR decreases. The ESR doesn't always follow the course of cancer.

ESR is called an acute-phase reactant test, meaning that it reacts to acute conditions in the body, such as infection or trauma. The rate increase follows a rise in temperature and white blood cells count, peaks after several days, and usually lasts longer than the elevated temperature or white blood cells count.

## Precautions

The ESR should not be used to screen healthy persons for disease.

## Description

The ESR test is a simple test dating back to the ancient Greeks. A specific amount of diluted, unclotted blood is placed in a special narrow tube and left undisturbed for exactly one hour. The red cells settle towards the bottom of the tube, and the pale yellow liquid (plasma) rises to the top. After 60 minutes, measurements are taken of the distance the red cells traveled to settle at the bottom of the tube. Two methods, the Westergren and the Wintrobe, are used by laboratories; each method produces slightly different results. Most laboratories use the Westergren method.

Normally red cells don't settle far toward the bottom of the tube. Many diseases make extra or abnormal proteins that cause the red cells to move close together, stack up, and form a column (rouleaux). In a group, red cells are heavier and fall faster. The faster they fall, the further they settle, and the higher the ESR.

The ESR test is covered by insurance when medically necessary. Results are usually available the same or following day.

## Preparation

This test requires 7mL–10 mL of blood. A health-care worker ties a tourniquet on the patient's upper arm, locates a vein in the inner elbow region, and inserts a needle into that vein. Vacuum action draws the blood through the needle into an attached tube. Collection of the sample takes only a few minutes.

## Aftercare

Discomfort or bruising may occur at the puncture site. Pressure applied to the puncture site until the bleeding stops reduces bruising. Warm packs to the puncture site relieve discomfort. The patient may feel dizzy or faint.

## Normal results

A normal value does not rule out disease. Normal values for the Westergren method are: Men 0 mm/hour–15 mm/hour; women 0 mm/hour–20 mm/hour; and children 0 mm/hour–10 mm/hour.

## Abnormal results

The highest ESR levels are usually seen in a cancer of a certain type of white blood cell (**multiple myeloma**)

## KEY TERMS

**Acute phase reactant**—A substance in the blood that increases as a response to an acute conditions such as infection, injury, tissue destruction, some cancers, burns, surgery, or trauma.

**Erythrocyte sedimentation rate (ESR)**—The distance that red blood cells settle in a tube of blood in one hour. It is an indication of inflammation.

**Rouleaux**—The stacking up of red blood cells, caused by extra or abnormal proteins in the blood that decrease the normal distance red cells maintain between each other.

and rheumatoid disease, such as **rheumatoid arthritis**. Many other diseases also increase the ESR: infection, kidney disease, anemia, diseases involving white blood cells, cancer, and autoimmune and inflammatory diseases.

Any disease that changes the shape and size of red blood cells decreases the ESR. Distorted cells, such as with **sickle cell disease**, do not stack, and consequently do not settle far, even in the presence of an ESR-associated disease. Diseases that cause the body to make less protein or extra red blood cells also decrease the ESR.

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Nancy J. Nordenson

## Erythromycins

### Definition

Erythromycins are medicines that kill bacteria or prevent their growth.

### Purpose

Erythromycins are **antibiotics**, medicines used to treat infections caused by microorganisms. Physicians

prescribe these drugs for many types of infections caused by bacteria, including **strep throat**, sinus infections, **pneumonia**, ear infections, **tonsillitis**, **bronchitis**, **gonorrhea**, **pelvic inflammatory disease** (PID), and urinary tract infections. Some medicines in this group are also used to treat **Legionnaires' disease** and ulcers caused by bacteria. These drugs will *not* work for colds, flu, and other infections caused by viruses.

## Description

The drugs described here include erythromycins (Erythrocin, Ery-C, E-Mycin, and other brands) and medicines that are chemically related to erythromycins, such as azithromycin (Zithromax) and clarithromycin (Biaxin). They are available only with a physician's prescription and are sold in capsule, tablet (regular and chewable), liquid, and injectable forms.

## Recommended dosage

The recommended dosage depends on the type of erythromycin, the strength of the medicine, and the medical problem for which it is being taken. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage.

Always take erythromycins exactly as directed. Never take larger, smaller, more frequent, or less frequent doses. To make sure the infection clears up completely, it is very important to take the medicine for as long as it has been prescribed. Do not stop taking the drug just because symptoms begin to improve. This is important with all types of infections, but it is especially important in "strep" infections, which can lead to serious heart problems if they are not cleared up completely.

Erythromycins work best when they are at constant levels in the blood. To help keep levels constant, take the medicine in doses spaced evenly through the day and night. Do not miss any doses. Some of these medicines are most effective when taken with a full glass of water on an empty stomach, but they may be taken with food if stomach upset is a problem. Others work equally well when taken with or without food. Check package directions or ask the physician or pharmacist for instructions on how to take the medicine.

## Precautions

Symptoms should begin to improve within a few days of beginning to take this medicine. If they do not, or if they get worse, check with the physician who prescribed the medicine.

Erythromycins may cause mild **diarrhea**, that usually goes away during treatment. However, severe diarrhea

could be a sign of a very serious side effect. Anyone who develops severe diarrhea while taking erythromycin or related drugs should stop taking the medicine and call a physician immediately.

## Special conditions

Taking erythromycins may cause problems for people with certain medical conditions or people who are taking certain other medicines. Before taking these drugs, be sure to let the physician know about any of these conditions:

**ALLERGIES.** Anyone who has had unusual reactions to erythromycins, azithromycin, or clarithromycin in the past 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.

**PREGNANCY.** Some medicines in this group may cause problems in pregnant women and have the potential to cause **birth defects**. Women who are pregnant or who may become pregnant should check with their physicians before taking these drugs.

**BREASTFEEDING.** Erythromycins pass into breast milk. Mothers who are breastfeeding and who need to take this medicine should check with their physicians.

**OTHER MEDICAL CONDITIONS.** Before using erythromycins, people with any of these medical problems should make sure their physicians are aware of their conditions:

- heart disease
- liver disease
- hearing loss

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

## Side effects

The most common side effects are mild diarrhea, nausea, vomiting, and stomach or abdominal cramps. These problems usually go away as the body adjusts to the drug and do not require medical treatment. Less common side effects, such as sore mouth or tongue and vaginal **itching** and discharge also may occur and do not need medical attention unless they persist or are bothersome.

More serious side effects are not common, but may occur. If any of the following side effects occur, check with a physician immediately:

- severe stomach **pain**, nausea, vomiting, or diarrhea

## KEY TERMS

**Bronchitis**—Inflammation of the air passages of the lungs.

**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.

**Legionnaire's disease**—A lung disease caused by a bacterium.

**Microorganism**—An organism that is too small to be seen with the naked eye.

**Pelvic inflammatory disease (PID)**—Inflammation of the female reproductive tract, caused by any of several microorganisms. Symptoms include severe abdominal pain, high fever, and vaginal discharge. Severe cases can result in sterility.

**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.

**Sinus**—Any of several air-filled cavities in the bones of the skull.

**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.

**Urinary tract**—The passage through which urine flows from the kidneys out of the body.

- fever
- skin rash, redness, or itching
- unusual tiredness or weakness

Although rare, very serious reactions to azithromycin (Zithromax) are possible, including extreme swelling of the lips, face, and neck, and **anaphylaxis** (a violent allergic reaction). Anyone who develops these symptoms after taking azithromycin should stop taking the medicine and get immediate medical help.

Other rare side effects may occur with erythromycins and related drugs. Anyone who has unusual symptoms after taking these medicines should get in touch with his or her physician.

### Interactions

Erythromycins may interact with many 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 erythromycins should let the physician know all other medicines he or she is taking. Among the drugs that may interact with erythromycins are:

- acetaminophen (Tylenol)
- medicine for overactive thyroid
- male hormones (androgens)
- female hormones (estrogens)
- other antibiotics

- blood thinners
- disulfiram (Antabuse), used to treat alcohol abuse
- antiseizure medicines such as valproic acid (Depakote, Depakene)
- caffeine
- the **antihistamines** astemizole (Hismanal)
- antiviral drugs such as (zidovudine) Retrovir

The list above does not include every drug that may interact with erythromycins. Be sure to check with a physician or pharmacist before combining erythromycins with any other prescription or nonprescription (over-the-counter) medicine.

Nancy Ross-Flanigan

Erythropoietin see **Cancer therapy, supportive; Immunologic therapies**

## Erythropoietin test

### Definition

Erythropoietin, also called EPO, is a type of protein called a glycoprotein that is formed mainly in the kidneys to stimulate the production of red blood cells.

## KEY TERMS

**Anemia**—A condition in which the hemoglobin concentration in the blood is below normal.

**Polycythemia vera**—A condition characterized by an unusually large number of red blood cells in the blood due to increased production by the bone marrow. Symptoms include headaches, blurred vision, high blood pressure, dizziness, and night sweats.

**Secondary polycythemia**—Secondary polycythemia occurs when the excess of red blood cells is caused by a condition other than polycythemia vera. For example, when low levels of oxygen in the blood stimulate the bone marrow to produce more red blood cells, as in chronic lung disease.

### Purpose

The erythropoietin (EPO) test is used to determine if hormonal secretion is causing changes in the red blood cells. The test has great value in evaluating low hemoglobin (anemia), and another disorder called polycythemia, in which unusually large numbers of red blood cells are found in the blood. The EPO test is also used to identify kidney tumors and to evaluate abuse by athletes who believe commercially prepared erythropoietin enhances performance.

### Precautions

Not every laboratory is equipped to evaluate EPO, so the reference laboratory (a large commercial lab that does tests for hospitals not equipped to do them) performing the test may require as many as four days to complete the analysis. It should also be noted that EPO values increase in **pregnancy**, in which significantly higher levels are found before the twenty-fourth week.

### Description

Erythropoietin is produced primarily in the kidneys but interacts with other factors in the bone marrow to increase red cell production. EPO is unique among the blood cell growth factors, because it is the only one that behaves like a hormone.

Erythropoietin acts as the principal regulator in the production of red blood cells (erythrocytes) by controlling the number, the kinds, and the survival of the cells. Because of this ability, it is being investigated for use in

cancer patients to prevent anemia (hemoglobin concentration in the blood is lower than normal), or to treat anemia that has been induced by **chemotherapy** and **bone marrow transplantation** (BMT).

The correction of anemia can result in reduced **transfusion** requirements, so the erythropoietin test is used to diagnose anemia, including the anemia of end-stage renal disease. Erythropoietin determination is also valuable in diagnosing a condition known as polycythemia, when increased numbers of red blood cells occur. Levels of erythropoietin are extremely low in **polycythemia vera** but are normal or high in **secondary polycythemia**. It happens rarely, but cysts in the liver or kidneys, as well as tumors in the kidneys or brain, can also produce erythropoietin. Patients with these conditions can have high levels of erythropoietin and may develop secondary polycythemia.

Some athletes use EPO to enhance performance, as the increased red cell volume adds more oxygen-carrying capacity to the blood. Adverse reactions to this practice can include clotting abnormalities, **headache**, seizures, high blood pressure, nausea, vomiting, **diarrhea**, and rash.

### Preparation

The EPO test requires a blood sample. The patient is to fast with nothing to eat or drink for at least eight hours before the test. It is also suggested that the patient lie down for 30 minutes before the test.

### Risks

Risks for this test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or feeling lightheaded after venipuncture, and hematoma (blood accumulating under the puncture site).

### Normal results

Reference values vary from laboratory to laboratory, but a general normal range is 11–48 mU/ml (milliunits per milliliter).

### Abnormal results

Low levels of EPO are found in anemic patients with inadequate or absent production of erythropoietin. Severe kidney disease may decrease production of EPO, and congenital absence of EPO can occur.

Elevated levels of EPO can be found in some **anemias** when the body tries to overcompensate for reduced blood volume. Elevated levels are also seen in polycythemia, and erythropoietin-secreting tumors.

## Resources

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**ESB** see **Electrical stimulation of the brain**

some instances **gangrene** in the colon. Other known *E. coli*-causing infections may include chronic renal failure, **pancreatitis**, and **diabetes mellitus**. Some neurological symptoms such as drowsiness, seizure and **coma** may occur. In infants, *E. coli* infections are present in cases of infantile gastroenteritis and neonatal meningitis.

Strains of *E. coli* that produce diarrhea were initially distinguished by their O (somatic) antigens found on the bacterial surface. Although there is an overlap in characteristics between strains, they may be classified into four main groups; enterohemorragic (O157), enteropathogenic (O55,O111), enterotoxigenic (O6,O78), and enteroinvasive (O124,O164).

### *E.coli* O157 (VTEC)

The O157:H7 strain is the member of the group most often associated with a particularly severe form of diarrhea. (The O indicates the somatic antigen, while the H denotes the flagellar antigen, both of which are found on the cell surface of the bacteria.) The bacterium was discovered in 1977, and first reports of infections followed in 1982. *E. coli* O157:H7, as it is frequently referred to by researchers, causes bloody diarrhea in many infected patients. It accounts for about 2% of all cases of diarrhea in the western world, and at least one-third of cases of hemorrhagic colitis, or about 20,000 cases per year.

*E. coli* O157:H7 is also the most common cause of unique syndromes, known as the **Hemolytic-Uremic Syndrome** (HUS) and thrombocytopenic purpura (TTP), which causes kidney failure, **hemolytic anemia**, and **thrombocytopenia**. Usually, infection with this strain of bacteria will subside without further complications. However, about 5% of people who are infected will develop HUS/TTP. This infection also accounts for the majority of episodes of HUS, especially in children.

This strain of bacteria produces a potent toxin called verotoxin, named for toxin's ability to kill green monkey kidney or "vero" cells. Bacteria that produce verotoxin are referred to as Verotoxin-producing *E. coli* (VTEC). The numbers of bacteria that are necessary to reproduce infectious levels of bacteria are quite small, estimated at 10-100 viable bacteria. These toxins are lethal for intestinal cells and those that line vessels (endothelial cells), inhibiting protein synthesis causing cell **death**. It is believed that the damage to blood vessels results in the formation of clots, which eventually leads to the Hemolytic-Uremic Syndrome. HUS/TTP is a serious, often fatal, syndrome that has other causes in addition to *E. coli* O157:H7; it is characterized by the breaking up of red blood cells (hemolysis) and kidney failure (uremia). The syndrome occurs most often in the very young and very old.

## Escherichia coli

### Definition

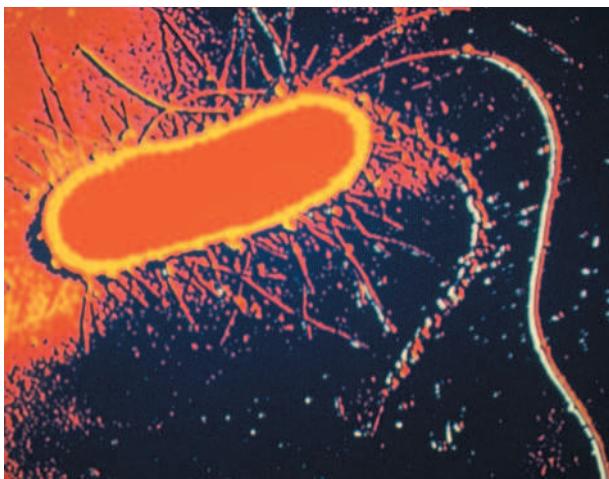
*E. coli* (*Escherichia coli*) is one of several types of bacteria that normally inhabit the intestine of humans and animals (commensal organism). Some strains of *E. coli* are capable of causing disease under certain conditions when the immune system is compromised or disease may result from an environmental exposure.

### Description

*E. coli* bacteria may give rise to infections in **wounds**, the urinary tract, biliary tract, and abdominal cavity (**peritonitis**). This organism may cause septicemia, neonatal **meningitis**, infantile **gastroenteritis**, tourist **diarrhea**, and hemorrhagic diarrhea. An *E. coli* infection may also arise due to environmental exposure. Infections with this type of bacteria pose a serious threat to public health with outbreaks arising from food and water that has been contaminated with human or animal feces or sewage. This type of bacteria has been used as a biological indicator for safety of drinking water since the 1890s. Exposure may also occur during hospitalization, resulting in **pneumonia** in immunocompromised patients or those on a ventilator.

### Causes and symptoms

The symptoms of infection and resulting complications are dependent upon the strain of *E. coli* and the site of infection. These bacteria produce toxins that have a wide range of effects. Symptoms caused by some *E. coli* infections range from mild to severe, bloody diarrhea, acute abdominal **pain**, vomiting, and **fever**. Gastrointestinal complications that can cause *E. coli* infections include **irritable bowel syndrome** (IBS) ischemic colitis, **appendicitis**, perforation of the large bowel, and in



**A magnified image of the *E. coli* bacterium.** (The Stock Market. Reproduced by permission.)

*E. coli* O157:H7 is commonly found in cattle and poultry, and outbreaks have of disease have been associated with cattle and bovine products. There are reports of contamination from unpasteurized apple juice, hamburger meat, radish sprouts, lettuce, and potatoes, as well as other food sources. Environmental contamination may occur in water drained from cattle pastures or water containing human sewage used for drinking or swimming. Human to human transmission, through contact with fecal matter, has also been identified in daycare centers.

After an incubation period of three to four days on average, watery diarrhea begins, which rapidly progresses to bloody diarrhea in many victims, in which case the bowel movement may be mostly blood. Nausea, vomiting, and low-grade fever are also frequently present. Gastrointestinal symptoms last for about one week, and recovery is often spontaneous. Symptomatic infection may occur in about 10% of infected individuals. About 5–10% of individuals, usually at the extremes of age or elevated leukocyte count, develop HUS/TTP, and ultimately, kidney failure. Patients taking **antibiotics** or medications for gastric acidity may also be at risk. Neurological symptoms can also occur as part of HUS/TTP and consist of seizures, **paralysis**, and coma. **Rectal prolapse** may also be a complication, and in some cases colitis, appendicitis, perforation of the large bowel, and gangrene in the bowel. Systemically, the most prevalent complications of *E. coli* 157 infections are HUS and TTP.

#### *E. coli* non-O157 (VTEC)

These strains of *E. coli* produce verotoxin, but are strains other than O157. There have been as many as one hundred different types implicated in the development of

disease. Strain OH111 was found to be involved in outbreaks in Australia, Japan, and Italy. The O128, O103, and O55 groups have also been implicated in diarrhea outbreaks. In Britain, cases of infantile gastroenteritis in maternity hospitals and neonatal units have been attributed to the *E. coli* non-O157 group. Many of these organisms have been identified in cattle.

#### *Enterotoxigenic E. coli*

Two toxins may be produced by this group, the heat-labile enterotoxin (LT) that can produce enteritis in infants, and a heat stable enterotoxin (ST), the action of which has yet to be determined.

#### *Enteroinvasive E. coli*

Some strains of the enteroinvasive *E. coli* have been involved in the development of gastroenteritis in infants. These organisms do not produce enterotoxin. The cells of the intestine are affected, with the development of symptoms that are typical of a shigellae infection.

#### **Diagnosis**

Diagnosis of a specific type of infection is dependent upon the characteristics of the particular strain of the organism.

#### *E. coli* O157:H7 (HUS)

This particular strain of *E. coli* is suspected when bloody diarrhea, bloody stools, lack of fever, elevated leukocyte count, and abdominal tenderness are present. Stool cultures are used to tentatively identify the bacteria. Unfortunately, cultures are often negative or inconclusive if done after 48 hours of symptoms. Further tests are usually needed, however, for confirmation of infection. This may include a full **blood count**, blood film, and tests to determine urea, electrolyte, and LDH (lactate dehydrogenase) levels. Damaged red blood cells, and elevated levels of creatinine, urea, and LDH with a drop in **platelet count** may indicate that HUS will develop. Immunomagnetic separation is now being used for diagnosis as well.

#### *E. coli* non-O157 (VTEC)

Diagnosis is often difficult for these types of bacteria, but production of enterohemolysin (Ehly) is used as an indicator. Other diagnostic tests are used to detect verotoxins, including ELISA (enzyme-linked immunosorbent assays), colony immunoblotting, and DNA-based tests.

#### *E. coli* O157 STEC

Methods for detection of this type of bacteria are under development, including culture growth media

selective for this organism. Immunomagnetic separation and specific ELISA, latex agglutination tests, colony immunoblot assays, and other immunological-based detection methods are being explored.

### Treatment

Uncomplicated cases of the *E. coli* O157:H7 the infection clear up within ten days. It is not certain that antibiotics are helpful in treating *E. coli* O157:H7 and there is some evidence that they may be harmful. **Dehydration** resulting from diarrhea must be treated with either Oral Rehydration Solution (ORS) or intravenous fluids. Anti-motility agents that decrease the intestines' ability to contract, should not be used in any patient with bloody diarrhea. Treatment of HUS, if it develops, involves correction of clotting factors, plasma exchange, and **kidney dialysis**. Blood transfusions may be required. Treatment methods for other *E. coli* infections are similar. Antibiotics are often used in the treatment of *E. coli* infections, but their role is controversial. Some antibiotics may enhance the development of HUS/TTP depending upon their action, as well as the use of anti diarrhea medications that should be avoided. Phosphoenolpyruvate analogues may be helpful. Gentamicin, ampicillin, ceftazidime, or beta-lactamase-stable cephalosporin may be administered for neonatal meningitis. Antibiotic therapy is further complicated by the presence of antibiotic resistant organisms.

### Alternative treatment

Studies have been conducted to determine if diarrhea symptoms can be reduced by alternative therapies such as the consumption of herbal teas, psyllium, and **acupuncture**. Patients should consult their doctors before using any alternative treatments, as *E. coli* can be life threatening and should be closely monitored.

### Prognosis

In most cases of O157:H7, symptoms last for about a week and recovery is often spontaneous. Ten percent of individuals with *E. coli* O157:H7 infection develop HUS; 5% of those will die of the disease. Some who recover from HUS will be left with some degree of kidney damage and possibly irritable bowel syndrome. Additionally, there is a possibility of chronic *E. coli* infection.

Infants that develop *E. coli* infections may be permanently affected. Gastroenteritis may leave the child with **lactose intolerance**. Neonates developing meningitis from *E. coli* strains have a high morbidity and mortality rate

### Prevention

Thorough cooking of all meat and poultry products and adhering to proper food preparation is the most

## KEY TERMS

**Antigen**—A substance, usually a protein, that causes the formation of an antibody and reacts specifically with that antibody.

**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.

**Colitis**—Inflammation of the colon or large intestine, usually causing diarrhea which may be bloody.

**Food irradiation methods**—A process using radiant energy to kill microorganisms in food, to extend the amount of time in that food can be sold and eaten safely.

**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.

**Urea**—Chemical formed during the body's metabolism of nitrogen and normally excreted by the kidney. Urea levels rise in the blood when kidney failure occurs.

effective way to avoid infection. More studies are needed to determine the appropriate safety margins for killing these bacteria. Food irradiation methods are also being developed to sanitize food. Vaccinations to *E. coli* O157 are under development, as are medications aimed at limiting the effects of the verotoxin. The enforcement of regulations for meat production and water are critical. Steam pasteurization is used in the United States and is being explored in other countries.

Prevention of *E. coli* gastroenteritis in infants is best achieved by breast-feeding. The breast milk contains antibodies that combat the infection. For bottle-fed infants, care should be taken in the preparation of the milk and bottles. Good hygiene of the umbilical cord area is important. Keeping this area clean and dry may reduce infection.

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Jill Granger  
David Kaminstein

Esophageal acidity test see **Esophageal function tests**

Esophageal aperistalsis see **Achalasia**

## Esophageal atresia

### Definition

Esophageal atresia is a serious birth defect in which the esophagus, the long tube that connects the mouth to the stomach, is segmented and closed off at any point.

This condition usually occurs with **tracheoesophageal fistula**, a condition in which the esophagus is improperly attached to the trachea, the nearby tube that connects the nasal area to the lungs. Esophageal atresia occurs in approximately 1 in 4,000 live births.

### Description

Failure of an unborn child (fetus) to develop properly results in **birth defects**. Many of these defects involve organs that do not function, or function only incidentally, before birth, and, as a result, go undetected until the baby is born. In this case, the digestive tract is unnecessary for fetal growth, since all **nutrition** comes from the mother through the placenta and umbilical cord.

During fetal development, the esophagus and the trachea arise from the same original tissue. Normally, the two tubes would form separately (differentiate); however, in cases of esophageal atresia and tracheoesophageal fistulas, they do not, resulting in various malformed configurations. The most common configuration is the "C" type, in which the upper part of the esophagus abruptly ends in a blind pouch, while the lower part attaches itself to the trachea. This configuration occurs in 85–90% of cases. Esophageal atresia without involvement of the trachea occurs in only 8% of cases.

### Causes and symptoms

The cause of esophageal atresia, like that of most birth defects, is unknown.

An infant born with this defect will at first appear all right, swallowing normally. However, the blind pouch will begin to fill with mucus and saliva that would normally pass through the esophagus to the stomach. These secretions back up into the mouth and nasal area, causing the baby to drool excessively. When fed, the baby will also immediately regurgitate what he or she has eaten. **Choking** and coughing may also occur as the baby breathes in the fluid backing up from the esophagus. Aspiration **pneumonia**, an infection of the respiratory system caused by inhalation of the contents of the digestive tract, may also develop.

### Diagnosis

Physicians who suspect esophageal atresia after being presented with the above symptoms diagnose the condition using x-ray imaging or by passing a catheter through the nose and into the esophagus. Esophageal atresia is indicated if the catheter hits an obstruction 4–5 in (10–13 cm) from the nostrils.

## KEY TERMS

**Fetal**—Refers to the fetus, also known in the first two months after conception as an embryo.

**Fistula**—Unnatural connection between two hollow organs or one organ and the outside.

## Treatment

Infants with esophageal atresia are unlikely to survive without surgery to reconnect the esophagus. The procedure is done as soon as possible; however, **prematurity**, the presence of other birth defects, or complications of aspiration pneumonia may delay surgery. Once diagnosed, the baby will be fed intraveneously until he or she has recovered sufficiently from the operation. Mucus and saliva will also be continuously removed via a catheter until recovery has occurred. When surgery is performed, the esophagus is reconnected and, if necessary, separated from the trachea. If the two ends of the esophagus are too far apart to be reattached, tissue from the large intestine is used to join them.

## Prognosis

Surgery to correct esophageal atresia is usually successful. Post-operative complications may include difficulty swallowing, since the esophagus may not contract efficiently, and gastrointestinal reflux, in which the acidic contents of stomach back up into the lower part of the esophagus, possibly causing ulcers.

## Resources

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J. Ricker Polsdorfer, MD

# Esophageal cancer

## Definition

Esophageal **cancer** is a malignancy that develops in tissues of the hollow, muscular canal (esophagus)

along which food and liquid travel from the throat to the stomach.

## Description

Esophageal cancer usually originates in the inner layers of the lining of the esophagus and grows outward. In time, the tumor can obstruct the passage of food and liquid, making swallowing painful and difficult. Since most patients are not diagnosed until the late stages of the disease, esophageal cancer is associated with poor quality of life and low survival rates.

Squamous cell carcinoma is the most common type of esophageal cancer, accounting for 95% of all esophageal cancers worldwide. The esophagus is normally lined with thin, flat squamous cells that resemble tiny roof **shingles**. Squamous cell carcinoma can develop at any point along the esophagus but is most common in the middle portion.

Adenocarcinoma has surpassed squamous cell carcinoma as the most common type of esophageal cancer in the United States. Adenocarcinoma originates in glandular tissue not normally present in the lining of the esophagus. Before adenocarcinoma can develop, glandular cells must replace a section of squamous cells. This occurs in Barrett's esophagus, a precancerous condition in which chronic acid reflux from the stomach stimulates a transformation in cell type in the lower portion of the esophagus.

A very small fraction of esophageal cancers are melanomas, **sarcomas**, or lymphomas.

There is great variability in the incidence of esophageal cancer with regard to geography, ethnicity, and gender. The overall incidence is increasing. About 13,000 new cases of esophageal cancer are diagnosed in the United States each year. During the same 12-month period, 12,000 people die of this disease. It strikes between five and ten North Americans per 100,000. In some areas of China the cancer is endemic.

Squamous cell carcinoma usually occurs in the sixth or seventh decade of life, with a greater incidence in African-Americans than in others. Adenocarcinoma develops earlier and is much more common in white patients. In general, esophageal cancer occurs more frequently in men than in women.

## Causes and symptoms

The exact cause of esophageal cancer is unknown, although many investigators believe that chronic irritation of the esophagus is a major culprit. Most of the identified risk factors represent a form of chronic irritation.

However, the wide variance in the distribution of esophageal cancer among different demographic groups raises the possibility that genetic factors also play a role.

Several risk factors are associated with esophageal cancer.

- Tobacco and alcohol consumption are the major risk factors, especially for squamous cell carcinoma. **Smoking** and alcohol abuse each increase the risk of squamous cell carcinoma by five-fold. The effects of the two are synergistic, in that the combination of smoking and alcohol increases the risk by 25- to 100- fold. It is estimated that drinking about 13 ounces of alcohol every day for an extended period of time raises the risk of developing esophageal cancer by 18%. That likelihood increases to 44% in individuals who also smoke one or two packs of cigarettes a day. Smokeless tobacco also increases the risk for esophageal cancer.
- Gastroesophageal reflux is a condition in which acid from the stomach refluxes backwards into the lower portion of the esophagus, sometimes causing symptoms of **heartburn**. In some cases of gastroesophageal reflux, the chronic exposure to acid causes the inner lining of the lower esophagus to change from squamous cells to glandular cells. This is called Barrett's esophagus. Patients with Barrett's esophagus are roughly 30 to 40 times more likely than the general population to develop adenocarcinoma of the esophagus.
- A diet low in fruits, vegetables, zinc, riboflavin, and other **vitamins** can increase risk of developing to esophageal cancer.
- Caustic injury to the esophagus inflicted by swallowing lye or other substances that damage esophageal cells can lead to the development of squamous cell esophageal cancer in later life.
- Achalasia is a condition in which the lower esophageal sphincter (muscle) cannot relax enough to let food pass into the stomach. Squamous cell esophageal cancer develops in about 6% of patients with achalasia.
- Tylosis is a rare inherited disease characterized by excess skin on the palms and soles. Affected patients have a much higher probability of developing esophageal cancer than the general population. They should have regular screenings to detect the disease in its early, most curable stages.
- Esophageal webs, which are protrusions of tissue into the esophagus, and diverticula, which are outpouchings of the wall of the esophagus, are associated with a higher incidence of esophageal cancer.

### Symptoms

Unfortunately, symptoms generally don't appear until the tumor has grown so large that the patient cannot be cured. **Dysphagia** (trouble swallowing or a sensation of having food stuck in the throat or chest) is the most common symptom. Swallowing problems may occur occasionally at first, and patients often react by eating more slowly and chewing their food more carefully and, as the tumor grows, switching to soft foods or a liquid diet. Without treatment, the tumor will eventually prevent even liquid from passing into the stomach. A sensation of burning or slight mid-chest pressure is a rare, often-disregarded symptom of esophageal cancer. Painful swallowing is usually a symptom of a large tumor obstructing the opening of the esophagus. It can lead to regurgitation of food, weight loss, physical wasting, and **malnutrition**. Anyone who has trouble swallowing, loses a significant amount of weight without dieting, or cannot eat solid food because it is too painful to swallow should see a doctor.

### Diagnosis

A barium swallow is usually the first test performed on a patient whose symptoms suggest esophageal cancer. After the patient swallows a small amount of barium, a series of x rays can highlight any bumps or flat raised areas on the normally smooth surface of the esophageal wall. It can also detect large, irregular areas that narrow the esophagus in patients with advanced cancer, but it cannot provide information about disease that has spread beyond the esophagus. A double contrast study is a barium swallow with air blown into the esophagus to improve the way the barium coats the esophageal lining. Endoscopy is a diagnostic procedure in which a thin lighted tube (endoscope) is passed through the mouth, down the throat, and into the esophagus. Cells that appear abnormal are removed for biopsy. Once a diagnosis of esophageal cancer has been confirmed through biopsy, staging tests are performed to determine whether the disease has spread (metastasized) to tissues or organs near the original tumor or in other parts of the body. These tests may include computed tomography, endoscopic ultrasound, **thoracoscopy**, **laparoscopy**, and **positron emission tomography**.

### Treatment

Treatment for esophageal cancer is determined by the stage of the disease and the patient's general health. The most important distinction to make is whether the cancer is curable. If the cancer is in the early stages, cure may be possible. If the cancer is advanced or if the patient will not tolerate major surgery, treatment is usually directed at palliation (relief of symptoms only) instead of cure.

## Staging

Stage 0 is the earliest stage of the disease. Cancer cells are confined to the innermost lining of the esophagus. Stage I esophageal cancer has spread slightly deeper, but still has not extended to nearby tissues, lymph nodes, or other organs. In Stage IIA, cancer has invaded the thick, muscular layer of the esophagus that propels food into the stomach and may involve connective tissue covering the outside of the esophagus. In Stage IIB, cancer has spread to lymph nodes near the esophagus and may have invaded deeper layers of esophageal tissue. Stage III esophageal cancer has spread to tissues or lymph nodes near the esophagus or to the trachea (windpipe) or other organs near the esophagus. Stage IV cancer has spread to distant organs like the liver, bones, and brain. Recurrent esophageal cancer is disease that develops in the esophagus or another part of the body after initial treatment.

## Surgery

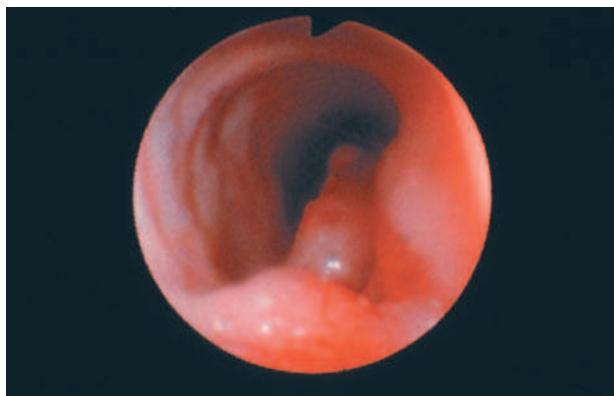
The most common operations for the treatment of esophageal cancer are esophagectomy and esophagogastrectomy. Esophagectomy is the removal of the cancerous part of the esophagus and nearby lymph nodes. This procedure is performed only on patients with very early cancer that has not spread to the stomach. Esophagogastrectomy is the removal of the cancerous part of the esophagus, nearby lymph nodes, and the upper part of the stomach. The resected esophagus is replaced with the stomach or parts of intestine so the patient can swallow. These procedures can significantly relieve symptoms and improve the nutritional status of more than 80% of patients with dysphagia. Although surgery can cure some patients whose disease has not spread beyond the esophagus, but more than 75% of esophageal cancers have spread to other organs before being diagnosed. Less extensive surgical procedures can be used for palliation.

## Chemotherapy

Oral or intravenous **chemotherapy** alone will not cure esophageal cancer, but pre-operative treatments can shrink tumors and increase the probability that cancer can be surgically eradicated. Palliative chemotherapy can relieve symptoms of advanced cancer but will not alter the outcome of the disease.

## Radiation

External beam or internal radiation, delivered by machine or implanted near cancer cells inside the body, is only rarely used as the primary form of treatment. Post-operative radiation is sometimes used to kill cancer cells that couldn't be surgically removed. Palliative radi-



**A close-up view of a cancerous esophageal tumor.** (Custom Medical Stock Photo. Reproduced by permission.)

ation is effective in relieving dysphagia in patients who cannot be cured. However, radiation is most useful when combined with chemotherapy as either the definitive treatment or preoperative treatment.

## Palliation

In addition to surgery, chemotherapy, and radiation, other palliative measures can provide symptomatic relief. Dilatation of the narrowed portion of the esophagus with soft tubes can provide short-term relief of dysphagia. Placement of a flexible, self-expanding stent within the narrowed portion is also useful in allowing more food intake.

## Follow-up treatments

Regular barium swallows and other imaging studies are necessary to detect recurrence or spread of disease or new tumor development.

## Alternative treatment

Photodynamic therapy (PDT) involves intravenously injecting a drug that is absorbed by cancer cells and kills them after they are exposed to specific laser beams. PDT can be used for palliation, but it also cured some early esophageal cancers during preliminary studies. Researchers are comparing its benefits with those of more established therapies.

Endoscopic laser therapy involves delivering short, powerful laser treatments to the tumor through an endoscope. It can improve dysphagia, but multiple treatments are required, and the benefit is seldom long-lasting.

## Prognosis

Since most patients are diagnosed when the cancer has spread to lymph nodes or other structures, the prog-

## KEY TERMS

**Computed tomography**—A radiology test by which images of cross-sectional planes of the body are obtained.

**Endoscopic ultrasound**—A radiology test utilizing high frequency sound waves, conducted via an endoscope.

**Laparoscopy**—Examination of the contents of the abdomen through a thin, lighted tube passed through a small incision.

**Positron emission tomography**—A radiology test by which images of cross-sectional planes of the body are obtained, utilizing the properties of the positron. The positron is a subatomic particle of equal mass to the electron, but of opposite charge.

**Synergistic**—The combined action of two or more processes is greater than the sum of each acting separately.

**Thoracoscopy**—Examination of the contents of the chest through a thin, lighted tube passed through a small incision.

nosis for esophageal cancer is poor. Generally, no more than half of all patients are candidates for curative treatment. Even if cure is attempted, the cancer can recur.

### Prevention

There is no known way to prevent esophageal cancer.

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National Coalition for Cancer Survivorship. 1010 Wayne Avenue, 5th Floor, Suite 300, Silver Spring, MD 20910. (888) 650-9127.

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Esophageal diverticula see **Esophageal pouches**

## Esophageal function tests

### Definition

The esophagus is the swallowing tube through which food passes on its way from the mouth to the stomach. The main function of this organ is to propel food down into the stomach. There is also a mechanism to prevent food from coming back up or "refluxing" from the stomach into the esophagus. Esophageal function tests are used to determine if these processes are normal or abnormal.

### Purpose

The esophagus is a long, muscular tube that also has two muscles (or sphincters) at the top and bottom. All of these muscular areas must contract in an exact sequence for swallowing to proceed normally. There are three main symptoms that occur when esophageal function is abnormal: difficulty with swallowing (dysphagia), **heartburn**, and **chest pain**.

Doctors perform a variety of tests to evaluate these symptoms. Endoscopy, which is not a test of esophageal function, is often used to determine if the lining of the esophagus has any ulcers, tumors, or areas of narrowing (strictures). Many times, however, endoscopy only shows the doctor if there is injury to the esophageal lining, and the procedure gives no information about the cause of the problem.

Therefore, in addition to endoscopy, several studies are available that measure esophageal function. There are three basic types of tests used to assess esophageal function:

- Manometry is used to study the way the muscles of the esophagus contract, and is most useful for the investigation of difficulty with swallowing.
- Esophageal pH monitoring measures changes in esophageal acidity, and is valuable for evaluating patients with heartburn or gastroesophageal reflux disease (GERD).
- X-ray studies investigate swallowing difficulties. They either follow the progress of barium during swallowing using a fluoroscope, or they use radioactive scanning techniques.

## Precautions

Pregnant patients undergoing x-ray exams should carefully review the risks and benefits with their doctors. Most x-ray exams of the gastrointestinal tract do not involve radiation levels that are harmful to the unborn baby.

## Description

### *Manometry*

This study is designed to measure the pressure changes produced by contraction of the muscular portions of the esophagus. An abnormality in the function of any one of the segments of the swallowing tube causes difficulty in swallowing. Doctors call this symptom dysphagia. This exam is most useful in evaluating those patients whose endoscopy is negative.

During manometry, the patient swallows a thin tube carrying a device that senses changes in pressures in the esophagus. Readings are taken at rest and during swallowing. Medications are sometimes given during the study to help in the diagnosis. The results are then transmitted to recording equipment. Manometry can best identify diseases that produce disturbances of motility or contractions of the esophagus.

### *Esophageal pH monitoring*

This procedure involves measuring the esophagus' exposure to acid that has "refluxed" from the stomach. The test is ideal for evaluating recurring heartburn or GERD. Too much acid produces not only heartburn, but also ulcers that can bleed or produce areas of narrowing (strictures) when they heal.

Normally, acid refluxes into the esophagus in only small amounts for short periods of time. A muscle called the lower esophageal sphincter prevents excessive reflux. Spontaneous contractions that increase esophageal emptying and production of saliva are other important protective mechanisms.

"pH" is the scientific term that tells just how acidic or alkaline a substance is. Researchers have shown that in the esophagus, the presence of acid is damaging only if it persists for prolonged periods. Therefore, the test has been designed to monitor the level of acidity over 24 hours, usually in the home. In this way, patients maintain their daily routine, documenting their symptoms, and at what point in their activities they occurred. During this period, a thin tube with a pH monitor remains in the esophagus to record changes. After the study, a computer is used to compare changes in acidity with symptoms reported by the patient.

Surgery is an effective and long-lasting treatment for symptoms of recurrent reflux and is the choice of many

patients and doctors. pH monitoring is usually performed before surgery to confirm the diagnosis and to judge the effects of drug therapy.

### *X-ray tests*

These fall into two categories: (1) those done with the use of barium and a fluoroscope; and (2) those performed with radioactive materials.

Studies performed with fluoroscopy are of greatest value in identifying a structural abnormality of the esophagus. Although this is not truly an esophageal function test, it does allow doctors to consider other diagnostic possibilities. Often a sandwich or marshmallow coated with barium is used to identify the site of an obstruction.

During fluoroscopy, the radiologist can observe the passage of material through the esophagus in real time, and video recordings can also be done. This is particularly useful when the swallowing symptoms appear to involve mainly the upper region of the esophagus. The most common cause of swallowing difficulties is a previous stroke, although other diseases of the neuromuscular system (like *myasthenia gravis*) can produce the same symptoms.

Scans using low-dose radioactive materials are useful because they are able not only to demonstrate that food passes through the esophagus more slowly than normal, but also how slow. These studies involve swallowing food coated with material that is followed by a nuclear medicine scanner. Scans are best used when other methods have failed to make a diagnosis, or if it is necessary to determine the degree of the abnormality. As of 1997, scans mainly served as research tools.

## Preparation

Patients should not eat or drink for several hours before the exam. Many medications affect the esophagus; doses sometimes need to be adjusted or even stopped for a while. Patients must inform doctors of all medications taken, including over-the-counter medications (purchased without a doctor's prescription), and any known allergies.

## Aftercare

For most of these studies, no special care is needed after the procedure. Patients can often go about normal daily activities following any of these tests. One exception is for those who undergo an x-ray exam with the use of barium. This can have a constipating effect and patients should ask about using a mild laxative later on.

## Risks

Exposure of a fetus to x rays, especially in the first three months, is a potential risk.

Other studies of esophageal function are essentially free of any significant risk. The tubes passed during these procedures are small, and most patients adjust to them quite well. However, since medications cannot be used to relax patients, some may not tolerate the exam.

### Abnormal results

Manometry is used to diagnose abnormalities related to contraction or relaxation of the various muscular regions of the esophagus. These studies cannot distinguish whether injury to either the muscle or nerves of the esophagus is producing the abnormal results. Only the final effect on esophageal muscle is identified. Results should be interpreted in light of the patient's entire medical history.

For example, there are many diseases that cause poor relaxation of the lower esophageal sphincter. When no cause is found, the disease is called **achalasia**.

Abnormal results of pH tests can confirm symptoms of heartburn or indicate a cause of chest pain (or rarely, swallowing difficulties). Doctors may want to start or change medications based on these results, or even repeat the test using different doses of medication. As noted above, these studies are indicated before surgical treatment of GERD.

X-ray tests can only serve to document an abnormality, and they are far from perfect. If they are negative, then other studies are often needed.

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David Kaminstein, MD

**Esophageal laceration** see **Mallory-Weiss syndrome**

**Esophageal manometry** see **Esophageal function tests**

## Esophageal pouches

### Definition

Esophageal pouches, also known as esophageal diverticula, are pocket-like structures formed when the interior space of the esophagus, the tube that connects the mouth to the stomach, protrudes into the walls that surround it.

### Description

The esophagus is a muscular tube that propels food into the stomach. A defect in the wall of the esophagus may allow the lining to herniate, creating a space where food can be caught. Pouches can appear anywhere between the throat and the stomach. They occur primarily in men and usually later in life.

Different names for the condition apply to different locations along the esophagus:

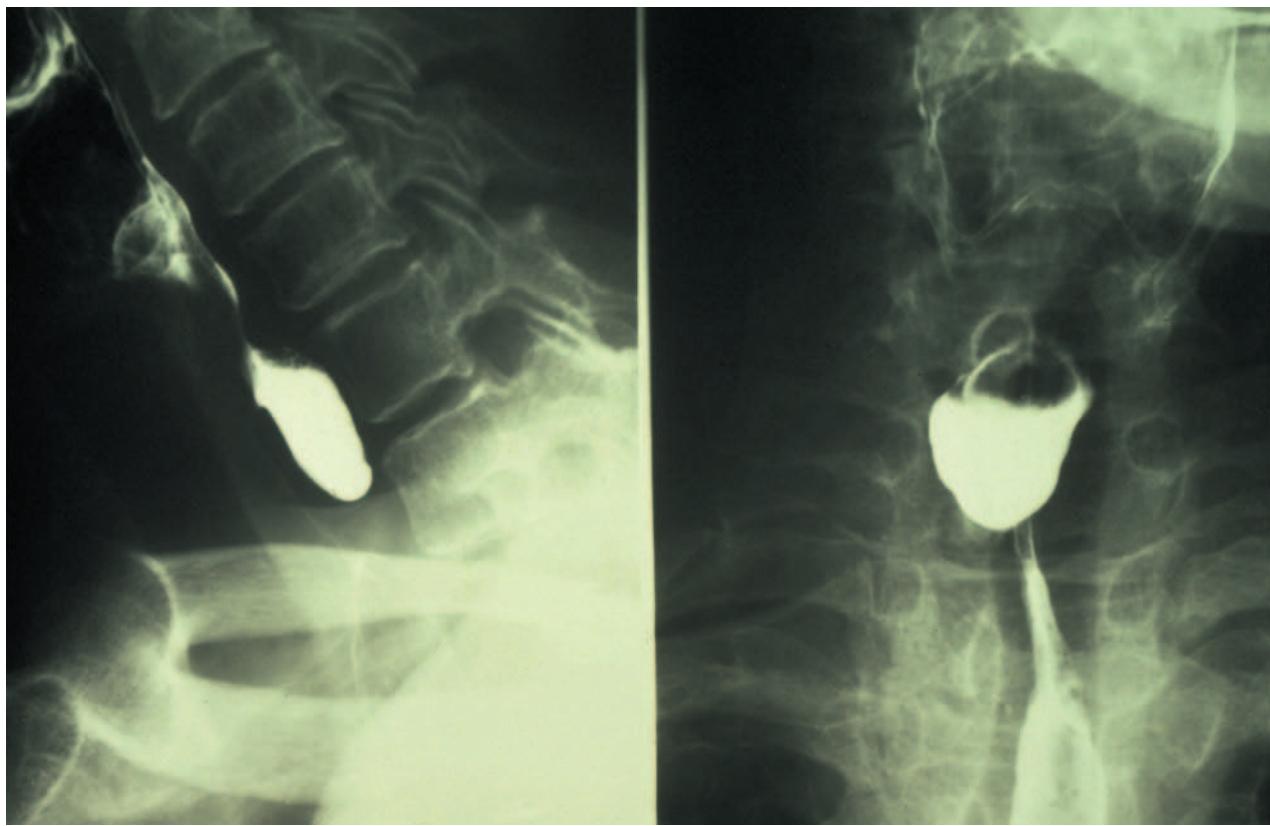
- Zenker's diverticula are pharyngeal pouches, or ones that occur in the upper neck area at the top of the esophagus.
- **Traction** diverticula are a type of mid-esophageal pouch.
- Epiphrenic diverticula occur at the bottom of the esophagus near where it enters the stomach.

### Causes and symptoms

To propel food into the stomach (or out of it during vomiting) the esophagus generates internal pressure just like the bowel. Under certain circumstances, that pressure can herniate the esophageal lining through a weakness in the wall, creating a pouch (a balloon squeezed in the hand will herniate through the fingers in the same way). Pouches are more common in people who have motility disorders of the esophagus, swallowing that is not well coordinated and may be spastic. A traction diverticulum can develop from a scar that pulls the esophagus out of shape. Food and saliva can collect in all of these pouches.

Pouches in the neck usually cause **bad breath** (halitosis) and the regurgitation of swallowed food and saliva. Some patients with Zenker's diverticula can push on their neck and make old food appear in their mouths. Pouches near the stomach may cause swallowing problems, conditions known as **achalasia** or **dysphagia**. Mid-esophageal pouches usually cause no symptoms.

In the most serious cases, a person may be unable to swallow because the esophagus is obstructed, or the esophagus may rupture, spilling its contents into the chest or neck.



A split x-ray image of the upper chest, neck, and esophagus (left), and chest and esophagus (right). (Custom Medical Stock Photo. Reproduced by permission.)

## Diagnosis

Difficulty swallowing, bad breath, or food reappearing in the back of the mouth are among the signs physicians look for when diagnosing this condition. Sometimes the patient may also experience **pain** in the chest resembling a **heart attack**. A series of x rays taken while swallowing a contrast agent usually demonstrates the diverticulum clearly. An esophagoscopy may also be needed to gather more detail. Manometry, measuring pressures inside the esophagus using a balloon that is passed down it, may help determine the cause of the diverticula.

## Treatment

Treatment for this condition is primarily aimed at alleviating symptoms. Physicians direct the patient to eat a bland diet, to chew his or her food thoroughly, and to drink water after eating to clean out the pouches. If the condition is severe, several types of surgery are available to remove the pouches and repair the defects. If a pouch is due to a stenosis (narrowing) in the esophagus it may be possible to relieve it by passing a dilator through it, a process called bougeinage.

## KEY TERMS

**Achalasia**—Failure of the lower end of the esophagus (or another tubular valve) to open, resulting in obstruction, either partial or complete.

**Contrast agent**—A substance that produces shadows on an x ray so that hollow structures can be more easily seen.

**Dysphagia**—Difficult swallowing.

**Esophagoscopy**—Looking down the esophagus with a flexible viewing instrument.

**Herniate**—To protrude beyond usual limits.

**Manometry**—Pressure measurement.

## Prognosis

The two complications that can render these nuisances dangerous, obstruction and rupture, are emergencies. Both require immediate medical attention. Other

than that, diverticula will usually grow slowly over the years, gradually increasing the symptoms they cause.

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Esophageal ulcers see **Ulcers (digestive)**

## KEY TERMS

**Pathologist**—A doctor who specializes in the anatomic (structural) and chemical changes that occur with diseases. These doctors function in the laboratory, examining biopsy specimens, and regulating studies performed by the hospital laboratories (blood tests, urine tests, etc). Pathologists also perform autopsies.

Biopsies (small tissue samples) of inflamed or "suspicious" areas can be obtained and examined by a pathologist. Cell scrapings can also be taken by the introduction of a small brush; this helps in the diagnosis of cancer or infections.

When treating conditions in the upper gastrointestinal tract, small instruments are passed through the endoscope that can stretch narrowed areas (strictures), or remove swallowed objects (such as coins or pins). In addition, bleeding from ulcers or vessels can be treated by a number of endoscopic techniques.

Recent studies have shown the usefulness of endoscopic removal of early tumors of the esophagus or stomach. This is done either with injection of certain materials (like alcohol), or with the use of instruments (like lasers) that burn the tumor. Other techniques combining medications and lasers also show promise.

## Precautions

Patients should inquire as to the doctor's expertise with these procedures, especially when therapy is the main goal. The doctor should be informed of any **allergies**, medication use, and medical problems.

## Description

First, a "topical" (local) medication to numb the gag reflex is given either by spray or is gargled. Patients are usually sedated for the procedure (though not always) by injection of medications into a vein. The endoscopist then has the patient swallow the scope, which is passed through the upper gastrointestinal tract. The lens or camera at the end of the instrument allows the endoscopist to examine each portion of the upper gastrointestinal tract; photos can be taken for reference. Air is pumped in through the instrument to allow proper observation. Biopsies and other procedures can be performed without any significant discomfort.

## Purpose

EGD is performed to evaluate or treat symptoms relating to the upper gastrointestinal tract, such as:

- upper abdominal or chest pain
- nausea or vomiting
- difficulty swallowing (dysphagia)
- bleeding from the upper intestinal tract
- anemia (low **blood count**). EGD can be used to treat certain conditions, such as an area of narrowing or bleeding in the upper gastrointestinal tract

Upper endoscopy is more accurate than x rays for detecting inflammation, ulcers, or tumors. It is used to diagnose early **cancer** and can frequently determine whether a growth is benign (not cancerous) or malignant (cancerous).



**Esophagogastroduodenoscopy (EGD)** is performed to evaluate or treat symptoms relating to the upper gastrointestinal tract. By inserting an endoscope into the mouth and guiding it through the gastrointestinal tract, the esophagus, stomach, and duodenum can be examined and abnormalities treated. (Illustration by Electronic Illustrators Group.)

## Preparation

The upper intestinal tract must be empty for the procedure, so it is necessary NOT to eat or drink for at least 6–12 hours before the exam. Patients need to inquire about taking their medications before the procedure.

## Aftercare

Someone should be available to take the person home after the procedure and stay with them for a while; patients will not be able to drive themselves due to **sedation**. Pain or any other unusual symptoms should be reported immediately.

It is important to recognize early signs of any possible complication. The doctor should be notified if the patient has **fever**, trouble swallowing, or increasing throat, chest, or abdominal pain.

## Risks

EGD is safe and well tolerated; however, complications can occur as with any procedure. These are most

often due to medications used during the procedure, or are related to endoscopic therapy. The overall complication rate of EGD is less than 2%, and many of these complications are minor (such as inflammation of the vein through which medication is given). However, serious ones can and do occur, and almost half of them are related to the heart or lungs. Bleeding or perforations (holes in the gastrointestinal tract) are also reported, especially when tumors or narrowed areas are treated or biopsied. Infections have also been rarely transmitted; improved cleaning techniques should be able to prevent them.

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Essential tremor see **Tremors**

Estradiol see **Hormone replacement therapy**

Estrogen see **Hormone replacement therapy**

Estrogen fractions test see **Sex hormones tests**

Estrogen replacement therapy see **Hormone replacement therapy**

Ethambutol see **Antituberculosis drugs**

Etodolac see **Nonsteroidal anti-inflammatory drugs**

- Auditory evoked potentials are used to diagnose hearing losses. They can distinguish damage to the acoustic nerve (which carries signals from the ear to the brain stem) from damage to the auditory pathways within the brain stem. Most auditory EPs record activity from the brain stem, and are therefore called "brainstem auditory evoked potentials." Disorders diagnosed with auditory EPs include **acoustic neuroma** (tumors of the inner ear) and multiple sclerosis (chronic disease in which nerves lose patches of their outer covering). They may also be used to assess high frequency hearing ability, to determine brain **death**, and to monitor brainstem function during surgery
- Somatosensory evoked potentials record transmission of nerve impulses from the limbs to the brain, and can be used to diagnose nerve damage or degeneration within the spinal cord or nerve roots from multiple sclerosis, trauma, or other degenerative disease. Somatosensory EPs can be used to distinguish central versus peripheral nerve disease, when combined with results from a nerve conduction velocity test, which measures nerve function in the extremities.

## Precautions

Evoked potential studies are painless, noninvasive, and without any significant risk. Somatosensory EP tests involve very mild electric shocks, usually felt as a tingling.

## Description

The person performing the test locates and marks specific spots on the patient's head for placement of electrodes. These spots are cleaned, and an adhesive conducting paste is applied. Cup electrodes are attached. For somatosensory EP, spots on the arm or leg are also marked and cleaned; electrodes may be taped in place. The patient sits or reclines in a chair throughout the tests.

For a visual EP, the patient focuses on a TV screen which displays a checkerboard pattern. The eye not being tested is covered with a patch. For children or others whose attention may wander, goggles are used which show the pattern to one eye at a time. Each eye is usually tested twice, and the entire procedure takes approximately 30–45 minutes.

For auditory EP, headphones are used to deliver a series of clicks to one ear at a time. A masking or static sound is played into the other ear. Each ear is usually tested twice, and the entire procedure takes approximately 30–45 minutes.

For somatosensory EP, mild electrical shocks are delivered to the arm or leg. This may cause some twitching and tingling. The stimulus lasts for about two minutes at a time, and the entire procedure takes approximately 30 minutes.

## Evoked potential studies

### Definition

Evoked potential studies are a group of tests of the nervous system that measure electrical signals along the nerve pathways.

### Purpose

Nerves convey information to the body by sending electrical signals down the length of the nerve. These signals can be recorded by wires placed over the nerves on the surface of the skin, in a procedure called an evoked potential (EP) study. The person conducting the test evokes the patient's neural activity by visual or auditory stimulation or using a mild electrical shock. This causes changes in the electrical potential in the nerves. Analysis of the signals can provide information about the condition of nerve pathways, especially those in the brain and spinal cord. They can indicate the presence of disease or degeneration, and can help determine the location of nerve lesions.

There are three major types of EP studies used regularly:

- Visual evoked potentials are used to diagnose visual losses due to optic nerve damage, especially from **multiple sclerosis**. They are also useful to diagnose "hysterical blindness," in which loss of vision is not due to any nerve damage.

## KEY TERMS

**Nerve conduction velocity test**—A test of the speed of conduction of nerves, performed on the nerves in the arm and leg.

After the tests, the electrodes are removed with acetone and the scalp is cleaned.

### Preparation

Hair must be clean, dry, and free of any braids, pins, or jewelry. The patient should shampoo before the test, and must not use any hair spray, gel, or other hair care products after shampooing. Clothing should be loose and comfortable. The patient may eat and take some medications as usual before the test, although sedative medications should be avoided on the day of the test, if possible. It is best to check with the physician supervising the test for specific instructions.

### Aftercare

This test is painless and has no residual effects. The patient may return to work or other activities immediately afterward.

### Normal results

EP test results are displayed as jagged electrical tracings (wave forms), which have characteristic shapes, heights, and lengths, indicating the speed and intensity of signal transmission. Results are read by someone trained in evoked potential studies.

### Abnormal results

Changes in the electrical tracings may indicate damage to or degeneration of nerve pathways to the brain from the eyes, ears, or limbs. Absence of any activity may mean complete loss of nerve function in that pathway. Other changes may provide evidence of the type and location of nerve damage.

### Resources

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Richard Robinson

Evoked responses see **Evoked potential studies**

Exanthema subitum see **Roseola**

## Exercise

### Definition

Exercise is physical activity that is planned, structured, and repetitive for the purpose of conditioning any part of the body. Exercise is utilized to improve health, maintain fitness and is important as a means of physical **rehabilitation**.

### Purpose

Exercise is useful in preventing or treating coronary heart disease, **osteoporosis**, weakness, diabetes, **obesity**, and depression. Range of motion is one aspect of exercise important for increasing or maintaining joint function. Strengthening exercises provide appropriate resistance to the muscles to increase endurance and strength. **Cardiac rehabilitation** exercises are developed and individualized to improve the cardiovascular system for prevention and rehabilitation of cardiac disorders and diseases. A well-balanced exercise program can improve general health, build endurance, and delay many of the effects of **aging**. The benefits of exercise not only improve physical health, but also enhance emotional well-being.

### Precautions

Before beginning any exercise program, an evaluation by a physician is recommended to rule out any potential health risks. Once health and fitness are determined, and any or all physical restrictions identified, an individual's exercise program should be under the supervision of a health care professional. This is especially the case when exercise is used as a form of rehabilitation. If symptoms of **dizziness**, nausea, excessive **shortness of breath**, or **chest pain** are present during any exercise program, an individual should stop the activity and inform a physician about these symptoms before resuming activity. Exercise equipment must be checked to determine if it can bear the weight of people of all sizes and shapes.

### Description

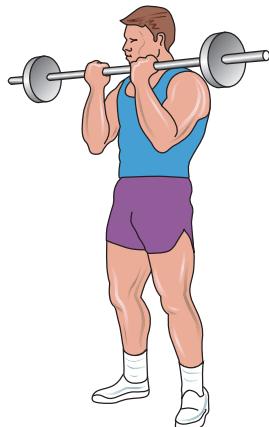
#### *Range of motion exercise*

Range of motion exercise refers to activity whose goal is improving movement of a specific joint. This

### THREE TYPES OF EXERCISE



Stretching, for flexibility



Weight-bearing, for strengthening muscles and bone mass



Aerobic, for the heart

**Exercise is utilized to improve health, maintain fitness, and is important as a means of physical rehabilitation.** (*Illustration by Electronic Illustrators Group.*)

motion is influenced by several structures: configuration of bone surfaces within the joint, joint capsule, ligaments, and muscles and tendons acting on the joint. There are three types of range of motion exercises: passive, active, and active assists. Passive range of motion is movement applied to a joint solely by another person or persons or a passive motion machine. When passive range of motion is applied, the joint of an individual receiving exercise is completely relaxed while the outside force moves the body part, such as a leg or arm, throughout the available range. Injury, surgery, or **immobilization** of a joint may affect the normal joint range of motion. Active range of motion is movement of a joint provided entirely by the individual performing the exercise. In this case, there is no outside force aiding in the movement. Active assist range of motion is described as a joint receiving partial assistance from an outside force. This range of motion may result from the majority of motion applied by an exerciser or by the person or persons assisting the individual. It may also be a half-and-half effort on the joint from each source.

#### **Strengthening exercise**

Strengthening exercise increases muscle strength and mass, bone strength, and the body's metabolism. It can help attain and maintain proper weight and improve body image and self-esteem. A certain level of muscle

strength is needed to do daily activities, such as walking, running and climbing stairs. Strengthening exercises increase this muscle strength by putting more strain on a muscle than it is normally accustomed to receiving. This increased load stimulates the growth of proteins inside each muscle cell that allow the muscle as a whole to contract. There is evidence indicating that strength training may be better than aerobic exercise alone for improving self-esteem and body image. Weight training allows one immediate feedback, through observation of progress in muscle growth and improved muscle tone. Strengthening exercise can take the form of isometric, isotonic and isokinetic strengthening.

**ISOMETRIC EXERCISE.** During isometric exercises, muscles contract. However, there is no motion in the affected joints. The muscle fibers maintain a constant length throughout the entire contraction. The exercises are usually performed against an immovable surface or object such as pressing one's hand against a wall. The muscles of the arm are contracting but the wall is not reacting or moving as a result of the physical effort. Isometric training is effective for developing total strength of a particular muscle or group of muscles. It is often used for rehabilitation since the exact area of muscle weakness can be isolated and strengthening can be administered at the proper joint angle. This kind of training can provide a relatively quick and convenient method

for overloading and strengthening muscles without any special equipment and with little chance of injury.

**ISOTONIC EXERCISE.** Isotonic exercise differs from isometric exercise in that there is movement of a joint during the muscle contraction. A classic example of an isotonic exercise is weight training with dumbbells and barbells. As the weight is lifted throughout the range of motion, the muscle shortens and lengthens. Calisthenics are also an example of isotonic exercise. These would include chin-ups, push-ups, and sit-ups, all of which use body weight as the resistance force.

**ISOKINETIC EXERCISE.** Isokinetic exercise utilizes machines that control the speed of contraction within the range of motion. Isokinetic exercise attempts to combine the best features of both isometrics and weight training. It provides muscular overload at a constant preset speed while a muscle mobilizes its force through the full range of motion. For example, an isokinetic stationary bicycle set at 90 revolutions per minute means that despite how hard and fast the exerciser works, the isokinetic properties of the bicycle will allow the exerciser to pedal only as fast as 90 revolutions per minute. Machines known as Cybex and Biodek provide isokinetic results; they are generally used by physical therapists and are not readily available to the general population.

### *Cardiac rehabilitation*

Exercise can be very helpful in prevention and rehabilitation of cardiac disorders and disease. With an individually designed exercise program set at a level considered safe for that individual, people with symptoms of **heart failure** can substantially improve their fitness levels. The greatest benefit occurs as muscles improve the efficiency of their oxygen use, which reduces the need for the heart to pump as much blood. While such exercise doesn't appear to improve the condition of the heart itself, the increased fitness level reduces the total workload of the heart. The related increase in endurance should also translate into a generally more active lifestyle. Endurance or aerobic routines, such as running, brisk walking, cycling, or swimming, increase the strength and efficiency of the muscles of the heart.

### **Preparation**

A **physical examination** by a physician is important to determine if strenuous exercise is appropriate or detrimental for an individual. Prior to the exercise program, proper stretching is important to prevent the possibility of soft tissue injury resulting from tight muscles, tendons, ligaments, and other joint-related structures.

## KEY TERMS

**Aerobic**—Exercise training that is geared to provide a sufficient cardiovascular overload to stimulate increases in cardiac output.

**Calisthenics**—Exercise involving free movement without the aid of equipment.

**Endurance**—The time limit of a person's ability to maintain either a specific force or power involving muscular contractions.

**Osteoporosis**—A disorder characterized by loss of calcium in the bone, leading to thinning of the bones. It occurs frequently in postmenopausal women.

### **Aftercare**

Proper cool down after exercise is important in reducing the occurrence of painful muscle spasms. It has been documented that proper cool down may also decrease frequency and intensity of muscle stiffness the day following any exercise program.

### **Risks**

Improper warm up can lead to muscle strains. Overexertion with not enough time between exercise sessions to recuperate can also lead to muscle strains, resulting in inactivity due to pain. **Stress fractures** are also a possibility if activities are strenuous over long periods of time without proper rest. Although exercise is safe for the majority of children and adults, there is still a need for further studies to identify potential risks.

### **Normal results**

Significant health benefits are obtained by including a moderate amount of physical exercise in the form of an exercise prescription. This is much like a drug prescription in that it also helps enhance the health of those who take it in the proper dosage. Physical activity plays a positive role in preventing disease and improving overall health status. People of all ages, both male and female, benefit from regular physical activity. Regular exercise also provides significant psychological benefits and improves quality of life.

### **Abnormal results**

There is a possibility of exercise burnout if an exercise program is not varied and adequate rest periods are

not taken between exercise sessions. Muscle, joint, and cardiac disorders have been noted among people who exercise. However, they often have had preexisting or underlying illnesses.

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### ORGANIZATIONS

- American Alliance for Health, Physical Education, Recreation & Dance. 1900 Association Dr., Reston, VA 20191-1598. (800) 213-7193. Fax: (703) 476-9527. <aahperd@aahperd.org>. <<http://www.aahperd.org>>.
- American College of Sports Medicine. 401 W. Michigan Street, Indianapolis, IN 46202-3233. (317) 637-9200. Fax: (317) 634-7817. <mkeckhaver@acsmed.org>. <<http://www.acsm.org>>.
- American Council on Exercise. 5820 Oberlin Drive, Suite 102, San Diego, CA 92121-3787. (800) 825-3636. Fax: (858) 535-1778. <<http://www.acefitness.org>>.
- American Medical Association. 515 N. State Street, Chicago, IL 60610. (312) 464-5000. <<http://www.ama-assn.org>>.
- American Physical Therapy Association. 1111 North Fairfax Street Alexandria, VA 22314. (703) 684-2782. <<http://www.apta.org>>.
- National Athletic Trainers' Association. 2952 Stemmons Freeway, Dallas, TX 75247-6916. (800) 879-6282. Fax: (214) 637-2206. <<http://www.nata.org>>.

## OTHER

- American Diabetes Association. <<http://www.diabetes.org/exercise/>>.
- American Heart Association. <<http://www.americanheart.org>>.
- American Orthopaedic Society for Sports Medicine. <<http://www.sportsmed.org>>.
- American Society of Exercise Physiologists. <<http://www.css.edu/asep/>>.

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Exercise electrocardiogram see **Stress test**

Exercise stress test see **Stress test**

Exhibitionism see **Sexual perversions**

Exocrine pancreatic cancer see **Pancreatic cancer, exocrine**

## Exophthalmos

### Definition

When there is an increase in the volume of the tissue behind the eyes, the eyes will appear to bulge out of the face. The terms exophthalmos and proptosis apply. Proptosis can refer to any organ that is displaced forward, while exophthalmos refers just to the eyes.

### Description

The eye socket (orbit) is made of bone and therefore will not yield to increased pressure within it. Only forward displacement of the eyeball (globe) will allow more room if tissue behind the eye is increasing.

### Causes and symptoms

The most common cause of exophthalmos is Graves' disease, overactivity of the thyroid gland. The contents of the orbits swell due to inflammation, forcing the eyes forward. The inflammation affects primarily the muscles. This combination of muscle impairment and forward displacement reduces eye movement, causing double vision and crossed eyes (**strabismus**). The optic nerves can also be affected, reducing vision, and the clear membrane (conjunctiva) covering the white part of the eyes and lining the inside of the eyelids can swell. Finally, the eyes may protrude so far that the eyelids cannot close over them, leading to corneal damage.

Exophthalmos from Graves' disease is bilateral (occurring on both sides), but not necessarily symmetri-



**A side view of the bulging eye (exophthalmos) of a person suffering from thyrotoxicosis. Exophthalmos is caused by swelling of the soft tissue in the eye socket, which forces the eyeball to be pushed forward and the eyelids stretched apart.** (Photograph by Dr. P. Marazzi, Photo Researchers, Inc. Reproduced by permission.)

cal. In contrast, exophthalmos from orbital tumors or a blood clot in the brain happens on only one side.

## Diagnosis

Exophthalmos is obvious when it is advanced enough to cause complications. When there is doubt in the early stages, a mechanical device called an exophthalmometer can measure the protrusion. **Computed tomography scans** (CT scans) are of great value in examining the bony components of the orbit. **Magnetic resonance imaging** (MRI) scanning is equally valuable for displaying the contents of the orbit, because it “sees through” the bone.

## Treatment

If a tumor is growing behind the eye, it needs to be removed. If Graves’ disease is the cause, it may subside with treatment of the overactive thyroid, but this is not guaranteed. Local care to the front of the eye to keep it moist is necessary if the eyelid cannot close.

## Prognosis

Exophthalmos can be progressive. Its progress must be carefully followed, treating complications as they occur.

## Prevention

Vision can usually be preserved with attentive treatment. There is currently no way to prevent any of the underlying conditions that lead to exophthalmos.

## KEY TERMS

**Conjunctivae**—The clear membranes that line the inside of the eyelids and cover the white part (sclera) of the eyeballs.

**Cornea**—The clear, dome-shaped part of the front of the eye, through which light first enters the eye. It is located in front of the colored part of the eye (iris).

**Inflammation**—The body’s reaction to invasion by foreign matter, particularly infection. The result is swelling and redness from an increase in water and blood, and pain from the chemical activity of the reaction.

**Strabismus**—Any deviation of the eyes from a common direction. Commonly called a turned eye.

**Thyroid**—A gland in the neck overlying the windpipe that regulates the speed of metabolic processes by producing a hormone, thyroxin.

## Resources

### BOOKS

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J. Ricker Polsdorfer, MD

## Expectorants

### Definition

Expectorants are drugs that loosen and clear mucus and phlegm from the respiratory tract.

### Purpose

The drug described here, guaifenesin, is a common ingredient in **cough** medicines. It is classified as an expectorant, a medicine that helps clear mucus and other secretions from the respiratory tract. However, some debate exists about how effectively guaifenesin does this. In addition, some cough medicines contain other ingredi-

## 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.
- Cough suppressant**—Medicine that stops or prevents coughing.
- 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.
- Phlegm**—Thick mucus produced in the air passages.
- Respiratory tract**—The air passages from the nose into the lungs.
- Secretion**—A substance, such as saliva or mucus, that is produced and given off by a cell or a gland.

ents that may cancel out guaifenesin's effects. **Cough suppressants** such as codeine, for example, work against guaifenesin because they discourage coughing up the secretions that the expectorant loosens.

There are other ways to loosen and clear the respiratory secretions associated with colds. These include using a humidifier and drinking six to eight glasses of water a day.

### Description

Guaifenesin is an ingredient in many cough medicines, such as Anti-Tuss, Dristan Cold & Cough, Guaifed, GuaiCough, and some Robitussin products. Some products that contain guaifenesin are available only with a physician's prescription; others can be bought without a prescription. They come in several forms, including capsules, tablets, and liquids.

### Recommended dosage

#### Adults and children 12 and over

200–400 mg every four hours. No more than 2,400 mg in 24 hours.

#### Children 6–11

100–200 mg every four hours. No more than 1,200 mg in 24 hours.

#### Children 2–5

50–100 mg every four hours. No more than 600 mg in 24 hours.

#### Children under two

Not recommended.

### Precautions

Do not take more than the recommended daily dosage of guaifenesin.

Guaifenesin is not meant to be used for coughs associated with **asthma**, **emphysema**, chronic **bronchitis**, or **smoking**. It also should not be used for coughs that are producing a large amount of 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.

Some studies suggest that guaifenesin causes **birth defects**. Women who are pregnant or plan to become pregnant should check with their physicians before using any products that contain guaifenesin. Whether guaifenesin passes into breast milk is not known, but no ill effects have been reported in nursing babies whose mothers used guaifenesin.

### Side effects

Side effects are rare, but may include vomiting, **diarrhea**, stomach upset, headache, skin rash, and **hives**.

### Interactions

Guaifenesin is not known to interact with any foods or other drugs. However, cough medicines that contain guaifenesin may contain other ingredients that do interact with foods or drugs. Check with a physician or pharmacist for details about specific products.

Nancy Ross-Flanigan

Exstrophy of the urinary bladder see  
**Congenital bladder anomalies**

External fetal monitoring see **Electronic fetal monitoring**

External otitis see **Otitis externa**

## External sphincter electromyography

### Definition

External sphincter **electromyography** helps physicians determine how well the external urinary sphincter muscle is working by measuring the electrical activity in it during contraction and relaxation.

### Purpose

The external sphincter muscle is the ring-like muscle that controls urine release from the bladder. When a patient cannot voluntarily control urination (incontinence), a physician may order this test to determine if the problem is caused by the failure of this muscle. The voluntary contraction or release of a muscle such as the external sphincter involves a complex process in which the nerves controlling the muscle signal it to move through the release and uptake of chemicals called neurotransmitters and the generation of electrical impulses. This test records the electrical impulses given off when the muscle contracts or relaxes and allows the physician to determine if the muscle is working properly, if it has been damaged by disease, or some other condition.

### Precautions

Patients who are taking **muscle relaxants** or drugs that act like or have an effect on the neurotransmitter acetylcholine (cholinergic or anti-cholinergic drugs) should tell the doctor since they will change the test results. The results will also be altered if the patient moves during the test or if the electrodes are improperly placed.

### Description

The patient puts on a surgical gown and lies down on the examining table. The procedure, which takes between 30–60 minutes, may be conducted one of three ways:

- Skin electrodes. This is the most commonly used method of recording information. The skin where the electrodes will be placed is cleaned and shaved and an electrically conductive paste is applied. The electrodes are then taped in place. For female patients, the electrodes are taped around the urethra, while for male patients they are placed between the scrotum and the anus.
- Needle electrodes. This is considered the most accurate method, since the electrodes are inserted directly into the muscle, using needles to guide placement. For male patients, a gloved finger is inserted in the rectum, then needles with wires attached are inserted through the skin between the anus and the scrotum. For female

### KEY TERMS

**Anti-cholinergic drug**—A medication that blocks or subdues the action of the neurotransmitter acetylcholine.

**Cholinergic drug**—A medication that mimics or enhances the action of the neurotransmitter acetylcholine.

**Sphincter**—A circular muscle that aids in the opening or closing of an opening in the body.

patients, the needles are inserted around the urethra. The discomfort of placing the needles is about the same as that of an injection. The needles are withdrawn, and the wires are taped to the thigh.

- Anal plug electrodes. The tip of an anal plug is lubricated and inserted into the rectum as the patient relaxes the anal sphincter. Electrodes are attached to the anal plug.

Once the electrodes are in place and attached to the recording device, the patient is asked to alternately contract and relax the external sphincter muscle. The electrical activity generated during these contractions and relaxations is recorded on a graph called an electromyogram.

### Preparation

Before the test, the patient should discuss with the doctor whether it is necessary to temporarily discontinue any medications, and follow the doctor's orders. No changes in diet or activity are necessary.

### Aftercare

Women may see some blood in their urine the first time they urinate after the test. Blood in the urine of men or blood in the urine of women after the first urination should be reported to the doctor. The patient should take a warm bath and drink plenty of fluids to ease any discomfort after the test.

### Risks

Complications of external sphincter electromyography are rare. Occasionally patients report blood in their urine after being tested with needle electrodes. Also, the urethra may become mildly irritated causing a change in the normal frequency of urination.

### Normal results

In a normally functioning external sphincter muscle, the electromyogram will show increased electrical activi-

ty when the patient tightens the muscle and a little or no electrical activity when it is relaxed.

### Abnormal results

A diseased external sphincter muscle will produce an abnormal pattern of electrical activity. Conditions that affect the external sphincter may include **multiple sclerosis**, **neurogenic bladder**, **Parkinson's disease**, **spinal cord injury**, and **stress incontinence**. However, additional tests must be done in order to confirm any of these diagnoses.

### Resources

#### BOOKS

"External Sphincter Electromyography." In *Illustrated Guide to Diagnostic Tests*, ed. J. A. Lewis. Springhouse, PA: Springhouse Corp. 1994.

Tish Davidson

## Extracorporeal membrane oxygenation

### Definition

Extracorporeal membrane oxygenation (ECMO) is a special procedure that uses an artificial heart-lung machine to take over the work of the lungs (and sometimes also the heart). ECMO is used most often in newborns and young children, but it also can be used as a last resort for adults whose heart or lungs are failing.

### Purpose

In newborns, ECMO is used to support or replace an infant's undeveloped or failing lungs by providing oxygen and removing carbon dioxide waste products so the lungs can rest. Infants who need ECMO may include those with:

- meconium aspiration syndrome (breathing in of a newborn's first stool by a fetus or newborn, which can block air passages and interfere with lung expansion)
- persistent **pulmonary hypertension** (a disorder in which the blood pressure in the arteries supplying the lungs is abnormally high)
- respiratory distress syndrome (a lung disorder usually of premature infants that causes increasing difficulty in breathing, leading to a life-threatening deficiency of oxygen in the blood)

- congenital diaphragmatic **hernia** (the profusion of part of the stomach through an opening in the diaphragm).
- pneumonia
- blood **poisoning**

ECMO is also used to support a child or adult patient's damaged, infected, or failing lungs for a few hours to allow treatment or healing. It is effective for those patients with severe, but reversible, heart or lung problems who haven't responded to treatment with a ventilator, drugs, or extra oxygen. Adults and children who need ECMO usually have one of these problems:

- heart failure
- pneumonia
- **respiratory failure** caused by trauma or severe infection

The ECMO procedure can help a patient's lungs and heart rest and recover, but it will not cure the underlying disease. Any patient who requires ECMO is seriously ill and will likely die without the treatment. Because there is some risk involved, this method is used only when other means of support have failed.

### Precautions

Typically, ECMO patients have daily chest x rays and blood work, and constant vital sign monitoring. They are usually placed on a special rotating bed that is designed to decrease pressure on the skin and help move secretions from the lungs.

After the patient is stable on ECMO, the breathing machine settings will be lowered to "rest" settings, which allows the lungs to rest without the risk of too much oxygen or pressure from the ventilator.

### Description

There are two types of ECMO: Venoarterial (V-A) ECMO supports the heart and lungs, and is used for patients with blood pressure or heart functioning problems in addition to respiratory problems. Venovenous (V-V) ECMO supports the lungs only.

V-A ECMO requires the insertion of two tubes, one in the jugular and one in the carotid artery. In the V-V ECMO procedure, the surgeon places a plastic tube into the jugular vein through a small incision in the neck.

Once in place, the tubes are connected to the ECMO circuit, and then the machine is turned on. The patient's blood flows out through the tube and may look very dark because it contains very little oxygen. A pump pushes the blood through an artificial membrane lung, where oxygen is added and carbon dioxide is removed. The size of the

artificial lung depends on the size of the patient; sometimes adults need two lungs. The blood is then warmed and returned to the patient. A steady amount of blood (called the flow rate) is pushed through the ECMO machine every minute. As the patient improves, the flow rate is lowered.

Many patients require heavy **sedation** while they are on ECMO to lessen the amount of oxygen needed by the muscles.

As the patient improves, the amount of ECMO support will be decreased gradually, until the machine is turned off for a brief trial period. If the patient does well without ECMO, the treatment is stopped.

Typically, newborns remain on ECMO for three to seven days, although some babies need more time (especially if they have a diaphragmatic hernia). Once the baby is off ECMO, he or she will still need a ventilator (breathing machine) for a few days or weeks. Adults may remain on ECMO for days to weeks, depending on the condition of the patient, but treatment may be continued for a longer time depending on the type of heart or lung disease, the amount of damage to the lungs before ECMO was begun, and the presence of any other illnesses or health problems.

## Preparation

Before ECMO is begun, the patient receives medication to ease **pain** and restrict movement.

## Aftercare

Because infants on ECMO may have been struggling with low oxygen levels before treatment, they may be at higher risk for developmental problems. They will need to be monitored as they grow.

## Risks

Bleeding is the biggest risk for ECMO patients, since blood thinners are given to guard against blood clots. Bleeding can occur anywhere in the body, but is most serious when it occurs in the brain. This is why doctors periodically perform ultrasound brain scans of anyone on ECMO. **Stroke**, which may be caused by bleeding or blood clots in the brain, has occurred in some patients undergoing ECMO.

If bleeding becomes a problem, the patient may require frequent blood transfusions or operations to control the bleeding. If the bleeding can't be stopped, ECMO will be withdrawn.

Other risks include infection or vocal cord injury. Some patients develop severe blood infections that cause irreversible damage to vital organs.

## KEY TERMS

**Carotid artery**—Two main arteries (passageway carrying blood from the heart to other parts of the body) that carry blood to the brain.

**Congenital diaphragmatic hernia**—The profusion of part of the stomach through an opening in the diaphragm.

**Meconium aspiration syndrome**—Breathing in of meconium (a newborn's first stool) by a fetus or newborn, which can block air passages and interfere with lung expansion.

**Membrane oxygenator**—The artificial lung that adds oxygen and removes carbon dioxide.

**Pulmonary hypertension**—A disorder in which the blood pressure in the arteries supplying the lungs is abnormally high.

**Respiratory distress syndrome**—A lung disorder usually of premature infants that causes increasing difficulty in breathing, leading to a life-threatening deficiency of oxygen in the blood.

**Venoarterial (V-A) bypass**—The type of ECMO that provides both heart and lung support, using two tubes (one in the jugular vein and one in the carotid artery).

**Venovenous (V-V) bypass**—The type of ECMO that provides lung support only, using a tube inserted into the jugular vein.

There is a small chance that some part of the complex equipment may fail, which could introduce air into the system or affect the patient's blood levels, causing damage or **death** of vital organs (including the brain). For this reason, the ECMO circuit is constantly monitored by a trained technologist.

## Normal results

Lungs and/or heart return to healthy functioning.

## Abnormal results

Lungs and/or heart do not improve while on ECMO.

## Resources

### PERIODICALS

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- American Society of Extra-Corporeal Technology. 11480 Sunset Hills Rd., Ste. 210E, Reston, VA 20190. (703) 435-8556. <<http://www.amsect.org>>.
- ECMO Moms and Dads. PO Box 53848, Lubbock, TX 79543. (806) 794-0259.
- Extracorporeal Life Support Organization. 1327 Jones Dr., Ste. 101, Ann Arbor, MI 48105. (734) 998-6600. <<http://www.elso.med.umich.edu>>.

Carol A. Turkington

Extracorporeal shock-wave see **Lithotripsy**

Extrinsic allergic alveolitis see  
**Hypersensitivity pneumonitis**

Eye and orbit sonograms see **Eye and orbit ultrasounds**

deeply lodged irritant (foreign body), or the effects of a previously unrecognized injury. When presented with general symptoms, ultrasound can speed diagnosis if a serious condition is suspected.

- Impaired vision. Fuzzy vision, poor night vision, restricted (tunnel) vision, blind spots, extreme light sensitivity, and even blindness can all stem from inner eye conditions ranging from **glaucoma** and **cataracts**, to retinitis, detached retina, tumors, or impaired blood circulation. Again, high resolution ultrasound can quickly identify causes and pinpoint their location. A special type of ultrasound, known as Doppler, can even perceive and measure circulation in the tiny blood vessels of the eye.
- Eye trauma. The eye can be damaged by a direct impact or a puncture wound, as a result of a general head trauma, or by intense light exposure. Even when the cause of injury is obvious, ultrasound can reveal the exact type, extent, and location of damage, from deformations and ruptures to internal bleeding, and help to guide emergency care efforts.
- Lens replacement surgery. Exact measurement of the eye's optical dimensions with ultrasound greatly improves the visual outcome for cataract patients receiving permanent synthetic lenses; and for severely myopic patients receiving implanted corrective lenses.

Ophthalmic ultrasound imaging is also used routinely to guide the precise placement of instruments during surgery, and can be used directly for the treatment of glaucoma and tumors of the eye.

#### Precautions

Ultrasound of the eye, properly performed by qualified personnel using appropriate equipment, has no risks. There is no evidence to suggest that the procedure itself poses any threat to a healthy eye, or worsens the condition of a diseased or injured eye.

#### Description

Ophthalmic ultrasound equipment sends high frequency pulses of sound into the eye, where they bounce off the boundaries between different structures in the eye and produce a distinctive pattern of echoes. This echo pattern is received and interpreted by a computer to produce an image on a television screen. The time it takes an echo to return to the receiver corresponds to the depth it traveled into the eye.

Single transducer (the sound transmitter/receiver) ultrasound is used to measure distances within the eye. This is A-mode ultrasound. A linear array of transduc-

#### Purpose

An ophthalmologist uses ultrasonic imaging to help diagnose the underlying cause(s) of a patient's symptoms, to assess the general condition of an injured eye, and to measure the eye prior to corrective surgery. Situations that may call for ultrasonic imaging include:

- Excessive tearing or visible infection. These external symptoms could indicate a serious underlying problem such as a tumor, an internal infection, the presence of a

ers in a single small probe, B-mode, provides a picture of a cross section through the eye. Doppler mode ultrasound combines B-mode with the ability to detect and measure the flow of blood in the tiny vessels of the eye.

As a direct treatment tool, the vibrations of high intensity A-mode ultrasound can be used to heat and erode tumors. The same technique can be used to control glaucoma by selectively destroying the cells which produce the fluid that causes the internal pressure of the eye to rise.

The procedure followed in a regular ultrasonic **eye examination** is relatively simple. The patient relaxes in a comfortable chair in a darkened room. Mild anesthetic eye drops are administered and the head is held secure. The ultrasonic probe, coated with a sterile gel to ensure good contact, is lightly pressed against the eye as the images are made. The probe may be applied to the eyelid or directly to the eye, as necessary. The patient feels nothing else, and the whole office procedure takes about 15 minutes.

## Preparation

Preparation by the patient is generally unnecessary, although under special circumstances an ophthalmologist may perform pretest procedures. The ophthalmologist and/or ultrasound technician will conduct all preparations at the time of the test.

## Aftercare

Patients may experience partial and temporary blurred vision, as well as "eye strain" headaches. These symptoms usually fade within an hour of the procedure, during which time patients should rest their eyes and avoid all activities that require good eyesight, like driving.

## Risks

Improperly focused, high-intensity ultrasound could burn and physically disrupt delicate eye tissue and cause injury. This risk is, however, slight and would arise only from improper use, or as a potential side effect of tumor or glaucoma treatment.

## Normal results

A normal ultrasound scan would indicate a fully healthy eye. For therapeutic ultrasound, a normal result would be an improvement in the targeted condition, such as shrinking of a tumor or lessening of pressure inside the eye of a glaucoma patient.

## KEY TERMS

**Cataracts**—A clouding of the lens of the eye or the material immediately surrounding it, causing blurred vision. For many people it occurs naturally with aging, but may also result from injury.

**Glaucoma**—A common eye disease characterized by increased fluid pressure in the eye that damages the optic nerve, which carries sensations to the brain. Glaucoma can be caused by another eye disorder, such as a tumor or congenital malformation, or appear without obvious cause, but if untreated it generally leads to blindness.

**Intraocular**—Literally, within the eye.

**Ophthalmologist**—A medical doctor specializing in eye care who is generally, but not necessarily, an eye surgeon.

**Retina**—The third and innermost membrane of the eye, which contains the light-sensitive nerve tissue that leads into the optic nerve and is the primary instrument of vision. Inflammation of the retina (retinitis) has many causes, including over-exposure to intense light, diabetes, and syphilis.

## Abnormal results

Because diagnostic ultrasound is generally used to investigate symptoms, the results of a scan will often be abnormal and they will detect evidence of an underlying condition.

## Resources

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

American Institute of Ultrasound in Medicine. 14750 Sweitzer Lane, Suite 100, Laurel, MD 20707-5906. (800) 638-5352. <<http://www.aium.org>>.

National Eye Institute. 2020 Vision Place, Bethesda, MD 20892-3655. (301) 496-5248. <<http://www.nei.nih.gov>>.

Kurt Richard Sternlof

# Eye cancer

### Definition

A cancerous growth in any part of the eye.

### Description

**Eye cancer** can occur in many parts of the eye where a tumor can occur. Because of this there are several types of ocular cancer. Their occurrence varies in the age of the affected individual. This article will focus on **retinoblastoma**, the most common eye cancer in children, and intraocular melanoma, the most common eye cancer in adults.

Retinoblastoma can occur at any age but is most often seen in children younger than five. About 200 children a year are diagnosed with it in the United States. Retinoblastoma starts with a small tumor in the retina, the very back of the eye. In growing children, the retina originates from cells called retinoblasts that grow and divide very quickly. These cells eventually become the mature cells of the retina when they stop growing. In the case of retinoblastoma the retinoblasts don't stop growing and form a tumor that can continue to grow and cause further complications if not treated quickly.

Retinoblastoma typically has three classifications: intraocular, extraocular and recurrent retinoblastoma. In the intraocular form the cancer can be found in one or both eyes but not in tissue external of the eye. In the extraocular form the cancer has spread outside the eye. It can spread to the tissue surrounding the eye or it can invade other areas of the body. In the recurrent form the cancer returns after already being treated. It may recur in the eye, its surrounding tissues, or elsewhere in the body.

Intraocular melanoma is a rare cancer overall, yet it is the most common eye cancer seen in adults. It is when cancer cells are found in the uvea of the eye. The uvea includes the iris (the colored portion of eye), the ciliary body (an eye muscle that focuses the lens) and the choroid (found in the back of the eye next to the retina).

Intraocular cancer of the iris usually grows slowly and usually doesn't spread. The tumor is seen on the iris as a darker spot than the surrounding area. Intraocular cancer of the choroid or ciliary body occurs in the back of the eye. They are classified by size with a small tumor being 2-3 mm or smaller and a medium or large tumor being bigger than 3 mm.

Intraocular cancer can spread and become extraocular as well. If not found and treated early enough it can spread to the surrounding tissues, the optic nerve or into the eye socket.

### Causes and symptoms

Genetics is thought to play a role in eye cancer. In regards to retinoblastoma, it is believed that if a tumor develops only in one eye then it isn't hereditary. However, if a tumor occurs in both eyes then it is hereditary. Those who have hereditary retinoblastoma have a rare risk of developing a tumor in the brain and should be monitored on a regular basis.

The cause of intraocular melanoma is still vague. Genetics could play a role, but age is also a factor. Interestingly enough, this type of cancer is seen most often in white people from a northern European descent.

The symptoms of this type of cancer usually begin with blurred vision and tenderness of the eye. Advanced symptoms may include loss of vision. If these symptoms persist a person should make an appointment with their ophthalmologist.

### Diagnosis

An ophthalmologist makes a diagnosis. The doctor is usually able to see the tumor through the pupil or directly on the iris if the cancer is intraocular melanoma of the iris. Because the doctor can usually readily see the tumor a biopsy is rarely needed.

An ultrasound or a fluorescein **angiography** are two tests doctors use to further diagnose eye cancers. In an ultrasound sound waves are pointed at the tumor and depending on how they reflect off the tumor the doctor can better diagnose it. In a fluorescein angiography a fluorescent dye is injected into the patients arm. When this dye circulates through the body and reaches the eye a series of rapid pictures are taken through the pupil. The tumor will show up in these photos.

Once a diagnosis has been made, the treatment can begin.

### Treatment

The treatment depends on how far advanced the tumor is. If the tumor is in the advanced stages and there is little hope of regaining vision the most effective treat-

ment is an enucleation, the removal of the eye. This obviously is a drastic treatment and is avoided if possible. Other eye surgeries include the following:

- choroidectomy—removal of part of the choroid,
- iridectomy—removal of part of the iris,
- iridocyclectomy—removal of parts of the ciliary body and parts of iris,
- iridotrabeculectomy—removal of parts of the supporting tissues around the cornea and iris.

In eye cancer where the tumor is small and there is a good chance that the vision will be restored less drastic measures than the above surgeries are taken. Radiation and **chemotherapy** are two courses of treatment that help in killing off the existing tumor and preventing its spread into other areas of the body.

Besides radiation and chemotherapy there are other methods of treating eye cancer. **Cryotherapy** uses extreme cold to destroy the cancer cells. Thermotherapy uses heat to destroy the cancer cells. Photocoagulation uses a laser to destroy blood vessels that supply the tumor with nutrients. If the tumor isn't advanced these are good options to treat it in order to avoid losing an eye.

A radiation/surgical treatment for eye cancer is brachytherapy. A small plaque with radioactive iodine on one side and gold on the other is stitched to the eye behind the tumor with the radioactive iodine facing the tumor. The gold is used to shield the other tissues from the radiation. It is left there for a period of time depending on the dosage of radiation needed and then it is removed. In this way the tumor is treated and hopefully will shrink and eventually die.

### Alternative Treatment

Other than the treatments above, there aren't any alternative treatments. New clinical trials are constantly under way to further the treatment of the disease in the future.

### Prognosis

All forms of retinoblastoma and intraocular melanoma are treatable. Enucleation can usually be avoided if found early enough. The outlook is positive for people with eye cancer.

### Prevention

A good healthy diet and lifestyle are always recommended to prevent cancer. Known carcinogens should always be avoided.

## KEY TERMS

**Carcinogen**—A substance that is known to cause cancer.

**Cornea**—The clear layer that covers the front part of the eye.

**Enucleation**—Surgical removal of the eye.

**Pupil**—The hole in the eye that allows light in.

## Resources

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## Eye examination

### Definition

An eye examination is a series of tests that measure a person's ocular health and visual status, to detect abnormalities in the components of the visual system, and to determine how well the person can see.

### Purpose

An eye examination is performed by an ophthalmologist M.D. or D.O. (doctor of **osteopathy**), or an optometrist (O.D.) to determine if there are any pre-existing or potential vision problems. Eye exams may also reveal the presence of many non-eye diseases. Many systemic diseases can affect the eyes, and since the blood vessels on the retina are observed during the exam, certain problems may be uncovered (e.g., high blood pressure or diabetes).

Infants should be examined by a physician to detect any physical abnormalities. Frequency of eye exams then generally differs with age and the health of the person. Eye exams can be performed in infants, and if a problem is

noted the infant can be seen, generally by a pediatric ophthalmologist. A child with no symptoms should have an eye exam at age three. Early exams are important because permanent decreases in vision (e.g., **amblyopia**, also called lazy eye) can occur if not treated early (usually by ages 6–9). Again, with no other symptoms, the second exam should take place before first grade. After first grade, the American Optometric Association recommends an eye exam every two years; ages 19–40, every two to three years; ages 41–60, every two years; and annually after that. However, these are recommendations for healthy people with no risk factors. Patients should ask their doctors how often they should come for exams. Some patients have risk factors for eye disease (e.g., people with diabetes or a family history of eye disease; African Americans, who are at higher risk for **glaucoma**) and may need more frequent checkups. Also, if children seem to be having trouble in school, problems with reading, rubbing their eyes when reading, etc., an eye exam may be necessary sooner.

### Precautions

The examiner needs to know if the patient is taking any medications or has any existing health conditions. Some medications, even over-the-counter (OTC) medications can affect vision or even interfere with the eyedrops the doctor may use during the exam. Certain eyedrops would not be used if the patient has **asthma**, heart problems, or other conditions.

The patient may need someone to drive them home in case the eyes were dilated. Bringing sunglasses to the exam may also help decrease the glare from light until the dilating drops wear off.

### Description

An eye examination, given by an ophthalmologist or optometrist, costs about \$100. It may or may not be covered by insurance. It begins with information from the patient (case history) and continues with a set of primary tests, plus additional specialized tests given as needed, dictated by the outcomes of initial testing and the patient's age. The primary tests can be divided into two groups, those that evaluate the physical state of the eyes and surrounding areas, and those that measure the ability to see.

The order of the tests for the exam may differ from doctor to doctor, however, most exams will include the following procedures:

#### *Information gathering and initial observations*

The examiner will take eye and medical histories that include the patient's chief complaint, any past eye disor-

ders, all medications being taken (e.g., OTC medications, **antibiotics**, and birth control pills), any blood relatives with eye disorders, and any systemic disorders the patient may have. The patient should also tell the doctor about hobbies and work conditions. This information helps in modifying prescriptions and lets the doctor know how the patient uses his or her eyes. For example, using a computer screen vs. construction work, the working distance of a computer screen may affect the prescription; the construction worker needs protective eyewear.

The patient should bring their current pair of glasses to the exam. The doctor can get the prescription from the glasses by using an instrument called a lensometer.

#### *Visual acuity examination*

Visual acuity measures how clearly the patient can see. It is measured for each eye separately, with and without the current prescription. It is usually measured with a Snellen eye chart, a poster with lines of different-sized letters, each line with a number at the side denoting the distance from which a person with normal vision can read that line. Other kinds of eye charts with identifiable figures are available for children or anyone unfamiliar with the Roman alphabet. These charts are made to be placed at a certain distance (usually 20 ft) from the person being tested. At this distance, people with normal vision can read a certain line (usually the lowest), marked the 20/20 line; these people are said to have 20/20 vision. For people who can't read the smallest line, the examiner assigns a ratio based on the smallest line they can read. The first number (numerator) of the ratio is the distance between the chart and the patient, and the second number (denominator) is the distance where a person with normal vision would be able to read that line. The ratio 20/40 means the patient can see at 20 ft. what people with normal vision can see at 40 ft. away.

When a patient is unable to read any lines on the chart, they are moved closer until they can read the line with the largest letters. The acuity is still measured the same way. A ratio of 5/200 means the person being tested can see at 5 ft what a normal person can see 200 ft.

When a patient can't read the chart at all, the examiner may hold up some fingers and ask the patient to count them at various distances, and records the result as "counting fingers" at the distance of recognition. If the patient cannot count the examiner's fingers at any distance, the examiner determines if the patient can see hand movements. If so, the result is recorded as "hand movements." If not, the examiner determines if the patient can detect light from a penlight. If the patient can detect the light but not its direction, the result is recorded as "light perception." If the patient can recognize its

direction, the result is recorded as “light projection.” If the patient cannot detect the light at all, the result is recorded as “no light perception.”

#### ***Eye movement examination and cover tests***

The examiner asks the patient to look up and down, and to the right and left to see if the patient can move the eyes to their full extent. The examiner asks the patient to stare at an object, then quickly covers one eye and notes any movement in the eye that remains uncovered. This procedure is repeated with the other eye. This, and another similar cover test, helps to determine if there is an undetected eye turn or problem with fixation. The doctor may also have the patient look at a pen and follow it as it is moved close to the eyes. This checks convergence.

#### ***Iris and pupil examination***

The doctor checks the pupil’s response to light (if it dilates and constricts appropriately). The iris is viewed for symmetry and physical appearance. The iris is checked more thoroughly later using a slit lamp.

#### ***Refractive error determination-Refraction***

The examiner will determine the refractive error and obtain a prescription for corrective lenses for people whose visual acuity is less than 20/20. An instrument called a phoropter, which the patient sits behind, is generally used (sometimes the refraction can be done with a trial frame that the patient wears). The phoropter is equipped with many lenses that allow the examiner to test many combinations of corrections to learn which correction allows the patient to see the eye chart most clearly. This is the part of the exam when the doctor usually says, “Which is better, one or two?” The phoropter also contains prisms, and sometimes the doctor will intentionally make the patient see double. This may help in determining a slight eye turn. The exam will check vision at distance and near (reading).

A prescription for corrective lenses can also be supplied by automated refracting devices, which measure the necessary refraction by shining a light into the eye and observing the reflected light. Another objective way to obtain a prescription is using a hand-held retinoscope. As in the automated method just mentioned, the doctor shines a light in the patient’s eyes and can determine an objective prescription. This is helpful in young children or infants.

Sometimes drops will be instilled in the patient’s eyes before this part of the exam. The drops may relax accommodation so that the refraction will be more accurate. This is helpful in children and people who are farsighted.



**A woman looking through a refractor.** (Photograph by John Greim, Photo Researchers. Reproduced by permission.)

After the refraction and other visual status tests, for example color tests or binocularity tests (can the patient see 3-D, or have depth perception), the doctor will check the health of the eyes and surrounding areas. The main instruments used are the ophthalmoscope and the slit lamp.

#### ***Ophthalmoscopic examination***

These observations are best accomplished after dilating the pupils and require an ophthalmoscope. The ophthalmoscope most frequently used is a called a *direct ophthalmoscope*. It is a hand-held illuminated 15X multi-lens magnifier that lets the examiner view the inside back area of the eye (fundus). The retina, blood vessels, optic nerve, and other structures are examined.

#### ***Slit lamp examination***

The slit lamp is a microscope with a light source that can be adjusted. This magnifies the external and some internal structures of the eyes. The lid and lid margin, cornea, iris, pupil, conjunctiva, sclera, and lens are examined. The slit lamp is also used in contact lens evaluations. A little probe called a tonometer may be used at this time to check the pressure of the eyes. A colored eyedrop may be instilled immediately prior to this test. The drop has a local anesthetic so the patient won’t feel the probe touch the eye. It is a quick procedure.

#### ***Visual field measurement***

A perimeter, the instrument for measuring visual fields, is a hollow hemisphere, equipped with a light source that projects dots of light over the inside surface. The patient’s head is positioned so that the eye being tested is at the center of the sphere and 13 in (about 33

## KEY TERMS

**Amblyopia**—Decreased visual acuity, usually in one eye, in the absence of any structural abnormality in the eye.

**Conjunctiva**—The mucous membrane that covers the white part of the eyes (sclera) and lines the eyelids.

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

**Floatters**—Translucent specks that float across the visual field, due to small objects floating in the vitreous humor.

**Fundus**—The inside of an organ. In the eye, refers to the back area that can be seen with the ophthalmoscope.

**Glaucoma**—There are many types of glaucoma. Glaucoma results in optic nerve damage and a decreased visual field and blindness if not treated. It is usually associated with increased IOP, but that is not always the case. The three factors associated with glaucoma are increased IOP, a change in the optic nerve head, and changes in the visual field.

**Gonioscope**—An instrument used to inspect the eye (e.g., the anterior chamber). It consists of a magnifier and a lens equipped with mirrors; it's placed on the patient's cornea.

**Iris**—The colored ring just behind the cornea and in front of the lens that controls the amount of light sent to the retina.

**Macula**—The central part of the retina where the rods and cones are densest.

**Ophthalmoscope**—An instrument designed to view structures in the back of the eye.

**Optic nerve**—The nerve that carries visual messages from the retina to the brain.

**Pupil**—The circular opening that looks like a black hole in the middle of the iris.

**Retina**—The inner, light-sensitive layer of the eye containing rods and cones; transforms the image it receives into electrical messages which are then sent to the brain via the optic nerve.

**Sclera**—The tough, fibrous, white outer protective covering that surrounds the eye.

**Slit lamp**—A microscope that projects a linear slit beam of light onto the eye; allows viewing of the conjunctiva, cornea, iris, aqueous humor, lens, and eyelid.

**Tonometer**—An instrument that measures intraocular pressure (IOP).

**Ultrasonography**—A method of obtaining structural information about internal tissues and organs where an image is produced because different tissues bounce back ultrasonic waves differently.

cm) from all points on the inside surface of the hemisphere. The patient stares straight ahead at an image on the center of the surface and signals whenever he or she detects a flash of light. The perimeter records which flashes are seen and which are missed and maps the patient's field of vision and blindspots.

### Intraocular pressure (IOP) measurement

Tonometers are used to measure IOP. Some tonometers measure pressure by expelling a puff of air (noncontact tonometer) towards the eyeball from a very short distance. Other tonometers are placed directly on the cornea. The noncontact tonometers are not as accurate as the contact tonometers and are sometimes used for screenings.

### Completing the evaluation with additional tests

Depending upon the results other tests may be necessary. These can include, but are not limited to

binocular indirect ophthalmoscopy, gonioscopy, color tests, contrast sensitivity testing, ultasonography, and others. The patient may have to return for additional visits.

## Results

### External observations

#### INITIAL OBSERVATIONS AND SLIT LAMP EXAM.

Some general observations the doctor may be looking for include: head tilt; drooping eyelids (**ptosis**); eye turns; red eyes (injection); eye movement; size, shape, and color of the iris; clarity of the cornea, anterior chamber, and lens. The anterior chamber lies behind the cornea and in front of the iris. If it appears cloudy or if cells can be seen in it during the slit lamp exam an inflammation may be present. A narrow anterior chamber may put the patient at risk for glaucoma. A clouding of the normally clear lens is called a cataract.

### **Internal observations**

**OPHTHALMOSCOPIC EXAM.** The observations include, but are not limited to the retina, blood vessels, and optic nerve. The optic nerve enters the back of the eye and can be checked for swelling or other problems. The blood vessels can be viewed as can the retina. The macula is a 3–5 mm area in the back of the eye and is responsible for central vision. The fovea is a small area located within the macula and is responsible for sharp vision. When a person looks at something, they are pointing the fovea at the object. Changes in the macular area can be observed with the ophthalmoscope. Retinal tears or detachments can also be seen.

### **Visual ability**

**VISUAL ACUITY.** The refraction will determine the refractive status for each eye for distance and for near. A prescription for glasses is made after taking many things into consideration. The eye doctor may alter a prescription based upon many factors. Different materials for glasses may be suggested. For example, polycarbonate may be suggested for children or people active in sports because it is very impact resistant. Bifocals, trifocals, single-vision spectacles, and contact lenses are also options.

**VISUAL FIELDS.** A normal visual field extends about 60° upward, about 75° downward, about 65° toward the nose, and about 100° toward the ear and has one blind spot close to the center. Defects in the visual field signify damage to the retina, optic nerve, or the neurological visual pathway.

Seeing clearly does not necessarily mean the eyes are healthy or that the eyes are working together as a team. Regular checkups can detect abnormalities, hopefully before a problem arises. The eye doctor can suggest ways to help protect the eyes and vision (e.g., safety goggles, ultraviolet (UV) coatings on lenses). A person should also have an eye exam if they notice a change in vision, eyestrain, blur, flashes of light, a sudden onset of floaters (little dots), distortion of objects, double vision, redness, **pain** or discharge.

### **Resources**

#### **BOOKS**

Chang, David F. "Ophthalmologic Examination." In *General Ophthalmology*. 14th ed. Ed. Daniel Vaughan. Stamford: Appleton & Lange, 1995.

#### **ORGANIZATIONS**

American Academy of Ophthalmology. 655 Beach Street, PO 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>>.

Lorraine Lica, PhD

Eye exercises see **Vision training**

## **Eye glasses and contact lenses**

### **Definition**

Eyeglasses and contact lenses are devices that correct refractive errors in vision. Eyeglass lenses are mounted in frames worn on the face, sitting mostly on the ears and nose, so that the lenses are positioned in front of the eyes. Contact lenses appear to be worn in direct contact with the cornea, but they actually float on a layer of tears that separates them from the cornea.

### **Purpose**

The purpose of eyeglasses and contact lenses is to correct or improve the vision of people with nearsightedness (**myopia**), farsightedness (**hyperopia**), **presbyopia**, and **astigmatism**.

### **Precautions**

People allergic to certain plastics should not wear contact lenses or eyeglass frames or lenses manufactured from that type of plastic. People allergic to nickel should not wear Flexon frames. People at risk of being in accidents that might shatter glass lenses should wear plastic lenses, preferably polycarbonate. (Lenses made from polycarbonate, the same type of plastic used for the space shuttle windshield, are about 50 times stronger than other lens materials.) Also, people at risk of receiving electric shock should avoid metal frames.

People employed in certain occupations may be prohibited from wearing contact lenses, or may be required to wear safety eyewear over the contact lenses. Some occupations, such as construction or auto repair, may require safety lenses and safety frames. Physicians and employers should be consulted for recommendations.

### **Description**

Eyes are examined by optometrists (O.D.) or by ophthalmologists (M.D. or D.O.—doctor of **osteopathy**). Prescriptions, if necessary, are then given to patients for

glasses. The glasses are generally made by an optician. A separate contact lens-fitting exam is necessary if the patient wants contact lenses, because an eyeglass prescription is not the same as a contact lens prescription.

### Eyeglasses

Over 140 million people in the United States wear eyeglasses. People whose eyes have refractive errors do not see clearly without glasses, because the light emitted from the objects they are observing does not come into focus on their retinas. For people who are farsighted, images come into focus behind the retina; for people who are nearsighted, images come into focus in front of the retina.

**LENSES.** Lenses work by changing the direction of light so that images come into focus on the retina. The greater the index of refraction of the lens material and the greater the difference in the curvature between the two surfaces of the lens, the greater the change in direction of light that passes through it, and the greater the correction.

Lenses can be unifocal, with one correction for all distances, or they can be correct for more than one distance (multifocal). One type of multifocal, the bifocal, has an area of the lens (usually at the bottom) that corrects for nearby objects (about 14 in from the eyes); the remainder of the lens corrects for distant objects (about 20 ft from the eyes). Another type of multifocal, a trifocal, has an area in-between that corrects for intermediate distances (usually about 28 in). Conventional bifocals and trifocals have visible lines between the areas of different correction; however, lenses where the correction gradually changes from one area to the other, without visible lines, have been available since the 1970s. Such lenses are sometimes called progressives or no-line bifocals.

To be suitable for eyeglass lenses, a material must be transparent, without bubbles, and have a high index of refraction. The greater the index of refraction, the thinner the lens can be. Lenses are made from either glass or plastic (hard resin). The advantage of plastic is that it is lightweight and more impact resistant than glass. The advantage of glass is that it is scratch resistant and provides the clearest possible vision.

Glass was the first material to be used for eyeglass lenses, and was used for several hundred years before plastic was introduced. The crown glass used for eyeglass lenses has an index of refraction of 1.52.

Optical-quality acrylic was introduced for eyeglass use in the early 1940s, but because it was easily scratched, brittle, and discolored rapidly, it did not supplant glass as the material of choice. Furthermore, it had a relatively low index of refraction, so it wasn't suitable

for people with large refractive errors. A plastic called CR-39, introduced in the 1960s, was more suitable. Today, eyeglass wearers can also choose between polycarbonate, which is the most impact-resistant material available for eyewear, and polyurethane, which has exceptional optical qualities and an index of refraction of up to 1.66, much higher than the conventional plastics used for lenses, and even higher than glass. Patients with high prescriptions should ask about high index material options for their lenses. Aspheric lenses are also useful for high prescriptions. They are flatter and lighter than conventional lenses.

There are many lenses and lens-coating options for individual needs, including coatings that block the ultraviolet (UV) light or UV and blue light which have been found to be harmful to the eyes. Such coatings are not needed on polycarbonate lenses, which already have UV protection. UV coatings are particularly important on sunglasses and ski goggles. Sunglasses, when nonprescription, should be labeled with an indication that they block out 99–100% of both UV-A and UV-B rays.

There are anti-scratch coatings that increase the surface hardness of lenses (an important feature when using plastic lenses) and anti-reflective (AR) coatings that eliminate almost all glare and allow other people to see the eyes of the wearer. AR coatings may be particularly helpful to people who use computers or who drive at night. Mirror coatings that prevent other people from seeing the wearer's eyes are also available. There is a whole spectrum of tints, from light tints to darker tints, used in sunglasses. Tint, however, does not block-out UV rays, so a UV coating is needed. Polaroid lenses that block out much of the reflected light also allow better vision in sunny weather and are helpful for people who enjoy boating. Photosensitive (photochromatic) lenses that darken in the presence of bright light are handy for people who don't want to carry an extra set of glasses. Photochromatic lenses are available in glass and plastic.

**FRAMES.** Frames can be made from metal or plastic, and they can be rimless. There is an almost unlimited variety of shapes, colors, and sizes. The type and degree of refractive correction in the lens determine to some extent the type of frame most suitable. Some lenses are too thick to fit in metal rims, and some large-correction prescriptions are best suited to frames with small-area lenses.

Rimless frames are the least noticeable type, and they are lightweight because the nosepiece and temples are attached directly to the lenses, eliminating the weight of the rims. They tend to not be as sturdy as frames with rims, so they are not a good choice for people who frequently

remove their glasses and put them on again. They are also not very suitable for lenses that correct a high degree of farsightedness, because such lenses are thin at the edges.

Metal frames are less noticeable than plastic, and they are lightweight. They are available in solid gold, gold-filled, anodized aluminum, nickel, silver, stainless steel, and now titanium and titanium alloy. Until the late 1980s, when titanium-nickel alloy and titanium frames were introduced, metal frames were, in general, more fragile than plastic frames. The titanium frames, however, are very strong and lightweight. An alloy of titanium and nickel, called Flexon, is not only strong and lightweight, but returns to its original shape after being twisted or dented. It is not perfect for everyone, though, because some people are sensitive to its nickel. Flexon frames are also relatively expensive.

Plastic frames are durable, can accommodate just about any lens prescription, and are available in a wide range of prices. They are also offered in a variety of plastics (including acrylic, epoxy, cellulose acetate, cellulose propionate, polyamide, and nylon) and in different colors, shapes, and levels of resistance to breakage. Epoxy frames are resilient and return to their original shape after being deformed, so they do not need to be adjusted as frequently as other types. Nylon frames are almost unbreakable. They revert to their original shape after extreme trauma and distortion; because of this property, though, they cannot be readjusted after they are manufactured.

**FIT.** The patient should have the distance between the eyes (PD) measured, so that the optical centers of the lenses will be in front of the patient's pupils. Bifocal heights also have to be measured with the chosen frame in place and adjusted on the patient. Again, this is so the lenses will be positioned correctly. If not positioned correctly, the patient may experience eyestrain or other problems. This can occur with over-the-counter reading glasses. The distance between the lenses is for a "standard" person. Generally, this will not be a problem, but if a patient is sensitive or has more closely set eyes, for example, it may pose a problem. Persons buying ready-made sunglasses or reading glasses should hold them up to see if they appear clear. They should also hold the lenses to see an object with straight lines reflected off of the lenses. If the lines don't appear straight, the lenses may be warped or inferior.

Patients may sometimes need a few days to adjust to a new prescription; however, problems should be reported, because the glasses may need to be rechecked.

### Contact lenses

Over 32 million people in the United States wear these small lenses that fit on top of the cornea. They pro-

vide a field of view unobstructed by eyeglass frames; they do not fog-up or get splattered, so it is possible to see well while walking in the rain; and they are less noticeable than any eyeglass style. On the other hand, they take time to get accustomed to; require more measurements for fitting; require many follow-up visits to the eye doctor; can lead to complications such as infections and corneal damage; and may not correct astigmatism as well as eyeglasses, especially if the astigmatism is severe.

Originally, hard contact lenses were made of a material called PMMA. Although still available, the more common types of contact lenses are listed below:

- Rigid gas-permeable (RGP) daily-wear lenses are made of plastic that does not absorb water but allows oxygen to get from the atmosphere to the cornea. (This is important because the cornea has no blood supply and needs to get its oxygen from the atmosphere through the film of tears that moves beneath the lens.) They must be removed and cleaned each night.
- Rigid gas-permeable (RGP) extended-wear lenses are made from plastic that also does not absorb water but is more permeable to oxygen than the plastic used for daily-wear lenses. They can be worn up to a week.
- Daily wear soft lenses are made of plastic that is permeable to oxygen and absorbs water; therefore, they are soft and flexible. These lenses must be removed and cleaned each night, and they do not correct all vision problems. Soft lenses are easier to get used to than rigid lenses, but are more prone to tears and do not last as long.
- Extended-wear soft lenses are highly permeable to oxygen, are flexible by virtue of their ability to absorb water, and can usually be worn for up to one week. They do not correct all vision problems. There is more of a risk of infection with extended-wear lenses than with daily-wear lenses.
- Extended-wear disposable lenses are soft lenses worn continually for up to six days and then discarded, with no need for cleaning.
- Planned-replacement soft lenses are daily wear lenses that are replaced on a regular schedule, which is usually every two weeks, monthly, or quarterly. They must also be cleaned.

Soft contact lenses come in a variety of materials. There are also different kinds of RGP and soft multifocal contact lenses available. Monovision, where one contact lens corrects for distance vision while the other corrects for near vision, may be an option for presbyopic patients. Monovision, however, may affect depth perception and may not be appropriate for everyone. Contact lenses also

## KEY TERMS

**Astigmatism**—Assymetric vision defects due to irregularities in the cornea.

**Cornea**—The clear outer covering of the front of the eye.

**Index of refraction**—A constant number for any material for any given color of light that is an indicator of the degree of the bending of the light caused by that material.

**Lens**—A device that bends light waves.

**Permeable**—Capable of allowing substances to pass through.

**Polycarbonate**—A very strong type of plastic often used in safety glasses, sport glasses, and children's eyeglasses. Polycarbonate lenses have approximately 50 times the impact resistance of glass lenses.

**Polymer**—A substance formed by joining smaller molecules. For example, plastic, acrylic, cellulose acetate, cellulose propionate, nylon, etc.

**Presbyopia**—A condition affecting people over the age of 40 where the system of accommodation that allows focusing of near objects fails to work because of age-related hardening of the lens of the eye.

**Retina**—The inner, light-sensitive layer of the eye containing rods and cones; transforms the image it receives into electrical messages sent to the brain via the optic nerve.

**Ultraviolet (UV) light**—Part of the electromagnetic spectrum with a wavelength just below that of visible light. It is damaging to living material, especially eyes and DNA.

come in a variety of tints. Soft contacts are available that can change dark-colored eyes a different color. Even though such lenses have no prescription, they must still be fitted and checked to make sure that an eye infection does not occur. People should NEVER wear someone else's contact lenses. This can lead to infection or damage to the eye.

### Aftercare

Contact lens wearers must be examined periodical-  
ly by their eye doctors to make sure that the lenses fit  
properly and that there is no infection. Both infection  
and lenses that do not fit properly can damage the  
cornea. Patients can be allergic to certain solutions that  
are used to clean or lubricate the lenses. For that reason,  
patients should not randomly switch products unless  
they speak with their doctor. Contact lens wearers  
should seek immediate attention if they experience eye  
**pain**, a burning sensation, red eyes, intolerable sensitiv-  
ity to light, cloudy vision, or an inability to keep the  
eyes open.

To avoid infection, it is important for contact lens  
wearers to exactly follow their instructions for lens inser-  
tion and removal, as well as cleaning. Soft contact lens  
wearers should never use tap water to rinse their lenses or  
to make-up solutions. All contact lens wearers should  
also always have a pair of glasses and a carrying case for  
their contacts with them, in case the contacts have to be  
removed due to eye irritation.

### Risks

Wearing contact lenses increases the risk of corneal  
damage and eye infections.

### Normal results

The normal expectation is that people will achieve  
20/20 vision while wearing corrective lenses.

### Resources

#### BOOKS

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#### ORGANIZATIONS

American Academy of Ophthalmology. 655 Beach Street, PO 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>>.

Optician Association of America. 7023 Little River Turnpike, Suite 207, Annandale, VA 22003. (703) 916-8856. <<http://www.opticians.org>>.

#### OTHER

*Contact Lens Council*. <<http://www.contactlenscouncil.org>>.

Lorraine Lica, PhD

## Eye muscle surgery

### Definition

Eye muscle surgery is surgery to weaken, strengthen, or reposition any of the muscles that move the eyeball (the extraocular muscles).

### Purpose

The purpose of eye muscle surgery is generally to align the pair of eyes so that they gaze in the same direction and move together as a team, either to improve appearance or to aid in the development of binocular vision in a young child. To achieve binocular vision, the goal is to align the eyes so that the location of the image on the retina of one eye corresponds to the location of the image on the retina of the other eye.

In addition, sometimes eye muscle surgery can help people with other eye disorders (**nystagmus** and Duane syndrome, for example).

### Precautions

Depth perception (stereopsis) develops around the age of three months old. For successful development of binocular vision and the ability to perceive three-dimensionally, the surgery should not be postponed past the age of four. The earlier the surgery the better the outcome, so an early diagnosis is important. Surgery may even be performed before two years old. After surgery, if binocular vision is to develop, corrective lenses and eye exercises (vision therapy) will probably be necessary.

### Description

The extraocular muscles attach via tendons to the sclera (the white, opaque, outer protective covering of the eyeball) at different places just behind an imaginary equator circling the top, bottom, left, and right of the eye. The other end of each of these muscles attaches to a part of the orbit (the eye socket in the skull). These muscles enable the eyes to move up, down, to one side or the other, or any angle in between.

Normally both eyes move together, receive the same image on corresponding locations on both retinas, and the brain fuses these images into one three-dimensional image. The exception is in **strabismus** which is a disorder where one or both eyes deviate out of alignment, most often outwardly (exotropia) or toward the nose (esotropia). The brain now receives two different images, and either suppresses one or the person sees double (diplopia). This deviation can be adjusted by weakening or strengthening the appropriate muscles to move the eyes

toward the center. For example, if an eye turns upward, the muscle at the bottom of the eye could be strengthened.

Rarely, eye muscle surgery is performed on people with nystagmus or Duane syndrome. Nystagmus is a condition where one or both eyes move rapidly or oscillate; it can sometimes be helped by moving the eyes to the position of least oscillation. Duane syndrome is a disorder where there is limited horizontal eye movement; it can sometimes be relieved by surgery to weaken an eye muscle.

There are two methods to alter extraocular muscles. Traditional surgery can be used to strengthen, weaken, or reposition an extraocular muscle. The surgeon first makes an incision in the conjunctiva (the clear membrane covering the sclera), then puts a suture into the muscle to prevent it from getting lost and loosens the muscle from the eyeball with a surgical hook. During a resection, the muscle is detached from the sclera, a piece of muscle is removed so the muscle is now shorter, and the muscle is reattached to the same place. This strengthens the muscle. In a recession, the muscle is made weaker by repositioning it. More than one extraocular eye muscle might be operated on at the same time.

Another way of weakening eye muscles, using botulinum toxin injected into the muscle, was introduced in the early 1980s. Although the botulinum toxin wears off, the realignment may be permanent, depending upon whether neurological connections for binocular vision were established during the time the toxin was active. This technique can also be used to adjust a muscle after traditional surgery.

The cost of eye muscle surgery is about \$2,000–\$4,000, and about 700,000 surgeries are performed annually in the United States.

### Preparation

Patients should make sure their doctors are aware of any medications that they are taking, even over-the-counter medications. Patients should not take **aspirin**, or any other blood-thinning medications for ten days prior to surgery, and should not eat or drink after midnight the night before.

### Aftercare

Patients will need someone to drive them home after their surgery. They should continue to avoid aspirin and other non-steroidal anti-inflammatory agents for an additional three days, but they can take **acetaminophen** (e.g., Tylenol). Patients should discuss this with the surgeon to be clear what medications they can or cannot take. **Pain** will subside after two to three days, and patients can resume most normal activities within a few days. Again,

## KEY TERMS

**Botulinum toxin (botulin)**—A neurotoxin made by *Clostridium botulinum*; causes paralysis in high doses, but is used medically in small, localized doses to treat disorders associated with involuntary muscle contraction and spasms, in addition to strabismus.

**Conjunctiva**—The mucous membrane that covers the eyes and lines the eyelids.

**Extraocular muscles**—The muscles (lateral rectus, medial rectus, inferior rectus, superior rectus, superior oblique, and inferior oblique) that move the eyeball.

**Orbit**—The cavity in the skull containing the eyeball; formed from seven bones: frontal, maxillary, sphenoid, lacrimal, zygomatic, ethmoid, and palatine.

**Retina**—The inner, light-sensitive layer of the eye containing rods and cones; transforms the image it receives into electrical messages sent to the brain via the optic nerve.

**Sclera**—The tough, fibrous, white outer protective covering of the eyeball.

**Strabismus**—A disorder where the two eyes do not point in the same direction.

this may vary with the patient and the patient should discuss returning to normal activity with the surgeon. They should not get their eyes wet for three to four days and should refrain from swimming for 10 days. Operated eyes will be red for about two weeks.

### Risks

As with any surgery, there are risks involved. Eye muscle surgery is relatively safe, but very rarely a cut muscle gets lost and can not be retrieved. This, and other serious reactions, including those caused by anesthetics, can result in vision loss in the affected eye. Occasionally, retinal or nerve damage occurs. Double vision is not uncommon after eye muscle surgery. As mentioned earlier, glasses or vision therapy may be necessary.

### Normal results

Cosmetic improvement is likely with success rate estimates varying from about 65–85%. According to the best statistics as of 1998, binocular vision is improved in

young children about 35% of the time. There is no improvement, or the condition worsens 15–35% of the time. A second operation may rectify less-than-perfect outcomes.

## Resources

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Olitsky, Scott E., and Leonard B. Nelson. *Strabismus Web Book*. 4 May 1998 <<http://www.smbs.buffalo.edu/oph/ped/webbook.htm>>.

Lorraine Lica, PhD

Eye training see **Vision training**

## Eyelid disorders

### Definition

An eyelid disorder is any abnormal condition that affects the eyelids.

### Description

Eyelids consist of thin folds of skin, muscle, and connective tissue. The eyelids protect the eyes and spread tears over the front of the eyes. The inside of the eyelids are lined with the conjunctiva of the eyelid (the palpebral conjunctiva), and the outside of the lids are covered with the body's thinnest skin. Some common lid problems include the following: stye, blepharitis, chalazion, entropion, ectropion, eyelid **edema**, and eyelid tumors.

### Stye

A stye is an infection of one of the three types of eyelid glands near the lid margins, at the base of the lashes.

### Chalazion

A chalazion is an enlargement of a meibomian gland (an oil-producing gland in the eyelid), usually not associated with an infectious agent. More likely, the gland opening is clogged. Initially, a chalazion may resemble a stye, but it usually grows larger. A chalazion may also be located in the middle of the lid and be internal.

### Blepharitis

Blepharitis is the inflammation of the eyelid margins, often with scales and crust. It can lead to eyelash loss, chalazia, styes, ectropion, corneal damage, excessive tearing, and chronic **conjunctivitis**.

### Entropion

Entropion is a condition where the eyelid margin (usually the lower one) is turned inward; the eyelashes touch the eye and irritate the cornea.

### Ectropion

Ectropion is a condition where one or both eyelid margins turn outward, exposing both the conjunctiva that covers the eye and the conjunctiva that lines the eyelid.

### Eyelid edema

Eyelid edema is a condition where the eyelids contain excessive fluid.

### Eyelid tumors

Eyelids are susceptible to the same skin tumors as the skin over the rest of the body, including noncancerous tumors and cancerous tumors (basal cell carcinoma, squamous cell carcinoma, **malignant melanoma**, and sebaceous gland carcinoma). Eyelid muscles are susceptible to sarcoma.

## Causes and symptoms

### Stye

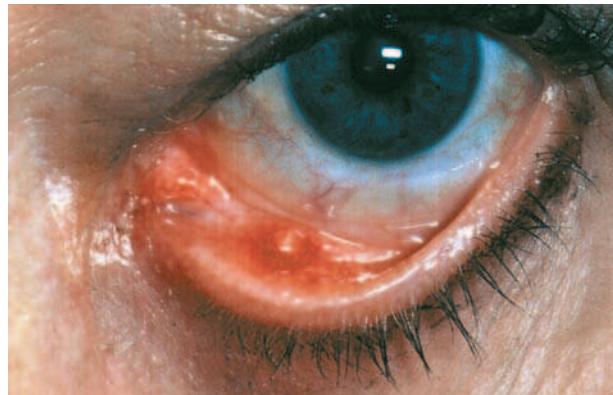
Styes are usually caused by bacterial **staphylococcal infections**. The symptoms are **pain** and inflammation in one or more localized regions near the eyelid margin.

### Chalazion

A chalazion is caused by a blockage in the outflow duct of a meibomian gland. Symptoms are inflammation and swelling in the form of a round lump in the lid that may be painful.

### Blepharitis

Some cases of blepharitis are caused by bacterial infection and some by head lice, but in some cases, the



**A chalazion on the eyelid.** This condition is caused by an obstruction of one of the meibomian glands which lubricate the edge of the eyelid. (Photo Researchers, Inc. Reproduced by permission.)

cause is unclear. It may also be caused by an overproduction of oil by the meibomian glands. Blepharitis can be a chronic condition that begins in early childhood and can last throughout life. Symptoms can include **itching**, burning, a feeling that something is in the eye, inflammation, and scales or matted, hard crusts surrounding the eyelashes.

### Entropion

Entropion usually results from **aging**, but sometimes can be due to a congenital defect, a spastic eyelid muscle, or a scar on the inside of the lid that could be from surgery, injury, or disease. It is accompanied by excessive tearing, redness, and discomfort.

### Ectropion

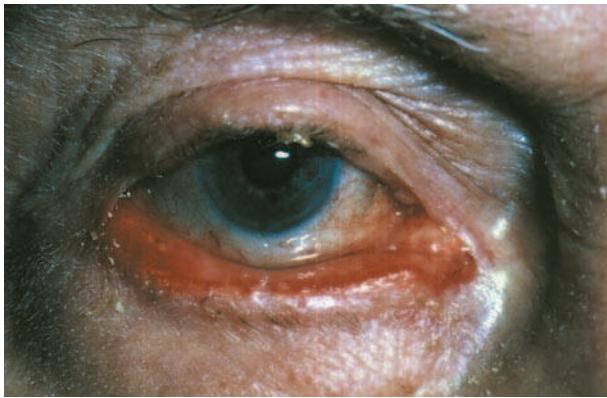
Similar to entropion, the usual cause of ectropion is aging. It also can be due to a spastic eyelid muscle or a scar, as in entropion. It also can be the result of **allergies**. Symptoms are excessive tearing and hardening of the eyelid conjunctiva.

### Eyelid edema

Eyelid edema is most often caused by allergic reactions, for example, allergies to eye makeup, eyedrops or other drugs, or plant allergens such as pollen. **Trichinosis**, a disease caused by eating undercooked meat, also causes eyelid edema. However, swelling can also be caused by more serious causes, such as infection, and can lead to orbital **cellulitis** which can threaten vision. Symptoms can include swelling, itching, redness, or pain.

### Eyelid tumors

Tumors found on the eyelids are caused by the same conditions that cause these tumors elsewhere on the



**A close-up of the eye of an elderly patient showing ectropion of the lower eyelid. Ectropion is a condition in which the eyelid turns away from the eye. The most common type is senile ectropion (seen here), in which the droop of the eyelid is due to loss of tissue elasticity in old age and weakness in the muscles surrounding the eye.** (Photograph by Dr. P. Marazzi, Photo Researchers, Inc. Reproduced by permission.)

body. They are usually painless and may or may not be pigmented. Some possible causes include AIDS (**Kaposi's sarcoma**) or increased exposure to ultraviolet (UV) rays which may lead to skin **cancer**.

## Diagnosis

An instrument called a slit lamp is generally used to magnify the structures of the eyes. The doctor may press on the lid margin to see if oil can be expressed from the meibomian glands. The doctor may invert the lid to see the inside of the lid. Biopsy is used to diagnose cancerous tumors.

## Treatment

### Stye

Styes are treated with warm-hot compresses for 10–15 minutes, three to four times a day. Sometimes topical **antibiotics** may be prescribed. If the initial treatment is ineffective, styes are lanced and drained.

### Chalazion

About 25% of chalazia will disappear spontaneously, but hot compresses may speed the process. Because chalazia are inside the lid, topical medications are generally of no benefit. Medication may need to be injected by the doctor into the chalazion or if that doesn't help the chalazion may need to be excised. If what appears to be a chalazion recurs on the same site as any previous one, the possibility of sebaceous gland carcinoma should be investigated by biopsy.

### Blepharitis

Blepharitis is treated with hot compresses, with antibiotic ointment, and by cleaning the eyelids with a moist washcloth and then with baby shampoo. Good hygiene is essential. If the blepharitis doesn't clear up with treatment or if it seems to be a chronic problem, the patient may have **acnerosacea**. These patients may need to see a dermatologist as well.

### Entropion and ectropion

Both entropion and ectropion can be surgically corrected. Prior to surgery, the lower lid of entropion can be taped down to keep the lashes off the eye, and both can be treated with lubricating drops to keep the cornea moist.

### Eyelid edema

Patients with swollen eyelids should contact their eye doctor. A severely swollen lid can press on the eye and possibly increase the intraocular pressure. An infection needs to be ruled out. Or, something as simple as an allergy to nail polish and then touching the eyes can cause swelling. The best treatment for allergic eyelid edema is to find and remove the substance causing the allergy. When that is not possible, as in the case of plant allergens, cold compresses and immunosuppressive drugs such as corticosteroid creams are helpful. However, steroids can cause **cataracts** and increase intraocular pressure and patients must be very careful not to get the cream in their eyes. This should not be done unless under a doctor's care. For edema caused by trichinosis, the trichinosis must be treated.

### Eyelid tumors

Cancerous tumors should be removed upon discovery, and noncancerous tumors should be removed before they become big enough to interfere with vision or eyelid function. Eyelid tumors require special consideration because of their sensitive location. It is important that treatment not compromise vision, eye movement, or eyelid movement. Accordingly, eyelid reconstruction will sometimes accompany tumor excision.

### Prognosis

The prognosis for styes and chalazia is good to excellent. With treatment, blepharitis, ectropion, and entropion usually have good outcomes. The prognosis for nonmalignant tumors, basal cell carcinoma, and squamous cell carcinoma is good once they are properly removed. Survival rate for malignant melanoma depends upon how early it was discovered and if it was completely removed. Sebaceous carcinomas are difficult to detect, so poor outcomes are more frequent.

All of these eyelid disorders, if not treated, can lead to other, possibly serious vision problems—dry eye, **astigmatism**, or even vision loss, for example. An ophthalmologist or optometrist should be consulted.

### Prevention

Good lid hygiene is very important. Regular eyelid washing with baby shampoo helps prevent styes, chalazia, blepharitis, and eyelid edema. To avoid these problems, it's also important to refrain from touching and rubbing the eyes and eyelids, especially with hands that have not just been washed.

Blepharitis is associated with dandruff, which is caused by a kind of bacteria that is one of the causes of blepharitis. Controlling dandruff by washing the hair, scalp, and eyebrows with shampoo containing selenium sulfide to kill the bacteria helps control the blepharitis. When using anything near the eyes, it is important to read the label or consult with a doctor first.

Avoiding allergens helps prevent allergic eyelid edema. Staying inside as much as possible when pollen counts are high and eliminating the use of, or at least removing eye makeup thoroughly, or using hypo-allergenic makeup may help if the person is sensitive to those substances.

Sunscreen, UV-blocking sunglasses, and wide brimmed hats can help prevent eyelid tumors.

Entropian and ectropian seem to be unpreventable.

### Resources

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Newell, Frank W. *Ophthalmology: Principles and Concepts*. 8th ed. St. Louis: Mosby, 1996.

#### ORGANIZATIONS

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

### KEY TERMS

**Allergen**—A substance capable of inducing an allergic response.

**Allergic reaction**—An immune system reaction to a substance in the environment; symptoms include rash, inflammation, sneezing, itchy watery eyes, and runny nose.

**Conjunctiva**—The mucous membrane that covers the white part of the eyes and lines the eyelids.

**Edema**—A condition where tissues contain excessive fluid.

**Meibomian gland**—Oil-producing glands in the eyelids that open near the eyelid margins.

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

American Society of Ophthalmic Plastic and Reconstructive Surgery. 1133 West Morse Blvd, #201, Winter Park, FL 32789. (407) 647-8839. <<http://www.asopr.org>>.

#### OTHER

“Eyelid Abnormalities.” *Eye Clinic of Fairbanks*. <<http://www.eyeclinicfbks.com/ECFLID.htm>>. RxMed. <<http://www.rxmed.com>>.

Lorraine Lica, PhD

Eyelid edema see **Eyelid disorders**

Eyelid plastic surgery see **Blepharoplasty**

# F

Fabry's disease see **Lipidoses**

## Face lift

### Definition

Face lift surgery is a cosmetic procedure that involves redirecting some of the skin and muscle tissue of the face and neck to counter signs of **aging** produced by gravity.

### Purpose

The purpose of face lift surgery, also known as facialplasty, rhytidoplasty, or cervicofacial rhytidectomy, is to improve the appearance of the face by repositioning the skin and tightening some of the underlying muscle and tissue. The procedure is designed to counter sagging and looseness in skin and muscle tissue caused by gravity as the patient ages. Face lift surgery will not erase all facial wrinkles, as the term rhytidectomy (which literally means "surgical removal of wrinkles") might imply. Wrinkles around the mouth and eyes, for example, may benefit little from face lift surgery. Other procedures, such as **blepharoplasty**, chemical peel, or dermabrasion, also may be necessary.

### Precautions

Patients with other medical conditions should consult with their primary physician before undergoing face lift surgery. Lung problems, heart disease, and certain other conditions can lead to a higher risk of complications. Patients who take medications that can alter the way their blood clots (including female hormones, **aspirin**, and some non-aspirin **pain** relievers) should stop these medications prior to surgery to lower the risk that a hematoma will form. A hematoma, a pocket of blood below the skin, is the most frequent complication of face lift surgery.

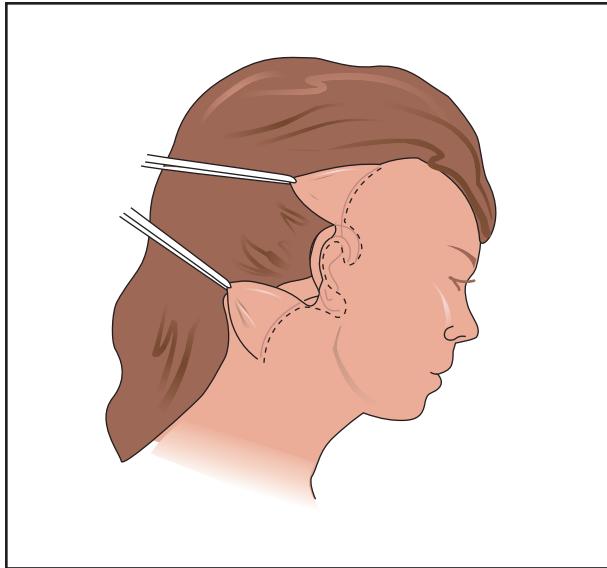
### Description

Face lift surgery can be performed on an outpatient basis with local anesthetics. Patients typically also receive "twilight anesthesia," an intravenous sedative that helps to lower their awareness of the procedure being performed.

There are a number of variations of face lift surgery. Which one is used will depend on the patient's facial structure, how much correction is needed, and the preferences of the surgeon performing the procedure. In a typical face lift surgery, the surgeon begins by making an incision within the hairline just above the ear. The incision continues down along the front edge of the ear, around the earlobe, and then up and behind the ear extending back into the hairline. The location of this incision is designed to hide any sign of the procedure later. The same procedure is repeated on the other side of the face. Using various instruments, the surgeon will then work to separate the skin of the face from its underlying tissue, moving down to the cheek and into the neck area and below the chin. Fat deposits over the cheeks and in the neck may be removed surgically or with **liposuction** at this time. The surgeon will then work to free up and tighten certain bands of muscle and tissue that extend up from the shoulder, below the chin, and up and behind the neck. If these muscles and tissue are not tightened, the looseness and sagging appearance of the skin will return. The surgeon then trims excess skin from the edges of the original incision, pulls the skin back, and staples or sutures it into place.

### Preparation

Prior to the procedure, patients meet with their surgeon to discuss the surgery, clarify the results that can be achieved, and discuss the potential problems that can occur. Having realistic expectations is important in any cosmetic procedure. Patients will learn, for example, that although face lift surgery can improve the contour of the face and neck, other procedures will be necessary to reduce the appearance of many wrinkles. As mentioned



In a typical face lift surgery, the surgeon begins by making an incision within the hairline just above the ear. The incision continues down along the front of the ear, around the earlobe, and then up and behind the ear extending back into the hairline, as shown above. The same procedure is repeated on the other side of the face. The surgeon will then separate the skin from the tissue, remove fat deposits over the cheeks and neck, tighten up muscles and tissues below the chin and upwards behind the neck. The surgeon then trims excess skin from the original incision, pulls the skin back, and sutures it into place. (Illustration by Electronic Illustrators Group.)

earlier, patients will stop taking aspirin, birth control or female hormones, and other medications affecting blood clotting about two weeks before the procedure. Some physicians prescribe vitamin C and K in the belief that this promotes healing. Patients will also be advised to stop **smoking** and to avoid exposure to passive smoke before the procedure and afterward. Some surgeons also recommend **antibiotics** be taken beforehand to limit the risk of infection. Some surgeons also use a steroid injection before or after the procedure, to reduce swelling.

## Aftercare

After the surgery, a pressure bandage will be applied to the face to reduce the risk of hematoma. The patient may spend a few hours resting in a recovery room to ensure no bleeding has occurred. The patient then returns home. Some surgeons recommend that the patient remain reclining for the next 24 hours, consuming a liquid diet, and avoiding any movements that lead the neck to flex. Ice packs for the first few days can help to reduce swelling and lower the risk of hematoma. Patients continue taking an antibiotic until the first stitches come out about five

## KEY TERMS

**Hematoma**—A complication of surgery in which a collection of blood forms below the skin.

**Rhytidectomy**—It literally means “wrinkle excision.” It is another, misleading, term for face lift surgery.

**Twilight anesthesia**—An intravenous mixture of sedatives and other medications that decreases patients’ awareness of the procedure being performed.

days after the procedure. The balance are removed seven to 10 days later. Many patients return to work and limited activities within two weeks of the procedure.

## Risks

The major complication seen following face lift surgery is a hematoma. If a hematoma forms, the patient may have to return to have the stitches reopened to find the source of the bleeding. Most hematomas form within 48 hours of surgery. The typical sign is pain or swelling affecting one side of the face but not the other.

Another risk of face lift surgery is nerve damage. Sometimes it can affect the patient’s ability to raise an eyebrow, or distort his smile, or leave him with limited feeling in his earlobe. Most of these nerve injuries, however, repair themselves within 2–6 months.

## Normal results

Some swelling and bruising is normal following face lift surgery. After these disappear, the patient should see a noticeable improvement in the contour of his face and neck. Some fine wrinkling of the skin may be improved, but deep wrinkles are likely to require another cosmetic procedure to improve their appearance.

## Abnormal results

In addition to the risks outlined above, other complications of face lift surgery include infection, scarring, and hair loss near incision lines.

## Resources

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Springer, Rachelle C. "Rhytidectomy: From Consultation to Recovery." *Plastic Surgical Nursing*, 16 (Spring 1996): 27-30.

#### ORGANIZATIONS

American Society for Dermatologic Surgery. 930 N. Meacham Road, PO Box 4014, Schaumburg, IL 60168-4014. (847) 330-9830. <<http://www.asds-net.org>>.

American Society of Plastic and Reconstructive Surgeons. 44 E. Algonquin Rd., Arlington Heights, IL 60005. (847) 228-9900. <<http://www.plasticsurgery.org>>.

Richard H. Camer

Facial nerve paralysis see **Bell's palsy**

Many persons with Munchausen go so far as to undergo major surgery repeatedly, and, to avoid detection, at several locations. Many have been employed in hospitals or in health care professions. The syndrome's onset is in early adulthood.

#### *Munchausen by proxy*

Munchausen by proxy is the name given to factitious disorders in children produced by parents or other caregivers. The parent may falsify the child's medical history or tamper with laboratory tests in order to make the child appear sick. Occasionally, they may actually injure the child to assure that the child will be treated.

#### *Ganser's syndrome*

Ganser's syndrome is an unusual dissociative reaction to extreme **stress** in which the patient gives absurd or silly answers to simple questions. It has sometimes been labeled as psychiatric malingering, but is more often classified as a factitious disorder.

#### Causes and symptoms

No single explanation of factitious disorders covers all cases. These disorders are variously attributed to underlying **personality disorders**; **child abuse**; the wish to repeat a satisfying childhood relationship with a doctor; and the desire to deceive or test authority figures. Also, the wish to assume the role of patient and be cared for is involved. In many cases, the suffering of a major personal loss has been implicated.

The following are regarded as indications of a factitious disorder:

- dramatic but inconsistent medical history
- extensive knowledge of medicine and/or hospitals
- negative test results followed by further symptom development
- symptoms that occur only when the patient is not being observed
- few visitors
- arguments with hospital staff or similar acting-out behaviors
- eagerness to undergo operations and other procedures

When patients with factitious disorders are confronted, they usually deny that their symptoms are intentional. They may become angry and leave the hospital. In many cases they enter another hospital, which has led to the nickname "hospital hoboies."

## Factitious disorders

### Definition

Factitious disorders are a group of mental disturbances in which patients intentionally act physically or mentally ill without obvious benefits. The name factitious comes from a Latin word that means artificial. These disorders are not **malingering**, which is defined as pretending illness when the "patient" has a clear motive, such as financial gain.

### Description

Patients with factitious disorders produce or exaggerate the symptoms of a physical or mental illness by a variety of methods, including contaminating urine samples with blood, taking hallucinogens, injecting themselves with bacteria to produce infections, and other similar behaviors.

There are no reliable statistics on the frequency of factitious disorders, but they are more common in men than in women. The following conditions are sometimes classified as factitious disorders:

#### *Munchausen syndrome*

**Munchausen syndrome** refers to patients whose factitious symptoms are dramatized and exaggerated.

## KEY TERMS

**Ganser's syndrome**—An unusual factitious disorder characterized by dissociative symptoms and absurd answers to direct questions.

**Malingering**—Pretending to be sick in order to be relieved of an unwanted duty or obtain some other obvious benefit.

**Munchausen by proxy**—A factitious disorder in children produced by a parent or other caregiver.

**Munchausen syndrome**—A factitious disorder in which the patient's symptoms are dramatized and exaggerated.

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Raney, David F. "Behavioral, Psychosocial & Psychiatric Pediatrics." In *Handbook of Pediatrics*, edited by Gerald B. Merenstein, et al. Norwalk, CT: Appleton & Lange, 1994.

Rebecca J. Frey

Factor IX deficiency see **Hemophilia**

Factor VIII deficiency see **Hemophilia**

## Diagnosis

Diagnosis of factitious disorders is usually based on the exclusion of bona fide medical or psychiatric conditions, together with a combination of the signs listed earlier. In some cases, the diagnosis is made on the basis of records from other hospitals.

## Treatment

Treatment of factitious disorders is usually limited to prompt recognition of the condition and the refusal to give unnecessary medications or to perform unneeded procedures. Factitious disorder patients do not usually remain in the hospital long enough for effective psychiatric treatment. Some clinicians have tried psychotherapeutic treatment for factitious disorder patients, and there are anecdotal reports that antidepressant or antipsychotic medications are helpful in certain cases.

## Prognosis

Some patients have only one or two episodes of factitious disorders; others develop a chronic form that may be lifelong. Successful treatment of the chronic form appears to be rare.

## Resources

### BOOKS

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- "Factitious Disorders." In *Diagnostic and Statistical Manual of Mental Disorders*. 4th ed. Washington, DC: The American Psychiatric Association, 1994.
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## Failure to thrive

### Definition

Failure to thrive (FTT) is used to describe a delay in a child's growth or development. It is usually applied to infants and children up to two years of age who do not gain or maintain weight as they should. Failure to thrive is not a specific disease, but rather a cluster of symptoms which may come from a variety of sources.

### Description

Shortly after birth most infants lose some weight. After that expected loss, babies should gain weight at a steady and predictable rate. When a baby does not gain weight as expected, or continues to lose weight, it is not thriving. Failure to thrive may be due to one or more conditions.

Organic failure to thrive (OFTT) implies that the organs involved with digestion and absorption of food are malformed or incomplete so the baby cannot digest its food. Non-organic failure to thrive (NOFTT) is the most common cause of FTT and implies the baby is not receiving enough food due to economic factors or parental neglect, or do to psychosocial problems.

### Causes and symptoms

Occasionally, there may be an underlying physical condition that inhibits the baby's ability to take in, digest, or process food. These defects can occur in the esophagus, stomach, small or large intestine, rectum or anus. Usually the defect is an incomplete development of the organ, and it must be surgically corrected. Most physical defects can be detected shortly after birth.

Failure to thrive may also result from lack of available food or the quality of the food offered. This can be due to economic factors in the family, parental beliefs and concepts of **nutrition**, or neglect of the child. In addition, if the baby is being breast fed, the quality or quantity of the mother's milk may be the source of the problem.

Psychosocial problems, often stemming from a lack of nurturing parent-child relations can lead to a failure to thrive. The child may exhibit poor appetite due to depression from insufficient attention from parents.

Infants and toddlers, whose growth is substantially less than expected, are considered to be suffering from FTT.

## Diagnosis

Most babies are weighed at birth and that weight is used as a base line for future well-baby check-ups. If the baby is not gaining weight at a predictable rate, the doctor will do a more extensive examination. If there are no apparent physical deformities in the digestive tract, the doctor will examine the child's environment. As part of that examination, the doctor will look at the family history of height and weight. In addition, the parents will be asked about feedings, illnesses, and family routines. If the mother is breastfeeding the doctor will also evaluate her diet, general health, and well being as it affects the quantity and quality of her milk.

Diagnosis of FTT is confirmed by a positive growth and behavioral response to increased nutrition.

## Treatment

If there is an underlying physical reason for failure to thrive, such as a disorder of swallowing mechanism or intestinal problems, correcting that problem should reverse the condition. If the condition is caused by environmental factors, the physician will suggest several ways parents may provide adequate food for the child. Maternal education and parental counseling may also be recommended. In extreme cases, hospitalization or a more nurturing home may be necessary.

## Prognosis

The first year of life is important as a foundation for growth and physical and intellectual development in the future. Children with extreme failure to thrive in the first year may never catch up to their peers even if their physical growth improves. In about one third of these extreme cases, mental development remains below normal and roughly half will continue to have psychosocial and eating problems throughout life.

## KEY TERMS

**Esophagus**—The muscular tube which connects the mouth and stomach.

**Psychosocial**—A term referring to the mind's ability to, consciously or unconsciously, adjust and relate the body to its social environment.

When failure to thrive is identified and corrected early, most children catch up to their peers and remain healthy and well developed.

## Prevention

Initial failure to thrive caused by physical defects cannot be prevented but can often be corrected before they become a danger to the child. Maternal education and emotional and economic support systems all help to prevent failure to thrive in those cases where there is no physical deformity.

## Resources

### ORGANIZATIONS

American Humane Association, Children's Division. 63 Inverness Drive East, Englewood, CO 80112-5117. (800) 227-4645. <[www.americanhumane.org](http://www.americanhumane.org)>.

Federation for Children With Special Needs. 1135 Tremont Street, Suite 420, Boston, MA 02120. (617) 236-7210. <<http://www.fcsn.org>>.

National Digestive Diseases Information Clearinghouse. 2 Information Way, Bethesda, MD 20892-3570. (800) 891-5389. <<http://www.niddk.nih.gov/health/digest/nddic.htm>>.

Dorothy Elinor Stonely

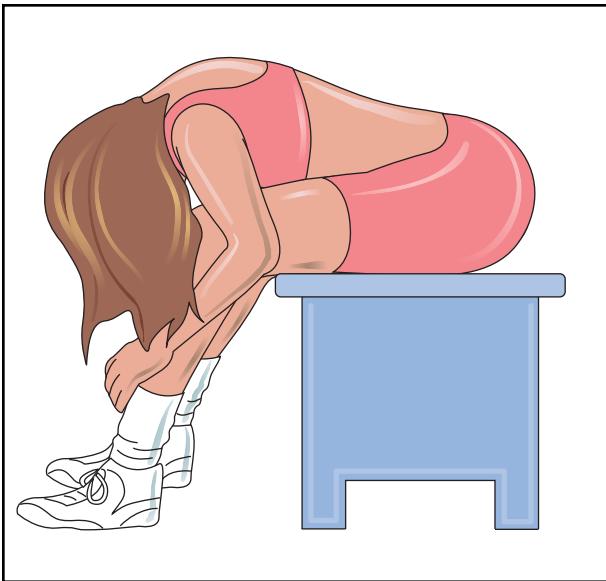
## Fainting

### Definition

Fainting is loss of consciousness caused by a temporary lack of oxygen to the brain. Known by the medical term "syncope," fainting may be preceded by **dizziness**, nausea, or a feeling of extreme weakness.

### Description

When a person faints, the loss of consciousness is brief. The person will wake up as soon as normal blood flow is restored to the brain. Blood flow is usually



If a person is feeling faint, unconsciousness may be prevented by sitting with the head between the knees, as shown in the illustration above, or by lying flat with the legs raised. Illustration by Electronic Illustrators Group.)

restored by lying flat for a short time. This position puts the head on the same level as the heart so that blood flows more easily to the brain.

A fainting episode may be completely harmless and of no significance, but it can be a symptom of a serious underlying disorder. No matter how trivial it seems, a fainting episode should be treated as a medical emergency until the cause is determined.

### Causes and symptoms

Extreme pain, fear, or stress may bring on fainting. This type of fainting is caused by overstimulation of the vagus nerve, a nerve connected to the brain that helps control breathing and circulation. In addition, a person who stands still or erect for too long may faint. This type of fainting occurs because blood pools in the leg veins, reducing the amount that is available for the heart to pump to the brain. This type of fainting is quite common in older people or those taking drugs to treat high blood pressure.

When an older person feels faint upon turning the head or looking upward suddenly, the cause could be osteoarthritis of the neck bones. Osteoarthritis damages the cartilage between the neck bones and causes pressure on blood vessels leading to the brain.

Fainting can be a symptom of a disease such as Stokes-Adams syndrome, a condition in which blood flow to the brain is temporarily reduced because of an irregular heartbeat. Some people may experience fainting

associated with weakness in the limbs or a temporary problem in speaking caused by obstructed blood flow in vessels passing through the neck to the brain. Pregnant women frequently feel faint. Fainting may also occur as a result of low blood sugar. Low blood sugar can occur if a person skips a meal or has diabetes.

Fainting can also be caused by:

- prolonged coughing
- straining to defecate or urinate
- blowing a wind instrument too hard
- remaining in a stuffy environment with too little oxygen

Sometimes fainting may be caused by a temporary drop in the blood supply to the brain caused by a **transient ischemic attack** (TIA). A TIA, sometimes called a mini-stroke, is a disruption in the blood supply to the brain caused by a blocked or burst blood vessel. Seek help immediately if a fainting spell is followed by one or more of the symptoms listed below:

- numbness or tingling in any body part
- blurred vision
- confusion
- difficulty speaking
- loss of movement in arms or legs

A few seconds before fainting, a person may sweat or become pale, feel nauseated or dizzy, and have blurred vision or racing heartbeat. Once the person loses consciousness, the pupils may dilate as the heart rate slows down. There may be abnormal movements. Muscles may tighten or the back may arch. These movements do not last long and they are not violent.

In most cases, the patient regains consciousness within a few minutes, but the fainting spell may be followed by nervousness, **headache**, nausea, dizziness, pallor or sweating. The person may faint again, especially if he or she stands up within 30 minutes.

### Diagnosis

Most episodes of fainting are a one-time occurrence. When a person experiences repeated fainting spells, a physician should be consulted.

### Treatment

Most of the time, a person who faints ends up lying on the floor. If this happens, the patient should be rolled onto his or her back. Because someone who faints often vomits, bystanders should keep the airway open. A person who is fainting should not be held upright or in a sitting position. These positions prevent blood flow to the brain and may bring on a seizure.

## KEY TERMS

**Osteoarthritis**—A disease characterized by damage to the cartilage in the joints. The joints become inflamed, deformed, and enlarged, and movement becomes painful.

**Stokes-Adams syndrome**—Recurrent episodes of temporary loss of consciousness (fainting) caused by an insufficient flow of blood from the heart to the brain. This syndrome is caused by a very rapid or a very slow heartbeat.

**Transient ischemic attack (TIA)**—A brief interruption of the blood supply to part of the brain that causes a temporary impairment of vision, speech, or movement. Usually, the episode lasts for just a few moments, but it may be a warning sign for a full-scale stroke.

**Vagus nerve**—A cranial nerve, that is, a nerve connected to the brain. The vagus nerve has branches to most of the major organs in the body, including the larynx, throat, windpipe, lungs, heart, and most of the digestive system.

Bystanders should check the patient's breathing and pulse rate. The pulse may be weak and slow. If there are no signs of breathing or heart rate, the problem is more serious than fainting, and **cardiopulmonary resuscitation (CPR)** must begin.

If breathing and pulse rates seem normal, the person's legs should be raised above the level of the head so that gravity can help the blood flow to the brain. Belts, collars or any other constrictive clothing should be loosened.

If the person does not regain consciousness within a minute or two after fainting, medical help should be summoned.

### Prognosis

After a fainting spell, the person should regain normal color but may continue to feel weak for a short time. Lying down quietly for a few moments may help.

In most cases, an attack of fainting is not serious. As soon as the underlying pain or stress passes, the danger of repeated episodes also is eliminated.

### Prevention

If a person is feeling faint, unconsciousness may be prevented by sitting with the head between the knees or lying flat with the legs raised.

A person who has fainted should lie flat for 10–15 minutes after regaining consciousness to give the system a chance to regain its balance. Standing up too soon may bring on another fainting spell.

### Resources

#### BOOKS

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Carol A. Turkington

Falciparum malaria see **Malaria**

Fallopian tube ligation see **Tubal ligation**

Fallopian tube removal see **Salpingectomy**

Fallopian tube x rays see

**Hysterosalpingography**

Famciclovir see **Antiviral drugs**

## Familial Mediterranean fever

### Definition

Familial Mediterranean fever (FMF) is an inherited disorder of the inflammatory response characterized by recurring attacks of fever, accompanied by intense pain in the abdomen, chest, or joints. Attacks usually last 12–72 hours, and can occasionally involve a skin rash. Kidney disease is a serious concern if the disorder is not treated. FMF is most prevalent in people of Armenian, Sephardic-Jewish, Arabic, and Turkish ancestry.

### Description

FMF could be described as a disorder of “inappropriate” inflammation. That is, an event that in a normal situation causes a mild or unnoticeable inflammation might cause a severe inflammatory response in someone with FMF. Certain areas of the body are at risk for FMF-related symptoms. A serosa is a serous (fluid-producing) membrane that can be found inside the abdominal cavity (peritoneum), around the lungs (pleura), around the heart (pericardium), and inside the joints (synovium). The symptoms of FMF are due to inflammation of one or more of the serosal membranes (serositis). Thus, FMF is also sometimes called recurrent polyserositis.

During an attack, large numbers of neutrophils, a type of white blood cell, move into the affected areas

causing painful inflammation and fever. These episodes may be accompanied by a skin rash or joint pain. In a few cases, chronic arthritis is a problem. **Amyloidosis** is a potentially serious condition in which proteins called amyloids are mistakenly produced and deposited in organs and tissues throughout the body. Left untreated, amyloidosis often leads to kidney failure, which is the major long-term health risk in FMF.

In most cases, the attacks of fever and pain are first noticed in childhood or adolescence. The interval between these episodes may be days or months, and is not predictable. However, during these intervals people with FMF typically lead normal lives. It is not entirely clear what brings on an attack, but people with FMF often report mild physical trauma, physical exertion, or emotional **stress** just prior to the onset of symptoms. Treatment for FMF involves an oral medication called colchicine, which is highly effective for the episodes of fever and pain, as well as for amyloidosis and the kidney disease that can result from it.

FMF is most common in certain ethnic groups from the eastern Mediterranean region, but cases in other ethnic groups in other parts of the world are increasingly being reported. FMF is also known by many other names. They include: recurrent hereditary polyserositis, benign paroxysmal **peritonitis**, familial paroxysmal polyserositis, paroxysmal polyserositis, familial recurrent polyserositis, periodic fever, periodic amyloid syndrome, periodic peritonitis syndrome, Reimann periodic disease, Reimann syndrome, Siegel-Cattan-Mamou syndrome, and Armenian syndrome.

Estimates of the incidence of FMF in specific eastern Mediterranean populations range from 1 in 2000 to 1 in 100, depending on the population studied. Specific mutations in the MEFV gene are more common in certain ethnic groups, and may cause a somewhat different course of the disease. A few mutations in the MEFV gene likely became common in a small population in the eastern Mediterranean several thousand years ago. It is postulated that carrying a single copy of a mutated gene produced a modified (but not abnormal) inflammatory response that may have been protective against some infectious agent at that time. Those who carried a single “beneficial” mutation in the MEFV gene were more likely to survive and reproduce, which may explain the high carrier frequency (up to one in five) in some populations. People of Armenian, Sephardic-Jewish, Arabic, and Turkish ancestry are at greatest risk for FMF. However, a better understanding and recognition of the symptoms of FMF in recent years has resulted in more reports of the condition in other ethnic groups, such as Italians and Armenian-Americans.

## Causes and symptoms

FMF is a genetic condition inherited in an autosomal recessive fashion. Mutations in the MEFV gene (short for Mediterranean Fever) on chromosome number 16 are the underlying cause of FMF. Autosomal recessive inheritance implies that a person with FMF has mutations in both copies of the MEFV gene. All genes come in pairs, and one copy of each pair is inherited from each parent. If neither parent of a child with FMF has the condition, it means they carry one mutated copy of the MEFV gene, but also one normal copy, which is enough to protect them from disease. If both parents carry the same autosomal recessive gene, there is a one in four chance in each **pregnancy** that the child will inherit both recessive genes, and thus have the condition.

The MEFV gene carries the instructions for production of a protein called pyrin, named for pyrexia, a medical term for fever. The research group in France that co-discovered the protein named it marenostrin, after ancient Latin words that referred to the Mediterranean Sea. The movement of neutrophils into an area of the body where trauma or infection has occurred is the major cause of inflammation, which is a normal process. Research has shown that pyrin has some function in controlling neutrophils. In a situation where minor trauma or stress occurs, some initial inflammation may follow, but a functional pyrin protein is responsible for shutting-down the response of neutrophils once they are no longer needed. An abnormal pyrin protein associated with FMF may be partly functional, but unstable. In some instances, the abnormal pyrin itself seems to be “stressed”, and loses its ability to regulate neutrophils and inflammation. Left unregulated, a normal, mild inflammation spirals out of control. Exactly what causes pyrin in FMF to lose its ability to control neutrophils in some situations is not known.

The recurrent acute attacks of FMF typically begin in childhood or adolescence. Episodes of fever and painful inflammation usually last 12–72 hours. About 90% of people with FMF have their first attack by age 20. The group of symptoms that characterizes FMF includes the following:

### Fever

An FMF attack is nearly always accompanied by a fever, but it may not be noticed in every case. Fevers are typically 100–104°F (38–40°C). Some people experience chills prior to the onset of fever.

### Abdominal pain

Nearly all people with FMF experience abdominal pain at one point or another, and for most it is the most

## KEY TERMS

**Acute phase reactants**—Blood proteins whose concentrations increase or decrease in reaction to the inflammation process.

**Amyloid**—A waxy translucent substance composed mostly of protein, that forms plaques (abnormal deposits) in the brain.

**Amyloidosis**—Accumulation of amyloid deposits in various organs and tissues in the body such that normal functioning of an organ is compromised.

**Colchicine**—A compound that blocks the assembly of microtubules—protein fibers necessary for cell division and some kinds of cell movements, including neutrophil migration. Side effects may include diarrhea, abdominal bloating, and gas.

**Leukocyte**—A white blood cell. The neutrophils are a type of leukocyte.

**Leukocytosis**—An increase in the number of leukocytes in the blood.

**Neutrophil**—The primary type of white blood cell involved in inflammation. Neutrophils are a type of granulocyte, also known as a polymorphonuclear leukocyte.

**Pericarditis**—Inflammation of the pericardium, the membrane surrounding the heart.

**Peritonitis**—Inflammation of the peritoneum, the membrane surrounding the abdominal contents.

**Pleuritis**—Inflammation of the pleura, the membrane surrounding the lungs.

**Pyrexia**—A medical term denoting fevers.

**Serositis**—Inflammation of a serosal membrane. Polyserositis refers to the inflammation of two or more serosal membranes.

**Synovitis**—Inflammation of the synovium, a membrane found inside joints.

common complaint. The pain can range from mild to severe, and can be diffuse or localized. It can mimic **appendicitis**, and many people with undiagnosed FMF have had appendectomies or exploratory surgery of the abdomen done, only to have the fever and abdominal pain return.

### Chest pain

Pleuritis, also called **pleurisy**, occurs in up to half of the affected individuals in certain ethnic groups. The pain is usually on one side of the chest. **Pericarditis** would also be felt as chest pain.

### Joint pain

About 50% of people with FMF experience joint pain during attacks. The pain is usually confined to one joint at a time, and often involves the hip, knee, or ankle. For some people, however, the recurrent joint pain becomes chronic arthritis.

### Myalgia

Up to 20% of individuals report muscle pain. These episodes typically last less than two days, and tend to occur in the evening or after physical exertion. Rare cases of muscle pain and fever lasting up to one month have been reported.

### Skin rash

A rash, described as erysipelas-like erythema, accompanies attacks in a minority of people, and most often occurs on the front of the lower leg or top of the foot. The rash appears as a red, warm, swollen area about 4–6 in (10–15 cm) in diameter.

### Amyloidosis

FMF is associated with high levels in the blood of a protein called serum amyloid A (SAA). Over time, excess SAA tends to be deposited in tissues and organs throughout the body. The presence and deposition of excess SAA is known as amyloidosis. Amyloidosis may affect the gastrointestinal tract, liver, spleen, heart, and testes, but effects on the kidneys are of greatest concern. The frequency of amyloidosis varies among the different ethnic groups, and its overall incidence is difficult to determine because of the use of colchicine to avert the problem. Left untreated, however, those individuals who do develop amyloidosis of the kidneys may require a renal transplant, or may even die of renal failure. The frequency and severity of a person's attacks of fever and serositis seem to have no relation to whether they will develop amyloidosis. In fact, a few people with FMF have been described who have had amyloidosis but apparently no other FMF-related symptoms.

### Other symptoms

A small percentage of boys with FMF develop painful inflammation around the testes, headaches are a common occurrence during attacks, and certain types of **vasculitis** (inflammation of the blood vessels) seem to be more common in FMF.

### Diagnosis

Individually, the symptoms that define FMF are common. Fevers occur for many reasons, and nonspecific pains in the abdomen, chest, and joints are also frequent ailments. Several infections can result in symptoms similar to FMF (Mallaret **meningitis**, for instance), and many people with FMF undergo exploratory abdominal surgery and ineffective treatments before they are finally diagnosed. Membership in a less commonly affected ethnic group may delay or hinder the correct diagnosis.

In general, symptoms involving one or more of the following broad groups should lead to suspicion of FMF: Unexplained recurrent fevers, polyserositis, skin rash, and/or joint pain; abnormal blood studies (see below); and renal or other disease associated with amyloidosis. A family history of FMF or its symptoms would obviously be an important clue, but the recessive nature of FMF means there usually is no family history. The diagnosis may be confirmed when a person with unexplained fever and pain responds to treatment with colchicine since colchicine is not known to have a beneficial effect on any other condition similar to FMF. Abnormal results on a blood test typically include **leukocytosis** (elevated number of neutrophils in the blood), an increased **erythrocyte sedimentation rate** (rate at which red blood cells form a sediment in a blood sample), and increased levels of proteins associated with inflammation (called acute phase reactants) such as SAA.

Direct analysis of the MEFV gene for FMF mutations is the only method to be certain of the diagnosis. However, it is not yet possible to detect all MEFV gene mutations that might cause FMF. Thus, if DNA analysis is negative, clinical methods must be relied upon. If both members of a couple were proven to be FMF carriers through **genetic testing**, highly accurate prenatal diagnosis would be available in any subsequent pregnancy.

Similar syndromes of periodic fever and inflammation include familial Hibernian fever and hyperimmunoglobulinemia D syndrome, but both are more rare than FMF.

### Treatment

Colchicine is a chemical compound that can be used as a medication, and is frequently prescribed for **gout**.

Some years ago, colchicine was discovered to also be effective in reducing the frequency and severity of attacks in FMF. Treatment for FMF at this point consists of taking colchicine daily. Studies have shown that about 75% of FMF patients achieve complete remission of their symptoms, and about 95% show marked improvement when taking colchicine. Lower effectiveness has been reported, but there is some question about the number of FMF patients who choose not to take their colchicine between attacks when they are feeling well, and thus lose some of the ability to prevent attacks. Compliance with taking colchicine every day may be hampered by its side effects, which include **diarrhea**, nausea, abdominal bloating, and gas. There is a theoretical risk that colchicine use could damage chromosomes in sperms and eggs, or in an embryo during pregnancy, or that it might reduce fertility. However, studies looking at reproduction in men and women who have used colchicine have so far not shown any increased risks. Colchicine is also effective in preventing, delaying, or reversing renal disease associated with amyloidosis.

Other medications may be used as needed to deal with the pain and fever associated with FMF attacks. Dialysis and/or renal transplant might become necessary in someone with advanced kidney disease. Given its genetic nature, there is no cure for FMF, nor is there likely to be in the near future. Any couple that has a child diagnosed with FMF, or anyone with a family history of the condition (especially those in high-risk ethnic groups), should be offered **genetic counseling** to obtain the most up-to-date information on FMF and testing options.

### Prognosis

For those individuals who are diagnosed early enough and take colchicine consistently, the prognosis is excellent. Most will have very few, if any, attacks of fever and polyserositis, and will likely not develop serious complications of amyloidosis. The problem of misdiagnosing FMF continues, but education attempts directed at both the public and medical care providers should improve the situation. Future research should provide a better understanding of the inflammation process, focusing on how neutrophils are genetically regulated. That information could then be used to develop treatments for FMF with fewer side effects, and might also assist in developing therapies for other diseases in which abnormal inflammation and immune response are a problem.

### Resources

#### BOOKS

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#### PERIODICALS

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- Samuels, Jonathan, et al. "Familial Mediterranean Fever at the Millennium: Clinical Spectrum, Ancient Mutations, and a Survey of 100 American Referrals to the National Institutes of Health." *Medicine* 77 (July 1998):268-97.

#### ORGANIZATIONS

- National Institute of Arthritis and Musculoskeletal and Skin Diseases. National Institutes of Health, One AMS Circle, Bethesda, MD 20892. <<http://www.nih.gov/niams>>.
- 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.rarediseases.org>>.
- National Society of Genetic Counselors. 233 Canterbury Dr., Wallingford, PA 19086-6617. (610) 872-1192. <<http://www.nsgc.org/GeneticCounselingYou.asp>>.

Scott J. Polzin, M.S.

## Familial polyposis

### Definition

Familial polyposis is an inherited condition which primarily affects the large intestine (colon and rectum). Large numbers of projecting masses of swollen and thickened or tumorous membrane (polyps) develop on the inner lining of this part of the bowel. The polyps eventually become malignant.

### Description

Familial polyposis (FP) is known by many synonyms, most include some combination of words which reflect what is known about the disease. As the disease is inherited, the word, family, is often included. Because these mushroom-like growths are the most obvious manifestation of the disorder, the word, polyp, is usually in the term as well. Adenoma refers to the particular kind of polyp that is typically discovered. Some of the names found in medical texts and journals include polyposis

coli, familial colonic polyposis, multiple familial polyposis, familial adenomatous colon polyposis, adenomatosis of the colon and rectum (ACR), and familial adenomatous polyposis (FAP). The last term and its abbreviation have been commonly used since the early 1990s. It will be used in this discussion.

Familial polyposis or familial adenomatous polyposis (FAP) is a premalignant disease. This means that a person with FAP, if left untreated, will invariably develop **cancer**. Individuals with this disorder grow hundreds of polyps throughout their large intestines. The polyps, which may also be called adenomas, commonly develop just after **puberty**. Approximately half of all FAP patients will have polyps by age 14. Ninety percent will have detectable polyps by age 25. Usually by age 35–40, one or more of these polyps will become cancerous.

FAP is a rare disease. One in 8,000 people in the United States have FAP. However, it may be very common in affected families. FAP is inherited in an autosomal dominant pattern. This means that a person with FAP has a 50% chance of passing the condition down to each of their children. FAP can also develop in someone with no family history of the disorder, due to a new genetic mutation in that individual. It is thought that approximately one percent of all colorectal cancers in the United States can be attributed to FAP.

### Causes and symptoms

FAP is caused by a portion of a gene that mutates or changes. The original cause of the mutation is unknown. Its exact role in FAP is not completely clear. Researchers theorize that the normal gene directs the manufacture of a protein which helps control cell growth. The mutated gene section in FAP generates an abnormal protein which does not perform its normal function. Cells grow out of control, causing the development of multiple, sometimes hundreds, of polyps. One or more of these eventually becomes cancerous.

Many individuals develop polyps without displaying any symptoms. Others experience such gastrointestinal problems as **diarrhea**, **constipation**, abdominal cramps, blood in the stool, or weight loss. FAP patients may also develop nonmalignant tumors (desmoid tumors), and/or some bone and dental abnormalities. In addition, they may exhibit a "spot" on the retina of the eye (congenital hypertrophy of the retinal pigment epithelium, or CHRPE).

Relatives of individuals with diagnosed FAP are at high risk of having the disease themselves. There are no other known risk factors for this condition.

## Diagnosis

The abnormal portion of the gene that causes FAP in most patients can be detected. A blood test can then be performed which identifies family members who have the same mutation. They will eventually develop the condition. Children who have a parent with FAP, and siblings of affected patients whose parental history is incomplete, should be evaluated. The polyps characteristic of FAP have been found in children as young as age five. Testing of appropriate individuals should take place as soon as the diagnosis of FAP is established in one member of a family.

Relatives of people with diagnosed FAP should **exercise** caution regarding where they seek advice and testing. One study of a commercially available blood test found that less than 20% of patients received any **genetic counseling**, and almost one third of their physicians misinterpreted the test results.

Registries for FAP patients can be found at many sites in the United States. Such a registry specializes in identification, assistance, and education of people with a particular disease, and is usually a separate department in a research hospital. A team of health professionals who have expertise in the disorder staff the registry.

Testing within a research setting and/or at a facility with a registry of patients with FAP is more likely to safeguard against problems, such as the misunderstanding of test results. As part of a research project, sometimes counseling as well as blood tests are available at no charge to the patient. Insurance coverage varies. Concerns about confidentiality, and future insurance and employment discrimination, may prompt individuals to pay for the examination out of pocket. Commercial blood tests cost approximately \$250 per sample.

If the abnormal gene is found in a family member, annual screening for colon polyps is recommended, beginning at age 11. Flexible **sigmoidoscopy** is used for this examination. It is usually done in a physician's office, or in a hospital department, most often by a gastroenterologist or a surgeon. Food intake may be restricted for 24 hours prior to the procedure. Before the study, the intestine is cleared of stool by one or more small **enemas**. Some physicians prefer to sedate the patient, to help them relax. Then a flexible, lighted, hollow tube (sigmoidoscope) is inserted into the anus and maneuvered into the large intestine. The physician examines the wall of the colon to look for polyps. If polyps are found, one or more may be removed for biopsy.

Most patients report little discomfort during the examination. The procedure itself takes five to fifteen minutes. The patient may be at the facility an hour, or more, if recovery from **sedation** is needed. If no medica-

tion was administered, driving and resumption of normal activities are permitted immediately. The cost of the procedure varies widely, but, as of 1997, it was covered by Medicare, indicating the likelihood of other types of insurance coverage.

In some cases the portion of the gene responsible for FAP cannot be identified. Family members of these patients cannot have a predictive blood test. The current recommendation is for these patients to have the same annual examination with flexible sigmoidoscopy as patients with a diagnosed FAP gene. A noninvasive screening **eye examination** to detect CHRPE, associated with FAP, may also be performed.

## Treatment

The only definitive treatment for FAP is surgical removal of the lower intestine. Since the goal is to prevent cancer, the operation is done as soon as adenomatous polyps are found on sigmoidoscopy. Waiting until a polyp becomes malignant is unsafe, as the cancer may invade surrounding tissues.

There are several choices about the type of surgery to treat this condition. Some authorities advocate removal of the colon, leaving the rectum or lowest portion of the intestine in place. The small intestine can be attached to the rectum, allowing normal bowel function. This is often called ileorectal anastomosis. Others argue that this section is also liable to develop polyps, needs to be monitored regularly, and may require eventual removal.

Excision of the entire lower intestine with preservation of normal bowel function is possible. This entails a more complex surgical procedure. The patient may experience more complications and a longer recovery period. However, the risk of polyp development in this area is very low. Periodic examination of the intestine may not be needed once healing is complete.

The more intricate surgery may be referred to as a J-pouch procedure, an ileal pouch-anal anastomosis, a restorative proctocolectomy, or an ileoanal reservoir procedure. It involves creating a "pouch" of tissue from the small intestine, which is attached to the anus. This serves as a reservoir or holding area for stool, much as the rectum does normally. The surgery is often done in several stages. A temporary ileostomy, which creates an opening of the small intestines onto the abdomen, is required. When all procedures are completed, and after a recuperation period, the patient regains normal bowel function through the anus.

Some researchers suggest that as **genetic testing** becomes more developed, the specific portion of the gene involved may dictate the type of surgery chosen. Those at high risk of developing **rectal polyps** may be

advised to have the more complex operation. FAP patients felt to be at lower risk for rectal polyps might be counseled to consider the less radical surgery.

Medical therapy to treat the adenomatous polyps has been attempted. Some **nonsteroidal anti-inflammatory drugs** have been effective in reducing the number and size of the polyps. It is possible that these agents will be used as an additional treatment for FAP, but they are unlikely to replace surgery.

Individuals with FAP are at increased risk for cancers of the upper digestive tract including the upper portion of the small bowel (duodenum) and the channels where bile flows (biliary tract). Cancers of the thyroid, pancreas, and adrenal gland are also more commonly found among FAP patients. Periodic examination for the development of malignancy in these areas is considered part of the treatment of FAP. In some cases, such as cancer involving the duodenum, the tests themselves carry a chance of complications. The risk of the study must be weighed against the potential benefits of knowing the results. Non-malignant growths, called desmoid tumors, also occur more frequently in patients with FAP. Although they are not malignant, they grow quickly into surrounding tissues, causing many difficulties, even **death** in some cases.

### Prognosis

The major cause of death in many patients with FAP remains colorectal cancer. One study suggested that even with improved disease recognition, social and emotional factors, such as fear of surgery, may significantly delay a patient's treatment. In recent years, the trend is towards mortality from other causes, such as desmoid tumors or cancers other than colorectal. It has been estimated that a patient with known FAP has a relative risk of dying over three times greater than that of the average population, at a given age.

### Prevention

FAP cannot be prevented. Aggressive diagnosis, treatment, and follow-up monitoring are keys to successful management of the disease.

### Resources

#### ORGANIZATIONS

Familial Polyposis Registry. Department of Colorectal Surgery.  
Cleveland Clinic Foundation. 9500 Euclid Ave., Cleveland OH 44195-5001. (216) 444-6470.  
National Organization for Rare Disorders. PO Box 8923, New Fairfield, CT 06812-8923. (800) 999-6673. <<http://www.rarediseases.org>>.

Ellen S. Weber, MSN

## KEY TERMS

**Gene**—The basic unit of heredity, made of DNA. Each gene occupies certain location on a chromosome.

**Mutation**—An alteration in a gene, especially one capable of producing a new trait, or a change in function.

## Family therapy

### Definition

Family therapy is a form of psychotherapy that involves all the members of a nuclear or extended family. It may be conducted by a pair or team of therapists. In many cases the team consists of a man and a woman in order to treat gender-related issues or serve as role models for family members. Although some forms of family therapy are based on behavioral or psychodynamic principles, the most widespread form is based on family systems theory. This approach regards the family, as a whole, as the unit of treatment, and emphasizes such factors as relationships and communication patterns rather than traits or symptoms in individual members.

Family therapy is a relatively recent development in psychotherapy. It began shortly after World War II, when doctors, who were treating schizophrenic patients, noticed that the patients' families communicated in disturbed ways. The doctors also found that the patients' symptoms rose or fell according to the level of tension between their parents. These observations led to considering a family as an organism or system with its own internal rules, patterns of functioning, and tendency to resist change. The therapists started to treat the families of schizophrenic patients as whole units rather than focusing on the hospitalized member. They found that in many cases the family member with **schizophrenia** improved when the "patient" was the family system. (This should not be misunderstood to mean that schizophrenia is caused by family problems, although family problems may worsen the condition.) This approach of involving the entire family in the treatment plan and therapy was then applied to families with problems other than the presence of schizophrenia.

Family therapy is becoming an increasingly common form of treatment as changes in American society are reflected in family structures. It has led to two further developments: couples therapy, which treats relationship

problems between marriage partners or gay couples; and the extension of family therapy to religious communities or other groups that resemble families.

## Purpose

Family therapy is often recommended in the following situations:

- Treatment of a family member with schizophrenia or **multiple personality disorder** (MPD). Family therapy helps other family members understand their relative's disorder and adjust to the psychological changes that may be occurring in the relative.
- Families with problems across generational boundaries. These would include problems caused by parents sharing housing with grandparents, or children being reared by grandparents.
- Families that deviate from social norms (common-law relationships, gay couples rearing children, etc.). These families may not have internal problems but may be troubled by outsiders' judgmental attitudes.
- Families with members from a mixture of racial, cultural, or religious backgrounds.
- Families who are scapegoating a member or undermining the treatment of a member in individual therapy.
- Families where the identified patient's problems seem inextricably tied to problems with other family members.
- Blended families with adjustment difficulties.

Most family therapists presuppose an average level of intelligence and education on the part of adult members of the family.

## Precautions

Some families are not considered suitable candidates for family therapy. They include:

- families in which one, or both, of the parents is psychotic or has been diagnosed with antisocial or paranoid personality disorder,
- families whose cultural or religious values are opposed to, or suspicious of, psychotherapy,
- families with members who cannot participate in treatment sessions because of physical illness or similar limitations,
- families with members with very rigid personality structures, (here, members might be at risk for an emotional or psychological crisis),
- families whose members cannot or will not be able to meet regularly for treatment,
- families that are unstable or on the verge of breakup.

## Description

Family therapy tends to be short-term treatment, usually several months in length, with a focus on resolving specific problems such as eating disorders, difficulties with school, or adjustments to bereavement or geographical relocation. It is not normally used for long-term or intensive restructuring of severely dysfunctional families.

In family therapy sessions, all members of the family and both therapists (if there is more than one) are present at most sessions. The therapists seek to analyze the process of family interaction and communication as a whole; they do not take sides with specific members. They may make occasional comments or remarks intended to help family members become more conscious of patterns or structures that had been previously taken for granted. Family therapists, who work as a team, also model new behaviors for the family through their interactions with each other during sessions.

Family therapy is based on family systems theory, which understands the family to be a living organism that is more than the sum of its individual members. Family therapy uses "systems" theory to evaluate family members in terms of their position or role within the system as a whole. Problems are treated by changing the way the system works rather than trying to "fix" a specific member. Family systems theory is based on several major concepts:

### *The identified patient*

The identified patient (IP) is the family member with the symptom that has brought the family into treatment. The concept of the IP is used by family therapists to keep the family from scapegoating the IP or using him or her as a way of avoiding problems in the rest of the system.

### *Homeostasis (balance)*

The concept of homeostasis means that the family system seeks to maintain its customary organization and functioning over time. It tends to resist change. The family therapist can use the concept of homeostasis to explain why a certain family symptom has surfaced at a given time, why a specific member has become the IP, and what is likely to happen when the family begins to change.

### *The extended family field*

The extended family field refers to the nuclear family, plus the network of grandparents and other members of the extended family. This concept is used to explain the intergenerational transmission of attitudes, problems, behaviors, and other issues.

### *Differentiation*

Differentiation refers to the ability of each family member to maintain his or her own sense of self, while

remaining emotionally connected to the family. One mark of a healthy family is its capacity to allow members to differentiate, while family members still feel that they are “members in good standing” of the family.

### ***Triangular relationships***

Family systems theory maintains that emotional relationships in families are usually triangular. Whenever any two persons in the family system have problems with each other, they will “triangle in” a third member as a way of stabilizing their own relationship. The triangles in a family system usually interlock in a way that maintains family homeostasis. Common family triangles include a child and its parents; two children and one parent; a parent, a child, and a grandparent; three siblings; or, husband, wife, and an in-law.

### **Preparation**

In some instances the family may have been referred to a specialist in family therapy by their pediatrician or other primary care provider. It is estimated that as many as 50% of office visits to pediatricians have to do with developmental problems in children that are affecting their families. Some family doctors use symptom checklists or psychological screeners to assess a family’s need for therapy.

Family therapists may be either psychiatrists, clinical psychologists, or other professionals certified by a specialty board in marriage and family therapy. They will usually evaluate a family for treatment by scheduling a series of interviews with the members of the immediate family, including young children, and significant or symptomatic members of the extended family. This process allows the therapist(s) to find out how each member of the family sees the problem, as well as to form first impressions of the family’s functioning. Family therapists typically look for the level and types of emotions expressed, patterns of dominance and submission, the roles played by family members, communication styles, and the locations of emotional triangles. They will also note whether these patterns are rigid or relatively flexible.

Preparation also usually includes drawing a genogram, which is a diagram that depicts significant persons and events in the family’s history. Genograms also include annotations about the medical history and major personality traits of each member. Genograms help in uncovering intergenerational patterns of behavior, marriage choices, family alliances and conflicts, the existence of family secrets, and other information that sheds light on the family’s present situation.

### **Risks**

The chief risk in family therapy is the possible unsettling of rigid personality defenses in individuals, or

## **KEY TERMS**

**Blended family**—A family formed by the remarriage of a divorced or widowed parent. It includes the new husband and wife, plus some or all of their children from previous marriages.

**Differentiation**—The ability to retain one’s identity within a family system while maintaining emotional connections with the other members.

**Extended family field**—A person’s family of origin plus grandparents, in-laws, and other relatives.

**Family systems theory**—An approach to treatment that emphasizes the interdependency of family members rather than focusing on individuals in isolation from the family. This theory underlies the most influential forms of contemporary family therapy.

**Genogram**—A family tree diagram that represents the names, birth order, sex, and relationships of the members of a family. Therapists use genograms to detect recurrent patterns in the family history and to help the members understand their problem(s).

**Homeostasis**—The tendency of a family system to maintain internal stability and resist change.

**Identified patient (IP)**—The family member in whom the family’s symptom has emerged or is most obvious.

**Nuclear family**—The basic family unit, consisting of father, mother, and their biological children.

**Triangling**—A process in which two family members lower the tension level between them by drawing in a third member.

couple relationships that had been fragile before the beginning of therapy. Intensive family therapy may also be difficult for psychotic family members.

### **Normal results**

Normal results vary, but in good circumstances, they include greater insight, increased differentiation of individual family members, improved communication within the family, loosening of previously automatic behavior patterns, and resolution of the problem that led the family to seek treatment.

### **Resources**

#### **BOOKS**

Clark, R. Barkley. “Psychosocial Aspects of Pediatrics & Psychiatric Disorders: Psychosocial Assessment of Children

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Rebecca J. Frey

Famine fever see **Relapsing fever**

## **Fanconi's syndrome**

### **Definition**

Fanconi's syndrome is a set of kidney malfunctions brought about by a variety of seemingly unrelated disorders. Kidney malfunction leads to excessive urine production and excessive thirst, resulting in deficits of water, calcium, potassium, magnesium, and other substances in the body. It often leads to bone disease and stunted growth.

### **Description**

Normally, kidneys cleanse the blood and keep its salt, water, and acidity in balance, leaving what the body needs in the blood and putting what the body doesn't need into the urine, which leaves the body. This task is performed in two steps. First, the blood is filtered through a kidney structure with small holes that keep the cells and large molecules in the blood. Second, some of the small molecules in the filtrate, needed by the body, are reabsorbed and returned to the bloodstream.

This reabsorption step is defective in Fanconi's syndrome. As a consequence, substances that are normally reabsorbed, like glucose, amino acids, small proteins, water, calcium, potassium, magnesium, bicarbonate, and phosphate, are lost and the body becomes overly acidic.

Fanconi's syndrome is also known as Fanconi syndrome, renal Fanconi syndrome, Fanconi renaltubular

syndrome, and Lignac-de Toni-Debré-Fanconi syndrome. Fanconi's anemia is, however, a totally different disease.

### **Causes and symptoms**

#### **Causes**

Fanconi's syndrome can be caused by a variety of genetic defects and by certain environmental assaults.

The genetic diseases known to give rise to Fanconi's syndrome are cystinosis (the most common cause in children), **galactosemia**, glycogen storage disease, **hereditary fructose intolerance**, Lowe syndrome, **Wilson disease**, tyrosinemia, medullary cystic disease, vitamin D dependency, and familial idiopathic Fanconi's syndrome.

Environmental assaults that cause Fanconi's syndrome include exposure to heavy metals (like cadmium, lead, mercury, platinum, uranium), certain drugs (like outdated tetracycline and gentamicin), other substances (like Lysol, paraquat, toluene, the amino acid lysine taken as a nutritional supplement), and **kidney transplantation**.

#### **Symptoms**

Fanconi's syndrome symptoms related directly to impaired absorption include excessive urine production and urination; excessive thirst; **dehydration**; **constipation**; **anorexia nervosa**; vomiting; elevated levels of glucose, phosphate, calcium, uric acid, amino acids, and protein (especially beta<sub>2</sub>-microglobulin and lysozyme) in the urine; elevated levels of chloride and decreased levels of phosphate and calcium in the blood; and excessively acidic blood.

The most noticeable indirect consequences of impaired reabsorption are the bone diseases, rickets and osteomalacia. Rickets affects children and is associated with bone deformities, failure to grow, and difficulty walking. If a person acquires Fanconi's syndrome as an adult, the bone disease is termed osteomalacia and is accompanied by severe bone **pain** and spontaneous **fractures**. Unlike rickets due to **malnutrition**, these diseases cannot be reversed with vitamin D. Muscle weakness and occasional **paralysis** are other indirect consequences of the ineffective reabsorption.

### **Diagnosis**

Diagnosis of Fanconi's syndrome can be made by urine and blood tests. It is also important to find the underlying cause to decide on the best treatment. Other symptoms specific to a particular patient will point to other useful diagnostic tests. For example, high levels of

blood galactose in conjunction with symptoms of Fanconi's syndrome indicate the patient is suffering from galactosemia, while high blood levels of cadmium indicate the patient is suffering from cadmium **poisoning**.

## Treatment

Fanconi's syndrome is best treated by attacking the underlying cause whenever possible. For example, when cystinosis is treated with the drug cysteamine to lower cystine levels in the body or Wilson disease is treated with penicillamine to lower the levels of copper, accompanying symptoms of Fanconi's syndrome will subside. If the patient has acquired the disease from a heavy metal or another toxic agent, all contact with the toxic agent should stop; the condition will then likely disappear.

Nevertheless, additional treatment will be necessary either when it's not possible to treat the underlying cause or while waiting for the kidneys to resume normal function. This is done by restricting sodium chloride (table salt), giving **antacids** to counteract the excessive acidity of the blood, and supplying potassium supplements.

Kidney transplant is the treatment of last resort, used for patients whose kidneys have failed.

## Prognosis

Fanconi's syndrome can be reversible. Fanconi's syndrome caused by kidney transplantation usually reverses itself within the first year after transplant surgery. When caused by a toxin in the environment, Fanconi's syndrome generally can be reversed by removing the causative agent from the patient's environment. If it is caused by a genetic disease, it can usually be reversed by treating the disease. However, if Fanconi's syndrome is not treated or if treatment is unsuccessful, the kidneys can fail.

## Prevention

Fanconi's syndrome caused secondarily by the genetic diseases galactosemia, glycogen storage disease, hereditary fructose intolerance, and tyrosinemia is prevented by appropriate dietary restrictions to treat the genetic disease, starting in infancy.

Fanconi's syndrome caused by heavy metals and other toxins can be prevented by avoiding these substances.

## Resources

### BOOKS

Bergeron, Michael, André Gouaux, and Patrick Vinay. "The Renal Fanconi Syndrome." In *The Metabolic and Molecular Bases of Inherited Disease*. 7th ed. Ed. Charles R. Scriver, et al. New York: McGraw-Hill, Inc., 1995.

## KEY TERMS

**Acidosis**—Condition where the body is more acidic than normal; associated with headache, nausea, vomiting, and visual disturbances.

**Fanconi's anemia**—An inherited form of aplastic anemia.

**Filtrate**—The part of filtered material that flows through the filter.

**Idiopathic**—Refers to a disease of unknown cause.

**Polydipsia**—Excessive thirst.

**Polyuria**—Excessive production of urine.

## ORGANIZATIONS

The American Society of Nephrology. 2025 M Street NW #800, Washington, DC 20036. (202) 367-1190. <<http://www ASN-online.com>>.

National Kidney Foundation. 30 East 33rd St., New York, NY 10016. (800) 622-9010. <<http://www.kidney.org>>.

## OTHER

"Online Mendelian Inheritance in Man." OMIM Homepage. <<http://www.ncbi.nlm.nih.gov/Omim>>.

Lorraine Lica, PhD

Farsightedness see **Hyperopia**

FAS see **Fetal alcohol syndrome**

## Fasciotomy

### Definition

Fasciotomy is a surgical procedure that cuts away the fascia to relieve tension or pressure

### Purpose

The fascia is thin connective tissue covering, or separating, the muscles and internal organs of the body. It varies in thickness, density, elasticity, and composition, and is different from ligaments and tendons.

The fascia can be injured either through constant strain or through trauma. Fasciitis is an inflammation of the fascia. The most common condition for which fasciotomy is performed is plantar fasciitis, an inflammation of the fascia on the bottom of the foot that is sometimes called a heel spur or stone bruise.

Plantar fasciitis is caused by long periods on the feet, being overweight, and wearing shoes that do not support the foot well. Teachers, mail carriers, runners, and others who make heavy use of their feet are especially likely to suffer from plantar fasciitis.

Plantar fasciitis results in moderate to disabling heel pain. If nine to twelve months of conservative treatment (reducing time on feet, non-steroid anti-inflammatory drugs, arch supports) under the supervision of a doctor does not result in pain relief, a fasciotomy may be done. Fasciotomy removes a small portion of the fascia to relieve tension and pain. Connective tissue grows back into the cut space left by the cut, effectively lengthening the fascia.

When a fasciotomy is performed on other parts of the body, it is usually done to relieve pressure from a compression injury to a limb. This type of injury often occurs during contact sports. The blood vessels of the limb are damaged. They swell and leak, causing inflammation. Fluid builds up in the area contained by the fascia. A fasciotomy is done to relieve this pressure and prevent tissue death. Similar injury occurs in high voltage electrical burns where deep tissue damage occurs.

### Precautions

In the case of injury, fasciotomy is done on an emergency basis, and the outcome of the surgery depends largely on the general health of the patient. Plantar fasciotomies are appropriate for most people whose foot problems cannot be resolved in any other way.

### Description

Fasciotomy in the limbs is usually done by a surgeon under general or regional anesthesia. An incision is made in the skin, and a small area of fascia is removed where it will best relieve pressure. Then the incision is closed.

Plantar fasciotomy is an endoscopic (performed with the use of an endoscope) procedure. It is done by a foot specialist in a doctor's office or outpatient surgical clinic under local anesthesia and takes 20 minutes to one hour. The doctor makes two small incisions on either side of the heel. An endoscope is inserted in one to guide the doctor in where to cut. A tiny knife is inserted in the other. A portion of the fascia is cut from near the heel; then the incisions are closed.

### Preparation

Little preparation is done before a fasciotomy. When the fasciotomy is related to burn injuries, the fluid and electrolyte status of the patient are constantly monitored.

## KEY TERMS

**Endoscope**—A tube that contains a tiny camera and light, that is inserted in the body to allow a doctor to see inside without making a large incision.

### Aftercare

Aftercare depends on the reason for the fasciotomy. People who have endoscopic plantar fasciotomy can walk without pain almost immediately, return to wearing their regular shoes within three to five days, and return to normal activities within three weeks. Most will need to wear arch supports in their shoes.

### Risks

In endoscopic plantar fasciotomy, the greatest risk is that the arch will drop slightly as a result of this surgery, causing other foot problems. Risks involved with other types of fasciotomy are those associated with the administration of anesthesia and the development of blood clots.

### Normal results

Fasciotomy in the limbs reduces pressure, thus reducing tissue death. Endoscopic plantar fasciotomy has a success rate of 90–95%.

### Resources

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Tish Davidson

## Fasting

### Definition

Fasting is voluntarily not eating food for varying lengths of time. Fasting is used as a medical therapy for many conditions. It is also a spiritual practice in many religions.

## Purpose

Fasting can be used for nearly every chronic condition, including **allergies**, **anxiety**, arthritis, **asthma**, depression, diabetes, headaches, heart disease, **high cholesterol**, low blood sugar, digestive disorders, mental illness, and **obesity**. Fasting is an effective and safe weight loss method. It is frequently prescribed as a **detoxification** treatment for those with conditions that may be influenced by environmental factors, such as **cancer** and **multiple chemical sensitivity**. Fasting has been used successfully to help treat people who have been exposed to high levels of toxic materials due to accident or occupation. Fasting is thought to be beneficial as a preventative measure to increase overall health, vitality, and resistance to disease. Fasting is also used as a method of mental and spiritual rejuvenation.

## Description

### Origins

Used for thousands of years, fasting is one of the oldest therapies in medicine. Many of the great doctors of ancient times and many of the oldest healing systems have recommended it as an integral method of healing and prevention. Hippocrates, the father of Western medicine, believed fasting enabled the body to heal itself. Paracelsus, another great healer in the Western tradition, wrote 500 years ago that “fasting is the greatest remedy, the physician within.” **Ayurvedic medicine**, the world’s oldest healing system, has long advocated fasting as a major treatment.

Fasting has also been used in nearly every religion in the world, including Christianity, Judaism, Buddhism, and Islam. Many of history’s great spiritual leaders fasted for mental and spiritual clarity, including Jesus, Buddha, and Mohammed. In one of the famous political acts of the last century, the Indian leader Mahatma Gandhi fasted for 21 days to promote peace.

Fasting has been used in Europe as a medical treatment for years. Many spas and treatment centers, particularly those in Germany, Sweden, and Russia, use medically supervised fasting. Fasting has gained popularity in American alternative medicine over the past several decades, and many doctors feel it is beneficial. Fasting is a central therapy in detoxification, a healing method founded on the principle that the build up of toxic substances in the body is responsible for many illnesses and conditions.

The principle of fasting is simple. When the intake of food is temporarily stopped, many systems of the body are given a break from the hard work of digestion. The extra energy gives the body the chance to heal and

restore itself, and burning stored calories gets rid of toxic substances stored in the body.

The digestive tract is the part of the body most exposed to environmental threats, including bacteria, viruses, parasites, and toxins. It requires the most immune system support. When food is broken down in the intestines, it travels through the blood to the liver, the largest organ of the body’s natural detoxification system. The liver breaks down and removes the toxic by-products produced by digestion, including natural ones and the chemicals now present in the food supply. During fasting, the liver and immune system are essentially freed to detoxify and heal other parts of the body.

Many healers claim that fasting is a particularly useful therapy for Americans and for the modern lifestyle, subjected to heavy **diets**, overeating, and constant exposure to food additives and chemicals. Some alternative practitioners have gone so far as to estimate that the average American is carrying 5-10 pounds of toxic substances in their bodies, for which fasting is the quickest and most effective means of removal.

### *Physiology of fasting*

Through evolution, the body became very efficient at storing energy and handling situations when no food was available. For many centuries, fasting was probably a normal occurrence for most people, and the body adapted to it. It is estimated that even very thin people can survive for 40 days or more without food. The body has a special mechanism that is initiated when no food is eaten. Fasting is not **starvation**, but rather the body’s burning of stored energy. Starvation occurs when the body no longer has any stored energy and begins using essential tissues such as organs for an energy source. Therapeutic fasts are stopped long before this happens.

Many physiological changes occur in the body during fasting. During the first day or so, the body uses its glycogen reserves, the sugars that are the basic energy supply. After these are depleted, the body begins using fat. However, the brain, which has high fuel requirements, still needs glucose (sugars converted from glycogen). To obtain glucose for the brain, the body begins to break down muscle tissue during the second day of the fast. Thus, during fasting some muscle loss will occur. To fuel the brain, the body would need to burn over a pound of muscle a day, but the body has developed another way to create energy that saves important muscle mass. This protein-sparing process is called ketosis, which occurs during the third day of a fast for men and the second day for women. In this highly efficient state, the liver begins converting stored fat and other nonessential tissues into ketones, which can be used by the brain, muscles, and

heart as energy. It is at this point in the fast that sensations of hunger generally go away, and many people experience normal or even increased energy levels. Hormone levels and certain functions become more stable in this state as well. The goal of most fasts is to allow the body to reach the ketosis state in order to burn excess fat and unneeded or damaged tissue. Thus, fasts longer than three days are generally recommended as therapy.

Weight loss occurs most rapidly during the first few days of a fast, up to 2 pounds per day. In following days, the figure drops to around 0.5 pound per day. An average weight loss of a pound a day for an entire fast can be expected.

### ***Performing a fast***

Fasts can be performed for varying lengths of time, depending on the person and his or her health requirements. For chronic conditions, therapists recommend from two to four weeks to get the most benefits. Seven-day fasts are also commonly performed. A popular fasting program for prevention and general health is a three-day fast taken four times per year, at the change of each season. These can be easily performed over long weekends. Preventative fasts of one day per week are used by many people as well.

Juice fasts are also used by many people, although these are not technically fasts. Juice fasts are less intensive than water fasts because the body doesn't reach the ketosis stage. The advantage of juice fasts is that fruit and vegetable drinks can supply extra energy and nutrients. People can fit a few days of juice fasting into their normal schedules without significant drops in energy. Juice fasts are also said to have cleansing and detoxifying effects. The disadvantage of juice fasts is that the body never gets to the ketosis stage, so these fasters are thought to lack the deep detoxification and healing effects of the water fast.

Medical supervision is recommended for any fast over three days. Most alternative medicine practitioners, such as homeopaths, naturopathic doctors, and ayurvedic doctors, can supervise and monitor patients during fasts. Those performing extended fasts and those with health conditions may require blood, urine, and other tests during fasting. There are many alternative health clinics that perform medically supervised fasts as well. Some conventional medical doctors may also supervise patients during fasts. Costs and insurance coverage vary, depending on the doctor, clinic, and requirements of the patient.

### ***Preparations***

Fasts must be entered and exited with care. To enter a fast, the diet should be gradually lightened over a few

days. First, heavy foods such as meats and dairy products should be eliminated for a day or two. Grains, nuts, and beans should then be reduced for several days. The day before a fast, only easily digested foods like fruits, light salads, and soups should be eaten. During the fast, only pure water and occasional herbal teas should be drunk.

Fasts should be ended as gradually as they are entered, going from lighter to heavier foods progressively. The diet after a fast should emphasize fresh, wholesome foods. Fasters should particularly take care not to overeat when they complete a fast.

### ***Precautions***

Fasting isn't appropriate for everyone and, in some cases, could be harmful. Any person undertaking a first fast longer than three days should seek medical supervision. Those with health conditions should always have medical support during fasting. Plenty of water should be taken by fasters since **dehydration** can occur. Saunas and sweating therapies are sometimes recommended to assist detoxification, but should be used sparingly. Those fasting should significantly slow down their lifestyles. Taking time off of work is helpful, or at least reducing the work load. Fasters should also get plenty of rest. **Exercise** should be kept light, such as walking and gentle stretching.

### ***Side effects***

Those fasting may experience side effects of **fatigue**, malaise, aches and pains, emotional duress, **acne**, headaches, allergies, swelling, vomiting, **bad breath**, and symptoms of colds and flu. These reactions are sometimes called *healing crises*, which are caused by temporarily increased levels of toxins in the body due to elimination and cleansing. Lower energy levels should be expected during a fast.

### ***Research and general acceptance***

The physiology of fasting has been widely studied and documented by medical science. Beneficial effects such as lowered cholesterol and improved general functioning have been shown. Fasting as a treatment for illness and disease has been studied less, although some studies around the world have shown beneficial results. A 1984 study showed that workers in Taiwan who had severe chemical **poisoning** had dramatic improvement after a ten-day fast. In Russia and Japan, studies have demonstrated fasting to be an effective treatment for mental illness. Fasting has been featured on the cover of medical journals, although mainstream medicine has generally ignored fasting and detoxification treatments as valid medical procedures.

## KEY TERMS

**Ayurvedic medicine**—A traditional healing system developed in India.

**Toxin**—A substance that has poisonous effects on the body.

The majority of research that exists on fasting is testimonial, consisting of individual personal accounts of healing without statistics or controlled scientific experiments. In the alternative medical community, fasting is an essential and widely accepted treatment for many illnesses and chronic conditions.

### Resources

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#### ORGANIZATIONS

Fasting Center International. 32 West Anapurna St., #360, Santa Barbara, CA 93101. <<http://www.fasting.com>>.

Douglas Dupler

Fasting blood sugar test see **Blood sugar tests**

Fasting plasma glucose test see **Blood sugar tests**

exhaustion that goes beyond normal sleepiness, it is usually a sign that something more serious is amiss.

Physically, fatigue is characterized by a profound lack of energy, feelings of muscle weakness, and slowed movements or central nervous system reactions. Fatigue can also trigger serious mental exhaustion. Persistent fatigue can cause a lack of mental clarity (or feeling of mental “fuzziness”), difficulty concentrating, and in some cases, memory loss.

### Causes and symptoms

Fatigue may be the result of one or more environmental causes such as inadequate rest, improper diet, work and home stressors, or poor physical conditioning, or one symptom of a chronic medical condition or disease process in the body. Heart disease, low blood pressure, diabetes, end-stage renal disease, iron-deficiency anemia, **narcolepsy**, and **cancer** can cause long-term, ongoing fatigue symptoms. Acute illnesses such as viral and bacterial infections can also trigger temporary feelings of exhaustion. In addition, mental disorders such as depression can also cause fatigue.

A number of medications, including **antihistamines**, **antibiotics**, and blood pressure medications, may cause drowsiness as a side-effect. Individuals already suffering from fatigue who are prescribed one of these medications may wish to check with their healthcare provider about alternative treatments.

Extreme fatigue which persists, unabated, for at least six months, is not the result of a diagnosed disease or illness, and is characterized by flu-like symptoms such as swollen lymph nodes, **sore throat**, and muscle weakness and/or **pain** may indicate a diagnosis of **chronic fatigue syndrome**. Chronic fatigue syndrome (sometimes called chronic fatigue immune deficiency syndrome), is a debilitating illness that causes overwhelming exhaustion and a constellation of neurological and immunological symptoms. Between 1.5 and 2 million Americans are estimated to suffer from the disorder.

## Fatigue

### Definition

Fatigue is physical and/or mental exhaustion that can be triggered by **stress**, medication, overwork, or mental and physical illness or disease.

### Description

Everyone experiences fatigue occasionally. It is the body's way of signaling its need for rest and sleep. But when fatigue becomes a persistent feeling of tiredness or

### Diagnosis

Because fatigue is a symptom of a number of different disorders, diseases, and lifestyle choices, diagnosis may be difficult. A thorough examination and patient history by a qualified healthcare provider is the first step in determining the cause of the fatigue. A physician can rule out physical conditions and diseases that feature fatigue as a symptom, and can also determine if prescription drugs, poor dietary habits, work environment, or other external stressors could be triggering the exhaustion. Several diagnostic tests may also be required to rule

out common physical causes of exhaustion, such as blood tests to check for iron-deficiency anemia.

Diagnosis of chronic fatigue syndrome is significantly more difficult. Because there is no specific biological marker or conclusive blood test to check for the disorder, healthcare providers must rely on the patient's presentation and severity of symptoms to make a diagnosis. In many cases, individuals with chronic fatigue syndrome go through a battery of invasive diagnostic tests and several years of consultation with medical professionals before receiving a correct diagnosis.

### Treatment

Conventional medicine recommends the dietary and lifestyle changes outlined above as a first line of defense against fatigue. Individuals who experience occasional fatigue symptoms may benefit from short term use of caffeine-containing central nervous stimulants, which make people more alert, less drowsy, and improve coordination. However, these should be prescribed with extreme caution, as overuse of the drug can lead to serious **sleep disorders**, like **insomnia**.

Another reason to avoid extended use of **caffeine** is its associated withdrawal symptoms. 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 an individual stops using caffeine abruptly, withdrawal symptoms may occur, including **headache**, fatigue, drowsiness, yawning, irritability, restlessness, vomiting, or runny nose. These symptoms can go on for as long as a week.

### Alternative treatment

The treatment of fatigue depends on its direct cause, but there are several commonly prescribed treatments for non-specific fatigue, including dietary and lifestyle changes, the use of essential oils and herbal therapies, deep breathing exercises, **traditional Chinese medicine**, and color therapy.

#### Dietary changes

Inadequate or inappropriate nutritional intake can cause fatigue symptoms. To maintain an adequate energy supply and promote overall physical well-being, individuals should eat a balanced diet and observe the following nutritional guidelines:

- Drink plenty of water. Individuals should try to drink 9 to 12 glasses of water a day. **Dehydration** can reduce blood volume, which leads to feelings of fatigue.

- Eat iron-rich foods (i.e., liver, raisins, spinach, apricots). Iron enables the blood to transport oxygen throughout the tissues, organs, and muscles, and diminished oxygenation of the blood can result in fatigue.
- Avoid high-fat meals and snacks. High fat foods take longer to digest, reducing blood flow to the brain, heart, and rest of the body while blood flow is increased to the stomach.
- Eat unrefined carbohydrates and proteins together for sustained energy.
- Balance proteins. Limiting protein to 15–20 grams per meal and two snacks of 15 grams is recommended, but not getting enough protein adds to fatigue. Pregnant or breastfeeding women should get more protein.
- Get the recommended daily allowance of B complex **vitamins** (specifically, pantothenic acid, **folic acid**, thiamine, and vitamin B<sub>12</sub>). Deficiencies in these vitamins can trigger fatigue.
- Get the recommended daily allowance of selenium, riboflavin, and niacin. These are all essential nutritional elements in metabolizing food energy.
- Control portions. Individuals should only eat when they're hungry, and stop when they're full. An overstuffed stomach can cause short-term fatigue, and individuals who are overweight are much more likely to regularly experience fatigue symptoms.

#### Lifestyle changes

Lifestyle factors such as a high-stress job, erratic work hours, lack of social or family support, or erratic sleep patterns can all cause prolonged fatigue. If stress is an issue, a number of relaxation therapies and techniques are available to help alleviate tension, including massage, **yoga**, **aromatherapy**, **hydrotherapy**, progressive relaxation exercises, **meditation**, and **guided imagery**. Some individuals may also benefit from individual or family counseling or psychotherapy sessions to work through stress-related fatigue that is a result of family or social issues.

Maintaining healthy sleep patterns is critical to proper rest. Having a set "bedtime" helps to keep sleep on schedule. A calm and restful sleeping environment is also important to healthy sleep. Above all, the bedroom should be quiet and comfortable, away from loud noises and with adequate window treatments to keep sunlight and streetlights out. Removing distractions from the bedroom such as televisions and telephones can also be helpful.

#### Essential oils

Aromatherapists, hydrotherapists, and other holistic healthcare providers may recommend the use of essential

oils of rosemary (*Rosmarinus officinalis*), eucalyptus blue gum (*Eucalyptus globulus*), peppermint, (*Mentha x piperata*), or scots pine oil (*Pinus sylvestris*) to stimulate the nervous system and reduce fatigue. These oils can be added to bathwater or massage oil as a topical application. Citrus oils such as lemon, orange, grapefruit, and lime have a similar effect, and can be added to a steam bath or vaporizer for inhalation.

### **Herbal remedies**

Herbal remedies that act as circulatory stimulants can offset the symptoms of fatigue in some individuals. An herbalist may recommend an infusion of ginger (*Zingiber officinale*) root or treatment with cayenne (*Capsicum annum*), balimony (*Chelone glabra*), damiana (*Turnera diffusa*), ginseng (*Panax ginseng*), or rosemary (*Rosmarinus officinalis*) to treat ongoing fatigue.

An infusion is prepared by mixing the herb with boiling water, steeping it for several minutes, and then removing the herb from the infusion before drinking. A strainer, tea ball, or infuser can be used to immerse loose herb in the boiling water before steeping and separating it. A second method of infusion is to mix the loose herbal preparation with cold water first, bringing the mixture to a boil in a pan or teapot, and then separating the tea from the infusion with a strainer before drinking.

Caffeine-containing **central nervous system stimulants** such as tea (*Camellia senensis*) and cola (*Cola nitida*) can provide temporary, short-term relief of fatigue symptoms. However, long-term use of caffeine can cause restlessness, irritability, and other unwanted side effects, and in some cases may actually work to increase fatigue after the stimulating effects of the caffeine wear off. To avoid these problems, caffeine intake should be limited to 300 mg or less a day (the equivalent of 4-8 cups of brewed, hot tea).

### **Traditional Chinese medicine**

Chinese medicine regards fatigue as a blockage or misalignment of *qi*, or energy flow, inside the human body. The practitioner of Chinese medicine chooses **acupuncture** and/or herbal therapy to rebalance the entire system. The Chinese formula Minot Bupleurum soup (or Xiao Chia Hu Tang) has been used for nearly 2,000 years for the type of chronic fatigue that comes after the flu. In this condition, the person has low-grade **fever**, nausea, and fatigue. There are other formulas that are helpful in other cases. Acupuncture involves the placement of a series of thin needles into the skin at targeted locations on the body known as acupoints in order to harmonize the energy flow within the human body.

## **KEY TERMS**

**Aromatherapy**—The therapeutic use of plant-derived, aromatic essential oils to promote physical and psychological well-being.

**Guided imagery**—The use of relaxation and mental visualization to improve mood and/or physical well-being.

**Hydrotherapy**—Hydrotherapy, or water therapy, is use of water (hot, cold, steam, or ice) to relieve discomfort and promote physical well-being.

### **Deep breathing exercises**

Individuals under stress often experience fast, shallow breathing. This type of breathing, known as chest breathing, can lead to **shortness of breath**, increased muscle tension, inadequate oxygenation of blood, and fatigue. Breathing exercises can both improve respiratory function and relieve stress and fatigue.

Deep breathing exercises are best performed while laying flat on the back on a hard surface, usually the floor. The knees are bent, and the body (particularly the mouth, nose, and face) is relaxed. One hand should be placed on the chest and one on the abdomen to monitor breathing technique. With proper breathing techniques, the abdomen will rise further than the chest. The individual takes a series of long, deep breaths through the nose, attempting to raise the abdomen instead of the chest. Air is exhaled through the relaxed mouth. Deep breathing can be continued for up to 20 minutes. After the **exercise** is complete, the individual checks again for body tension and relaxation. Once deep breathing techniques have been mastered, an individual can use deep breathing at any time or place as a quick method of relieving tension and preventing fatigue.

### **Color therapy**

Color therapy, also known as chromatherapy, is based on the premise that certain colors are infused with healing energies. The therapy uses the seven colors of the rainbow to promote balance and healing in the mind and body. Red promotes energy, empowerment, and stimulation. Physically, it is thought to improve circulation and stimulate red blood cell production. Red is associated with the seventh chakra, located at the root; or base of spine. In yoga, the chakras are specific spiritual energy centers of the body.

Therapeutic color can be administered in a number of ways. Practitioners of Ayurvedic, or traditional Indian

medicine, wrap their patients in colored cloth chosen for its therapeutic hue. Individuals suffering from fatigue would be wrapped in reds and oranges chosen for their uplifting and energizing properties. Patients may also be bathed in light from a color filtered light source to enhance the healing effects of the treatment.

Individuals may also be treated with color-infused water. This is achieved by placing translucent red colored paper or colored plastic wrap over and around a glass of water and placing the glass in direct sunlight so the water can soak up the healing properties and vibrations of the color. Environmental color sources may also be used to promote feelings of stimulation and energy. Red wall and window treatments, furniture, clothing, and even food may be recommended for their energizing healing properties.

Color therapy can be used in conjunction with both hydrotherapy and aromatherapy to heighten the therapeutic effect. Spas and holistic healthcare providers may recommend red color baths or soaks, which combine the benefits of a warm or hot water soak with energizing essential oils and the fatigue-fighting effects of bright red hues used in color therapy.

### Prognosis

Fatigue related to a chronic disease or condition may last indefinitely, but can be alleviated to a degree through some of the treatment options outlined here. Exhaustion that can be linked to environmental stressors is usually easily alleviated when those stressors are dealt with properly.

There is no known cure for chronic fatigue syndrome, but steps can be taken to lessen symptoms and improve quality of life for these individuals while researchers continue to seek a cure.

### Prevention

Many of the treatments outlined above are also recommended to prevent the onset of fatigue. Getting adequate rest and maintaining a consistent bedtime schedule are the most effective ways to combat fatigue. A balanced diet and moderate exercise program are also important to maintaining a consistent energy level.

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Paula Ford-Martin

## Fatty liver

### Definition

Fatty liver is the collection of excessive amounts of triglycerides and other fats inside liver cells.

### Description

Also called steatosis, fatty liver can be a temporary or long-term condition, which is not harmful itself, but may indicate some other type of problem. Left untreated, it can contribute to other illnesses. It is usually reversible once the cause of the problem is diagnosed and corrected. The liver is the organ responsible for changing fats eaten in the diet to types of fat that can be stored and used by the body. Triglycerides are one of the forms of fat stored by the body and used for energy and new cell formation. The break down of fats in the liver can be disrupted by **alcoholism, malnutrition, pregnancy, or poisoning**. In fatty liver, large droplets of fat, containing mostly triglycerides, collect within cells of the liver. The condition is generally not painful and may go unnoticed for a long period of time. In severe cases, the liver can increase to over three times its normal size and may be painful and tender.

### Causes and symptoms

The most common cause of fatty liver in the United States is alcoholism. In alcoholic fatty liver, over consumption of alcohol changes the way that the liver breaks down and stores fats. Often, people with chronic alcoholism also suffer from malnutrition by eating irregularly and not consuming a balanced diet. Conditions that can also cause fatty liver are other forms of malnutrition (especially when there is not enough protein in the diet), **obesity, diabetes mellitus, and Reye's syndrome** in children. Pregnancy can cause a rare, but serious form of fatty liver that starts late in pregnancy and may be associated with **jaundice** and liver failure. Some drug overdoses or toxic chemical poisonings, such as carbon tetrachloride, can also cause fatty liver.

Often, there are no symptoms associated with fatty liver. If there are symptoms, they can include **pain** under the rib cage on the right side of the body, swelling of the

abdomen, jaundice, and **fever**. Symptoms that occur less often in alcoholic fatty liver, but more often in pregnancy related fatty liver, are nausea, vomiting, loss of appetite, and abdominal pain.

## Diagnosis

During a **physical examination**, a doctor might notice that the liver is enlarged and tender when the abdomen is palpated (examined with the tips of the fingers while the patient lies flat). Blood tests may be used to determine if the liver is functioning properly. A **liver biopsy**, where a small sample of liver tissue is removed with a long needle or through a very small incision, can be used to confirm fatty liver. In pregnant women, the fatty liver condition is usually associated with another serious complication, pre-eclampsia or eclampsia. In this condition, the mother has seriously high blood pressure, swelling, and possibly, seizures. Laboratory abnormalities include elevations of the SGOT (serum glutamic-oxaloacetic transaminase) and SGPT (serum glutamic pyruvic transaminase). In many cases the alkaline phosphatase will be significantly elevated due to **cholestasis** produced by the fatty infiltration.

## Treatment

Treatment involves correcting the condition that caused fatty liver and providing supportive care. In fatty liver caused by alcoholism, the treatment is to give up drinking alcohol and to eat a healthy, well balanced diet. In fatty liver associated with pregnancy, the recommended treatment is to deliver the baby, if the pregnancy is far enough along. Vitamin and mineral supplements along with nutritional support may be useful.

## Prognosis

Fatty liver is usually reversible if recognized and treated. There may be some long-term tendency toward other types of liver problems depending on how long and how severe the fatty liver condition was. In pregnant women with the condition, the situation can be life threatening for both the mother and the infant. Left untreated, there is a high risk of **death** for both the mother and baby. Severe liver damage that may require a liver transplant can occur in the mother if the condition is not recognized early.

## Prevention

Prevention consists of maintaining a well balanced diet and healthy lifestyle with moderate or no alcohol consumption. Pregnant women require good prenatal care so that symptoms can be recognized and treated as early as possi-

## KEY TERMS

**Jaundice**—A condition where the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood.

**Reye's syndrome**—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. In fatal cases, there is evidence of accumulation of fat in the liver.

**Triglycerides**—A type of fat consumed in the diet and produced by and stored in the body as an energy source.

ble. To prevent Reye's syndrome, children should not be given **aspirin** to treat symptoms of the flu or other viruses.

## Resources

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Altha Roberts Edgren

Febrile agglutination tests see **Fever evaluation tests**

Fecal fat test see **Stool fat test**

## Fecal incontinence

### Definition

Fecal incontinence is the inability to control the passage of gas or stools (feces) through the anus. For some

people fecal incontinence is a relatively minor problem, as when it is limited to a slight occasional soiling of underwear, but for other people it involves a considerable loss of bowel control and has a devastating effect on quality of life and psychological well-being. Fortunately, professional medical treatment is usually able to restore bowel control or at least substantially reduce the severity of the condition.

## Description

Fecal incontinence, also called bowel incontinence, can occur at any age, but is most common among people over the age of 65, who sometimes have to cope with **urinary incontinence** as well. It was reported in 1998 that about 2% of adults experience fecal incontinence at least once a week whereas for healthy independent adults over the age of 65 the figure is about 7%. An extensive American survey, published in 1993, found fecal soiling in 7.1% of the surveyed population, with gross incontinence in 0.7%. For men and women the incidence of soiling was the same, but women were almost twice as likely to suffer from gross incontinence.

The wider public health impact of fecal incontinence is considerable. In the United States, more than \$400 million is spent each year on disposable underwear and other incontinence aids. Fecal incontinence is the second most common reason for seeking a nursing home placement. One-third of the institutionalized elderly suffer from this condition. Incontinence sufferers, however, often hesitate to ask their doctors for help because they are embarrassed or ashamed. The 1993 American survey discovered that only one-sixth of those experiencing soiling had sought medical advice, and only one-half of those afflicted with gross incontinence.

## Causes and symptoms

Fecal incontinence can result from a wide variety of medical conditions, including childbirth-related anal injuries, other causes of damage to the anus or rectum, and nervous system problems.

Vaginal-delivery **childbirth** is a major cause of fecal incontinence. In many cases, childbirth results in damage to the anal sphincter, which is the ring of muscle that closes the anus and keeps stools within the rectum until a person can find an appropriate opportunity to defecate. Nerve injuries during childbirth may also be a factor in some cases. An ultrasound study of first-time mothers found sphincter injuries in 35%. About one-third of the injured women developed fecal incontinence or an uncontrollable and powerful urge to defecate (urgency) within six weeks of giving birth. Childbirth-related

incontinence is usually restricted to gas, but for some women involves the passing of liquid or solid stools.

The removal of **hemorrhoids** by surgery or other techniques (hemorrhoidectomies) can also cause anal damage and fecal incontinence, as can more complex operations affecting the anus and surrounding areas. Anal and rectal infections as well as **Crohn's disease** can lead to incontinence by damaging the muscles that control defecation. For some people, incontinence becomes a problem when the anal muscles begin to weaken in midlife or old age.

**Dementia, mental retardation, strokes, brain tumors, multiple sclerosis,** and other conditions that affect the nervous system can cause fecal incontinence by interfering with muscle function or the normal rectal sensations that trigger sphincter contraction and are necessary for bowel control. One study of multiple sclerosis patients discovered that about half were incontinent. Nerve damage caused by long-lasting **diabetes mellitus (diabetic neuropathy)** is another condition that can give rise to incontinence.

## Diagnosis

Medical assessments in cases of fecal incontinence typically involve three steps: asking questions about the patient's past and current health (the medical history); a **physical examination** of the anal region; and testing for objective information regarding anal and rectal function.

### Patient history

The medical history relies on questions that allow the doctor to evaluate the nature and severity of the problem and its effect on the patient's life. The doctor asks, for instance, how long the patient has been suffering from incontinence; how often and under what circumstances incontinence occurs; whether the patient has any control over defecation; and whether the patient has obstacles to defecation in his or her everyday surroundings, such as a toilet that can be reached only by climbing a long flight of stairs. For women who have given birth, a detailed obstetric history is also necessary.

### Physical examination

The physical examination begins with a visual inspection of the anus and the area lying between the anus and the genitals (the perineum) for hemorrhoids, infections, and other conditions that might explain the patient's difficulties. During this phase of the examination the doctor asks the patient to bear down. Bearing down enables the doctor to check whether **rectal pro-**

**lapse** or certain other problems exist. Rectal prolapse means that the patient's rectum has been weakened and drops down through the anus. Next, the doctor uses a pin or probe to **stroke** the perianal skin. Normally this touching causes the anal sphincter to contract and the anus to pucker; if it does not, nerve damage may be present. The final phase of the examination requires the doctor to examine internal structures by carefully inserting a gloved and lubricated finger into the anal canal. This allows the doctor to judge the strength of the anal sphincter and a key muscle (the puborectalis muscle) in maintaining continence; to look for abnormalities such as scars and rectal masses; and to learn many other things about the patient's medical situation. At this point the doctor performs the anal wink test again and asks the patient to squeeze and bear down.

#### Laboratory tests

Information from the medical history and physical examination usually needs to be supplemented by tests that provide objective measurements of anal and rectal function. Anorectal manometry, a common procedure, involves inserting a small tube (catheter) or balloon device into the anal canal or rectum. Manometry measures, among other things, pressure levels in the anal canal, rectal sensation, and anal and rectal reflexes. Tests are also available for assessing nerve damage. An anal ultrasound probe can supply accurate images of the anal sphincter and reveal whether injury has occurred. **Magnetic resonance imaging**, which requires the insertion of a coil into the anal canal, is useful at times.

#### Treatment

Fecal incontinence arising from an underlying condition such as diabetic neuropathy can sometimes be helped by treating the underlying condition. When that does not work, or no underlying condition can be discovered, one approach is to have the patient use a suppository or enema to stimulate defecation at the same time every day or every other day. The goal is to restore regular bowel habits and keep the bowels free of stools. Medications such as loperamide (Imodium) and codeine phosphate are often effective in halting incontinence, but only in less severe cases involving liquid stools or urgency. Dietary changes and exercises done at home to strengthen the anal muscles may also help.

Good results have been reported for **biofeedback** training, although the subject has not been properly researched. In successful cases, patients regain complete control over defecation, or at least improve their control, by learning to contract the external part of the anal sphincter whenever stools enter the rectum. All healthy

#### KEY TERMS

**Anus**—The opening at the lower end of the rectum.

**Colostomy**—A surgical procedure in which an opening is made in the wall of the abdomen to allow a part of the large intestine (the colon) to empty outside the body.

**Crohn's disease**—A disease marked by inflammation of the intestines.

**Defecation**—Passage of stools through the anus.

**Hemorrhoids**—Enlarged veins in the anus or rectum. They are sometimes associated with fecal incontinence.

**Rectum**—The lower section of the large intestine that holds stools before defecation.

**Sphincter**—A circular band of muscle that surrounds and encloses an opening to the body or to one of its hollow organs. Damage to the sphincter surrounding the anus can cause fecal incontinence.

**Stools**—Undigested food and other waste that is eliminated through the anus.

**Suppository**—A solid medication that slowly dissolves after being inserted into the rectum or other body cavity.

people have this ability. Biofeedback training begins with the insertion into the rectum of a balloon manometry device hooked up to a pressure monitor. The presence of stools in the rectum is simulated by inflating the balloon, which causes pressure changes that are recorded on the monitor. The monitor also records sphincter contraction. By watching the monitor and following instructions from the equipment operator, the patient gradually learns to contract the sphincter automatically in response to fullness in the rectum. Sometimes one training session is enough, but often several are needed. Biofeedback is not an appropriate treatment in all cases, however. It is used only with patients who are highly motivated; who are able, to some extent, to sense the presence of stools in the rectum; and who have not lost all ability to contract the external anal sphincter. One specialist suggests that possibly two-thirds of incontinence sufferers are candidates for biofeedback.

Some people may require surgery. Sphincter damage caused by childbirth is often effectively treated with surgery, however, as are certain other kinds of incontinence-related sphincter injuries. Sometimes surgical treat-

ment requires building an artificial sphincter using a thigh muscle (the gracilis muscle). At one time a **colostomy** was necessary for severe cases of incontinence, but is now rarely performed.

### Prognosis

Fecal incontinence is a problem that usually responds well to professional medical treatment, even among elderly and institutionalized patients. If complete bowel control cannot be restored, the impact of incontinence on everyday life can still be lessened considerably in most cases. When incontinence remains a problem despite medical treatment, disposable underwear and other commercial incontinence products are available to make life easier. Doctors and nurses can offer advice on coping with incontinence, and people should never be embarrassed about seeking their assistance. Counseling and information are also available from support groups.

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International Foundation for Functional Gastrointestinal Disorders. PO Box 17864, Milwaukee, WI 53217. (888) 964-2001. <<http://www.iffgd.org>>.

National Association for Continence. PO Box 8310, Spartanburg, SC 29305-8310. (800) 252-3337. <<http://www.nafc.org>>.

National Digestive Diseases Information Clearinghouse. 2 Information Way, Bethesda, MD 20892-3570. (800) 891-5389. <<http://www.niddk.nih.gov/health/digest/nddic.htm>>.

Howard Baker

**Fecal lipids test** see **Stool fat test**

## Fecal occult blood test

### Definition

The fecal occult blood test (FOBT) is performed as part of the routine **physical examination** during the

examination of the rectum. It is used to detect microscopic blood in the stool and is a screening tool for colorectal cancer.

### Purpose

FOBT uses chemical indicators on stool samples to detect the presence of blood not otherwise visible. (The word "occult" in the test's name means that the blood is hidden from view.) Blood originating from or passing through the gastrointestinal tract can signal many conditions requiring further diagnostic procedures and, possibly, medical intervention. These conditions may be benign or malignant and some of them include:

- colorectal and gastric cancers
- ulcers
- hemorrhoids
- polyps
- inflammatory bowel disease
- irritations or lesions of the gastrointestinal tract caused by medications (such as **nonsteroidal anti-inflammatory drugs**, also called NSAIDs)
- irritations or lesions of the gastrointestinal tract caused by stomach acid disorders, such as reflux esophagitis

The FOBT is used routinely (in conjunction with a **rectal examination** performed by a physician) to screen for colorectal cancer, particularly after age 50. The ordering of this test should not be taken as an indication that cancer is suspected. The FOBT must be combined with regular screening endoscopy (such as a **sigmoidoscopy**) to detect cancers at an early stage.

### Precautions

Certain foods and medicines can influence the test results. Some fruits contain chemicals that prevent the guaiac, the chemical in which the test paper is soaked, from reacting with the blood. **Aspirin** and some NSAIDs irritate the stomach, resulting in bleeding and should be avoided prior to the examination, along with red meat and many vegetables and fruits containing vitamin C. All of these factors could result in a false-positive test.

### Description

Feces for the stool samples is obtained either by the physician at the rectal examination or by the patient at home, using a small spatula or a collection device. In most cases, the collection of stool samples can easily be done at home, using a kit supplied by the physician. The standard kit contains a specially prepared card on which

a small sample of stool will be spread, using a stick provided in the kit. The sample is placed in a special envelope and either mailed or brought in for analysis. When the physician applies hydrogen peroxide to the back of the sample, the paper will turn blue if an abnormal amount of blood is present.

### *Types of fecal occult blood tests*

Hemoccult is one type of fecal occult blood test, and it is the most commonly used. The Hemoccult test takes less than five minutes to perform and may be performed in the physician's office or in the laboratory. The Hemoccult blood test can detect bleeding from the colon as low as 0.5 mg per day.

Tests that use anti-hemoglobin antibodies (or immunochemical tests) to detect blood in the stool are also used. Immunochemical tests can detect up to 0.7 mg of hemoglobin in the stool and do not require dietary restrictions. Immunochemical tests

- are not accurate for screening for stomach cancer
- are more sensitive than Hemoccult tests in detecting colorectal cancer
- are more expensive than Hemoccult tests

Hemoquant, another fecal occult blood test, is used to detect as much as 500 mg/g of blood in the stool. Like the Hemoccult, the Hemoquant test is affected by red meat. It is not affected by chemicals in vegetables.

Fecal blood may also be measured by measuring the chromium in the red blood cells in the feces. The stool is collected for three to ten days. The test is used in cases where the exact amount of the blood loss is required and it is the only test that can exclude blood loss from the gastrointestinal tract with accuracy.

### **Preparation**

For 72 hours prior to collecting samples, patients should avoid red meats, NSAIDs (including aspirin), **antacids**, steroids, iron supplements, and vitamin C, including citrus fruits and other foods containing large amounts of vitamin C. Foods like uncooked broccoli, uncooked turnips, cauliflower, uncooked cantaloupe, uncooked radish and horseradish and parsnips should be avoided and not eaten during the 72 hours prior to the examination. Fish, chicken, pork, fruits (other than melons) and many cooked vegetables are permitted in the diet.

### **Results**

Many factors can result in false-positive and false-negative findings.

## **KEY TERMS**

**Occult**—Not visible or easily detected.

### *Positive results*

It is important to note that a true-positive finding only signifies the presence of blood—it is not an indication of cancer. The National Cancer Institute states that, in its experience, less than 10% of all positive results were caused by cancer. The FOBT is positive in 1–5% of the unscreened population and 2–10% of those are found to have cancer. The physician will want to follow up on a positive result with further tests, as indicated by other factors in the patient's history or condition.

### *Negative results*

Alternatively, a negative result (meaning no blood was detected) does not guarantee the absence of **colon cancer**, which may bleed only occasionally or not at all. (Only 50% of colon cancers are FOBT-positive.)

### *Conclusions*

Screening using the FOBT has been demonstrated to reduce colorectal cancer. However, because only half of colorectal cancers are FOBT-positive, FOBT must be combined with regular screening endoscopy to increase the detection of pre-malignant colorectal polyps and cancers. Since, through FOBT, cancer may be detected early, the benefits of possible early detection must be considered along with the likelihood of complications and costs for additional studies.

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National Cancer Institute (NCI). (800) 422-6237. <<http://rex.nci.nih.gov>>.

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## Feldenkrais method

### Definition

The Feldenkrais method is an educational system that allows the body to move and function more efficiently and comfortably. Its goal is to re-educate the nervous system and improve motor ability. The system can accomplish much more, relieving pressure on joints and weak points, and allowing the body to heal repetitive strain injuries. Continued use of the method can relieve **pain** and lead to higher standards of achievement in sports, the martial arts, dancing and other physical disciplines.

Pupils are taught to become aware of their movements and to become aware of how they use their bodies, thus discovering possible areas of **stress** and strain. The goal of Feldenkrais is to take the individual from merely functioning, to functioning well, free of pain and restriction of movement. Feldenkrais himself stated that his goal was, “To make the impossible possible, the possible easy, and the easy, elegant.”

### Purpose

This method of re-educating the nervous system can be beneficial to a wide range of people, including athletes, children, the elderly, martial artists, those who are handicapped, people with special needs, and those suffering from degenerative diseases. It has also proved popular with artists, particularly musicians, a number of whom have used Feldenkrais to improve their performance.

The Feldenkrais Guild of North America (FGNA) states that over half of the those who turn to Feldenkrais practitioners are seeking relief from pain. Many people who have pain from an injury compensate by changing

their movements to limit pain. Often these changed movements remain after the pain from the original injury is gone, and new pain may occur. Feldenkrais helps students become aware of the changed movements and allows them to learn new movements that relieve their pain. Apart from the obvious physical benefits of more efficient movement and freedom from pain and restriction, Feldenkrais practitioners assert that there are other positive benefits for overall physical and mental health. Feldenkrais can result in increased awareness, flexibility, and coordination, and better relaxation. Feldenkrais practitioners have also noted other benefits in their students, including improvements in awareness, flexibility, coordination, breathing, digestion, sleep, mood, mental alertness, energy, and range of motion, as well as reduced stress and **hypertension**, and fewer headaches and backaches.

Musicians and athletes can improve their performance in many ways when they learn to use their bodies more efficiently. Feldenkrais can also help injured athletes regain lost potential and free them from pain and restriction of movement.

There are numerous accounts of the remarkable results obtained when Feldenkrais is taught to handicapped children so that they can learn to function despite their limitations. Handicapped people can learn to make full use of whatever potential they have, and to have more confidence in their abilities. Practitioners who specialize in teaching Feldenkrais to those who have handicaps have in many cases allowed the patient to discover ways of performing tasks which were previously thought to be impossible for them.

The elderly, whose movements are often restricted by pain and stiffness, can learn to overcome these obstacles with Feldenkrais instruction. In some instances even severe cases of arthritis have been conquered. Theoretically, Feldenkrais can make possible renewed levels of energy and freedom from restriction.

### Description

#### Origins

Moshe Feldenkrais (1904–1984) was a Russian-born Israeli physicist and engineer who was also an active soccer player and judo master. He devised his system in response to his own recurring knee injury, which had restricted his movement and caused him great pain over a long period of time. Feldenkrais believed that repeated muscle patterns cause the parts of the brain controlling those muscles to stay in a fixed pattern as well. He thought that the more the muscles are used, the more parts of the brain can be activated.

He devised a method of re-educating the neuromuscular system and re-evaluating movement to increase efficiency and reduce stress, using his knowledge of mechanics and engineering, and applying some of his martial arts training.

Feldenkrais is described as being a dual system, with two components: "Awareness Through Movement" and "Functional Integration." The system aims to re-educate the body so that habitual movements that cause strain or pain can be relearned to improve efficiency and eliminate dangerous or painful action.

Feldenkrais helps to translate intention into action. In practice, an individual can learn to achieve his or her highest potential, while at the same time learning to avoid and eliminate stresses, strains, and the possibility of injury.

### ***Functional integration***

During this session, the patient wears comfortable clothing, and may sit, stand, walk, or lie on a low padded table. The practitioner helps the pupil by guiding him or her through a number of movements. The practitioner may use touch to communicate with the student, but touch is not used to correct any movements. The purpose of this session is to increase a student's awareness of his or her own movement and become open to different possibilities for movement. The instruction can be focused on a particular activity that the student does every day, or that causes him or her pain. The student can learn to alter habitual movements and re-educate the neuromuscular system. This type of session is particularly useful for those who suffer from limitations originating from misuse, stress, illness, or accident. It can also help athletes and musicians perform to the best of their ability by increasing their possibilities for movement. It offers students the potential for improving their physical and mental performance in addition to heightening the sense of well-being.

### ***Awareness through movement***

Feldenkrais's martial arts background can be clearly identified in many of the aspects of Awareness Through Movement (ATM). During group sessions, pupils are taught to become acutely aware of all their movements and to imagine them, so that they can improve the efficiency of their actions in their minds, and put them into practice. Pupils are encouraged to be disciplined about practicing their exercises, to achieve maximum benefit.

Awareness through movement is described as an exploratory, nonjudgmental process through which pupils

## **MOSHE FELDENKRAIS (1904–1984)**

Moshe Feldenkrais was born on the border between Russia and Poland. When he was only a boy of 13, he traveled to Palestine on foot. The journey took a year, and once there, young Feldenkrais worked as a laborer and cartographer, also tutoring others in mathematics. Moving to France in 1933, he graduated in mechanical and electrical engineering from the Ecole des Travaux Publics de Paris.

Feldendrais became the first person to open a Judo center in Paris after meeting with Jigaro Kano. He was also one of the first Europeans to become a black belt in Judo, in 1936.

Obtaining his Ph.D. at the Sorbonne, he went on to assist Nobel Prize laureate, Frédéric Joliot-Curie at the Curie Institute. During World War II in England, he worked on the new sonar anti-submarine research.

Prompted by a recurring leg injury, he applied his knowledge of the martial arts and his training as an engineer to devise a method of re-integrating the body. The concept was that more efficient movement would allow for the treatment of pain or disability, and the better-functioning of the body as a whole. Later on, he would begin to teach what he had learned to others in Tel Aviv.

In addition to many books about judo, including *Higher Judo*, he wrote six books on his method.

**Patricia Skinner**

are encouraged to observe and learn about themselves and their movements. The range of this therapy is wide, and there are thousands of different lessons designed to help specific areas.

### **Preparations**

No preparation is necessary for the practice of Feldenkrais, and all are encouraged to seek help from this system. No condition is considered a preclusion to the benefits of Feldenkrais.

### **Precautions**

As with any therapy or treatment, care should be taken to choose a qualified practitioner. Feldenkrais practitioners stress that the body must not be forced to do anything, and if any movement is painful, or even uncomfortable, it should be discontinued immediately and the patient should seek professional help.

## KEY TERMS

**Neuromuscular**—The body system of nerves and muscles as they function together.

**Repetitive strain injury**—Injury resulting from a repeated movement such as typing or throwing a ball.

### Side effects

No known side effects are associated with the practice of Feldenkrais.

### Research and general acceptance

Since Moshe Feldenkrais began to teach his method, it has gradually gained acceptance as an education system. Published research using the method can be found in United States and foreign publications.

### Resources

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#### ORGANIZATIONS

Feldenkrais Guild of North America. 3611 SW Hood Ave., Suite 100, Portland, OR 97201. (800) 775-2118. (503) 221-6612. Fax: (503) 221-6616. <<http://www.feldenkrais.com/>>.

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Female circumcision see **Female genital mutilation**

Female condom see **Condom**

## Female genital mutilation

### Definition

Female genital mutilation (FGM) is the cutting, or partial or total removal, of the external female genitalia for cultural, religious, or other non-medical reasons. It is usually performed on girls between the ages of four and 10. It is also called female **circumcision**.

### Purpose

FGM results in the cutting or removal of the tissues around the vagina that give women pleasurable sexual feelings. This procedure is used for social and cultural control of women's sexuality. In its most extreme form, infibulation, where the girl's vagina is sewn shut, the procedure ensures virginity. In some cultures where female circumcision has been a tradition for hundreds of years, this procedure is considered a rite of passage for young girls. Families fear that if their daughters are left uncircumcised, they may not be marriageable. As in most cultures, there is also the fear that the girl might bring shame to the family by being sexually active and becoming pregnant before marriage.

### Precautions

It is illegal to perform FGM in many countries, including the United States, Canada, France, Great Britain, Sweden, Switzerland, Egypt, Kenya, and Senegal. This procedure is usually done in the home or somewhere other than a medical setting. Often, it is performed by a family member or by a local "circumciser," using knives, razor blades, or other tools that may not be sterilized before use.

### Description

Female circumcision includes a wide range of procedures. The simplest form involves a small cut to the clitoris or labial tissue. A Sunna circumcision removes the prepuce (a fold of skin that covers the clitoris) and/or the tip of the clitoris. A clitoridectomy removes the entire clitoris and some or all of the surrounding tissue; this procedure occurs in approximately 80% of cases. The most extreme form of genital mutilation is excision and infibulation, in which the clitoris and all of the surrounding tissue are cut away and the remaining skin is sewn together. Only a small opening is left for the passage of

urine and menstrual blood. Infibulation accounts for approximately 15% of FGM procedures.

The World Health Organization (WHO) estimates that between 100 million and 140 million girls and women have undergone some form of FGM. As a very deeply rooted cultural and religious tradition still practiced in over 28 African and Asian countries, up to two million girls per year are at risk. The following countries have the highest number of occurrences of FGM: Djibouti (98%), Egypt (97%), Eritrea (95%), Guinea (99%), Mali (94%), Sierra Leone (90%), and Somalia (98–100%). As more people move to Western countries from countries where female circumcision is performed, the practice has come to the attention of health professionals in the United States, Canada, Europe, and Australia.

In an effort to integrate old customs with modern medical care, some immigrant families have requested that physicians perform the procedure. While trying to be sensitive to cultural traditions, health care providers are sometimes put in the difficult position of choosing to perform this procedure in a medical facility under sanitary conditions, or refusing the request, knowing that it may be done anyway with no medical supervision. Some families who are intent on having this procedure done will take their daughters back to the country they immigrated from in order to have the girls circumcised.

Many national and international medical organizations including the American Medical Association (AMA), Canadian medical organizations, and WHO oppose the practice of female genital mutilation. The United Nations (UN) considers female genital mutilation a violation of human rights. WHO has undertaken a number of projects aimed at decreasing the incidence of FGM. These include the following activities:

- publishing a statement addressing the regional status of FGM and encouraging the development of national policy against its practice,
- organizing training for regional community workers,
- developing educational materials for local health care workers,
- providing alternative occupations for individuals who perform FGM procedures.

### Aftercare

A girl or young woman who has recently had the procedure performed may require supportive care to control bleeding and **antibiotics** to prevent infection. Women who were circumcised as children may require medical care to treat complications. Pregnant women

## KEY TERMS

**Circumcision**—A procedure, usually with religious or cultural significance, where the prepuce or skin covering the tip of the penis on a boy, or the clitoris on a girl, is cut away.

**Clitoridectomy**—A procedure where the clitoris and possibly some of the surrounding labial tissue at the opening of the vagina is cut away.

**Infibulation**—A procedure where the tissue around the vagina is sewn shut, leaving only a small opening for the passage of urine and menstrual blood.

who have been infibulated may have to have the labial tissue cut open to allow the baby to be delivered. After-care should be provided with a supportive and nonjudgmental approach towards the girls and women who have undergone this procedure.

### Risks

The immediate risks after the procedure are hemorrhage (excessive bleeding), severe **pain**, and infection (including abscesses, **tetanus**, and **gangrene**). The most severe consequence is **death** due to excessive blood loss. Long term complications include scarring, interference with the drainage of urine and menstrual blood, chronic urinary tract infections, pelvic and back pain, and **infertility**. Sexual intercourse can be painful. Complications of **childbirth** are also a risk. It is unclear whether it is related to the procedure itself, or related to the general condition of medical practice, but infant and maternal death rates are generally higher in those communities where female circumcision is practiced.

### Resources

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"Female Genital Mutilation." *The World Health Organization.* <<http://www.who.int/frh-whd/FGM/index.htm>>.

Altha Roberts Edgren

## Female sexual arousal disorder

### Definition

Female sexual arousal disorder (FSAD) occurs when a woman is continually unable to attain or maintain arousal and lubrication during intercourse, is unable to reach orgasm, or has no desire for sexual intercourse.

### Description

The disorder typically affects up to 25 percent of all American women, or an estimated 47 million women. Three-fourths of women with FSAD are postmenopausal. Women describe it as being "unable to get turned on," or being continually disinterested in sex. It is also called "frigidity." Other terms for the disorder include dyspareunia and vaginismus, both of which involve **pain** during intercourse.

### Causes and symptoms

There are numerous causes of this disorder. They include:

- physical problems, such as **endometriosis**, **cystitis**, or **vaginitis**
- systemic problems, such as diabetes, high blood pressure, or **hypothyroidism**. Even **pregnancy** or the postpartum period (time after delivery of a child) may affect desire. **Menopause** is also known to reduce sexual desire.
- medications, including **oral contraceptives**, anti-depressants, antihypertensives, and tranquilizers
- surgery, such as **mastectomy** or **hysterectomy** which may affect how a woman feels about her sexual self.
- stress
- depression
- use of alcohol, drugs, or cigarette **smoking**

Symptoms vary. A woman may have no desire for sex, or may not be able to maintain arousal, or may be

unable to reach orgasm. She may also have pain during sex or orgasm, which interferes with her desire for intercourse.

### Diagnosis

To make a diagnosis, a woman's physician - either family doctor, gynecologist, or even urologist — takes a complete medical history to determine when the problem started, how it presents, how severe it is, and what the patient thinks may be causing it. The doctor will also conduct a complete **physical examination**, looking for any abnormalities in the genital region

### Treatment

The physician should start by providing education about the disorder and recommending various non-medical treatment strategies. These include:

- use of erotic materials, such as vibrators, books, magazines and videos
- sensual massage, avoiding the genitals
- position changes to reduce pain
- use of lubricants to moisten the vagina and genital area
- kegel exercises to strengthen the vagina and clitoris
- therapy to overcome any relationship or sexual **abuse** issues

Medical treatments include:

- estrogen replacement therapy, which may help with vaginal dryness, pain and arousal
- testosterone therapy in women who have low levels of this male hormone (side effects, however, may include deepening voice, hair growth, and acne)
- the EROS clitoral therapy device (EROS-CTD), recently approved by the Food and Drug Administration; a small vacuum pump, placed over the clitoris and gently activated to provide a gentle suction designed to increase blood flow to the region, which, in turn, helps with arousal
- using the herb yohimbine combined with nitric oxide has been found to increase vaginal blood flow in post-menopausal women and thus help with some forms of FSAD

### Alternative treatment

Natural estrogens, such as those found in soy products and flax, may be effective. Herbal remedies include belladonna, gingko, and motherwort. However, there is no scientific evidence to prove these herbs actually help.

## KEY TERMS

**Dyspareunia**— pain in the pelvic area during or after sexual intercourse.

**Vaginismus** —An involuntary spasm of the muscles surrounding the vagina, making penetration painful or impossible.

Some women squirt vitamin E in their vagina to increase lubrication.

Women may also want to see a sexual therapist for additional help.

### Prognosis

Generally, once women seek the appropriate help they are quite likely to find a way to resolve their problems. Often, a holistic approach, using physical as well as emotional therapies, is required for success.

### Prevention

Maintaining a close and open relationship with a partner is one way to avoid the emotional pain and isolation that can lead to **sexual dysfunction**. Additionally, women should learn if any medications they take affect sexual function, and should refrain from alcohol and drugs and quit smoking. Women who have anxieties and fears about sexual intercourse, whether because of earlier abuse, rape, or a prudish upbringing, should deal with those issues through therapy.

### Resources

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#### PERIODICALS

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- "Consumer Update: Female Sexual Problems." *American Association for Marriage and Family Therapy* (14 June 1999).

## ORGANIZATIONS

Female Sexual Medicine Center UCLA Medical Center 924 Westwood Blvd., Suite 520 Los Angeles, CA 90024.

(310) 825-0025 <[www.newshe.com](http://www.newshe.com)>.

National Women's Health Resource Center, 120 Albany Street Suite 820 New Brunswick, NJ 08901. (877) 986-9472. <[www.healthywomen.org](http://www.healthywomen.org)>.

Debra Gordon

Female infertility see **Infertility**

Femoral hernia see **Hernia**

## Fetal alcohol syndrome

### Definition

Fetal alcohol syndrome (FAS) is a pattern of **birth defects**, learning, and behavioral problems affecting individuals whose mothers consumed alcohol during pregnancy.

### Description

FAS is the most common preventable cause of **mental retardation**. This condition was first recognized and reported in the medical literature in 1968 in France and in 1973 in the United States. Alcohol is a teratogen, the term used for any drug, chemical, maternal disease or other environmental exposure that can cause birth defects or functional impairment in a developing fetus. Some features may be present at birth including low birth weight, **prematurity**, and microcephaly. Characteristic facial features may be present at birth, or may become more obvious over time. Signs of brain damage include delays in development, behavioral abnormalities, and mental retardation, but affected individuals exhibit a wide range of abilities and disabilities. It has only been since 1991 that the long-term outcome of FAS has been known. Learning, behavioral, and emotional problems are common in adolescents and adults with FAS. Fetal Alcohol Effect (FAE), a term no longer favored, is sometimes used to describe individuals with some, but not all, of the features of FAS. In 1996, the Institute of Medicine suggested a five-level system to describe the birth defects, learning and behavioral difficulties in offspring of women who drank alcohol during pregnancy. This system contains criteria including confirmation of maternal alcohol exposure, characteristic facial features, growth problems, learning and behavioral problems, and birth defects known to be associated with prenatal alcohol exposure.

The incidence of FAS varies among different populations studied, and ranges from approximately one in 200 to one in 2000 at birth. However, a recent study reported in 1997, utilizing the Institute of Medicine criteria, estimated the prevalence in Seattle, Washington from 1975–1981 at nearly one in 100 live births. Avoiding alcohol during pregnancy, including the earliest weeks of the pregnancy can prevent FAS. There is no amount of alcohol use during pregnancy that has been proven to be completely safe.

There is no racial or ethnic predilection for FAS. Individuals from different genetic backgrounds exposed to similar amounts of alcohol during pregnancy may exhibit different signs or symptoms of FAS. Several studies have estimated that between 25–45% of chronic alcoholic women will give birth to a child with FAS if they continue to drink during pregnancy. The risk of FAS appears to increase as a chronic alcoholic woman progresses in her childbearing years and continues to drink. That is, a child with FAS will often be one of the last born to a chronic alcoholic woman, although older siblings may exhibit milder features of FAS. Binge drinking, defined as sporadic use of five or more standard alcoholic drinks per occasion, and “moderate” daily drinking (two to four 12 oz bottles of beer, eight to 16 ounces of wine, two to four ounces of liquor) can also result in offspring with features of FAS.

### Causes and symptoms

FAS is not a genetic or inherited disorder. It is a pattern of birth defects, learning, and behavioral problems that are the result of maternal alcohol use during the pregnancy. The alcohol freely crosses the placenta and causes damage to the developing embryo or fetus. Alcohol use by the father cannot cause FAS. If a woman who has FAS drinks alcohol during pregnancy, then she may also have a child with FAS. Not all individuals from alcohol exposed pregnancies have obvious signs or symptoms of FAS; individuals of different genetic backgrounds may be more or less susceptible to the damage that alcohol can cause. The dose of alcohol, the time during pregnancy that alcohol is used, and the pattern of alcohol use all contribute to the different signs and symptoms that are found.

Classic features of FAS include short stature, low birthweight and poor weight gain, microcephaly, and a characteristic pattern of facial features. These facial features in infants and children may include small eye openings (measured from inner corner to outer corner), epicanthal folds (folds of tissue at the inner corner of the eye), small or short nose, low or flat nasal bridge, smooth or poorly developed philtrum (the area of the upper lip above

the colored part of the lip and below the nose), thin upper lip, and small chin. Some of these features are nonspecific, meaning they can occur in other conditions, or be appropriate for age, racial, or family background. Other major and minor birth defects that have been reported include cleft palate, congenital heart defects, **strabismus**, **hearing loss**, defects of the spine and joints, alteration of the hand creases, small fingernails, and toenails. Since FAS was first described in infants and children, the diagnosis is sometimes more difficult to recognize in older adolescents and adults. Short stature and microcephaly remain common features, but weight may normalize, and the individual may actually become overweight for his/her height. The chin and nose grow proportionately more than the middle part of the face and dental crowding may become a problem. The small eye openings and the appearance of the upper lip and philtrum may continue to be characteristic. Pubertal changes typically occur at the normal time.

Newborns with FAS may have difficulties with feeding due to a poor suck, have irregular sleep-wake cycles, decreased or increased muscle tone, seizures or **tremors**. Delays in achieving developmental milestones such as rolling over, crawling, walking and talking may become apparent in infancy. Behavior and learning difficulties typical in the preschool or early school years include poor attention span, hyperactivity, poor motor skills, and slow language development. Attention deficit-hyperactivity disorder is a common associated diagnosis. Learning disabilities or mental retardation may be diagnosed during this time. Arithmetic is often the most difficult subject for a child with FAS. During middle school and high school years the behavioral difficulties and learning difficulties can be significant. Memory problems, poor judgment, difficulties with daily living skills, difficulties with abstract reasoning skills, and poor social skills are often apparent by this time. It is important to note that animal and human studies have shown that neurologic and behavioral abnormalities can be present without characteristic facial features. These individuals may not be identified as having FAS, but may fulfill criteria for alcohol-related diagnoses, as set forth by the Institute of Medicine.

In 1991, Streissguth and others reported some of the first long-term follow-up studies of adolescents and adults with FAS. In the approximate 60 individuals they studied, the average IQ was 68, with 70 being the lower limit of the normal range. However, the range of IQ was quite large, as low as 20 (severely retarded) to as high as 105 (normal). The average achievement levels for reading, spelling, and arithmetic were fourth grade, third grade and second grade, respectively. The Vineland Adaptive Behavior Scale was used to measure adaptive functioning in these individuals. The composite score for this group showed functioning at the level of a seven-

year-old. Daily living skills were at a level of nine years, and social skills were at the level of a six-year-old.

In 1996, Streissguth and others published further data regarding the disabilities in children, adolescents and adults with FAS. Secondary disabilities, that is, those disabilities not present at birth and that might be preventable with proper diagnosis, treatment, and intervention, were described. These secondary disabilities include: mental health problems; disrupted school experiences; trouble with the law; incarceration for mental health problems, drug abuse, or a crime; inappropriate sexual behavior; alcohol and drug abuse; problems with employment; dependent living; and difficulties parenting their own children. In that study, only seven out of 90 adults were living and working independently and successfully. In addition to the studies by Streissguth, several other authors in different countries have now reported on long term outcome of individuals diagnosed with FAS. In general, the neurologic, behavioral and emotional disorders become the most problematic for the individuals. The physical features change over time, sometimes making the correct diagnosis more difficult in older individuals, without old photographs and other historical data to review. Mental health problems including attention deficit, depression, panic attacks, **psychosis** and suicide threats and attempts, and overall were present in over 90% of the individuals studied by Streissguth. A 1996 study in Germany reported more than 70% of the adolescents they studied had persistent and severe developmental disabilities and many had psychiatric disorders, the most common of which were emotional disorders, repetitive habits, **speech disorders**, and hyperactivity disorders.

## Diagnosis

FAS is a clinical diagnosis, which means that there is no blood, x ray or psychological test that can be performed to confirm the suspected diagnosis. The diagnosis is made based on the history of maternal alcohol use, and detailed **physical examination** for the characteristic major and minor birth defects and characteristic facial features. It is often helpful to examine siblings and parents of an individual suspected of having FAS, either in person or by photographs, to determine whether findings on the examination might be familial, or if other siblings may also be affected. Sometimes, genetic tests are performed to rule out other conditions that may present with developmental delay or birth defects. Individuals with developmental delay, birth defects or other unusual features are often referred to a clinical geneticist, developmental pediatrician, or neurologist for evaluation and diagnosis of FAS. Psychoeducational testing to determine IQ and/or the presence of learning disabilities may also be part of the evaluation process.

## KEY TERMS

**Cleft palate**—A congenital malformation in which there is an abnormal opening in the roof of the mouth that allows the nasal passages and the mouth to be improperly connected.

**Congenital**—Refers to a disorder which is present at birth.

**IQ**—Abbreviation for Intelligence Quotient. Compares an individual's mental age to his/her true or chronological age and multiplies that ratio by 100.

**Microcephaly**—An abnormally small head.

**Miscarriage**—Spontaneous pregnancy loss.

**Placenta**—The organ responsible for oxygen and nutrition exchange between a pregnant mother and her developing baby.

**Strabismus**—An improper muscle balance of the ocular muscles resulting in crossed or divergent eyes.

**Teratogen**—Any drug, chemical, maternal disease, or exposure that can cause physical or functional defects in an exposed embryo or fetus.

## Treatment

There is no treatment for FAS that will reverse or change the physical features or brain damage associated with maternal alcohol use during the pregnancy. Most of the birth defects associated with prenatal alcohol exposure are correctable with surgery. Children should have psychoeducational evaluation to help plan appropriate educational interventions. Common associated diagnoses such as attention deficit-hyperactivity disorder, depression, or **anxiety** should be recognized and treated appropriately. The disabilities that present during childhood persist into adult life. However, some of the secondary disabilities mentioned above may be avoided or lessened by early and correct diagnosis, better understanding of the life-long complications of FAS, and intervention. Streissguth has describe a model in which an individual affected by FAS has one or more advocates to help provide guidance, structure and support as the individual seeks to become independent, successful in school or employment, and develop satisfying social relationships.

## Prognosis

The prognosis for FAS depends on the severity of birth defects and the brain damage present at birth. **Miscarriage, stillbirth or death** in the first few weeks of

life may be outcomes in very severe cases. Major birth defects associated with FAS are usually treatable with surgery. Some of the factors that have been found to reduce the risk of secondary disabilities in FAS individuals include diagnosis before the age of six years, stable and nurturing home environments, never having experienced personal violence, and referral and eligibility for disability services. The long-term data helps in understanding the difficulties that individuals with FAS encounter throughout their lifetime and can help families, caregivers and professionals provide the care, supervision, education and treatment geared toward their special needs.

Prevention of FAS is the key. Prevention efforts must include public education efforts aimed at the entire population, not just women of child bearing age, appropriate treatment for women with high-risk drinking habits, and increased recognition and knowledge about FAS by professionals, parents, and caregivers.

## Resources

### BOOKS

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## ORGANIZATIONS

Arc's Fetal Alcohol Syndrome Resource Guide. The Arc's Publication Desk, 3300 Pleasant Valley Lane, Suite C, Arlington, TX 76015. (888) 368-8009. <<http://www.thearc.org/mis/faslist.html>>.

Fetal Alcohol Syndrome Family Resource Institute. PO Box 2525, Lynnwood, WA 98036. (800) 999-3429. <<http://www.fetalalcoholsyndrome.org>>.

Institute of Medicine. National Academy Press, Washington, DC. <<http://www.come-over.to/FAS/IOMsummary.htm>>.

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. (888) 663-4637. <[resourcecenter@modimes.org](mailto:resourcecenter@modimes.org)>. <<http://www.modimes.org>>.

Nofas. 216 G St. NE, Washington, DC 20002. (202) 785-4585. <<http://www.nofas.org>>.

Laurie Heron Seaver

**Ferritin test** see **Iron tests**

**Fetal death** see **Stillbirth**

## Fetal hemoglobin test

### Definition

Fetal hemoglobin (Hemoglobin F), Alkali-resistant hemoglobin, HBF (or Hb F), is the major hemoglobin component in the bloodstream of the fetus. After birth, it decreases rapidly until only traces are found in normal children and adults.

### Purpose

The determination of fetal hemoglobin is an aid in evaluating low concentrations of hemoglobin in the blood (anemia), as well as the hereditary persistence of fetal hemoglobin, and a group of inherited disorders affecting hemoglobin, among which are the thalassemias and sickle cell anemia.

### Description

At birth, the newborn's blood is comprised of 60%-90% of fetal hemoglobin. The fetal hemoglobin then rapidly decreases to 2% or less after the second to fourth years. By the time of adulthood, only traces (0.5% or less) are found in the bloodstream.

In some diseases associated with abnormal hemoglobin production (see Hemoglobinopathy, below), fetal hemoglobin may persist in larger amounts. When this occurs, the elevation raises the question of possible underlying disease.

For example, HBF can be found in higher levels in hereditary hemolytic **anemias**, in all types of leukemias, in **pregnancy**, diabetes, thyroid disease, and during anti-convulsant drug therapy. It may also reappear in adults when the bone marrow is overactive, as in the disorders of **pernicious anemia**, **multiple myeloma**, and metastatic **cancer** in the marrow. When HBF is increased after age four, it should be investigated for cause.

### **Hemoglobinopathy**

Hemoglobin is the oxygen-carrying pigment found in red blood cells. It is a large molecule made in the bone marrow from two components, heme and globin.

Defects in hemoglobin production may be either genetic or acquired. The genetic defects are further subdivided into errors of heme production (porphyria), and those of globin production (known collectively as the **hemoglobinopathies**).

There are two categories of hemoglobinopathy. In the first category, abnormal globin chains give rise to abnormal hemoglobin molecules. In the second category, normal hemoglobin chains are produced but in abnormal amounts. An example of the first category is the disorder of sickle cell anemia, the inherited condition characterized by curved (sickle-shaped) red blood cells and chronic **hemolytic anemia**. Disorders in the second category are called the thalassemias, which are further divided into types according to which amino acid chain is affected (alpha or beta), and whether there is one defective gene (**thalassemia minor**) or two defective genes (**thalassemia major**).

### **Preparation**

This test requires a blood sample. The patient is not required to be in a **fasting** state (nothing to eat or drink for a period of hours before the test).

### **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**

Reference values vary from laboratory to laboratory but are generally found within the following ranges:

- six months to adult: up to 2% of the total hemoglobin
- newborn to six months: up to 75% of the total hemoglobin

## **KEY TERMS**

**Anemia**—A disorder characterized by a reduced blood level of hemoglobin, the oxygen-carrying pigment of blood.

**Hemolytic anemia**—A form of anemia caused by premature destruction of red cells in the blood stream (a process called hemolysis). Hemolytic anemias are classified according to whether the cause of the problem is inside the red blood cell (in which case it is usually an inherited condition), or outside the cell (usually acquired later in life).

### **Abnormal results**

Greater than 2% of total hemoglobin is abnormal.

## **Resources**

### **BOOKS**

- Cahill, Mathew. *Handbook of Diagnostic Tests*. Springhouse, PA: Springhouse Corporation, 1995.  
 Jacobs, David S., et al. *Laboratory Test Handbook*. 4th ed. New York: Lexi-Comp Inc., 1996.  
 Pagana, Kathleen Deska. *Mosby's Manual of Diagnostic and Laboratory Tests*. St. Louis: Mosby, Inc., 1998.

Janis O. Flores

Fetishes see **Sexual perversions**

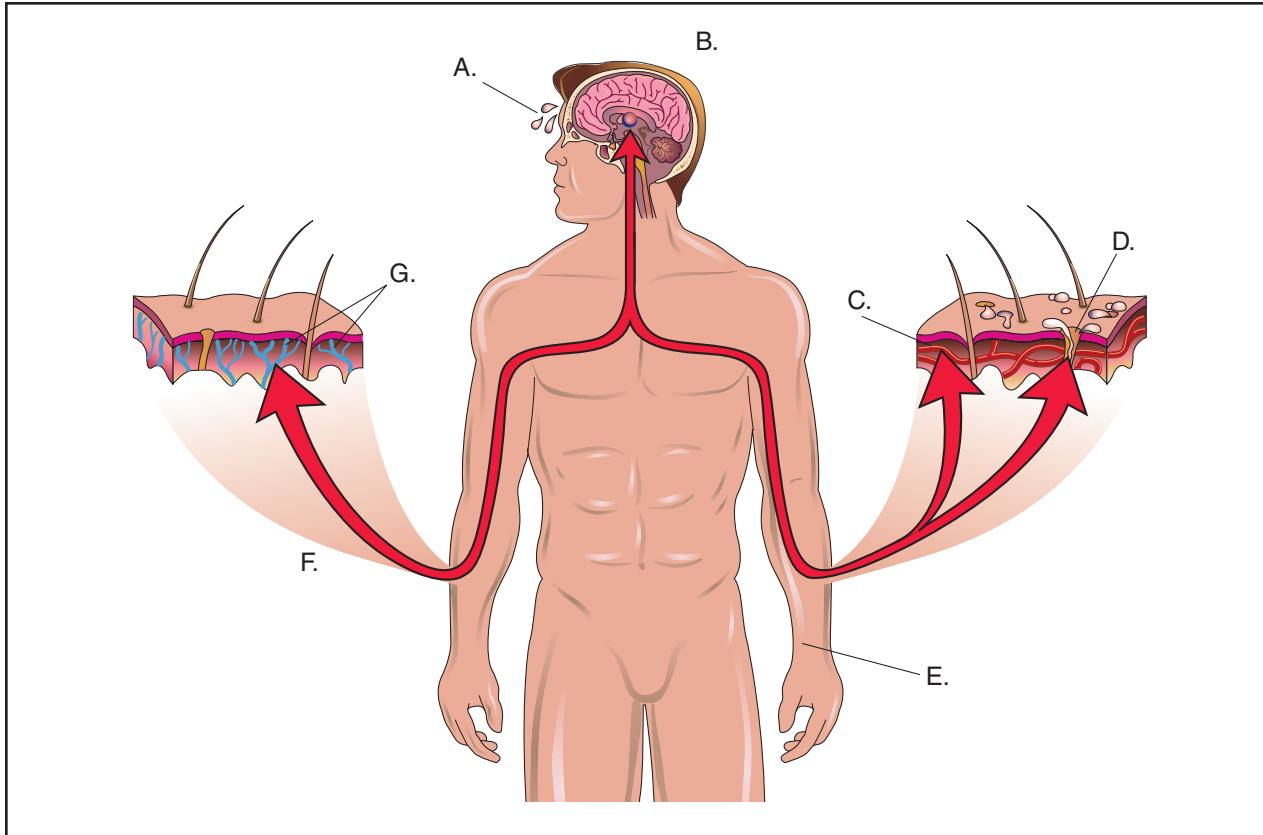
## **Fever**

### **Definition**

A fever is any body temperature elevation over 100°F (37.8°C).

### **Description**

A healthy person's body temperature fluctuates between 97°F (36.1°C) and 100°F (37.8°C), with the average being 98.6°F (37°C). The body maintains stability within this range by balancing the heat produced by the metabolism with the heat lost to the environment. The "thermostat" that controls this process is located in the hypothalamus, a small structure located deep within the brain. The nervous system constantly relays information about the body's temperature to the thermostat, which in turn activates different physical responses designed to



A dramatic rise in body temperature often includes the following symptoms: A. Loss of fluid results in dehydration. B. The hypothalamic set-point is increased, raising metabolism. C. Blood vessels in skin dilate. D. Sweat glands produce excess perspiration. E. Increased pulse rate. F. Increased hypothalamic set-point may introduce chills and shivering to promote heat production from muscles. G. Skin becomes more heat-sensitive. (Illustration by Electronic Illustrators Group.)

cool or warm the body, depending on the circumstances. These responses include: decreasing or increasing the flow of blood from the body's core, where it is warmed, to the surface, where it is cooled; slowing down or speeding up the rate at which the body turns food into energy (metabolic rate); inducing shivering, which generates heat through muscle contraction; and inducing sweating, which cools the body through evaporation.

A fever occurs when the thermostat resets at a higher temperature, primarily in response to an infection. To reach the higher temperature, the body moves blood to the warmer interior, increases the metabolic rate, and induces shivering. The "chills" that often accompany a fever are caused by the movement of blood to the body's core, leaving the surface and extremities cold. Once the higher temperature is achieved, the shivering and chills stop. When the infection has been overcome or drugs such as **aspirin** or **acetaminophen** (Tylenol) have been taken, the thermostat resets to normal and the body's cooling mechanisms switch on: the blood moves to the surface and sweating occurs.

Fever is an important component of the immune response, though its role is not completely understood. Physicians believe that an elevated body temperature has several effects. The immune system chemicals that react with the fever-inducing agent and trigger the resetting of the thermostat also increase the production of cells that fight off the invading bacteria or viruses. Higher temperatures also inhibit the growth of some bacteria, while at the same time speeding up the chemical reactions that help the body's cells repair themselves. In addition, the increased heart rate that may accompany the changes in blood circulation also speeds the arrival of white blood cells to the sites of infection.

### Causes and symptoms

Fevers are primarily caused by viral or bacterial infections, such as **pneumonia** or **influenza**. However, other conditions can induce a fever, including allergic reactions; autoimmune diseases; trauma, such as breaking a bone; **cancer**; excessive exposure to the sun; intense

**exercise**; hormonal imbalances; certain drugs; and damage to the hypothalamus. When an infection occurs, fever-inducing agents called pyrogens are released, either by the body's immune system or by the invading cells themselves, that trigger the resetting of the thermostat. In other circumstances, the immune system may overreact (allergic reactions) or become damaged (autoimmune diseases), causing the uncontrolled release of pyrogens. A **stroke** or tumor can damage the hypothalamus, causing the body's thermostat to malfunction. Excessive exposure to the sun or intensely exercising in hot weather can result in heat stroke, a condition in which the body's cooling mechanisms fail. Malignant hyperthermia is a rare, inherited condition in which a person develops a very high fever when given certain anesthetics or **muscle relaxants** in preparation for surgery.

How long a fever lasts and how high it may go depends on several factors, including its cause, the age of the patient, and his or her overall health. Most fevers caused by infections are acute, appearing suddenly and then dissipating as the immune system defeats the infectious agent. An infectious fever may also rise and fall throughout the day, reaching its peak in the late afternoon or early evening. A low-grade fever that lasts for several weeks is associated with autoimmune diseases such as lupus or with some cancers, particularly leukemia and lymphoma.

## Diagnosis

A fever is usually diagnosed using a thermometer. A variety of different thermometers are available, including traditional glass and mercury ones used for oral or rectal temperature readings and more sophisticated electronic ones that can be inserted in the ear to quickly register the body's temperature. For adults and older children, temperature readings are usually taken orally. Younger children who cannot or will not hold a thermometer in their mouths can have their temperature taken by placing an oral thermometer under their armpit. Infants generally have their temperature taken rectally using a rectal thermometer.

As important as registering a patient's temperature is determining the underlying cause of the fever. The presence or absence of accompanying symptoms, a patient's medical history, and information about what he or she may have ingested, any recent trips taken, or possible exposures to illness help the physician make a diagnosis. Blood tests can aid in identifying an infectious agent by detecting the presence of antibodies against it or providing samples for growth of the organism in a culture. Blood tests can also provide the doctor with white blood cell counts. Ultrasound tests, **magnetic resonance imaging** (MRI) tests, or computed tomography (CT) scans

## KEY TERMS

**Antipyretic**—A drug that lowers fever, like aspirin or acetaminophen.

**Autoimmune disease**—Condition in which a person's immune system attacks the body's own cells, causing tissue destruction.

**Febrile seizure**—Convulsions brought on by fever.

**Malignant hyperthermia**—A rare, inherited condition in which a person develops a very high fever when given certain anesthetics or muscle relaxants in preparation for surgery.

**Meningitis**—A potentially fatal inflammation of the thin membrane covering the brain and spinal cord.

**Metabolism**—The chemical process by which the body turns food into energy, which can be given off as heat.

**Pyrogen**—A chemical circulating in the blood that causes a rise in body temperature.

**Reye's syndrome**—A disorder principally affecting the liver and brain, marked by the rapid development of life-threatening neurological symptoms.

may be ordered if the doctor cannot readily determine the cause of a fever.

## Treatment

Physicians agree that the most effective treatment for a fever is to address its underlying cause, such as through the administration of **antibiotics**. Also, because a fever helps the immune system fight infection, it usually should be allowed to run its course. Drugs to lower fever (antipyretics) can be given if a patient (particularly a child) is uncomfortable. These include aspirin, acetaminophen (Tylenol), and ibuprofen (Advil). Aspirin, however, should not be given to a child or adolescent with a fever since this drug has been linked to an increased risk of **Reye's syndrome**. Bathing a patient in cool water can also help alleviate a high fever.

A fever requires emergency treatment under the following circumstances:

- newborn (three months or younger) with a fever over 100.5°F (38°C)
- infant or child with a fever over 103°F (39.4°C)
- fever accompanied by severe **headache**, neck stiffness, mental confusion, or severe swelling of the throat

A very high fever in a small child can trigger seizures (febrile seizures) and therefore should be treated immediately. A fever accompanied by the above symptoms can indicate the presence of a serious infection, such as **meningitis**, and should be brought to the immediate attention of a physician.

## Prognosis

Most fevers caused by infection end as soon as the immune system rids the body of the pathogen and do not produce any lasting effects. The prognosis for fevers associated with more chronic conditions, such as autoimmune disease, depends upon the overall outcome of the disorder.

## Resources

### BOOKS

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Gelfand, Jeffrey. "Fever, Including Fever of Unknown Origin." In *Harrison's Principles of Internal Medicine*, ed. Anthony S. Fauci, et al. New York: McGraw-Hill, 1997.

Bridget Travers

Fever blister see **Cold sore**

## I Fever evaluation tests

### Definition

**Fever** evaluation tests, better known as febrile agglutinins tests, are performed to detect the presence of antibodies in the blood that are sensitive to temperature changes. Antibodies are proteins produced by the immune system in response to specific infectious agents, such as viruses or bacteria. Febrile agglutinins are antibodies that cause red blood cells to clump, but only when the blood is warmed to temperatures higher than the average body temperature of 98.6°F (37°C).

### Purpose

The febrile agglutinins test is used to confirm the diagnosis of certain infectious diseases that stimulate the body to produce febrile agglutinins. The disease most commonly diagnosed by this test is **brucellosis**, an infection caused by bacteria belonging to the genus *Brucella*.

and characterized by intermittent fever, sweating, chills, aches, and mental depression. The test is also used to diagnose certain other infectious diseases: salmonellosis, caused by *Salmonella* bacteria and marked by nausea and severe **diarrhea**; rickettsial infections, a group of diseases caused by the bacteria *Rickettsia*; and **tularemia**, also called rabbit fever, a bacterial infection characterized by a high fever and swollen lymph nodes. The febrile agglutinins test can also be used to confirm the presence of two types of **cancer**, leukemia and lymphoma; however, doctors rarely use the test for this purpose, since other diagnostic tests are more reliable.

### Description

A febrile agglutinins test can be performed at a doctor's office or a hospital. A nurse or technician will collect a few drops of blood (about 7ml) in a small tube that has been cooled slightly. The specimen is then taken to a laboratory where it is heated and examined for clumping. If the cells clump after warming and unclump as they cool, a febrile agglutinin titer (concentration) of greater than 1:80 is present.

### Normal results

The results of febrile agglutinins tests require a doctor's interpretation. In general, however, a normal value is lower than 1:32.

### Abnormal results

An value higher than 1:80 suggests a diagnosis for brucellosis or one of the other conditions indicated by this test.

Jill S. Lasker

## I Fever of unknown origin

### Definition

**Fever** of unknown origin (FUO) refers to the presence of a documented fever for a specified time, for which a cause has not been found after a basic medical evaluation. The classic criteria developed in 1961 included: temperature greater than 101°F (38.3°C), for at least three weeks, and inability to find a cause after one week of study. Within the past decade, a revision has been proposed that categorizes FUO into classic, hospital acquired FUO, FUO associated with low white blood counts, and HIV associated FUO (AIDS related).

## Description

Fever is a natural response of the body that helps in fighting off foreign substances, such as microorganisms, toxins, etc. Body temperature is set by the thermoregulatory center, located in an area in the brain called hypothalamus. Body temperature is not constant all day, but actually is lowest at 6 A.M. and highest around 4–6 P.M. In addition, temperature varies in different regions of the body; for example, rectal and urine temperatures are about one degree Fahrenheit higher than oral temperature and rectal temperature is higher than urine. It is also important to realize that certain normal conditions can effect body temperature, such as **pregnancy**, food ingestion, age, and certain hormonal changes.

Substances that cause fever are known as “pyrogens.” There are two types of pyrogens; exogenous and endogenous. Those that originate outside the body, such as bacterial toxins, are called “exogenous” pyrogens. Pyrogens formed by the body’s own cells in response to an outside stimulus (such as a bacterial toxin) are called “endogenous” pyrogens.

Researchers have discovered that there are several “endogenous” pyrogens. These are made up of small groups of amino acids, the building blocks of proteins. These natural pyrogens have other functions in addition to inducing fever; they have been named “cytokines”. When cytokines are injected into humans, fever and chills develop within an hour. Interferon, tumor necrosis factor, and various interleukins are the major fever producing cytokines.

The production of fever is a very complex process; somehow, these cytokines cause the thermoregulatory center in the hypothalamus to reset the normal temperature level. The body’s initial response is to conserve heat by vasoconstriction, a process in which blood vessels narrow and prevent heat loss from the skin and elsewhere. This alone will raise temperature by two to three degrees. Certain behavioral activities also occur, such as adding more clothes, seeking a warmer environment, etc. If the hypothalamus requires more heat, then shivering occurs.

Fever is a body defense mechanism. It has been shown that one of the effects of temperature increase is to slow bacterial growth. However, fever also has some downsides; the body’s metabolic rate is increased and with it, oxygen consumption. This can have a devastating effect on those with poor circulation. In addition, fever can lead to seizures in the very young.

When temperature elevation occurs for an extended period of time and no cause is found, the term FUO is then used. The far majority of these patients are eventually found to have one of several diseases.

## KEY TERMS

**AIDS**—Acquired immune deficiency syndrome is often represented by these initials. The disease is associated with infection by the human immunodeficiency virus (HIV), and has the main feature of repeated infections, due to failure of certain parts of the immune system. Infection by HIV damages part of the body’s natural immunity, and leads to recurrent illnesses.

**Antibiotic**—A medication that is designed to kill or weaken bacteria.

**Computed tomography scan (CT Scan)**—A specialized x-ray procedure in which cross-sections of the area in question can be examined in detail. This allows physicians to examine organs such as the pancreas, bile ducts, and others which are often the site of hidden infections.

**Magnetic Resonance Imaging (MRI)**—This is a new technique similar to CT Scan, but based on the magnetic properties of various areas of the body to compose images.

**NSAID**—Nonsteroidal anti-inflammatory drugs are medications such as aspirin and ibuprofen that decrease pain and inflammation. Many can now be obtained without a doctor’s prescription.

**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 is very useful for diagnosing diseases of the gallbladder, liver, and hidden infections, such as abscesses.

## Causes and symptoms

The most frequent cause of FUO is still infection, though the percentage has decreased in recent years. **Tuberculosis** remains an important cause, especially when it occurs outside the lungs. The decrease in infections as a cause of FUO is due in part to improved culture techniques. In addition, technological advances have made it easier to diagnose non-infectious causes. For example, tumors and autoimmune diseases in particular are now easier to diagnose. (An autoimmune disease is one that arises when the body tolerance for its own cell antigenic cell markers disappears.)

**Allergies** to medications can also cause prolonged fever; sometimes patients will have other symptoms suggesting an allergic reaction, such as a rash.

There are many possible causes of FUO; generally though, a diagnosis can be found. About 10% of patients will wind up without a definite cause, and about the same percentage have “factitious fevers” (either self induced or no fever at all).

Some general symptoms tend to occur along with fever; these are called constitutional symptoms and consist of myalgias (muscle aches), chills, and **headache**.

## Diagnosis

Few symptoms in medicine present such a diagnostic challenge as fever. Nonetheless, if a careful, logical, and thorough evaluation is performed, a diagnosis will be found in most cases. The patient's past medical history as well as travel, social, and family history should be carefully searched for important clues.

Usually the first step is to search for an infectious cause. Skin and other screening tests for diseases such as tuberculosis, and examination of blood, urine, and stool, are generally indicated. Antibody levels to a number of infectious agents can be measured; if these are rising, they may point to an active infection.

Various x-ray studies are also of value. In addition to standard examinations, recently developed radiological techniques using ultrasound, computed tomography scan (CT scan) and **magnetic resonance imaging** (MRI) scans are now available. These enable physicians to examine areas that were once accessible only through surgery. Furthermore, new studies using radioactive materials (nuclear medicine), can detect areas of infection and inflammation previously almost impossible to find, even with surgery.

Biopsies of any suspicious areas found on an x-ray exam can be performed by either traditional or newer surgical techniques. Material obtained by biopsy is then examined by a pathologist to look for clues as to the cause of the fever. Evidence of infection, tumor or other diseases can be found in this way. Portions of the biopsy are also sent to the laboratory for culture in an attempt to grow and identify an infectious organism.

Patients with HIV are an especially difficult problem, as they often suffer from many unusual infections. HIV itself is a potential cause of fever.

## Treatment

Most patients who undergo evaluation for FUO do not receive treatment until a clear-cut cause is found. **Antibiotics** or medications designed to suppress a fever (such as NSAIDs) will only hide the true cause. Once physicians are satisfied that there is no infectious cause, they may use medications such as NSAIDs, or **corticosteroids** to decrease inflammation and diminish constitutional symptoms.

The development of FUO in certain settings, such as that acquired by patients in the hospital or in those with a low white **blood count**, often needs rapid treatment to avoid serious complications. Therefore, in these instances patients may be placed on antibiotics after a minimal number of diagnostic studies. Once test results are known, treatment can be adjusted as needed.

## Prognosis

The outlook for patients with FUO depends on the cause of the fever. If the basic illness is easily treatable and can be found rather quickly, the potential for a cure is quite good. Some patients continue with temperature elevations for six months or more; if no serious disease is found, medications such as NSAIDs are used to decrease the effects of the fever. Careful follow-up and reevaluation is recommended in these cases.

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Fiber-modified diet see **Diets**

Fibrin degradation products see **Fibrin split products**

## Fibrin split products

### Definition

Fibrin split products (FSP) are fragments of protein released from a dissolving clot. The fibrin split products test is one of several tests done to evaluate a person with

blood clotting problems (coagulation), particularly disseminated intravascular coagulation (DIC).

## Purpose

High levels of FSP in a person's blood are associated with DIC, a serious medical condition that develops when the normal balance between bleeding and clotting is disturbed. Excessive bleeding and clotting injures body organs, and causes anemia or **death**.

## Description

Coagulation begins typically with an injury to some part of the body. The injury sets in motion a cascade of biochemical activities (the coagulation cascade) to stop the bleeding, by forming a clot from a mixture of the blood protein fibrin and platelets.

Once bleeding is stopped, another blood protein dissolves the clot by breaking down the fibrin into fragments. Measurement of these fragments gives information about the clot dissolving portion of coagulation, called fibrinolysis.

In DIC, the coagulation cascade is triggered in an abnormal way. A blood infection, a **transfusion** reaction, a large amount of tissue damage, such as a burn, a dead fetus, and some cancers can begin the chain of biochemical events leading to blood clots. The coagulation cascade becomes overwhelmed with excessive clotting followed by excessive bleeding. As the large number of clots dissolve, fibrin split products accumulate in the blood and encourage even more bleeding.

Laboratory tests for FSP are done on the yellow liquid portion left over after blood clots (serum). A person's serum is mixed with a substance that binds to FSP. This bound complex is measured, and the original amount of FSP is determined. Some test methods give an actual measurement of FSP; some give a titer, or dilution. Methods that provide a titer look for the presence or absence of FSP. If the serum is positive for FSP, the serum is diluted, or titered, and the test is done again. These steps are repeated until the serum is so dilute that it no longer gives a positive result. The last dilution that gives a positive result is the titer reported.

The FSP test is covered by insurance when medically necessary. Results are usually available within one to two hours. Other names for this test are fibrin degradation products, fibrin breakdown products, or FDP.

## Preparation

This test requires 0.17 oz (5 ml) of blood. A healthcare worker ties a tourniquet on the patient's upper arm,

## KEY TERMS

**Coagulation**—The entire process of blood clotting.

**Coagulation cascade**—A sequence of biochemical activities to stop bleeding by forming a clot.

**Disseminated intravascular coagulation (DIC)**—A serious medical condition that develops when the normal balance between bleeding and clotting is disturbed. Excessive bleeding and clotting injures body organs, and causes anemia or death.

**Fibrin split products (FSP)**—Pieces of the protein fibrin released from a dissolving clot.

**Fibrinolysis**—The clot dissolving portion of the coagulation process.

**Titer**—A dilution of a substance with an exact known amount of fluid. For example, one part of serum diluted with four parts of saline is a titer of 1:4.

locates a vein in the inner elbow region, and inserts a needle into that vein. Vacuum action draws the blood through the needle into an attached tube. Collection of the sample takes only a few minutes.

## Aftercare

Discomfort or bruising may occur at the puncture site. Pressure applied to the puncture site until the bleeding stops reduces bruising. Warm packs to the puncture site relieve discomfort. The patient may feel dizzy or faint.

## Risks

People with coagulation problems may bleed longer than normal. The healthcare provider must make sure bleeding has stopped before leaving the patient unattended.

## Normal results

Negative at a less than or equal to 1:4 dilution or less than 10 g/mL.

## Abnormal results

High levels of FSP indicate DIC. Results of the test must be interpreted by the physician according to the person's clinical symptoms and medical history. Other conditions that increase blood clotting activity also increase FSP: venous thrombosis, surgery and trans-

plants, blood clots in the lung, certain cancers, and **heart attack** (myocardial infarction).

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## Fibrinogen test

### Definition

Fibrinogen (Factor I) is a protein that originates in the liver. It is converted to fibrin during the blood-clotting process (coagulation).

### Purpose

The fibrinogen test aids in the diagnosis of suspected clotting or bleeding disorders caused by fibrinogen abnormalities.

### Precautions

This test is not recommended for patients with active bleeding, acute infection or illness, or in those patients who have received blood transfusions within four weeks.

Drugs that may increase fibrinogen levels include estrogens and **oral contraceptives**. Drugs that may cause decreased levels include anabolic steroids, androgens, phenobarbital, urokinase, streptokinase, and valproic acid.

### Description

Fibrinogen plays two essential roles in the body: it is a protein called an acute-phase reactant that becomes elevated with tissue inflammation or tissue destruction, and it is also a vital part of the "common pathway" of the coagulation process.

In order for blood to clot, fibrinogen must be converted to fibrin by the action of an enzyme called thrombin. Fibrin molecules clump together to form long filaments, which trap blood cells to form a solid clot.

## KEY TERMS

**Fibrin**—The last step in the coagulation process. Fibrin forms strands that add bulk to a forming blood clot to hold it in place and help "plug" an injured blood vessel wall.

**Platelet**—An irregularly shaped cell-like particle in the blood that is an important part of blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process.

**Prothrombin**—A type of protein called a glycoprotein that is converted to thrombin during the clotting process.

**Thrombin**—An enzyme that converts fibrinogen into strands of fibrin.

The conversion of fibrinogen to fibrin is the last step of the "coagulation cascade," a series of reactions in the blood triggered by tissue injury and platelet activation. With each step in the cascade, a coagulation factor in the blood is converted from an inactive to an active form. The active form of the factor then activates several molecules of the next factor in the series, and so on, until the final step, when fibrinogen is converted into fibrin.

The factors involved in the coagulation cascade are numbered I, II, and V through XIII. Factor I is fibrinogen, while factor II (fibrinogen's immediate precursor) is called prothrombin. Most of the coagulation factors are made in the liver, which needs an adequate supply of vitamin K to manufacture the different clotting factors.

When fibrinogen acts as an "acute-phase reactant," it rises sharply during tissue inflammation or injury. When this occurs, high fibrinogen levels may be a predictor for an increased risk of heart or circulatory disease. Other conditions in which fibrinogen is elevated are cancers of the stomach, breast, or kidney, and inflammatory disorders like **rheumatoid arthritis**.

Reduced fibrinogen levels can be found in liver disease, **prostate cancer**, lung disease, bone marrow lesions, malnourishment, and certain bleeding disorders. The low levels can be used to evaluate disseminated intravascular coagulation (DIC), a serious medical condition that develops when there is a disturbed balance between bleeding and clotting. Other conditions related to decreased fibrinogen levels are those in which fibrino-

gen is completely absent (congenital afibrinogenemia), conditions in which levels are low (hypofibrinogenemia), and conditions of abnormal fibrinogen (dysfibrinogenemia). Obstetric complications or trauma may also cause low levels. Large-volume blood transfusions cause low levels because banked blood does not contain fibrinogen.

## Preparation

This test is performed with a blood sample, which can be drawn at any time of day. The patient does not have to be **fasting** (nothing to eat or drink).

## Aftercare

Because a fibrinogen test is often ordered when a bleeding disorder is suspected, the patient should apply pressure or a pressure dressing to the blood-drawn site for a period of time after blood is drawn, and then reexamine the site for bleeding.

## Risks

Risks for this test are minimal, but may include slight bleeding from the blood-drawing site, **fainting** or feeling lightheaded after procedure, or the seeing the accumulation of blood under the puncture site (hematoma).

## Normal results

Normal reference ranges are laboratory-specific, but are usually within the following:

- adult: 200 mg/dL–400 mg/dL
- newborn: 125 mg/dL–300 mg/dL

## Abnormal results

Spontaneous bleeding can occur with values less than 100 mg/dL.

## Resources

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Fibrocystic breast disease see **Fibrocystic condition of the breast**

# Fibroadenoma

## Definition

Fibroadenomas are benign breast tumors commonly found in young women. Fibroadenoma means “a tumor composed of glandular (related to gland) and fibrous (containing fibers) tissues.”

## Description

Breast fibroadenomas, abnormal growths of glandular and fibrous tissues, are most common between the ages of 15 and 30, and are found in 10% of all women (20% of African-American women). They are found rarely in postmenopausal women.

Described as feeling like marbles, these firm, round, movable, and “rubbery” lumps range from 1–5 cm in size. Giant fibroadenomas are larger, lemon-sized lumps. Usually single, from 10–15% of women have more than one.

While some types of breast lumps come and go during the menstrual cycle, fibroadenomas typically do not disappear after a woman’s period, and should be checked by a doctor.

## Causes and symptoms

The cause of breast fibroadenomas is unknown. They may be dependent upon estrogen, because they are common in premenopausal women, can be found in postmenopausal women taking estrogen, and because they grow larger in pregnant women.

Fibroadenomas usually cause no symptoms and may be discovered during **breast self-examination**, or during a routine check-up.

## Diagnosis

When the doctor takes a complete medical history, they will ask when the lump was first noticed, if there were any symptoms or changes in lump size, and if there is any personal or family history of breast disease.

The doctor thoroughly feels the breasts (palpates). Tests are done, usually including **mammography** or ultrasound scans, or surgical removal of cells or tissue for examination under a the microscope (biopsy).

Diagnostic tests include:

- Mammogram. An x-ray examination of the breast.
- Ultrasound scan. A technique that uses sound waves to display a two-dimensional image of the breast, showing whether a lump is solid or fluid-filled (cystic).
- Fine-needle aspiration biopsy. A minor procedure wherein fluid or cells are drawn out of the lump through a small needle (aspirated).

## KEY TERMS

**Aspiration** —To withdraw material with a needle and syringe.

**Biopsy** —To remove cells or tissue for microscopic examination.

**Estrogen** —Female sex hormone produced by the ovaries.

- **Core biopsy.** A procedure wherein a larger piece of tissue is withdrawn from the lump through a larger needle.
- **Incisional biopsy.** A surgical procedure wherein a piece of the lump is removed through an cut (incision).
- **Excisional biopsy.** A surgical procedure wherein the entire lump is removed through an cut (incision).

Most insurance plans cover the costs of diagnosing and treating fibroadenomas.

## Treatment

Performed usually in outpatient settings, breast fibroadenomas are removed by **lumpectomy**, or surgical excision under local or general anesthesia. Sometimes lumps in younger women are not removed but are monitored by self-examination, yearly doctor check-ups, and mammograms. Surgery is generally recommended for women over 30, and for lumps that are painful or enlarging.

## Alternative treatments

Alternative treatments for breast fibroadenomas include a low-fat, high-fiber, vegetarian-type diet; a reduction in **caffeine** intake; supplementation with evening primrose oil (*Oenothera biennis*), flax oil, or fish oil and **vitamins E** and **C**; and the application of hot compresses to the breast. In addition, a focus on liver cleansing is important to assist the body in conjugation and elimination of excess estrogens. Botanical remedies can be useful in hormone balancing, as can **acupuncture** and **homeopathy**. Massaging the breasts with castor oil, straight or infused with herbs or essential oils, can help fibroadenomas reduce and dissipate, as well as keep women in touch with changes in their breast tissue.

## Prognosis

Breast fibroadenomas are not cancerous. The lumps recur in up to 20% of women. A small number of lumps disappear on their own.

## Prevention

Breast fibroadenomas cannot be prevented. They can be discovered early by regular breast self-examination.

## Resources

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### ORGANIZATIONS

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## Fibrocystic condition of the breast

### Definition

Fibrocystic condition of the breast is a term that may refer to a variety of symptoms: breast lumpiness or tenderness, microscopic breast tissue, and/or the x ray or ultrasound picture of the breast. It has been called a

“wastebasket” diagnosis because a wide range of vaguely defined benign breast conditions may be labeled as fibrocystic condition. It is not a **cancer**, and the majority of types of fibrocystic conditions do not increase the risk of **breast cancer**.

## Description

There is no such thing as a normal or typical female breast. Breasts come in all shapes and sizes, with varying textures from smooth to extremely lumpy. The tissues of the female breast change in response to hormone levels, normal **aging**, nursing (**lactation**), weight fluctuations, and injury. To further complicate matters, the breast has several types of tissue; each of these tissue types may respond differently to changes in body chemistry.

Fibrocystic breast condition may be called fibrocystic disease, although it is clearly not a single, specific disease process. Variations or changes in the way the breast feels or looks on x ray may cause the condition to be called “fibrocystic change.” Other names have been used to refer to this imprecise and ill-defined term: mammary dysplasia, mastopathy, chronic cystic **mastitis**, indurative mastopathy, mastalgia, lumpy breasts, or physiologic nodularity.

Estimates vary, but 40–90% of all women have some evidence of “fibrocystic” condition, change, or disease. It is most common among women between the ages 30 and 50, but may be seen at other ages.

## Causes and symptoms

Fibrocystic condition of the breast refers to technical findings on diagnostic testing (signs); however, this discussion focuses on symptoms that may fall under the general category of the fibrocystic condition. First, a brief review of the structure and function of the breast may be useful.

The breast is not supposed to be a soft, smooth organ. It is actually a type of sweat gland. Milk, the breasts’ version of sweat, is secreted when the breast receives appropriate hormonal and environmental stimulation.

The normal breast contains milk glands, with their accompanying ducts, or pipelines, for transporting the milk. These complex structures may not only alter in size, but can increase or decrease in number as needed. Fibrous connective tissue, fatty tissue, nerves, blood and lymph vessels, and lymph nodes, with their different shapes and textures, lie among the ever-changing milk glands. It is no wonder that a woman’s breasts may not feel uniform in texture and that the “lumpiness” may wax and wane.

The fibrocystic condition refers to the tenderness, enlargement, and/or changing “lumpiness” that many

women encounter just before or during their menstrual periods. At this time, female hormones are preparing the breasts for **pregnancy**, by stimulating the milk-producing cells, and storing fluid. Each breast may contain as much as three to six teaspoons of excess fluid. Swelling, with increased sensitivity or **pain**, may result. If pregnancy does not occur, the body reabsorbs the fluid, and the engorgement and discomfort are relieved.

Symptoms of fibrocystic breast condition range from mildly annoying in some women to extremely painful in others. The severity of discomfort may vary from month to month in the same woman. Although sometimes distressing, this experience is the body’s normal response to routine hormonal changes.

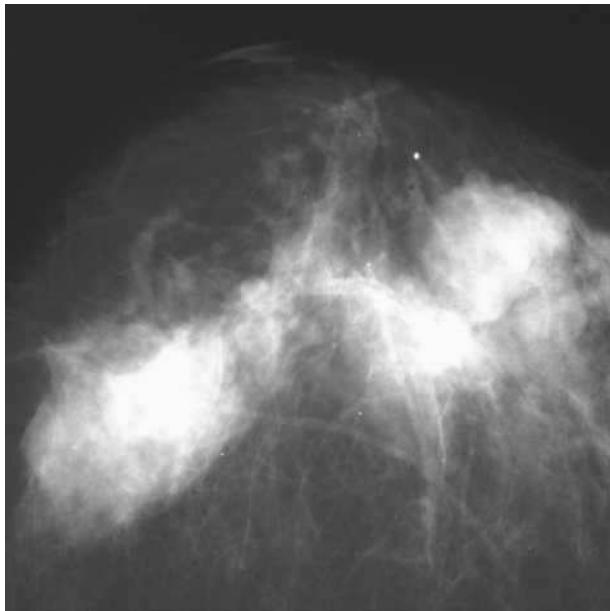
This cycle of breast sensitivity, pain and/or enlargement, can also result from medications. Some hormone replacement therapies (estrogen and progesterone) used for postmenopausal women can produce these effects. Other medications, primarily, but not exclusively those with hormones may also provoke these symptoms.

Breast pain unrelated to hormone shifts is called “noncyclic” pain. “Trigger-zone breast pain” is a term that may also be used to describe this area-specific pain. This type of pain may be continuous, or it may be felt intermittently. Trauma, such as a blow to the chest area, a prior **breast biopsy**, or sensitivity to certain medications may also underlie this type of pain. Fibrocystic condition of the breast may be cited as the cause of otherwise unexplained breast pain.

Lumps, apart from those clearly associated with hormone cycles, may also be placed under the heading of fibrocystic condition. These lumps stand out from enlarged general breast tissue. Although noncancerous lumps may occur, the obvious concern with such lumps is cancer.

Noncancerous breast lumps include:

- Adenosis. This condition refers to the enlargement of breast lobules, which contain a greater number of glands than usual. If a group of lobules are found near each other, the affected area may be large enough to be felt.
- Cysts. These are fluid-filled sacs in the breast and probably develop as ducts that become clogged with old cells in the process of normal emptying and filling. Cysts usually feel soft and round or oval. However a cyst deep within the breast may feel hard, as it pushes up against firmer breast tissue. A woman with a cyst may experience pain, especially if it increases in size before her menstrual cycle, as is often the case. Women between the age of 30 and 50 are most likely to develop cysts.
- Epithelial hyperplasia. Also called proliferative breast disease, this condition refers to an overgrowth of cells lining either the ducts or the lobules.



**A mammogram of a female breast indicating multiple cysts.**  
(Custom Medical Stock Photo. Reproduced by permission.)

- Fibroadenomas. These are tumors that form in the tissues outside the milk ducts. The cause of fibroadenomas is unknown. They generally feel smooth and firm, with a somewhat rubber-like texture. Typically a **fibroadenoma** is not attached to surrounding tissue and moves slightly when touched. They are most commonly found in adolescents and women in their early twenties but can occur at any age.
- Fibrosis. Sometimes one area of breast tissue persistently feels thicker or more prominent than the rest of the breast. This feeling may be caused by old hardened scar tissue and/or dead fat tissue as a result of surgery or trauma. Often the cause of this type of breast tissue is unknown.
- Miscellaneous disorders. A number of other benign (noncancerous) breast problems may be placed under the heading of “fibrocystic condition.” These problems include disorders that may lead to breast inflammation (mastitis), infection, and/or nipple discharge.

#### *Atypical ductal hyperplasia*

The condition known as atypical ductal hyperplasia (ADH) is a condition in which the cells lining the milk ducts of the breast are growing abnormally. This condition may appear as spots of calcium salts, or calcifications, on the mammogram. A biopsy removed from the breast would confirm the diagnosis. Atypical ductal hyperplasia is not a cancer. In most women, this condition will cause no problems. However, for some women, especially women with family histories of breast cancer,

the risk of developing breast cancer is increased. (One study with over 3,000 female participants indicated that about 20% of the participants with atypical hyperplasia and a family history of breast cancer developed breast cancer, as compared to the 8% of participants who developed the disease with atypical hyperplasia and no family history of breast cancer.) For women with ADH and a family history of breast cancer, more frequent mammograms and closer monitoring may be required.

#### **Diagnosis**

Breast cancer is the most common concern of women who feel a breast lump or experience an abnormal breast symptom. For peace of mind, and to rule out any possibility of cancer, any newly discovered breast lumps should be brought to the attention of a family physician or an obstetrician-gynecologist. He or she will obtain a history and conduct thorough **physical examination** of the area. Depending on the findings on physical examination, the patient is usually referred for tests. The most common of these tests include:

- **Mammography.** A mammogram is an x-ray examination of the breasts. The two major types of abnormalities doctors look for are masses and calcifications; either abnormality may be benign or malignant. The size, shape, and edges of these masses help doctors determine whether or not cancer is present. Sometimes, however, this test may be difficult to interpret, however, due to dense breast tissue.
- **Ultrasonography.** If a suspicious lump is detected during mammography, an ultrasound (the use of high-frequency sound waves to outline the shape of various organs and tissues in the body) is useful (although not definitive) in distinguishing benign from cancerous growths.
- **Ductography.** A ductogram (also called a galactogram) is a test that is sometimes useful in evaluating nipple discharge. A very fine tube is threaded into the opening of the duct onto the nipple. A small amount of dye is injected, outlining the shape of the duct on an x ray, and indicates whether or not there is a mass in the duct.
- **Biopsy.** If a lump cannot be proven benign by mammography and ultrasound, a breast biopsy may be considered. Usually a tissue sample is removed through a needle (fine-needle aspiration biopsy, or FNAB) to obtain a sample of the lump. The sample is examined under the microscope by a pathologist, and a detailed diagnosis regarding the type of benign lesion or cancer is established. In some cases, however, FNAB may not provide a clear diagnosis, and another type of biopsy (such as a surgical biopsy, core-needle biopsy, or other stereotactic biopsy methods—such as the Mammotome or Advanced Breast Biopsy Instrument) may be required.

Other breast conditions such as inflammation or infection are usually recognized on the basis of suspicious history, breastfeeding, or characteristic symptoms such as pain, redness, and swelling. A positive response to appropriate therapies often confirms the diagnosis.

## Treatment

Once a specific disorder within the broad category of fibrocystic condition is identified, treatment can be prescribed. There are a number of treatment options for women with a lump that has been diagnosed as benign. If it is not causing a great deal of pain, the growth may be left in the breast. However, some women may choose to have a lump such as a fibroadenoma surgically removed, especially if it is large. Another option to relieve the discomfort of a painful benign lump is to have the cyst suctioned, or drained. If there is any uncertainty regarding diagnosis, the fluid may be sent to the lab for analysis.

Symptoms of cycle breast sensitivity and engorgement may also be treated with diet, medication, and/or physical modifications. For example,

- Although there is no scientific data to support this claim, many women have reported relief of symptoms when **caffeine** was reduced or eliminated from their **diets**. Decreasing salt before and during the period when breasts are most sensitive may also ease swelling and discomfort. Low-fat diets and elimination of dairy products also appear to decrease soreness for some women. However, it may take several months to realize the effects of these various treatments.
- Over-the-counter **analgesics** such as **acetaminophen** (Tylenol) or ibuprofen (Advil) may be recommended. In some cases, treatment with prescription drugs such as hormones or hormone blockers may prove successful. **Oral contraceptives** may also be prescribed.
- Warm soaks or ice packs may provide comfort. A well-fitted support bra can minimize physical movement and do much to relieve breast discomfort. Breast massage may promote removal of excess fluid from tissues and alleviate symptoms. Massaging the breast with castor oil, straight or infused with herbs or essential oils, can help reduce and dissipate fibroadenomas as well as keep women in touch with changes in their breast tissue.
- Infections are often treated with warm compresses and **antibiotics**. Lactating women are encouraged to continue breastfeeding because it promotes drainage and healing. However, a serious infection may progress to form an **abscess** that may need surgical drainage.
- Some studies of alternative or complementary treatments, although controversial, have indicated that **vitamins A, B complex and E**, and mineral supplements

may reduce the risk of developing fibrocystic condition of the breast. Evening primrose oil (*Oenothera biennis*), flaxseed oil, and fish oils have been reported to be effective in relieving cyclic breast pain for some women.

## Prognosis

Most benign breast conditions carry no increased risk for the development of breast cancer. However, a small percentage of biopsies uncover overgrowth of tissue in a particular pattern in some women; this pattern indicates a 15–20% increased risk of breast cancer over the next 20 years. Strict attention to early detection measures, such as annual mammograms, is especially important for these women.

## Prevention

There is no proven method of preventing the various manifestations of fibrocystic condition from occurring. Some alternative health care practitioners believe that eliminating foods high in methyl xanthines (primarily coffee and chocolate) can decrease or reverse fibrocystic breast changes.

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## KEY TERMS

- Advanced Breast Biopsy Instrument (ABBI)**—Uses a rotating circular knife and thin heated electrical wire to remove a large cylinder of abnormal breast tissue.
- Lobules**—A small lobe or subdivision of a lobe (often on a gland) that may be seen on the surface of the gland by bumps or bulges.
- Lymph nodes**—Rounded, encapsulated bodies consisting of an accumulation of lymphatic tissue.
- Mammotome**—A method for removing breast biopsies using suction to draw tissue into an opening in the side of a cylinder inserted into the breast tissue. A rotating knife then cuts tissue samples from the rest of the breast; also known as a vacuum-assisted biopsy
- Stereotactic biopsy**—A biopsy taken by precisely locating areas of abnormal growth through the use of delicate instruments.

### ORGANIZATIONS

- American Cancer Society. 1599 Clifton Rd. NE, Atlanta, GA 30329. (800) ACS-2345 <<http://www.cancer.org>>.
- American College of Obstetricians and Gynecologists. 409 12th St., S.W., P.O. Box 96920, Washington, DC 20090-6920. <<http://www.acog.org>>.
- Cancer Information Service (CIS). 9000 Rockville Pike, Building 31, Suite 10A18, Bethesda, MD 20892. (800) 4-CANCER.

### OTHER

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Fibroids see **Uterine fibroids**

## Fibromyalgia

### Definition

Fibromyalgia is described as inflammation of the fibrous or connective tissue of the body. Widespread muscle **pain**, **fatigue**, and multiple tender points charac-

terize these conditions. Fibrositis, fibromyalgia, and fibromyositis are names given to a set of symptoms believed to be caused by the same general problem.

### Description

Fibromyalgia is more common than previously thought, with as many as 3–6% of the population affected by the disorder. Fibromyalgia is more prevalent in adults than children, with more women affected than men, particularly women of childbearing age.

### Causes and symptoms

The exact cause of fibromyalgia is not known. Sometimes it occurs in several members of a family, suggesting that it may be an inherited disorder. People with fibromyalgia are most likely to complain of three primary symptoms: muscle and joint pain, stiffness, and fatigue.

Pain is the major symptom with aches, tenderness, and stiffness of multiple muscles, joints, and soft tissues. The pain also tends to move from one part of the body to another. It is most common in the neck, shoulders, chest, arms, legs, hips, and back. Although the pain is present most of the time and may last for years, the severity of the pain is changeable and dependent on individual patient perception.

Symptoms of fatigue may result from the individual's chronic pain coupled with **anxiety** about the problem and how to find relief. The inflammatory process also produces chemicals that are known to cause fatigue. Other common symptoms are tension headaches, difficulty swallowing, recurrent abdominal pain, **diarrhea**, and numbness or tingling of the extremities. **Stress**, anxiety, depression, or lack of sleep can increase symptoms. Intensity of symptoms is variable ranging from gradual improvement to episodes of recurrent symptoms.

### Diagnosis

Diagnosis is difficult and frequently missed because symptoms of fibromyalgia are vague and generalized. Coexisting nerve and muscle disorders such as **rheumatoid arthritis**, spinal arthritis, or **Lyme disease** may further complicate the diagnostic process. Presently, there are no tests available to specifically diagnose fibromyalgia. The diagnosis is usually made after ruling out other medical conditions with similar symptoms.

Because of the emotional distress experienced by people with this condition and the influence of stress on the symptoms themselves, fibromyalgia has often been labeled a psychological problem. Recognition of the

underlying inflammatory process involved in fibromyalgia has helped promote the validity of this disease.

In 1990, the America College of Rheumatology developed standards for fibromyalgia that health care practitioners can use to diagnose this condition. According to these standards, a person is thought to have fibromyalgia if he or she has widespread pain in combination with tenderness in at least 11 of the 18 sites known as trigger points. Trigger point sites include the base of the neck, along the backbone, in front of the hip and elbow, and at the rear of the knee and shoulder.

## Treatment

There is no known cure for fibromyalgia. Therefore, the goal of treatment is successful symptom management. Treatment usually requires a combination of therapies, **exercise**, proper rest, and diet. A patient's clear understanding of his or her role in the recovery process is imperative for successful management of this condition.

Treatments found to be helpful include heat and occasionally cold applications. A regular stretching program is often useful. Aerobic activities focusing on increasing the heart rate are the preferred forms of exercise over most other forms of exertion. Exercise programs need to include good warm-up and cool-down sessions, with special attention given to avoiding exercises causing joint pain. The diet should include a large variety of fruits and vegetables which provide the body with trace elements and **minerals** that are necessary for healthy muscles.

Adequate rest is essential in the treatment of fibromyalgia. Avoidance of stimulating foods or drinks (such as coffee) and medications like **decongestants** prior to bedtime is advised. If diet, exercise, and adequate rest do not relieve the symptoms of fibromyalgia, medications may be prescribed. Medications prescribed and found to have some benefit include **antidepressant drugs**, **muscle relaxants**, and anti-inflammatory drugs.

People with fibromyalgia often need a rheumatology consultation (a meeting with a doctor who specializes in disorders of the joints, muscles, and soft tissue) to decide the cause of various rheumatic symptoms, to be educated about fibromyalgia and its treatment, and to exclude other rheumatic diseases. A treatment program must be individualized to meet the patient's needs. The rheumatologist, as the team leader, enlists and coordinates the expertise of other health professionals in the care of the patient.

## Alternative treatment

**Massage therapy** can be helpful, especially when a family member is instructed on specific massage techniques to manage episodes of increased symptoms. Spe-

## KEY TERMS

**Connective tissue**—Tissue that supports and binds other body tissue and parts.

**Lyme disease**—An acute recurrent inflammatory disease involving one or a few joints, believed to be transmitted by a tickborne virus. The condition was originally described in the community of Lyme, Connecticut, but has also been reported in other parts of the United States and other countries. Knees, other large joints are most commonly involved with local inflammation and swelling.

**Rheumatology**—The study of disorders characterized by inflammation, degeneration of connective tissue, and related structures of the body. These disorders are sometimes collectively referred to as rheumatism.

cific attention to mental health, including psychological consultation, may also be important, since depression may precede or accompany fibromyalgia. Other alternative therapies, including **hellerwork**, **rolfing**, homeopathic medicine, Chinese traditional medicine (both **acupuncture** and herbs), **polarity therapy**, and Western botanical medicine, can assist the person with fibromyalgia to function day to day and can contribute to healing.

## Prognosis

Fibromyalgia is a chronic problem. The symptoms sometimes improve and at other times worsen, but they often continue for months to years.

## Prevention

There is no known or specific way to prevent fibromyalgia. However, similar to many other medical conditions, remaining as healthy as possible with a good diet, safe exercise, and adequate rest is the best prevention.

## Resources

### BOOKS

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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>>.

Arthritis Foundation. 1300 W. Peachtree St., Atlanta, GA 30309. (800) 283-7800. <<http://www.arthritis.org>>.

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Fibromyomas see **Uterine fibroids**  
 Fibrous breast lumps see **Fibroadenoma**

## I Fifth disease

### Definition

Fifth disease is a mild childhood illness caused by the human parvovirus B19 that causes flu-like symptoms and a rash. It is called fifth disease because it was fifth on a list of common childhood illnesses that are accompanied by a rash, including **measles**, **rubella** or German measles, **scarlet fever** (or scarlatina), and scarlatinella, a variant of scarlet fever.

### Description

The Latin name for the disease is *erythema infectiosum*, meaning infectious redness. It is also called the “slapped cheek disease” because, when the bright red rash first appears on the cheeks, it looks as if the face has been slapped. Anyone can get the disease, but it occurs more frequently in school-aged children. The disease is usually mild, and both children and adults usually recover quickly without complications. In fact, some individuals exhibit no symptoms and never even feel ill. Outbreaks most often occur in the winter and spring.

### Causes and symptoms

Fifth disease is caused by the human parvovirus B19, a member of the Parvoviridae family of viruses, that lives in the nose and throat of the infected person. The virus is spread through the air by coughing and sneezing. Because the virus needs a rapidly dividing cell in order to multiply, it attacks the red blood cells of the body. Once infected, a person is believed to be immune to reinfection.

Symptoms may appear four to 21 days after being exposed to the virus. Initial symptoms are flu-like and include **headache**, body ache, **sore throat**, a mild fever of 101°F (38.3°C), and chills. It is at this time, prior to the development of the rash, that individuals are contagious. These symptoms last for two to three days. In children, a bright red rash that looks like a slap mark develops suddenly on the cheeks. The rash may be flat or raised and may or may not be itchy. Sometimes, the rash spreads to the arms, legs, and trunk, where it has a lace-like or net-like appearance. The rash can also involve the palms of the hands and soles of the feet. By the time the rash appears, individuals are no longer infectious. On average, the rash lasts for 10–11 days, but may last for as



This infant has a rash caused by Fifth disease, or erythema infectiosum. (Custom Medical Stock Photo. Reproduced by permission.)

long as five to six weeks. The rash may fade away and then reappear upon exposure to sunlight, hot baths, emotional distress, or vigorous exercise.

Adults generally do not develop a rash, but instead may have swollen and painful joints, especially in the hands and feet. In adults, symptoms such as sore throat, headache, muscle and joint pain, abdominal pain, **diarrhea**, and vomiting occur more frequently than in children and are usually more severe. The joint pain can be arthritis-like and last for several months, especially in women, but the disease does not appear to progress to **rheumatoid arthritis**.

The virus causes the destruction of red blood cells and, therefore, a deficiency in the oxygen-carrying capacity of the blood (anemia) can result. In healthy people, the anemia is mild and only lasts a short while. In people with weakened immune systems, however, either because they have a chronic disease like **AIDS** or **cancer** (immunocompromised), or are receiving medication to suppress the immune system (immunosuppressed), such as organ transplant recipients, this anemia can be severe and last long after the infection has subsided. Symptoms of anemia include **fatigue**, lack of color, lack of energy, and **shortness of breath**. Some individuals with sickle cell anemia, iron deficiency, a number of different hereditary blood disorders, and those who have received bone marrow transplants may be susceptible to developing a potentially life-threatening complication called a transient aplastic crisis where the body is temporarily unable to form new red blood cells.

In very rare instances, the virus can cause inflammation of different areas of the body, including the brain (**encephalitis**), the covering of the brain and spinal cord (**meningitis**), the lungs (pneumonitis), the liver (hepatitis), and the heart muscle (**myocarditis**). The virus can also aggravate symptoms for people with an autoimmune disease called **systemic lupus erythematosus**.

There is some concern about fifth disease in pregnant women. Although no association with an increased number of **birth defects** has been demonstrated, there is concern that infection during the first three months of **pregnancy** may lead to a slight increase in the number of miscarriages. There is also some concern that infection later in pregnancy may involve a very small risk of premature delivery or stillbirths. As a result, women who get fifth disease while they are pregnant should be monitored closely by a physician.

## Diagnosis

Fifth disease is usually suspected based on a patient's symptoms, including the typical appearance of the bright red rash on the cheeks, patient history, age, and the time of year. The physician will also exclude other potential causes for the symptoms and rash, including rubella, **infectious mononucleosis**, bacterial infections like **Lyme disease**, allergic reactions, and lupus.

In addition, there is a blood test for fifth disease, but it is generally used only for pregnant women and for people who have weakened immune systems or who suffer from blood disorders, such as sickle cell anemia. The test involves measuring for a particular antibody or protein that the body produces in response to infection with the human parvovirus B19. The test is 92–97% specific for this disease.

Because fifth disease can pose problems for an unborn fetus exposed to the disease through the mother, testing may also be conducted while a fetus is still in the uterus. This test uses fluid collected from the sac around the fetus (amniotic fluid) instead of blood to detect the viral DNA.

## Treatment

In general, no specific treatment for fifth disease is required. The symptoms can be treated using over-the-counter medications, such as **acetaminophen** (Tylenol) or ibuprofen (Motrin, Advil). If the rash itches, calamine lotion can be applied. **Aspirin** is usually not given to children under the age of 18 to prevent the development of a serious illness called **Reye's syndrome**.

Patients who are receiving medications to suppress the immune system in the treatment of some other condition may be allowed to temporarily decrease the medications in order to allow the immune system to combat the infection and recover from the anemia. Those with weak-

## KEY TERMS

**Anemia**—A congenital or acquired deficiency in the iron-carrying capacity of the blood.

**Antibody**—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

**Immunocompromised**—A state in which the immune system is weakened or is not functioning properly due to chronic disease.

**Immunosuppressed**—A state in which the immune system is suppressed by medications during the treatment of other disorders, like cancer, or following an organ transplantation.

**Reye's syndrome**—A very serious, rare disease, most common in children, that involves an upper respiratory tract infection followed by brain and liver damage.

**Sickle cell anemia**—A hereditary blood disorder in which the red blood cells are misshapen into crescent or sickle shapes resulting in the reduced oxygen-carrying capacity of the lungs.

ened (not suppressed) immune systems, such as AIDS patients, may be given immunoglobulin intravenously to help the immune system fight the infection. People with severe anemia or who experience an aplastic crisis may require hospitalization and blood transfusions.

## Prognosis

Generally, fifth disease is mild, and patients tend to improve without any complications. In cases where the patient is either immunocompromised or immunosuppressed, a life-threatening aplastic crisis can occur. With prompt treatment, however, the prognosis is good. Mothers who develop the infection while pregnant can pass the infection on to their fetus, and as such, stand an increased risk of **miscarriage** and **stillbirth**. There are tests and treatments, however, that can be performed on the fetus while still in the uterus that can reduce the risk of anemia or other complications.

## Prevention

Currently, there is no vaccine against fifth disease. Avoiding contact with persons who exhibit symptoms of a cold and maintaining good personal hygiene by regularly washing hands may minimize the chances of an

infection. Pregnant women should avoid exposure to persons infected with the disease and notify their obstetrician immediately if they are exposed so that they can be tested and monitored closely.

## Resources

### BOOKS

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Lata Cherath, PhD

# Filariasis

### Definition

Filariasis is the name for a group of tropical diseases caused by various thread-like parasitic round worms (nematodes) and their larvae. The larvae transmit the disease to humans through a mosquito bite. Filariasis is characterized by **fever**, **chills**, **headache**, and **skin lesions** in the early stages and, if untreated, can progress to include gross enlargement of the limbs and genitalia in a condition called **elephantiasis**.

### Description

Approximately 170 million people in the tropical and subtropical areas of southeast Asia, South America, Africa, and the islands of the Pacific are affected by this debilitating parasitic disease. While filariasis is rarely fatal, it is the second leading cause of permanent and long-term disability in the world. The World Health Organization (WHO) has named filariasis one of only six "potentially eradicable" infectious diseases and has embarked upon a 20-year campaign to eradicate the disease.

In all cases, a mosquito first bites an infected individual then bites another uninfected individual, transferring some of the worm larvae to the new host. Once within the body, the larvae migrate to a particular part of the body and mature to adult worms. Filariasis is classified into three distinct types according to the part of the body that becomes infected: lymphatic filariasis affects the circulatory system that moves tissue fluid and immune cells (lymphatic system); subcutaneous filariasis infects the

areas beneath the skin and whites of the eye; and serous cavity filariasis infects body cavities but does not cause disease. Several different types of worms can be responsible for each type of filariasis, but the most common species include the following: *Wuchereria bancrofti*, *Brugia malayi* (lymphatic filariasis), *Onchocerca volvulus*, *Loa loa*, *Mansonella streptocerca*, *Dracunculus medinensis* (subcutaneous filariasis), *Mansonella pustans*, and *Mansonella ozzardi* (serous cavity filariasis).

The two most common types of the disease are Bancroftian and Malayan filariasis, both forms of lymphatic filariasis. The Bancroftian variety is found throughout Africa, southern and southeastern Asia, the Pacific islands, and the tropical and subtropical regions of South America and the Caribbean. Malayan filariasis occurs only in southern and southeastern Asia. Filariasis is occasionally found in the United States, especially among immigrants from the Caribbean and Pacific islands.

A larvae matures into an adult worm within six months to one year and can live between four and six years. Each female worm can produce millions of larvae, and these larvae only appear in the bloodstream at night, when they may be transmitted, via an insect bite, to another host. A single bite is usually not enough to acquire an infection, therefore, short-term travelers are usually safe. A series of multiple bites over a period of time is required to establish an infection. As a result, those individuals who are regularly active outdoors at night and those who spend more time in remote jungle areas are at an increased risk of contracting the filariasis infection.

### Causes and symptoms

In cases of lymphatic filariasis, the most common form of the disease, the disease is caused by the adult worms actually living in the lymphatic vessels near the lymph nodes where they distort the vessels and cause local inflammation. In advanced stages, the worms can actually obstruct the vessels, causing the surrounding tissue to become enlarged. In Bancroftian filariasis, the legs and genitals are most often involved, while the Malayan variety affects the legs below the knees. Repeated episodes of inflammation lead to blockages of the lymphatic system, especially in the genitals and legs. This causes the affected area to become grossly enlarged, with thickened, coarse skin, leading to a condition called elephantiasis.

In conjunctiva filariasis, the worms' larvae migrate to the eye and can sometimes be seen moving beneath the skin or beneath the white part of the eye (conjunctiva). If untreated, this disease can cause a type of blindness known as onchocerciasis.

Symptoms vary, depending on what type of parasitic worm has caused the infection, but all infections usually

begin with chills, headache, and fever between three months and one year after the insect bite. There may also be swelling, redness, and **pain** in the arms, legs, or scrotum. Areas of pus (abscesses) may appear as a result of dying worms or a secondary bacterial infection.

## Diagnosis

The disease is diagnosed by taking a patient history, performing a **physical examination**, and by screening blood specimens for specific proteins produced by the immune system in response to this infection (antibodies). Early diagnosis may be difficult because, in the first stages, the disease mimics other bacterial skin infections. To make an accurate diagnosis, the physician looks for a pattern of inflammation and signs of lymphatic obstruction, together with the patient's possible exposure to filariasis in an area where filariasis is common. The larvae (microfilariae) can also be found in the blood, but because mosquitos, which spread the disease, are active at night, the larvae are usually only found in the blood between about 10 pm and 2 am.

## Treatment

Either ivermectin, albendazole, or diethylcarbamazine is used to treat a filariasis infection by eliminating the larvae, impairing the adult worms' ability to reproduce, and by actually killing adult worms. Unfortunately, much of the tissue damage may not be reversible. The medication is started at low doses to prevent reactions caused by large numbers of dying parasites.

While effective, the medications can cause severe side effects in up to 70% of patients as a result either of the drug itself or the massive **death** of parasites in the blood. Diethylcarbamazine, for example, can cause severe allergic reactions and the formation of pus-filled sores (abscesses). These side effects can be controlled using **antihistamines** and anti-inflammatory drugs (**corticosteroids**). Rarely, treatment with diethylcarbamazine in someone with very high levels of parasite infection may lead to a fatal inflammation of the brain (**encephalitis**). In this case, the fever is followed by headache and confusion, then stupor and **coma** caused when massive numbers of larvae and parasites die. Other common drug reactions include **dizziness**, weakness, and nausea.

Symptoms caused by the death of the parasites include fever, headache, muscle pain, abdominal pain, **nausea and vomiting**, weakness, dizziness, lethargy, and **asthma**. Reactions usually begin within two days of starting treatment and may last between two and four days.

No treatment can reverse elephantiasis. Surgery may be used to remove surplus tissue and provide a way to drain

the fluid around the damaged lymphatic vessels. Surgery may also be used to ease massive enlargement of the scrotum. Elephantiasis of the legs can also be helped by elevating the legs and providing support with elastic bandages.

## Prognosis

The outlook is good in early or mild cases, especially if the patient can avoid being infected again. The disease is rarely fatal, and with continued WHO medical intervention, even gross elephantiasis is now becoming rare.

## Prevention

The best method of preventing filariasis is to prevent being repeatedly bitten by the mosquitoes that carry the disease. Some methods of preventing insect bites include the following:

- limit outdoor activities at night, particularly in rural or jungle areas
- wear long sleeves and pants and avoid dark-colored clothing that attracts mosquitoes
- avoid perfumes and colognes
- treat one or two sets of clothing ahead of time with permethrin (Duramon, Permanone)
- wear DEET insect repellent or, especially for children, try citronella or lemon eucalyptus, to repel insects
- if sleeping in an open area or in a room with poor screens, use a bed net to avoid being bitten while asleep
- use air conditioning, the cooler air makes insects less active

In addition, filariasis can be controlled in highly infested areas by taking ivermectin preventatively before being bitten. Currently, there is no vaccine available, but scientists are working on a preventative vaccine at this time.

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## KEY TERMS

**Abscess**—An area of inflamed and injured body tissue that fills with pus.

**Antibody**—A specific protein produced by the immune system in response to a specific foreign protein or particle called an antigen.

**Conjunctiva**—The mucous membrane that lines the inside of the eyelid and the exposed surface of the eyeball.

**Elephantiasis**—A condition characterized by the gross enlargement of limbs and/or the genitalia that is also accompanied by a hardening and stretching of the overlying skin. Often a result of an obstruction in the lymphatic system caused by infection with a filarial worm.

**Encephalitis**—Inflammation of the brain.

**Lymphatic system**—The circulatory system that drains and circulates fluid containing nutrients, waste products, and immune cells, from between cells, organs, and other tissue spaces.

**Microfilariae**—The larvae and infective form of filarial worms.

**Nematode**—Round worms.

**Subcutaneous**—The area directly beneath the skin.

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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>>.

National Institute of Allergies and Infectious Diseases, Division of Microbiology and Infectious Diseases. Building 31, Room. 7A-50, 31 Center Drive MSC 2520, Bethesda, MD 20892. <<http://www.niaid.nih.gov>>.

World Health Organization, Division of Emerging and Other Communicable Diseases Surveillance and Control. Avenue Appia 20, 1211 Geneva 27, Switzerland. (+00 41 22) 791 21 11. <<http://www.who.int>>.

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Carol A. Turkington

Filgras see **Cancer therapy, supportive; Immunologic therapies**

Fingernail removal see **Nail removal**

## Fingertip injuries

### Definition

Fingertip trauma covers cuts, accumulation of blood (hematoma), bone breakage, or **amputation** in the fingertip.

### Description

The fingertips are specialized areas of the hand with highly developed sensory and manipulative functions. Large sensory and motor areas located in the brain regulate the precise and delicate functions of fingertips. The fingertip is the site where extensor and flexor tendons insert. Fingertip injuries are extremely common since the hands hold a wide array of objects. In 2001, the approximately 10% of all accidents in the United States referred for Emergency Room consults involve the hand. Hand injuries are frequently the result of job injuries and account for 11–14% of on-the-job injuries and 6% of compensation paid injuries. Injury to the nail bed occurs in approximately 15–24% of fingertip injuries.

Fingertip injuries can result in amputation or tissue loss. The injury is assessed whether the bone and underlying tissue are intact and the size of the wound area. The pulp is the area of skin opposite the fingernail and is usually very vulnerable to injury. Pulp injuries commonly occur in persons who use or are in close contact with fast moving mechanical devices. These injuries can crush, cut, and puncture. The fingertips can also be injured by common crushing accidents. This could cause the development of a subungual hematoma (an accumulation of blood under the nail). At the base of the distal phalanx (the first circular skin fold from the tip) injuries can occur that can fracture the underlying bone in the area. Quite commonly

a hammer, closing a door, or sport accidents usually cause these injuries. These **fractures** can be simple, requiring little treatment or more complicated involving the joint. The accident may involve the point of insertion of a tendon. Usually this occurs when the terminal joint is being forced to flex while held straight. This motion typically occurs when tucking in sheets during bed making, a common cause of tendon injury. This injury causes a loss of extension (straightening the finger) ability.

### Causes and symptoms

Accidental amputations will usually result in profuse bleeding and tissue loss. Injuries to the pulp can occur as from fast moving mechanical instruments, such as drills. These injuries may puncture the pulp. Injuries such as a subungual hematoma are caused by a crushing type injury. Fractures typically occur as the result of crushing injuries or tendon avulsion. These crushing injuries are frequently caused during sport injury and can be treated by simple interventions such as **immobilization** or more complex procedures if tendons are affected (the trauma is then treated as a tendon injury). Fractures can cause **pain** and, depending on the extent of swelling, there may be some restriction of movement. Tendon injuries can be caused when the terminal joint is exposed to force flexing motion (moving the finger toward the palm) while held straight.

### Diagnosis

The attending clinician should evaluate the injury in a careful and systematic manner. The appearance of the hand can provide valuable information concerning presence of fractures, vascular status, and tendon involvement. Bones and joints should be evaluated for motion and tenderness. Nerves should be examined for sensory (feeling sensations) and motor (movement) functioning. Amputations usually profusely bleed and there is tissue loss. The wound is treated based on loss of tissue, bone, and wound area. Injuries to the pulp can be obvious during inspection. Subungual hematoma usually present a purplish-black discoloration under the nail. This is due to a hematoma underneath the nail. Radiographs may be required to assess the alignment of fractures or detect foreign bodies. Patients usually suffer from pain since injuries to the fingertip bone are usually painful and movement may be partially restricted due to swelling of the affected area. Tendon injuries usually result in the loss of ability to straighten or bend the finger.

### Treatment

Amputation with bone and underlying tissue intact and a wound area 1 cm or less should be cleaned and treated with a dressing. With these types of **wounds**

### KEY TERMS

**Distal**—Movement away from the origin.

**Flex**—To bend.

**Laceration**—A cut in the skin

**Phalanx**—A bone of the fingers or toes.

**Tendon**—A structure that connects a skeletal muscle to bone.

healthy tissue will usually grow and replace the injured area. Larger wounds may require surgical intervention. Puncture wounds should be cleaned and left open to heal. Patients typically receive **antibiotics** to prevent infection. A procedure called trephining treats subungual hematomas. This procedure is usually done with a straight cutting needle positioned over the nail. The clinician spins the needle with forefinger and thumb until a hole is made through the nail.

Patients who have extensive crush injuries or subungual hematomas involving laceration to skin folds or nail damage should have the nail removed to examine the underlying tissue (called the matrix). Patients who have a closed subungual hematoma with an intact nail and no other damage (no nail disruption or laceration) are treated conservatively. If the fracture is located two-thirds below the fingertip immobilization using a splint may be needed. Conservative treatment is recommended for crush injuries that fracture the terminal phalanx if a subungual hematoma is not present. Severe fractures near the fist circular skin crease may require surgical correction to prevent irregularity of the joint surface, which can cause difficulty with movement. Injury to a flexor tendon usually requires surgical repair. If this is not possible, the finger and wrist should be placed in a splint with specific positioning to prevent further damage.

### Prognosis

Prognosis depends on the extent of traumatic damage to the affected area. Nail lacerations that are not treated may cause nail deformities. When amputation is accompanied with loss of two-thirds of the nail, half of the fingers develop beaking, or a curved nail. Aftercare and follow up are important components of treatment. The patient is advised to keep the hand elevated, check with a clinician two days after treatment, and to splint fractures for two weeks in the extended position. Usually a nail takes about 100 days to fully grow. Healing for an amputation takes about 21 to 27 days. This markedly

decreases in elderly patients, primarily due to a compromised circulation normally part of advancing age.

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### ORGANIZATION

- American Society of Plastic Surgeons. 444 E. Algonquin Rd., Arlington Heights, IL. (888) 475-2784.

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watery **diarrhea**. The most characteristic symptoms of the illness are those involving the nervous system. These include **numbness and tingling** around the lips, tongue, and mouth; **itching**; **dry mouth**; metallic taste in the mouth; and blurry vision. In more prominent cases, patients may complain of temporary blindness, a slow pulse, and a feeling that their teeth are loose. Patients may also have the strange symptom of reversal of hot and cold sensations on the skin, where cold things feel very hot or painful to the touch. In very severe cases, there may be difficulties in breathing or low blood pressure.

## Diagnosis

Ciguatera diagnosis is based on the typical combination of symptoms after eating fish. There are no readily available blood or urine tests to detect the poisoning, but some researchers have developed a test for the toxin left on any remaining fish. A person does not have to be in a tropical area to get ciguatera. Fish can be caught from one of these distant areas, and can then be shipped and eaten locally. It is important to report suspected cases to local public health officials because more cases may occur from other contaminated fish.

## Treatment

The treatment for this illness is general. Patients are given fluids (by mouth or through a vein) and medications to decrease the itching or to treat vomiting and/or diarrhea. The neurological symptoms can cause discomfort and treatment with amitriptyline (a medicine that has been used for depression) may be useful. Other medications may also be given.

## Prognosis

Although **death** can occur, almost all patients diagnosed with ciguatera will recover. Recovery, however, can be slow and some symptoms can last for weeks or even months. Symptoms can also be aggravated by other illnesses or alcohol.

## Prevention

Knowing the kinds of fish linked to ciguatera can help a person avoid eating high-risk fish. However, over 400 different kinds of fish have been linked to the disease, even salmon. A particular fish in a given area may be more likely to cause ciguatera than other fish. For example, red snapper is most often the source of ciguatera in the Pacific, while barracuda is more likely to contain the toxin in Florida. This is why it is illegal to sell barracuda in Florida for human consumption. Cooking the fish does not prevent ciguatera.

# Fish and shellfish poisoning

## Definition

Fish and shellfish **poisoning** is a common but often unrecognized group of illnesses related to food. Three of these illnesses include ciguatera, scombroid, and paralytic shellfish poisoning.

## Ciguatera

### Definition

Ciguatera (from the Spanish word for a poisonous snail) is a food-related illness that causes abdominal and neurological symptoms.

### Causes and symptoms

Ciguatera is caused by eating fish that have a toxin called ciguatoxin. Scientists believe this toxin is acquired by the fish through the food chain, and is originally produced by small algae microorganisms (dinoflagellates). The fish most likely contaminated with ciguatoxin are those that feed close to tropical reefs, including red snapper, grouper, and barracuda. Larger fish are more likely to contain the toxin. Although not as common in the United States, ciguatera is commonly diagnosed on many of the islands in the Pacific Ocean.

Illness from ciguatera can occur in just a few minutes to about 30 hours after eating. Most cases occur one to six hours after eating the contaminated fish. Initial symptoms are abdominal cramps, nausea, vomiting, or

## Scombroid

### *Definition*

Scombroid is a fish-associated illness caused by eating improperly handled fish. Fish linked to this disease are usually in the Scombridae family, which includes yellowfin tuna, skipjack, bonito, and mackerel.

### *Causes and symptoms*

Scombroid occurs after eating fish that has not been properly refrigerated after capture. Unlike ciguatera, the toxins linked with scombroid are not contracted by the fish from its surroundings. Bacteria that are normally found in fish act directly on a chemical (called histidine) in the flesh of fish that are not properly cooled when stored. This interaction produces histamine and other chemicals that cause the illness when the fish is eaten.

Symptoms of scombroid occur quickly after eating the fish, as soon as 10 minutes. Since histamine is released by certain cells in the body during an allergic reaction, scombroid can be confused with a fish allergy. Scombroid causes flushing of the face, sweating, a burning feeling in the mouth or throat, vomiting, diarrhea, and headaches. A rash that looks like a **sunburn** may occur, and a small number of patients have **hives**. Some patients have a metallic or peppery taste in their mouths. In more severe cases, rapid pulse, blurred vision, and difficulty breathing can occur. Symptoms usually last about four hours.

### *Diagnosis*

Like ciguatera, scombroid poisoning is diagnosed based on typical symptoms occurring after eating fish. There are usually no available tests for the patient. Experimentally, however, elevated levels of histamine-related products have been found in the urine. It may be possible for public health officials to test any remaining fish flesh for histamine levels. Improperly refrigerated fish caught in both temperate and tropical waters have been linked to the illness. An outbreak of similar cases may be helpful in correctly diagnosing the problem.

### *Treatment*

The treatment for scombroid is usually general. **Antihistamines** like diphenhydramine (Benadryl) may shorten the duration of the illness, but the illness will go away on its own. Some doctors have found that cimetidine (Tagamet) given through a vein may be helpful as well. In rare, more severe cases, epinephrine (adrenaline) may be used.

## KEY TERMS

**Algae**—Plants that have one cell.

**Histamine**—A chemical found naturally in the body that produces inflammation and increases blood flow; the uncomfortable symptoms of an allergy attack or an allergic reaction are generally caused by the release of histamine.

**Toxin**—A poisonous substance usually produced by a living thing.

### *Prognosis*

Although sometimes dramatic and alarming symptoms can occur, scombroid is usually not serious. The patient should be reassured that scombroid is not a fish allergy.

### *Prevention*

Adequate storage of the target fish will always prevent scombroid. Since the fish does not appear spoiled or smell bad, the consumer cannot detect the risk of the illness before eating the fish. Cooking the fish does not prevent scombroid. Suspected cases should be reported to public health officials.

## Paralytic shellfish poisoning

### *Definition*

Paralytic shellfish poisoning (PSP) is a nervous system disease caused by eating cooked or raw shellfish that contain environmental toxins. These toxins are produced by a group of algae (dinoflagellates). It is unclear whether these toxins are related to the “blooming” of the algae, also called red tide because the algae can turn the water reddish brown. PSP occurs mostly in May through November.

### *Causes and symptoms*

PSP develops usually within minutes after eating a contaminated shellfish, most commonly a mussel, clam, or oyster. Symptoms include **headache**, a floating feeling, **dizziness**, lack of coordination, and tingling of the mouth, arms, or legs. Muscle weakness causing difficulty swallowing or speaking may occur. Abdominal symptoms such as nausea, vomiting, and diarrhea can also occur. Unlike ciguatera and scombroid, PSP may have a much more serious outcome. PSP may cause difficulty breathing related to weakness or **paralysis** of the breathing muscle. The symptoms may last for six to 12 hours, but a patient may continue to feel weak for a week or more.

### Diagnosis

PSP diagnosis is based on symptoms after eating shellfish, even if the shellfish are adequately cooked. No blood or urine test is available to diagnose the illness, but tests in mice to detect the toxin from the eaten fish can be done by public health officials.

### Treatment

The treatment of PSP is mostly supportive. If early symptoms are recognized, the doctor will try to flush the toxin from the gastrointestinal tract with medications that create diarrhea. Vomiting may be induced if the patient has no signs of weakness. In cases where the muscles of breathing are weakened, the patient may be placed on a respirator until the weakness goes away. However, this measure is not usually needed. Likewise, the use of a machine to clean the blood (dialysis) has been used in severe cases.

### Prognosis

The prognosis for PSP is quite good, especially if the patient has passed the initial 12 hours of illness without needing breathing support. Most deaths occur during this period if breathing help is not available.

### Prevention

Measures to control PSP require detecting rising numbers of algae in coastal waters by periodic microscopic examination. By law, shellfish beds are closed when levels of the toxin-producing organisms are above acceptable standards. Cooking the shellfish does not prevent this disease. Suspected cases should be reported to public health officials.

### Resources

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5p- syndrome see **Cri du chat syndrome**

## Flesh-eating disease

### Definition

Flesh-eating disease is more properly called necrotizing fasciitis, a rare condition in which bacteria destroy tissues underlying the skin. This tissue **death**, called necrosis or **gangrene**, spreads rapidly. This disease can be fatal in as little as 12 to 24 hours.

### Description

Although the term is technically incorrect, flesh-eating disease is an apt descriptor: the infection appears to devour body tissue. Media reports increased in the middle and late 1990s, but the disease is not new. Hippocrates described it more than three millennia ago and thousands of reports exist from the Civil War. Approximately 500 to 1,500 cases of necrotizing fasciitis occur in the United States each year.

Flesh-eating disease is divided into two types. Type I is caused by anaerobic bacteria, with or without the presence of aerobic bacteria. Type II, also called hemolytic streptococcal gangrene, is caused by group A streptococci; other bacteria may or may not be present. The disease may also be called synergistic gangrene.

Type I fasciitis typically affects the trunk, abdomen, and genital area. For example, Fournier's gangrene is a "flesh-eating" disease in which the infection encompasses the external genitalia. The arms and legs are most often affected in type II fasciitis, but the infection may appear anywhere.

### Causes and symptoms

The two most important factors in determining whether or not a person will develop flesh-eating disease are: the virulence (ability to cause disease) of the bacteria and the susceptibility (ability of a person's immune system to respond to infection) of the person who becomes infected with this bacteria.

In nearly every case of flesh-eating disease, a skin injury precedes the disease. As bacteria grow beneath the skin's surface, they produce toxins. These toxins destroy superficial fascia, subcutaneous fat, and deep fascia. In some cases, the overlying dermis and the underlying muscle are also affected.

Initially, the infected area appears red and swollen and feels hot. The area is extremely painful, which is a prominent feature of the disease. Over the course of hours or days, the skin may become blue-gray, and fluid-filled blisters may form. As nerves are destroyed the area

becomes numb. An individual may go into **shock** and develop dangerously low blood pressure. Multiple organ failure may occur, quickly followed by death.

## Diagnosis

The appearance of the skin, paired with **pain** and **fever** raises the possibility of flesh-eating disease. An x ray, **magnetic resonance imaging** (MRI), or **computed tomography scans** (CT scans) of the area reveals a feathery pattern in the tissue, caused by accumulating gas in the dying tissue. Necrosis is evident during exploratory surgery, during which samples are collected for bacterial identification.

## Treatment

Rapid, aggressive medical treatment, specifically, antibiotic therapy and surgical **debridement**, is imperative. **Antibiotics** may include penicillin, an aminoglycoside or third-generation cephalosporin, and clindamycin or metronidazole. **Analgesics** are employed for pain control. During surgical debridement, dead tissue is stripped away. After surgery, patients are rigorously monitored for continued infection, shock, or other complications. If available, hyperbaric oxygen therapy has also been used.

## Prognosis

Flesh-eating disease has a fatality rate of about 30%. Diabetes, arteriosclerosis, immunosuppression, kidney disease, **malnutrition**, and **obesity** are connected with a poor prognosis. Older individuals and intravenous drug users may also be at higher risk. The infection site also has a role. Survivors may require plastic surgery and may have to contend with permanent physical disability and psychological adjustment.

## Prevention

Flesh-eating disease, which occurs very rarely, cannot be definitively prevented. The best ways to lower the risk of contracting flesh-eating disease are:

- take care to avoid any injury to the skin that may give the bacteria a place of entry
- when skin injuries do occur, they should be promptly washed and treated with an antibiotic ointment or spray
- people who have any skin injury should rigorously attempt to avoid people who are infected with streptococci bacteria. A bacteria that causes a simple **strep throat** in one person may cause flesh-eating disease in another
- have any areas of unexplained redness, pain, or swelling examined by a doctor, particularly if the affected area seems to be expanding

## KEY TERMS

**Aerobic bacteria**—Bacteria that require oxygen to live and grow.

**Anaerobic bacteria**—Bacteria that require the absence of oxygen to live and grow.

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

**Debridement**—Surgical procedure in which dead or dying tissue is removed.

**Dermis**—The deepest layer of skin.

**Fascia, deep**—A fibrous layer of tissue that envelopes muscles.

**Fascia, superficial**—A fibrous layer of tissue that lies between the deepest layer of skin and the subcutaneous fat.

**Gangrene**—An extensive area of dead tissue.

**Hyperbaric oxygen therapy**—A treatment in which the patient is placed in a chamber and breathes oxygen at higher-than-atmospheric pressure. This high-pressure oxygen stops bacteria from growing and, at high enough pressure, kills them.

**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.

**Necrosis**—Abnormal death of cells, potentially caused by disease or infection.

**Subcutaneous**—Referring to the area beneath the skin.

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#### ORGANIZATIONS

National Necrotizing Fascitis Foundation. PO Box 145, Niantic, CT 06357. (616) 261-2538. <<http://www.nnff.org/>>.

Paul A. Johnson

Flight medicine see **Aviation medicine**

Floppy mitral valve see **Mitral valve prolapse**

devote his life to this research. It is known that at this point, he ceased to dispense the mixture of homeopathy and allopathic medicine that he had been using. Instead, he began investigating the healing properties of plant essences and discovered that he possessed an "intuition" for judging the properties of each flower. Accordingly, he developed the system of treatment that bears his name, and is also the foundation for all other flower-remedy systems.

The Bach Flower Remedies were ostensibly the only system of significance from the 1920s until in the 1970s, when there was a renewed interest in the subject by doctors working in the field of natural medicine. Perhaps the most notable was Dr. Richard Katz, who was seeking new methods of dealing with modern **stress** and the resulting ailments. He focused on the concept of a psychic, psychological effect and chose to pursue this line of research.

In 1979, Katz founded the Flower Essence Society in California, (FES). This society pledged to further the research and development of Bach's principles. As of 2000, FES hosts a database of over 100 flower essences from more than 50 countries. FES is now an international organization of health practitioners, researchers, students, and others concerned with flower essence therapy.

The Society has connections with an estimated 50,000 active practitioners from around the world, who use flower essence therapy as part of their treatment. FES encourages the study of the plants themselves to determine the characteristics of flower essences. They are compiling an extensive database of case studies and practitioner reports of the use of essences therapeutically, allowing verification and development of the original definitions. They are also engaged in the scientific study of flower essence therapy.

FES says they have developed the theories of Paracelsus and Goethe who researched the "signatures" and "gestures" of botanical specimens, on the premise that the human body and soul are a reflection of the system of nature. FES plant research interprets the therapeutic properties of flower essences according to these insights.

In this regard, they have devised 12 "windows of perception" for monitoring the attributes of plants. Each of these windows reveals an aspect of the plant's qualities, although they maintain that what they are seeking is a "whole which is greater than the sum of its parts." The 12 windows are not considered independent classifications, but more of a blended tapestry of views of the qualities that each plant possesses.

The first window is concerned with the "form" of a plant—its shape classification. The second focuses on its "gesture" or spatial relationship. The third window is a plant's botanical classification; the Flower Essence Society maintains that considering a plant's botanical family

## Flower remedies

### Definition

Flower remedies are specially prepared flower essences, containing the healing energy of plants. They are prescribed according to a patient's emotional disposition, as ascertained by the therapist, doctor, or patients themselves.

### Purpose

Flower remedies are more homeopathic than herbal in the way they work, effecting energy levels rather than chemical balances. They have been described as "liquid energy." The theory is that they encapsulate the flowers' healing energy, and are said to deal with and overcome negative emotions, and so relieve blockages in the flow of human energy that can cause illness.

### Description

#### Origins

Perhaps the most famous and widely used system is the Bach flower remedies. This system originated in the 1920s when British physician and bacteriologist, Dr. Edward Bach (1886–1936), noticed that patients with physical complaints often seemed to be suffering from **anxiety** or some kind of negative emotion. He concluded that assessing a patient's emotional disposition and prescribing an appropriate flower essence could treat the physical illness. Bach was a qualified medical doctor, but he also practiced **homeopathy**.

As a result of his own serious illness in 1917, Bach began a search for a new and simple system of medicine that would treat the whole person. In 1930, he gave up his flourishing practice on Harley Street at the Royal London Homeopathic Hospital and moved to the countryside to

is essential to obtaining an overview of its properties as a flower essence. The fourth window concerns the time orientation of a particular specimen regarding the daily and seasonal cycles. Why do some flowers bloom at different times of the day, while others, such as the evening primrose, respond to the moon? The fifth window observes a plant's relationship to its environment. Where a plant chooses to grow, and where it cannot survive, reveals much about its qualities. The sixth window observes a plant's relationship to the Four Elements and the Four Ethers, as FES maintains that plants exist in one of the elemental or etheric forces in addition to their physical life. "Elements" refers to those developed by the Greeks, as opposed to the modern concept of "molecular building blocks." It seems that commonly, two elements predominate in a plant, indicating a polarity of qualities, while two can be said to be recessive. The seventh window relates to a plant's relationship with the other kingdoms of nature: mineral, animal and human, while the eighth relates to the color and color variations of a plant. Katz explains how the language of color tells us so much about the "soul qualities" of a plant. The ninth window concerns all other sensory perceptions of a plant, such as fragrance, texture, and taste. The tenth window involves assessing the chemical substances and properties; the eleventh studies medicinal and herbal uses, as by studying the physical healing properties of plants, we can also understand something of their more subtle effects on the soul. Finally, the twelfth window involves the study of the lore, mythology, folk wisdom, and spiritual and ritual qualities associated with a particular plant. Katz relates how in the past, human beings were more in touch with the natural world, and the remnants of this unconscious plant wisdom live on in the form of folklore, mythology, and so on.

Because flower remedies operate on approximately the same principles as homeopathy, practitioners quite often prescribe the two therapies in conjunction with each other. They can also be used concurrently with allopathic medicine.

The system consists of 38 remedies, each for a different disposition. The basic theory is that if the remedy for the correct disposition is chosen, the physical illness resulting from the present emotional state can then be cured. There is a rescue remedy made up of five of the essences—cherry plum, clematis, impatiens, rock star, and star of Bethlehem—that is recommended for the treatment of any kind of physical or emotional shock. Therapists recommended that rescue remedy be kept on hand to help with all emergencies.

#### The 38 Bach Remedies

- agrimony: puts on a cheerful front, hides true feelings, and worries or problems

## EDWARD BACH (1886–1936)

Edward Bach was a graduate of University College Hospital (M.B., B.S., M.R.C.S.) in England. He left his flourishing Harley Street practice in favor of homeopathy, seeking a more natural system of healing than allopathic medicine. He concluded that healing should be as simple and natural as the development of plants, which were nourished and given healing properties by earth, air, water, and sun.

Bach believed that he could sense the individual healing properties of flowers by placing his hands over the petals. His remedies were prepared by floating summer flowers in a bowl of clear stream water exposed to sunlight for three hours.

He developed 38 remedies, one for each of the negative states of mind suffered by human beings, which he classified under seven group headings: fear, uncertainty, insufficient interest in present circumstances, loneliness, over-sensitivity to influences and ideas, despondency or despair, and overcare for the welfare of others. The Bach remedies can be prescribed for plants, animals, and other living creatures as well as human beings.

- aspen: feelings of apprehension, dark foreboding, and premonitions
- beech: critical, intolerant, picky
- centaury: easily comes under the influence of others, weak willed
- cerato: unsure, no confidence in own judgement, intuition, and seeks approval from others
- cherry plum: phobic, fear of being out of control, and tension
- chestnut bud: repeats mistakes, does not learn from experience
- chicory: self-centered, possessive, clingy, demanding, self pity
- clematis: absent minded, dreamy, apathetic, and lack of connection with reality
- crab apple: a "cleanser" for prudishness, self-disgust, feeling unclean
- elm: a sense of being temporarily overwhelmed in people who are usually capable and in control
- gentian: discouraged, doubting, despondent
- gorse: feelings of pessimism, accepting defeat
- heather: need for company, talks about self, and concentrates on own problems

- holly: jealousy, envy, suspicion, anger, and hatred
- honeysuckle: reluctance to enter the present and let the past go
- hornbeam: reluctant to face a new day, weary, can't cope (mental fatigue)
- impatiens: impatience, always in a hurry, and resentful of constraints
- larch: feelings of inadequacy and apprehension, lack of confidence and will to succeed
- mimulus: fearful of specific things, shy, and timid
- mustard: beset by "dark cloud" and gloom for no apparent reason
- oak: courageous, persevering, naturally strong but temporarily overcome by difficulties
- olive: for physical and mental renewal, to overcome exhaustion from problems of long-standing
- pine: for self-reproach, always apologizing, assuming guilt
- red chestnut: constant worry and concern for others
- rock rose: panic, intense alarm, dread, horror
- rock water: rigid-minded, self-denial, restriction
- scleranthus: indecision, uncertainty, fluctuating moods
- star of Bethlehem: consoling, following shock or grief or serious news
- sweet chestnut: desolation, despair, bleak outlook
- vervain: insistent, fanatical, over-enthusiastic
- vine: dominating, overbearing, autocratic, tyrannical
- walnut: protects during a period of adjustment or vulnerability
- water violet: proud, aloof, reserved, enjoys being alone
- white chestnut: preoccupation with worry, unwanted thoughts
- wild oat: drifting, lack of direction in life
- wild rose: apathy, resignation, no point in life
- willow bitter: resentful, dissatisfied, feeling life is unfair

Originally, Bach collected the dew from chosen flowers by hand to provide his patients with the required remedy. This became impractical when his treatment became so popular that production could not keep up with demand. He then set about finding a way to manufacture the remedies, and found that floating the freshly picked petals on the surface of spring water in a glass bowl and leaving them in strong sunlight for three hours produced the desired effect. Therapists explain that the water is "potentized" by the essence of the flowers. The potentized water can then be bottled and sold. For more woody specimens, the procedure is to boil them in a sterilized pan of water for 30 minutes. These two methods

### Bach Flower Remedies

Name	Remedy
Agrimony	Upset by arguments, nonconfrontational, conceals worry and pain
Aspen	Fear of the unknown, anxiety, prone to nightmares, and apprehension
Beech	Critical, intolerant, and negative
Centaury	Submissive and weak-willed
Erato	Self doubting and overly dependent
Cherry Plum	Emotional thoughts and desperation
Chestnut	Repeats mistakes and has no hindsight
Chicory	Selfish, controlling, attention-seeking, and possessive
Clematis	Absorbed, impractical, and indifferent
Crab Apple	Shame and self-loathing
Elm	Overwhelmed and feelings of inadequacy
Gentian	Negative, doubt, and depression
Gorse	Pessimism, hopelessness, and despair
Heather	Self-centered and self-absorbed
Holly	Jealousy, hatred, suspicion, and envy
Honeysuckle	Homesick, living in the past, and nostalgic
Hornbeam	Procrastination, fatigue, and mental exhaustion
Impatiens	Impatience, irritability, and impulsive
Larch	No confidence, inferiority complex, and dependency
Mimulus	Timid, shy, and fear of the unknown
Mustard	Sadness and depression of unknown origin
Oak	Obstinate, inflexible, and overachieving
Olive	Exhaustion
Pine	Guilt and self blame
Red Chestnut	Fear and anxiety for loved ones
Rock Rose	Nightmares, hysteria, terror, and panic
Rock Water	Obsessive, repression, perfectionism, and self denial
Scleranthus	Indecision, low mental clarity, and confusion
Star-of-Bethlehem	Grief and distress
Sweet Chestnut	Despair and hopelessness
Vervain	Overbearing and fanatical
Vine	Arrogant, ruthless, and inflexible
Walnut	Difficulty accepting change
Water Violet	Pride and aloofness
White Chestnut	Worry, preoccupation, and unwanted thoughts
Wild Oat	Dissatisfaction
Wild Rose	Apathy and resignation
Willow	Self pity and bitterness

produce "mother tinctures" and the same two methods devised by Bach are still used today. Flower essences do not contain any artificial chemical substances, except for alcohol preservative.

Bach remedies cost around \$10 each, and there is no set time limit for treatment. It may take days, weeks, or in some cases months. Flower essences cost around \$6 each, and there is also no set time for the length of treatment, or the amount of essences that may be taken. These treatments are not generally covered by medical insurance.

### Precautions

Bach remedies and flower essences are not difficult to understand, and are considered suitable for self administration. The only difficulty may be in finding the correct remedy, as it can sometimes be tricky to pinpoint an

individual's emotional disposition. They are even safe for babies, children, and animals. An important aspect of treatment with flower remedies, is that if you feel instinctively that you need a particular remedy, you are encouraged to act on that instinct. However, it is advisable not to continue a particular remedy once you feel you no longer need it, and to try a different one if you feel that progress is not being made.

The remedies are administered from a stoppered bottle and need to be diluted. Individuals sensitive to alcohol can apply the concentrate directly to temples, wrists, behind the ears, or underarms. They should be kept in a cool dark place; like this they should last indefinitely. However, a diluted remedy should not be kept longer than three weeks. Two drops of each diluted remedy should be taken four times a day, including first thing in the morning and last thing at night. If the rescue remedy is being used, four drops should be used instead. Most therapists recommend that they be taken in spring water, but the remedy can be taken directly from the bottle, if care is taken that the dropper does not touch the tongue, as this would introduce bacteria that would spoil the remedy.

It is not recommended that more than six or seven Bach remedies be used at any one time. Instead, it is preferable to divide a larger amount up into two lots to ensure the optimum effectiveness of the remedies. No combination, or amount of combinations of the remedies can cause any harm, rather they become less effective.

Unlike FES, the Bach Centre does not encourage research to "prove" that the remedies work, preferring that people find out for themselves. They strive to keep the use of the Bach remedies as simple as possible, and to this end they do not keep case records. Bach warned before he died that others would try to change his work and make it more complicated. He was determined to keep it simple so that anyone could use it, and that is why he limited the system to only 38 remedies. The Centre points out that many who have used Bach's research as a starting point have added other remedies to the list, even some that Bach himself rejected.

### Side effects

Flower remedies or essences are generally regarded as being totally safe, and there are no known side effects apart from the rare appearance of a slight rash, which is not a reason to discontinue treatment, says the Bach Centre.

### Research and general acceptance

Bach flower remedies and flower essences have not yet officially won the support of allopathic medicine, despite the fact that more and more medical doctors are referring

## KEY TERMS

**Aura**—Emanation of light from living things (plants and animals) that can be recorded by Kirlian photography.

**Essence**—The constituent of a plant that determines its characteristics.

**Potentize**—The process of transferring the healing energy of a plant into spring water.

**Window**—A perspective adopted to assess the property of a given plant.

patients for such treatments on the strength of personal conviction. However, it is difficult to discount the scores of testimonials. Some practitioners refer skeptics to the research that has been done regarding the "auras" of living things. Theoretically, the stronger the aura, the more alive an organism is. Flower essences have very strong auras.

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### ORGANIZATIONS

The Dr. Edward Bach Centre, Mount Vernon, Bakers Lane, Sotwell, Oxon, OX10 OPX, UK. <[centre@bachcentre.com](mailto:centre@bachcentre.com)>. <<http://www.bachcentre.com>>.

The Flower Essence Society. P.O. Box 459, Nevada City, CA 95959. (800) 736-9222. Fax: (530) 265-0584. <[mail@flowersociety.org](mailto:mail@flowersociety.org)>. <<http://www.flowersociety.org>>.

Patricia Skinner

Flu see **Influenza**

Fluconazole see **Antifungal drugs, systemic**

## Fluke infections

### Definition

Fluke infections are diseases of the digestive tract and other organ systems caused by several different



**A micrograph of adult intestinal blood flukes, *Schistosoma mansoni*.** Humans can become infected while bathing or working in contaminated water. (Photo Researchers, Inc. Reproduced by permission.)

species of parasitic flatworms (Trematodes) that have complex life cycles involving hosts other than human beings. Trematode comes from a Greek word that means having holes and refers to the external suckers that adult flukes use to draw nourishment from their hosts. Fluke infections are contracted by eating uncooked fish, plants, or animals from fluke-infected waters. Symptoms vary according to the type of fluke infection.

## Description

In humans, fluke infections can be classified according to those diseases caused by liver flukes and those caused by lung flukes. Diseases caused by liver flukes include fascioliasis, opisthorchiasis, and clonorchiasis. Cases of liver fluke infection have been reported in Europe and the United States, as well as the Middle East, China, Japan, and Africa. Diseases caused by lung flukes include paragonimiasis. Paragonimiasis is a common infection in the Far East, Southeast Asia, Africa, Central and South America, Indonesia, and the Pacific Islands. It is estimated that between 40 million and 100 million people worldwide suffer from either liver or lung fluke infections.

In their adult stage, liver and lung flukes are symmetrical in shape, ranging between 1/4–1 in in length, and look somewhat like long, plump leaves or blades of grass. They enter through the mouth and can infect any person at any age.

## Causes and symptoms

The symptoms of fluke infection differ somewhat according to the type of fluke involved. All forms of liver and lung fluke infection, however, have the following characteristics:

- most persons who get infected do not develop symptoms (asymptomatic)
- the early symptoms of an acute fluke infection are not unique to these diseases alone (nonspecific symptoms)
- infection does not confer immunity against re-infection by the same species or infection by other species of flukes
- infection is usually associated with eating uncooked fish, plants, or animals that live in fresh water

### Fascioliasis

Fascioliasis is caused by *Fasciola hepatica*, the sheep liver fluke. The fluke has a three-part life cycle that begins when eggs from a host's feces are deposited in water. The eggs release free-swimming larvae (miracidia) that infect snails. The snails then release free-swimming larvae with tails (cercariae) that form cysts containing larvae in the infective stage (metacercariae) on vegetation growing in fresh water. Humans become infected when they eat watercress, water chestnuts, or other plants covered with the encysted metacercariae.

When a person eats contaminated plants, the cysts are broken open in the digestive system, and the metacercariae leave their cysts, pass through the wall of the intestine, and enter the liver, where they cause inflammation and destroy tissue. After a period of 10–15 weeks in the liver, the adult flukes move to the bile ducts and produce eggs. Acute fascioliasis is marked by abdominal pain with headache, loss of appetite, anemia, and vomiting. Some patients develop hives, muscle pains, or a yellow-color to the skin and whites of the eyes (jaundice). Chronic forms of the disease may produce complications, including blockage of the bile ducts or the migration of adult flukes to other parts of the body.

### Opisthorchiasis and clonorchiasis

These infections are caused by *Clonorchis sinensis*, the Chinese liver fluke, and *Opisthorchis viverrini* or *O. felineus*. The diseases are widespread, affecting more than 20 million people in Japan, China, Southeast Asia, and India. The life cycle of these liver flukes is similar to that of *F. hepatica* except that the cercariae are encysted in freshwater fish rather than on plants. Dogs, cats, and other mammals that eat raw fish can be infected with opisthorchiasis and clonorchiasis.

## KEY TERMS

**Aspirator**—A medical instrument that uses suction to withdraw fluids from the lungs, digestive tract, or other parts of the body for laboratory testing.

**Asymptomatic**—Persons who carry a disease and are usually capable of transmitting the disease but, who do not exhibit symptoms of the disease are said to be asymptomatic.

**Cercaria (plural, cercariae)**—An intermediate-stage of the fluke larva, released into water by infected snails.

**Cross-reaction**—A reaction that occurs in blood testing when a disease agent reacts to the specific antibody for another disease agent. Cross-reactions are common in blood tests for fluke infections because the different species are closely related.

**Encysted**—Enclosed in a cyst or capsule. Flukes spend part of their life cycle as encysted larvae.

**Fluke**—A parasitic flatworm that has external suckers. Flukes are sometimes called trematodes.

**Host**—The living animal that supplies nutrition to a parasite.

**Jaundice**—Yellowing of the skin and the whites of the eyes as a result of excess bile in the blood due to an improperly functioning liver.

**Metacercaria (plural, metacercariae)**—The encysted stage of a fluke larva that produces infection in human beings.

**Miracidium (plural, miracidia)**—The free-swimming larval form in the life cycle of the liver fluke.

**Parasite**—An organism that lives on or inside an animal of a different species and feeds on it or draws nutrients from it.

**Trematode**—Parasitic flatworms or another name for fluke, taken from a Greek word that means having holes.

The symptoms of opisthorchiasis and clonorchiasis are similar to those of fascioliasis and include both acute and chronic forms. In acute infection, the patient may be tired, have a low-grade fever, pains in the joints, a swollen liver, abdominal pain, and a skin rash. The acute syndrome may be difficult to diagnose because the fluke eggs do not appear in the patient's stool for three to four weeks after infection. Patients with the chronic form of the disease experience a loss of appetite, fatigue, low-grade fever, diarrhea, and an enlarged liver that feels sore when the abdomen is pressed.

### Paragonimiasis

Paragonimiasis is caused by a lung fluke, either *Paragonimus westermani* or *P. skrjabini*. These flukes are larger than liver flukes and infect meat- or fish-eating animals as well as humans. Their life cycle is similar to that of liver flukes except that their encysted larvae infect crabs and crayfish rather than plants or fish. Humans can ingest the encysted metacercariae from drinking contaminated water or eating raw or undercooked crabs and crayfish.

In humans, the metacercariae are released from their cysts in the small intestine and migrate to the lungs or the brain in 1% of cases. In the lungs, the flukes lay their eggs and form areas of inflammation covered with a thin layer of fibrous tissue. These areas of infection may eventually rupture, causing the patient to cough up fluke eggs, blood, and inflamed tissue. The period between the

beginning of the infection and the appearance of the eggs during coughing is about six weeks. Patients with lung infections may have chest pain and fever as well as rust-colored or bloody sputum. Lung infections can lead to lung abscess, pneumonia, or bronchitis. Patients with fluke infections of the brain may experience seizures or a fatal inflammation of brain tissue called encephalitis. Some patients also develop diarrhea and abdominal pain or lumps under the skin that contain adult flukes.

### Diagnosis

Diagnosis of fluke infections is based on a combination of the patient's history, particularly travel or residence in areas known to have flukes, and identification of the fluke's eggs or adult forms. In some patients, the eggs are found in fluid from the lungs, bile duct, or small intestine. Samples of these fluids can be obtained with a suction instrument (aspirator). Because most types of fluke infections are rare in the United States, stool specimens or body fluid samples may need to be sent to a laboratory with experts in unusual diseases or conditions to identify the specific parasite. In some cases, adult flukes may be found in the patient's stools, vomit, sputum, or skin lumps (for lung flukes). In the case of lung flukes, it is important for the doctor to rule out tuberculosis as a possible diagnosis. A tuberculosis skin test and chest x-ray will usually be sufficient to do this.

Blood tests may be useful in diagnosing fluke infections, but their usefulness is limited because of cross-reactions. A cross-reaction occurs in blood testing when a particular disease agent reacts with antibodies specific to another disease agent. This result means that the doctor may know that the person is infected by flukes but cannot tell from the blood test alone which specific type of fluke is causing the disease. In addition, blood tests for fluke infections cannot distinguish between past and current infections. In some cases, sophisticated imaging techniques, such as **computed tomography scans** (CT scans) or ultrasound scans of the patient's chest or brain (for lung flukes) or abdomen (for liver flukes), are useful in confirming a diagnosis of fluke infection.

### Treatment

Liver and lung fluke infections are treated with medications. These include triclabendazole, praziquantel, bithionol, albendazole, and mebendazole. Praziquantel works by paralyzing the flukes' suckers, forcing them to drop away from the walls of the host's blood vessels. In the United States, bithionol is available only from the Centers for Disease Control (CDC). Depending on the species of fluke and the severity of infection, the course of treatment can vary from several days to several weeks. Cure rates vary from 50–95%. Most patients experience mild temporary side effects from these drugs, including diarrhea, **dizziness**, or headache.

### Prognosis

The prognosis for recovery from liver fluke infections is good, although patients with serious infections may be more vulnerable to other diseases, particularly if significant liver damage has occurred. Most patients with lung fluke infections also recover, however, severe infections of the brain can cause **death** from the destruction of central nervous system or brain tissue.

### Prevention

No vaccines have been developed that are effective against lung or liver fluke infections. Prevention of these infections includes the following measures:

- boiling or purifying drinking water
- avoiding raw or undercooked fish or salads made from fresh aquatic plants; all food eaten in areas with fluke infestations should be cooked thoroughly; pickling or **smoking** will not kill fluke cysts in fish or shellfish
- control or eradication of the snails that serve as the flukes' intermediate hosts

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Rebecca J. Frey

## Fluoroquinolones

### Definition

Fluoroquinolones are medicines that kill bacteria or prevent their growth.

### Purpose

Fluoroquinolones are **antibiotics**, medicines used to treat infections caused by microorganisms. Physicians prescribe these drugs for bacterial infections in many parts of the body. For example, they are used to treat bone and joint infections, skin infections, urinary tract infections, inflammation of the prostate, serious ear infections, **bronchitis**, **pneumonia**, **tuberculosis**, some **sexually transmitted diseases** (STD), and some infections that affect people with **AIDS**.

### Description

Fluoroquinolones are available only with a physician's prescription and are sold in tablet and injectable forms. Examples of these medicines are moxifloxacin (Avelox), ciprofloxacin (Cipro), ofloxacin (Floxin), levofloxacin (Levaquin), lomefloxacin (Maxaquin), nor-

floxacin (Noroxin), enoxacin (Penetrex), gatifloxacin (Tequin), and sparfloxacin (Zagam).

### Recommended dosage

The recommended dosage depends on the type and strength of fluoroquinolone, and the kind of infection for which it is being taken. Check with the physician who prescribed the drug or the pharmacist who filled the prescription for the correct dosage.

To make sure the infection clears up completely, take the medicine for as long as it has been prescribed. Do not stop taking the drug just because symptoms begin to improve. Symptoms may return if the drug is stopped too soon.

Fluoroquinolones work best when they are at constant levels in the blood. To help keep levels constant, take the medicine in doses spaced evenly through the day and night. Do not miss any doses. For best results, take this medicine with a full glass of water and drink several more glasses throughout the day, every day during treatment with the drug. The extra water will help prevent some side effects. Some fluoroquinolones should be taken on an empty stomach; others may be taken with meals. Check package directions or ask the physician or pharmacist for instructions on how to take the medicine.

### Precautions

Research suggests that fluoroquinolones may cause bone development problems in children and teenagers. Infants, children, teenagers, pregnant women, and women who are breastfeeding should not take this medicine unless directed to do so by a physician.

Although such side effects are rare, some people have had severe and life-threatening reactions to fluoroquinolones. Call a physician immediately if any of these signs of a dangerous reaction occur:

- swelling of the face and throat
- swallowing problems
- shortness of breath
- rapid heartbeat
- tingling of fingers or toes
- **itching or hives**
- loss of consciousness

Some fluoroquinolones may weaken the tendons in the shoulder, hand, or heel, making the tendons more likely to tear. Anyone who notices **pain** or inflammation in these or other tendon areas should stop taking the medicine immediately and call a physician. Rest and avoid **exercise** until the physician determines whether

### KEY TERMS

**Bacteria**—Tiny, one-celled forms of life that cause many diseases and infections.

**Bronchitis**—Inflammation of the air passages of the lungs.

**Digestive tract**—The stomach, intestines, and other parts of the body through which food passes.

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

**Microorganism**—An organism that is too small to be seen with the naked eye.

**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.

**Prostate**—A donut-shaped gland in males below the bladder that contributes to the production of semen.

**Sexually transmitted disease (STD)**—A disease that is passed from one person to another through sexual intercourse or other intimate sexual contact.

**Tendon**—A tough band of tissue that connects muscle to bone.

**Tuberculosis**—An infectious disease that usually affects the lungs, but may also affect other parts of the body. Symptoms include fever, weight loss, and coughing up blood.

**Urinary tract**—The passage through which urine flows from the kidneys out of the body.

the tendons are damaged. If the tendons are torn, surgery may be necessary to repair them.

These medicines make some people feel drowsy, dizzy, lightheaded, or less alert. 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.

This medicine may increase sensitivity to sunlight. Even brief exposure to sun can cause a severe **sunburn** or a rash. While being treated with fluoroquinolones, avoid being in direct sunlight, especially between 10 A.M. and 3 P.M.; wear a hat and tightly woven clothing that covers the arms and legs; use a sunscreen with a skin protection factor (SPF) of at least 15; protect the lips with a sun block lipstick; and do not use tanning beds, tanning booths, or sunlamps.

Do not take **antacids** that contain aluminum, calcium, or magnesium at the same time as fluoroquinolones. The antacids may keep the fluoroquinolones from working as they should. If antacids are needed, take them at least two hours before or two hours after taking norfloxacin or ofloxacin, at least four hours before or two hours after taking ciprofloxacin. Follow the same instructions for taking sucralfate (Carafate), a medicine used to treat stomach ulcers and other irritation in the digestive tract and mouth.

Anyone who has had unusual reactions to fluoroquinolones or related medicines such as cinoxacin (Cinobac) or nalidixic acid (NegGram) in the past 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.

Before using fluoroquinolones, people with any of these medical problems should make sure their physicians are aware of their conditions:

- kidney disease
- liver disease with kidney disease
- diseases of the brain or spinal cord, including hardening of the arteries in the brain, epilepsy, and other seizure disorders

Taking fluoroquinolones with certain other drugs may affect the way the drugs work or may increase the chance of side effects.

## Side effects

The most common side effects are mild **diarrhea**, nausea, vomiting, stomach or abdominal pain, **dizziness**, drowsiness, lightheadedness, nervousness, sleep problems, and **headache**. These problems usually go away as the body adjusts to the drug and do not require medical treatment unless they are bothersome.

More serious side effects are not common, but may occur. If any of the following side effects occur, check with a physician immediately:

- skin rash or other skin problems such as itching, peeling, hives, or redness
- **fever**
- agitation or confusion
- hallucinations
- shakiness or **tremors**
- seizures or convulsions
- tingling of fingers or toes
- pain where the medicine was injected (lasting after the injection)

- pain in the calves, spreading to the heels
- swelling of the calves or lower legs
- swelling of the face or neck
- swallowing problems
- rapid heartbeat
- shortness of breath
- loss of consciousness

Other rare side effects may occur. Anyone who has unusual symptoms after taking fluoroquinolones should get in touch with his or her physician.

## Interactions

Fluoroquinolones 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 fluoroquinolones should let the physician know all other medicines he or she is taking. Among the drugs that may interact with fluoroquinolones are:

- antacids that contain aluminum, calcium, or magnesium
- medicines that contain iron or zinc, including multivitamin and mineral supplements
- sucralfate (Carafate)
- caffeine
- blood thinning drugs such as warfarin (Coumadin)
- airway opening drugs (**bronchodilators**) such as aminophylline, theophylline (Theo-Dur and other brands), and oxtriphylline (choledyl and other brands)
- didanosine (Videx), used to treat HIV infection.

The list above does not include every drug that may interact with fluoroquinolones. Be sure to check with a physician or pharmacist before combining fluoroquinolones with any other prescription or nonprescription (over-the-counter) medicine.

## Resources

### OTHER

"Fluoroquinolones (Systemic)." National Library of Medicine.  
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Rosalyn Carson-DeWitt

**Fluoxetine** see **Selective serotonin reuptake inhibitors**

**Flurbiprofen** see **Nonsteroidal anti-inflammatory drugs**

Focal glomerulosclerosis see **Nephrotic syndrome**

## Folic acid

### Definition

Folic acid is a water-soluble vitamin belonging to the B-complex group of **vitamins**. These vitamins help the body break down complex carbohydrates into simple sugars to be used for energy. Excess B vitamins are excreted from the body rather than stored for later use. This is why sufficient daily intake of folic acid is necessary.

### Description

Folic acid is also known as folate, or folacin. It is one of the nutrients most often found to be deficient in the Western diet, and there is evidence that deficiency is a problem on a worldwide scale. Folic acid is found in leafy green vegetables, beans, peas and lentils, liver, beets, brussel sprouts, poultry, nutritional yeast, tuna, wheat germ, mushrooms, oranges, asparagus, broccoli, spinach, bananas, strawberries, and cantaloupes. In 1998, the U.S. Food and Drug Administration (FDA) required food manufacturers to add folic acid to enriched bread and grain products to boost intake and to help prevent neural tube defects (NTD).

### Purpose

Folic acid works together with vitamin B<sub>12</sub> and vitamin C to metabolize protein in the body. It is important for the formation of red and white blood cells. It is necessary for the proper differentiation and growth of cells and for the development of the fetus. It is also used to form the nucleic acid of DNA and RNA. It increases the appetite and stimulates the production of stomach acid for digestion and it aids in maintaining a healthy liver. A deficiency of folic acid may lead to anemia, in which there is decreased production of red blood cells. This reduces the amounts of oxygen and nutrients that are able to get to the tissues. Symptoms may include **fatigue**, reduced secretion of digestive acids, confusion, and forgetfulness. During **pregnancy**, a folic acid deficiency may lead to preeclampsia, premature birth, and increased bleeding after birth.

People who are at high risk of strokes and heart disease may greatly benefit by taking folic acid supplements. An elevated blood level of the amino acid homocysteine has been identified as a risk factor for some of

these diseases. High levels of homocysteine have also been found to contribute to problems with **osteoporosis**. Folic acid, together with vitamins B<sub>6</sub> and B<sub>12</sub>, helps break down homocysteine, and may help reverse the problems associated with elevated levels.

Pregnant women have an increased need for folic acid, both for themselves and their child. Folic acid is necessary for the proper growth and development of the fetus. Adequate intake of folic acid is vital for the prevention of several types of **birth defects**, particularly NTDs. The neural tube of the embryo develops into the brain, spinal cord, spinal column, and the skull. If this tube forms incompletely during the first few months of pregnancy a serious, and often fatal, defect results in **spina bifida** or anencephaly. Folic acid, taken from one year to one month before conception through the first four months of pregnancy, can reduce the risk of NTDs by 50–70%. It also helps prevent a **cleft lip and palate**.

Research shows that folic acid can be used to successfully treat cervical dysplasia, a condition diagnosed by a Pap smear, of having abnormal cells in the cervix. This condition is considered to be a possible precursor to **cervical cancer**, and is diagnosed as an abnormal Pap smear. Daily consumption of 1,000 mcg of folic acid for three or more months has resulted in improved cervical cells upon repeat Pap smears.

Studies suggest that long-term use of folic acid supplements may also help prevent lung and **colon cancer**. Researchers have also found that alcoholics who have low folic acid levels face a greatly increased possibility of developing colon cancer.

### Preparations

To correct a folic acid deficiency, supplements are taken in addition to food. Since the functioning of the B vitamins is interrelated, it is generally recommended that the appropriate dose of B-complex vitamins be taken in place of single B vitamin supplements. The Recommended Dietary Allowances (RDA) for folate is 400 mcg per day for adults, 600 mcg per day for pregnant women, and 500 mcg for nursing women. Medicinal dosages of up to 1,000–2,000 mcg per day may be prescribed.

### Precautions

Folic acid is not stable. It is easily destroyed by exposure to light, air, water, and cooking. Therefore, the supplement should be stored in a dark container in a cold, dry place, such as a refrigerator. Many medications interfere with the body's absorption and use of folic acid. This includes sulfa drugs, sleeping pills, estrogen, anti-convulsants, birth control pills, **antacids**, quinine, and

## KEY TERMS

**Homocysteine**—An amino acid involved in the breakdown and absorption of protein in the body.

**Preeclampsia**—A serious disorder of late pregnancy in which the blood pressure rises, there is a large amount of retained fluids, and the kidneys become less effective and excrete proteins directly into the urine.

**Raynaud's disease**—A symptom of various underlying conditions affecting blood circulation in the fingers and toes and causing them to be sensitive to cold.

**Recommended Daily Allowance (RDA)**—Guidelines for the amounts of vitamins and minerals necessary for proper health and nutrition established by the National Academy of Sciences in 1989.

**Water-soluble vitamins**—Vitamins that are not stored in the body and are easily excreted. They must, therefore, be consumed regularly as foods or supplements to maintain health.

some **antibiotics**. Using large amounts of folic acid (e.g., over 5,000 mcg per day) can mask a vitamin B<sub>12</sub> deficiency and thereby risk of irreversible nerve damage.

### Side effects

At levels of 5,000 mcg or less, folic acid is generally safe for use. Side effects are uncommon. However, large doses may cause nausea, decreased appetite, bloating, gas, decreased ability to concentrate, and **insomnia**. Large doses may also decrease the effects of phenytoin (Dilantin), a seizure medication.

### Interactions

As with all B-complex vitamins, it is best to take folic acid with the other B vitamins. Vitamin C is important to the absorption and functioning of folic acid in the body.

### Resources

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### ORGANIZATIONS

- Centers for Disease Control and Prevention. 4770 Buford Highway NE, MSF-45, Atlanta, GA 30341-3724.  
(888)232-6789. Flo@cdc.gov. <<http://www.cdc.gov/nchh/programs/cddh/folic/folicfaqs.htm>>.

### OTHER

- Adams, Suzanne L. *The Art of Cytology: Folic Acid/ B-12 Deficiency* suzann@concentric.net. <<http://www.concentric.net/~Suza2/page22.htm>>.  
"Folic Acid." <<http://www.cybervitamins.com/folicacid.htm>>.  
"Folic Acid: Coming to A Grocery Store Near You." <<http://www.mayohealth.org/mayo/9710/htm/folic.htm>>.  
"Folic acid (oral/injectible)." Dr. Koop.com.Inc. 700 N. Mopac, Suite 400, Austin, TX 48731. <<http://www.drkoop.com/hcr/drugstore/pharmacy/leaflets/english/d00241a1.asp>>.  
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Patience Paradox

## Folic acid deficiency anemia

### Definition

**Folic acid deficiency**, an abnormally low level of one of the B **vitamins**, results in anemia characterized by red blood cells that are large in size but few in number.

### Description

Folic acid is necessary for growth and cellular repair, since it is a critical component of DNA and RNA as well as essential for the formation and maturation of red blood cells. Folic acid deficiency is one of the most common of all vitamin deficiencies. Although it occurs in both males and females, folic acid deficiency anemia most often affects women over 30. It becomes increasingly common as age impedes the body's ability to absorb folic acid, a water-soluble vitamin that is manufactured by intestinal bacteria and stored for a short time in the liver. Folic acid deficiency has also been implicated

ed as a cause of neural tube defects in the developing fetus. Recent research has shown that adequate amounts of folic acid can prevent up to half of these **birth defects**, if women start taking folic acid supplements shortly before conception.

A healthy adult needs at least 400 mcg of folic acid every day. Requirements at least double during **pregnancy**, and increase by 50% when a woman is breastfeeding. The average American diet, high in fats, sugar, and white flour, provides about 200 mcg of folic acid, approximately the amount needed to maintain tissue stores of the substance for six to nine months before a deficiency develops. Most of the folic acid in foods (with the exception of the folic acid added to enriched flour and breakfast cereals) occurs as folate. Folate is only about half as available for the body to use as is the folic acid in pills and supplements. Folate also is easily destroyed by sunlight, overcooking, or the storing of foods at room temperature for an extended period of time.

Good dietary sources of folate include:

- leafy green vegetables
- liver
- mushrooms
- oatmeal
- peanut butter
- red beans
- soy
- wheat germ

### Causes and symptoms

This condition usually results from a diet lacking in foods with high folic acid content, or from the body's inability to digest foods or absorb foods having high folic acid content. Other factors that increase the risk of developing folic acid deficiency anemia are:

- age
- alcoholism
- birth-control pills, anticonvulsant therapy, sulfa **antibiotics**, and certain other medications
- illness
- smoking
- stress

**Fatigue** is often the first sign of folic acid deficiency anemia. Other symptoms include:

- anorexia nervosa
- pale skin

- paranoia
- rapid heart beat
- sore, inflamed tongue
- weakness
- weight loss

### Diagnosis

Diagnostic procedures include blood tests to measure hemoglobin, an iron-containing compound that carries oxygen to cells throughout the body. Symptoms may be reevaluated after the patient has taken prescription folic acid supplements.

### Treatment

Folic acid supplements are usually prescribed, and self-care includes avoiding:

- alcohol
- non-herbal tea, **antacids**, and phosphates (contained in beer, ice cream, and soft drinks), which restrict iron absorption
- tobacco

A person with folic acid deficiency anemia should rest as often as necessary until restored energy levels make it possible to resume regular activities. A doctor should be seen if **fever**, chills, muscle aches, or new symptoms develop during treatment, or if symptoms do not improve after two weeks of treatment.

### Alternative treatment

Alternative therapies for folic acid deficiency anemia may include **reflexology** concentrated on areas that influence the liver and spleen. Increasing consumption of foods high in folate is helpful. Eating a mixture of yogurt (8 oz) and turmeric (1 tsp) also may help resolve symptoms. A physician should be contacted if the tongue becomes slick or smooth or the patient:

- bruises or tires easily
- feels ill for more than five days
- feels weak or out of breath
- looks pale or jaundiced

### Prognosis

Although adequate folic acid intake usually cures this condition in about three weeks, folic acid deficiency anemia can make patients infertile or more susceptible to infection. Severe deficiencies can result in congestive **heart failure**.

## Prevention

Eating raw or lightly cooked vegetables every day will help maintain normal folic acid levels, as will taking a folic acid supplement containing at least 400 mcg of this vitamin. Because folic acid deficiency can cause birth defects, all women of childbearing age who can become pregnant should consume at least 400 mcg of folic acid daily; a woman who is pregnant should have regular medical checkups, and take a good prenatal vitamin.

## Resources

### BOOKS

*New Choices in Natural Healing.* Ed. Bill Gottlieb, et al.

Emmaus, PA: Rodale Press, 1995.

*Professional Guide to Diseases.* 5th ed. Springhouse, PA: Springhouse Corporation, 1995.

### OTHER

"Folic-Acid Deficiency Anemia." *ThriveOnline.* 6 Apr. 1998 <<http://thriveonline.oxygen.com>>.

"Gastrointestinal Disorders: Malabsorption Syndromes." *The Meck Page.* 13 Apr. 1998 <<http://www.merck.com>>.

Maureen Haggerty

# Follicle-stimulating hormone test

## Definition

The follicle-stimulating hormone (FSH) test measures the amount of FSH in the blood. FSH is a hormone that regulates the growth and development of eggs and sperm, and this test is used to diagnose or evaluate disorders involving the pituitary gland and reproductive system.

## Purpose

FSH testing is performed if a physician suspects the patient may have a disorder involving the reproductive system or pituitary gland. The pituitary gland produces FSH, which stimulates the growth of the sacks (follicles) that surround the eggs in a woman's ovaries. This is important for the process of ovulation, in which the egg is released. In men, FSH stimulates production of sperm. If there are abnormal levels of FSH in the blood it may mean that one of several disorders are present. Normal fluctuations occur as a result of **puberty**, the menstrual cycle, **pregnancy**, and **menopause**.

The FSH test is performed more often on women than on men. In women, it is used to determine if menopause has begun, to diagnose **infertility** and **menstrual disor-**

**ders** (such as anovulatory bleeding), to measure hormone levels in children who enter puberty at an early age, and to diagnose other disorders. In men, it can be used to determine early puberty, abnormal tissue growth on one or more of the hormone-secreting (endocrine) glands (called multiple endocrine neoplasia), or to diagnose other disorders.

## Description

The FSH test is a blood test. Blood will be drawn from the patient and analyzed in a laboratory.

## Preparation

In preparation for the test, there are no food or fluid intake restrictions. Patients may be advised to discontinue certain medications for 48 hours before the test. A menstruating woman having hot flashes or irregular periods should be tested on the second or third day of her menstrual cycle. A woman who has missed a period and is having other menopausal symptoms can be tested at any time.

## Aftercare

No aftercare is necessary.

## Risks

There are no risks associated with this test.

## Normal results

Normal FSH test results vary according to age and sexual maturity. The phase of a woman's menstrual cycle or use of birth-control pills also affects test results.

For an adult male, normal results range from about 4–25 units of FSH in every liter of blood (U/L) or about 5–20 micro-international units in every milliliter.

For a premenopausal woman, normal values range from 4–30 U/L or 5–20 micro-international units per milliliter. In a pregnant woman, FSH levels are too low to measure. After menopause, normal values range from 40–250 U/L or 50–100 micro-international units per milliliter.

FSH levels fluctuate during premenopause. If no other symptoms are present, an elevated FSH level should not be interpreted as proof that menopause has begun.

## Abnormal results

**Anorexia nervosa** and disorders of the hypothalamus or pituitary gland can result in abnormally low FSH levels.

Abnormal levels can also indicate:

- infertility
- hypopituitarism
- klinefelter syndrome (in men)

## KEY TERMS

- Anovulatory bleeding**—Bleeding without release of an egg from an ovary.
- Hypopituitarism**—Underactivity of the pituitary gland.
- Hypothalamus**—The part of the brain that controls the endocrine system.
- Klinefelter's syndrome**—Chromosomal abnormality characterized by small testes and male infertility.
- Multiple endocrine neoplasia**—Abnormal tissue growth on one or more of the endocrine (hormone-secreting) glands.
- Polycystic ovary disease**—A condition in which a woman has little or no menstruation, is infertile, has excessive body hair, and is obese. The ovaries may contain several cysts.
- Turner syndrome**—Chromosomal abnormality characterized by immature reproductive organs in women.

- turner syndrome
- ovarian failure
- polycystic ovary syndrome

## Resources

### BOOKS

*Everything You Need to Know About Medical Tests*. Ed. Michael Shaw, et al. Springhouse, PA: Springhouse Corporation, 1996.

### OTHER

“Follicle-Stimulating Hormone Test.” *ThriveOnline*. 24 Feb. 1998 <<http://thriveonline.oxygen.com>>.  
 “Follicle-Stimulating Hormone Test.” *HealthAnswers.com* 25 Feb. 1998 <<http://www.healthanswers.com>>.

Maureen Haggerty

Follicular cysts see **Ovarian cysts**

## Folliculitis

### Definition

Folliculitis is inflammation or infection of one or more hair follicles (openings in the skin that enclose hair).

## Description

Folliculitis can affect both women and men at any age. It can develop on any part of the body, but is most likely to occur on the scalp, face, or parts of the arms, armpits, or legs not usually covered by clothing.

Small, yellowish-white blister-like lumps (pustules) surrounded by narrow red rings are usually present with both bacterial folliculitis and fungal folliculitis. Hair can grow through or alongside of the pustules, which sometimes ooze blood-stained pus.

Folliculitis can cause **boils** and, in rare instances, serious skin infections. Bacteria from folliculitis can enter the blood stream and travel to other parts of the body.

## Causes and symptoms

Folliculitis develops when bacteria, such as *Staphylococcus*, or a fungus enters the body through a cut, scrape, surgical incision, or other break in the skin near a hair follicle. Scratching the affected area can trap fungus or bacteria under the fingernails and spread the infection to hair follicles on other parts of the body.

The bacteria that cause folliculitis are contagious. A person who has folliculitis can infect others who live in the same household.

Factors that increase the risk of developing folliculitis include:

- dermatitis
- diabetes
- dirty, crowded living conditions
- eczema
- exposure to hot, humid temperatures
- infection in the nose or other recent illness
- tight clothing

## Diagnosis

Diagnosis is based on the patient’s medical history and observations. Laboratory analysis of the substance drained from a pustule can be used to distinguish bacterial folliculitis from fungal folliculitis.

## Treatment

Bacterial folliculitis may disappear without treatment, but is likely to recur. Non-prescription topical **antibiotics** like Bacitracin, Mycitracin, or Neomycin, gently rubbed on to affected areas three or four times a day, can clear up a small number of bacterial folliculitis pustules. Oral antibiotics such as erythromycin (Ery-



**Acne folliculitis.** (Custom Medical Stock Photo. Reproduced by permission.)

thocin) may be prescribed if the infection is widespread. The drug griseofulvin (Fulvicin) and topical antifungal medications are used to treat fungal folliculitis.

A doctor should be notified if:

- pustules spread after treatment has begun or reappear after treatment is completed
- the patient's **fever** climbs above 100°F (37.8°C)
- the patient develops boils or swollen ankles
- redness, swelling, warmth, or **pain** indicate that the infection has spread
- unexplained new symptoms appear

### Alternative treatment

Eating a balanced diet, including protein, complex carbohydrates, healthy fats, fresh fruits and vegetables, and drinking eight to 10 glasses of water a day may stimulate the body's immune system and shorten the course of the infection. Garlic (*Allium sativum*) and goldenseal (*Hydrastis canadensis*), both antiseptic agents against staph infections, may be taken. The daily dosage would vary from person to person and is based on the severity of the infection. **Echinacea** (*Echinacea spp.*) is helpful in modulating immune function. Again, the dosage would vary.

Daily doses of 30–50 mg zinc and 1,000–5,000 mg Vitamin C (taken in equal amounts at several times during the day), and 300–2,000 mg bioflavonoids can also strengthen the body's infection-fighting ability. High

doses of **vitamins** and **minerals** should not be used without a doctor's approval.

### Prognosis

If properly treated, the symptoms of bacterial folliculitis generally disappear in about two weeks. Fungal folliculitis should clear up within six weeks. But it can worsen if the condition is misdiagnosed and inappropriately treated with steroid creams.

### Prevention

Anyone who has a tendency to develop folliculitis should cleanse the skin with antibacterial soap twice a day and before shaving and should not use oily skin lotions. Men should not shave while the beard area is infected. When they begin shaving again, they should use a new blade each time. Women who have had fungal folliculitis should use depilatory creams instead of razors. Daily shampooing can help prevent folliculitis in the scalp. The spread of infection can be prevented by not sharing towels or washcloths

### Resources

#### BOOKS

The Editors of Time-Life Books. *The Medical Advisor: The Complete Guide to Alternative and Conventional Treatments*. Alexandria, VA: Time Life, Inc., 1996.

#### OTHER

- “Bacterial folliculitis.” *ThriveOnline*. 5 Apr. 1998 <<http://thriveonline.oxygen.com>>.
- “Folliculitis.” *ThriveOnline*. 5 Apr. 1998 <<http://thriveonline.oxygen.com>>.
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Maureen Haggerty

Foot acupressure see **Reflexology**

## Food poisoning

### Definition

Food **poisoning** is a general term for health problems arising from eating contaminated food. Food may be contaminated by bacteria, viruses, environmental toxins, or toxins present within the food itself, such as the poisons in some mushrooms or certain seafood. Symptoms of food poisoning usually involve nausea, vomiting

and/or **diarrhea**. Some food-borne toxins can affect the nervous system.

## Description

Every year millions of people suffer from bouts of vomiting and diarrhea each year that they blame on "something I ate." These people are generally correct. Each year in the United States, one to two bouts of diarrheal illness occur in every adult. The Centers for Disease Control and Prevention (CDC) estimates that there are from six to 33 million cases of food poisoning in the United States annually. Many cases are mild and pass so rapidly that they are never diagnosed. Occasionally a severe outbreak creates a newsworthy public health hazard.

Classical food poisoning, sometimes incorrectly called ptomaine poisoning, is caused by a variety of different bacteria. The most common are *Salmonella*, *Staphylococcus aureus*, *Escherichia coli* O157:H7 or other *E. coli* strains, *Shigella*, and *Clostridium botulinum*. Each has a slightly different incubation period and duration, but all except *C. botulinum* cause inflammation of the intestines and diarrhea. Sometimes food poisoning is called bacterial **gastroenteritis** or infectious diarrhea. Food and water can also be contaminated by viruses (such as the Norwalk agent that causes diarrhea and the viruses of **hepatitis A** and **E**), environmental toxins (heavy metals), and poisons produced within the food itself (**mushroom poisoning** or **fish and shellfish poisoning**).

Careless food handling during the trip from farm to table creates conditions for the growth of bacteria that make people sick. Vegetables that are eaten raw, such as lettuce, may be contaminated by bacteria in soil, water, and dust during washing and packing. Home canned and commercially canned food may be improperly processed at too low a temperature or for too short a time to kill the bacteria.

Raw meats carry many food-borne bacterial diseases. The United States Food and Drug Administration (FDA) estimates that 60% or more of raw poultry sold at retail carry some disease-causing bacteria. Other raw meat products and eggs are contaminated to a lesser degree. Thorough cooking kills the bacteria and makes the food harmless. However, properly cooked food can become re-contaminated if it comes in contact with plates, cutting boards, countertops, or utensils that were used with raw meat and not cleaned and sanitized.

Cooked foods can also be contaminated after cooking by bacteria carried by food handlers or from bacteria in the environment. It is estimated that 50% of healthy people have the bacteria *Staphylococcus aureus* in their nasal passages and throat, and on their skin and hair. Rubbing a runny nose, then touching food can introduce

the bacteria into cooked food. Bacteria flourish at room temperature, and will rapidly grow into quantities capable of making people sick. To prevent this growth, food must be kept hot or cold, but never just warm.

Although the food supply in the United States is probably the safest in the world, anyone can get food poisoning. Serious outbreaks are rare. When they occur, the very young, the very old, and those with immune system weaknesses have the most severe and life-threatening cases. For example, this group is 20 times more likely to become infected with the *Salmonella* bacteria than the general population.

Travel outside the United States to countries where less attention is paid to sanitation, water purification, and good food handling practices increases the chances that a person will get food poisoning. People living in institutions such as nursing homes are also more likely to get food poisoning.

## Causes and symptoms

The symptoms of food poisoning occur because food-borne bacteria release toxins or poisons as a byproduct of their growth in the body. These toxins (except those from *C. botulinum*) cause inflammation and swelling of the stomach, small intestine and/or large intestine. The result is abdominal muscle cramping, vomiting, diarrhea, **fever**, and the chance of **dehydration**. The severity of symptoms depends on the type of bacteria, the amount consumed, and the individual's general health and sensitivity to the bacterial toxin.

## Salmonella

According to a 2001 report from the CDC, *Salmonella* caused almost 50,000 culture-confirmed cases of food poisoning in the United States annually. However, between two and four million probably occur each year. *Salmonella* is found in egg yolks from infected chickens, in raw and undercooked poultry and in other meats, dairy products, fish, shrimp, and many more foods. The CDC estimates that one out of every 50 consumers is exposed to a contaminated egg yolk each year. However, thorough cooking kills the bacteria and makes the food harmless. *Salmonella* is also found in the feces of pet reptiles such as turtles, lizards, and snakes.

About one out of every 1,000 people get food poisoning from *Salmonella*. Of these, two-thirds are under age 20, with the majority under age nine. Most cases occur in the warm months between July and October.

Symptoms of food poisoning begin eight to 72 hours after eating food contaminated with *Salmonella*. These include traditional food poisoning symptoms of abdomi-

nal pain, diarrhea, vomiting, and fever. The symptoms generally last one to five days. Dehydration can be a complication in severe cases. People generally recover without antibiotic treatment, although they may feel tired for a week after the active symptoms subside.

### **Staphylococcus aureus**

*Staphylococcus aureus* is found on humans and in the environment in dust, air, and sewage. The bacteria is spread primarily by food handlers using poor sanitary practices. Almost any food can be contaminated, but salad dressings, milk products, cream pastries, and any food kept at room temperature, rather than hot or cold are likely candidates.

It is difficult to estimate the number of cases of food poisoning from *Staphylococcus aureus* that occur each year, because its symptoms are so similar to those caused by other foodborne bacteria. Many cases are mild and the victim never sees a doctor.

Symptoms appear rapidly, usually one to six hours after the contaminated food is eaten. The acute symptoms of vomiting and severe abdominal cramps without fever usually last only three to six hours and rarely more than 24 hours. Most people recover without medical assistance. Deaths are rare.

### **Escherichia coli (E. coli)**

There are many strains of *E. coli*, and not all of them are harmful. The strain that causes most severe food poisoning is *E. coli O157:H7*. Food poisoning by *E. coli* occurs in three out of every 10,000 people. Foodborne *E. coli* is found and transmitted mainly in food derived from cows such as raw milk, raw or rare ground beef and fruit or vegetables that are contaminated.

Symptoms of food poisoning from *E. coli* are slower to appear than those caused by some of the other foodborne bacteria. *E. coli* produces toxins in the large intestine rather than higher up in the digestive system. This accounts for the delay in symptoms and the fact that vomiting rarely occurs in *E. coli* food poisoning.

One to three days after eating contaminated food, the victim with *E. coli O157:H7* begins to have severe abdominal cramps and watery diarrhea that usually becomes bloody within 24 hours. There is little or no fever, and rarely does the victim vomit. The bloody, watery diarrhea lasts from one to eight days in uncomplicated cases.

### **Campylobacter jejuni (C. jejuni)**

According to the FDA, *C. jejuni* is the leading cause of bacterial diarrhea in the United States. It is responsible for more cases of bacterial diarrhea than *Shigella* and *Salmo-*

*nella* combined. Anyone can get food poisoning from *C. jejuni*, but children under five and young adults between the ages of 15 and 29 are more frequently infected.

*C. jejuni* is carried by healthy cattle, chickens, birds, and flies. It is not carried by healthy people in the United States or Europe. The bacteria is also found ponds and stream water. The ingestion of only a few hundred *C. jejuni* bacteria can make a person sick.

Symptoms of food poisoning begin two to five days after eating food contaminated with *C. jejuni*. These symptoms include fever, abdominal pain, nausea, **headache**, muscle pain, and diarrhea. The diarrhea can be watery or sticky and may contain blood. Symptoms last from seven to 10 days, and relapses occur in about one quarter of people who are infected. Dehydration is a common complication. Other complications such as arthritis-like joint pain and **hemolytic-uremic syndrome (HUS)** are rare.

### **Shigella**

*Shigella* is a common cause of diarrhea in travelers to developing countries. It is associated with contaminated food and water, crowded living conditions, and poor sanitation. The bacterial toxins affect the small intestine.

Symptoms of food poisoning by *Shigella* appear 36–72 hours after eating contaminated food. These symptoms are slightly different from those associated with most foodborne bacteria. In addition to the familiar watery diarrhea, nausea, vomiting, abdominal cramps, chills and fever occur. The diarrhea may be quite severe with cramps progressing to classical dysentery. Up to 40% of children with severe infections show neurological symptoms. These include seizures caused by fever, confusion, headache, lethargy, and a stiff neck that resembles **meningitis**.

The disease runs its course usually in two to three days but may last longer. Dehydration is a common complication. Most people recover on their own, although they may feel exhausted, but children who are malnourished or have weakened immune systems may die.

### **Clostridium botulinum (C. botulinum)**

*C. botulinum*, which causes both adult **botulism** and infant botulism, is unlike any of the other foodborne bacteria. First, *C. botulinum* is an anaerobic bacterium in that it can only live in the absence of oxygen. Second, the toxins from *C. botulinum* are neurotoxins. They poison the nervous system, causing **paralysis** without the vomiting and diarrhea associated with other foodborne illnesses. Third, toxins that cause adult botulism are released when the bacteria grows in an airless environment outside the body. They can be broken down and made harm-

less by heat. Finally, botulism is much more likely to be fatal even in tiny quantities.

Adult botulism outbreaks are usually associated with home canned food, although occasionally commercially canned or vacuum packed foods are responsible for the disease. *C. botulinum* grows well in non-acidic, oxygen-free environments. If food is canned at too low heat or for too brief a time, the bacteria is not killed. It reproduces inside the can or jar, releasing its deadly neurotoxin. The toxin can be made harmless by heating the contaminated food to boiling for ten minutes. However, even a very small amount of the *C. botulinum* toxin can cause serious illness or death.

Symptoms of adult botulism appear about 18–36 hours after the contaminated food is eaten, although there are documented times of onset ranging from four hours to eight days. Initially a person suffering from botulism feels weakness and **dizziness** followed by double vision. Symptoms progress to difficulty speaking and swallowing. Paralysis moves down the body, and when the respiratory muscles are paralyzed, death results from asphyxiation. People who show any signs of botulism poisoning must receive immediate emergency medical care to increase their chance of survival.

Infant botulism is a form of botulism first recognized in 1976. It differs from food-borne botulism in its causes and symptoms. Infant botulism occurs when a child under the age of one year ingests the spores of *C. botulinum*. These spores are found in soil, but a more common source of spores is honey.

The *C. botulinum* spores lodge in the baby's intestinal tract and begin to grow, producing their neurotoxin. Onset of symptoms is gradual. Initially the baby is constipated. This is followed by poor feeding, lethargy, weakness, drooling, and a distinctive wailing cry. Eventually, the baby loses the ability to control its head muscles. From there the paralysis progresses to the rest of the body.

## Diagnosis

One important aspect of diagnosing food poisoning is for doctors to determine if a number of people have eaten the same food and show the same symptoms of illness. When this happens, food poisoning is strongly suspected. The diagnosis is confirmed when the suspected bacteria is found in a **stool culture** or a fecal smear from the person. Other laboratory tests are used to isolate bacteria from a sample of the contaminated food. Botulism is usually diagnosed from its distinctive neurological symptoms, since rapid treatment is essential. Many cases of food poisoning go undiagnosed, since a definite diagnosis is not necessary to effectively treat the symptoms. Because it takes time for symptoms to develop, it is not

## Common Pathogens Causing Food Poisoning

Pathogen	Common Host(s)
Campylobacter E.coli 0157:H7	Poultry Undercooked, contaminated ground beef
Listeria	Found in a variety of raw foods, such as uncooked meats and vegetables, and in processed foods that become contaminated after processing
Salmonella	Poultry, eggs, meat, and milk
Shigella	This bacteria is transmitted through direct contact with an infected person or from food or water that become contaminated by an infected person
Vibrio	Contaminated seafood

necessarily the most recent food one has eaten that is the cause of the symptoms.

## Treatment

Treatment of food poisoning, except that caused by *C. botulinum*, focuses on preventing dehydration by replacing fluids and electrolytes lost through vomiting and diarrhea. Electrolytes are salts and **minerals** that form electrically charged particles (ions) in body fluids. Electrolytes are important because they control body fluid balance and are important for all major body reactions. Pharmacists can recommend effective, pleasant-tasting, electrolytically balanced replacement fluids that are available without a prescription. When more fluids are being lost than can be consumed, dehydration may occur. Dehydration more likely to happen in the very young, the elderly, and people who are taking **diuretics**. To prevent dehydration, a doctor may give fluids intravenously.

In very serious cases of food poisoning, medications may be given to stop abdominal cramping and vomiting. Anti-diarrheal medications are not usually given. Stopping the diarrhea keeps the toxins in the body longer and may prolong the infection.

People with food poisoning should modify their diet. During period of active vomiting and diarrhea they should not try to eat and should drink only clear liquids frequently but in small quantities. Once active symptoms stop, they should eat bland, soft, easy to digest foods for two to three days. One example is the BRAT diet of bananas, rice, applesauce, and toast, all of which are easy to digest. Milk products, spicy food, alcohol and fresh fruit should be avoided for a few days, although babies should continue to breastfeed. These modifications are often all the treatment that is necessary.

Severe bacterial food poisonings are sometimes treated with **antibiotics**. Trimethoprim and sulfamethox-

## KEY TERMS

**Diuretic**—Medication that increases the urine output of the body.

**Electrolytes**—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

**Lactobacillus acidophilus**—This bacteria is found in yogurt and changes the balance of the bacteria in the intestine in a beneficial way.

**Platelets**—Blood cells that help the blood to clot.

azole (Septra, Bactrim), ampicillin (Amcill, Polycill) or ciprofloxacin (Ciloxan, Cipro) are most frequently used.

Botulism is treated in a different way from other bacterial food poisonings. Botulism antitoxin is given to adults, but not infants, if it can be administered within 72 hours after symptoms are first observed. If given later, it provides no benefit.

Both infants and adults require hospitalization, often in the intensive care unit. If the ability to breathe is impaired, patients are put on a mechanical ventilator to assist their breathing and are fed intravenously until the paralysis passes.

### Alternative treatment

Alternative practitioners offer the same advice as traditional practitioners concerning diet modification. In addition they recommend taking charcoal tablets, *Lactobacillus acidophilus*, *Lactobacillus bulgaricus*, and citrus seed extract. An electrolyte replacement fluid can be made at home by adding one teaspoon of salt and four teaspoons of sugar to one quart of water. For food poisoning other than botulism, two homeopathic remedies, either *Arsenicum album* or *Nux vomica*, are strongly recommended.

### Prognosis

Most cases of food poisoning (except botulism) clear up on their own within one week without medical assistance. The ill person may continue feel tired for a few days after active symptoms stop. So long as the ill person does not become dehydrated, there are few complications. Deaths are rare and usually occur in the very

young, the very old and people whose immune systems are already weakened.

Complications of *Salmonella* food poisoning include arthritis-like symptoms that occur three to four weeks after infection. Although deaths from *Salmonella* are rare, they do occur. Most deaths caused by *Salmonella* food poisoning have occurred in elderly people in nursing homes.

Adults usually recover without medical intervention, but many children need to be hospitalized as the result of *E. coli* food poisoning. *E. coli* toxins may be absorbed into the blood stream where they destroy red blood cells and platelets. Platelets are important in blood clotting. About 5% of victims develop hemolytic-uremic syndrome which results in sudden kidney failure and makes dialysis necessary. (Dialysis is a medical procedure used to filter the body's waste product when the kidneys have failed).

Botulism is the deadliest of the bacterial food-borne illnesses. With prompt medical care, the death rate is less than 10%.

### Prevention

Food poisoning is almost entirely preventable by practicing good sanitation and good food handling techniques. These include:

- keep hot foods hot and cold foods cold
- cook meat to the recommended internal temperature, use a meat thermometer to check and cook eggs until they are no longer runny
- refrigerate leftovers promptly, do not let food stand at room temperature
- avoid contaminating surfaces and other foods with the juices of uncooked meats
- wash fruits and vegetables before using
- purchase pasteurized dairy products and fruit juices
- throw away bulging or leaking cans or any food that smells spoiled
- wash hands well before and during food preparation and after using the bathroom
- sanitize food preparation surfaces regularly

### Resources

#### PERIODICALS

U. S. Food and Drug Administration. "The Unwelcome Dinner Guest: Preventing Food-Borne Illness." In *FDA Consumer* (December 1997).

#### ORGANIZATION

Centers for Disease Control and Prevention. <<http://www.cdc.gov>>.

**OTHER**

U. S. Food and Drug Administration. Center for Food Safety and Applied Nutrition. *Bad Bug Book*. <<http://vm.cfsan.fda.gov>>.

Suzanne M. Lutwick, MPH

## Foot care

### Definition

Foot care involves all aspects of preventative and corrective care of the foot and ankle. Doctors specializing in foot care are called podiatrists.

### Purpose

During an average lifetime, each person walks about 115,000 miles and three-quarters of people have foot problems at some point in their lives.

Foot problems can arise from wearing ill-fitting shoes, from general wear and tear, as a result of injury, or as a complication of disease. People with **diabetes mellitus** or circulatory diseases are 20 times more likely to have foot problems than the general public.

Podiatrists are doctors who specialize in treating the foot and ankle. Other doctors who have experience with foot problems are family physicians, orthopedists, sports medicine specialists, and those who care for diabetics. Problems with the feet include foot **pain**, joint inflammation, plantar **warts**, fungal infections (like **athlete's foot**), nerve disorders, torn ligaments, broken bones, bacterial infections, and tissue injuries (like frostbite).

### Precautions

People with diabetes or circulatory disorders should be alert to even small foot problems. In these people, a break in the skin can lead to infection, **gangrene**, and **amputation**.

### Description

Daily foot care for people likely to develop foot problems includes washing the feet in tepid water with mild soap and oiling the feet with vegetable oil or a lanolin-based lotion. Toenails should be cut straight across above the level of the skin after soaking the feet in tepid water. **Corns and calluses** should not be cut. If they need removal, it should be done under the care of a doctor. Athletes foot and plantar warts should also be treated by a doctor if they develop in high risk patients.

Many people with diabetes or circulatory disorders have problems with cold feet. These problems can be reduced by avoiding **smoking** tobacco (smoking constricts the blood vessels), wearing warm socks, not crossing the legs while sitting or not sitting in one position too long, or avoiding constricting stockings.

People with circulatory problems should not use heating pads or hot water bottles on their feet, as even moderate heat can damage the skin if circulation is impaired.

### Preparation

No special preparation other than an understanding of the nature of foot problems is necessary to begin routine foot care.

### Aftercare

Foot care is preventative and should be ongoing throughout a person's life.

### Risks

There are no risks associated with foot care. The risks are in ignoring the feet and allowing problems to develop.

### Normal results

With regular care, foot disorders such as infections, skin ulcers, and gangrene can be prevented.

### Resources

#### BOOKS

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#### ORGANIZATIONS

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

American Podiatry Association. 20 Chevy Chase Circle, NW, Washington, D.C. 20015.

Tish Davidson

Foreign bodies see **Foreign objects**

## Foreign objects

### Definition

"Foreign" means "originating elsewhere" or simply "outside the body." Foreign bodies typically become



**X ray of swallowed spoon and blade in the intestine.** (Photo Researchers. Reproduced by permission.)

lodged in the eyes, ears, nose, airways, and rectum of human beings.

## Description

Both children and adults experience problems caused by foreign objects getting stuck in their bodies. Young children, in particular, are naturally curious and may intentionally put shiny objects, such as coins or button batteries, into their mouths. They also like to stick things in their ears and up their noses. Adults may accidentally swallow a non-food object or inhale a foreign body that gets stuck in the throat. Even if an object like a toothpick successfully passes through the esophagus and into the stomach, it can get stuck inside the rectum. Airborne particles can lodge in the eyes of people at any age.

Foreign bodies can be in hollow organs (like swallowed batteries) or in tissues (like bullets). They can be inert or irritating. If they irritate they will cause inflam-

mation and scarring. They can bring infection with them or acquire it and protect it from the body's immune defenses. They can obstruct passageways either by their size or by the scarring they cause. Some can be toxic.

## Causes and symptoms

### Eyes

Dust, dirt, sand, or other airborne material can lodge in the eyes, causing minor irritation and redness. More serious damage can be caused by hard or sharp objects that penetrate the surface and become embedded in the cornea or conjunctiva (the mucous membranes around the inner surface of the eyelids). Swelling, redness, bleeding from the surface blood vessels, sensitivity to light, and sudden vision problems are all symptoms of foreign matter in the eyes.

### Ears and nose

Children will sometimes put things into their noses, ears, and other openings. Beans, popcorn kernels, raisins, and beads are just a few of the many items that have been found in these bodily cavities. On occasion, insects may also fly into the ears and nose. **Pain, hearing loss**, and a sense of something stuck in the ear are symptoms of foreign bodies in the ears. A smelly, bloody discharge from one nostril is a symptom of foreign bodies in the nose.

### Airways and stomach

At a certain age children will eat anything. A very partial list of items recovered from young stomachs includes the following: Coins, chicken bones, fish bones, beads, rocks, plastic toys, pins, keys, round stones, marbles, nails, rings, batteries, ball bearings, screws, staples, washers, a heart pendant, a clothespin spring, and a toy soldier. Some of these items will pass right on through and come out the other end. The progress of metal objects has been successfully followed with a metal detector. Others, like sharp bones, can get stuck and cause trouble. Batteries are corrosive and must be removed immediately.

Children eat things and stick things into their bodily openings of their own volition. But they inhale them unwittingly. The most commonly inhaled item is probably a peanut. A crayon and a cockroach have been found in a child's windpipes. These items always cause symptoms (difficulty swallowing and spitting up saliva, for instance) and may elude detection for some time while the child is being treated for **asthma** or recurring **pneumonia**.

Adults are not exempt from unorthodox inedibles. Dental devices are commonly swallowed. Adults with mental illness or subversive motives may swallow inappropriate objects, such as toothbrushes.

## *Rectum*

Sometimes a foreign object will successfully pass through the throat and stomach only to get stuck at the juncture between the rectum and the anal canal. Items may also be self-introduced to enhance sexual stimulation and then get stuck. Sudden sharp pain during elimination may signify that an object is lodged in the rectum. Other symptoms vary depending upon the size of the object, its location, how long it has been in place, and whether or not infection has set in.

## **Diagnosis**

The symptoms are as diverse as the objects and their locations. The most common manifestation of a foreign object anywhere in the body is infection. Even if the object started out sterile, germs still seem to find it and are able to hide from the body's defenses there. Blockage of passageways—breathing, digestive or excretory—is another result. Pain is common.

## **Treatment**

### *Eyes*

Small particles like sand may be removable without medical help, but if the object is not visible or cannot be retrieved, prompt emergency treatment is necessary. Trauma to the eyes can lead to loss of vision and should never be ignored. Before attempting any treatment, the person should move to a well-lighted area where the object can be more easily spotted. Hands should be washed and only clean, preferably sterile, materials should make contact with the eyes. If the particle is small, it can be dislodged by blinking or pulling the upper lid over the lower lid and flushing out the speck. A clean cloth can also be used to pick out the offending particle. Afterwards, the eye should be rinsed with clean, lukewarm water or an ophthalmic wash.

If the foreign object cannot be removed at home, the eye should be lightly covered with sterile gauze to discourage rubbing. A physician will use a strong light and possibly special eyedrops to locate the object. Surgical tweezers can effectively remove many objects. An antibiotic sterile ointment and a patch may be prescribed. If the foreign body has penetrated the deeper layers of the eye, an ophthalmic surgeon will be consulted for emergency treatment.

### *Ears and nose*

A number of ingenious extraction methods have been devised for removing foreign objects from the nose and ears. A bead in a nostril, for example, can be popped

## **KEY TERMS**

**Bronchoscope**—An illuminated instrument that is inserted into the airway to inspect and retrieve objects from the bronchial tubes.

**Conjunctiva**—Mucous membranes around the inner surface of the eyelid.

**Cornea**—The rounded, transparent portion of the eye that covers the pupil and iris and lets light into the interior

**Endoscopy**—The surgical use of long, thin instruments that have both viewing and operating capabilities.

**Heimlich maneuver**—An emergency procedure for removing a foreign object lodged in the airway that is preventing the person from breathing. To perform the Heimlich maneuver on a conscious adult, the rescuer stands behind the victim and encircles his waist. The rescuer makes a fist with one hand and places the other hand on top, positioned below the rib cage and above the waist. The rescuer then applies pressure by a series of upward and inward thrusts to force the foreign object back up the victim's trachea.

out by blowing into the mouth while holding the other nostril closed. Skilled practitioners have removed peas from the ears by tiny improvised corkscrews; marbles by q-tips with super glue. Tweezers often work well, too. Insects can be floated out of the ear by pouring warm (not hot) mineral oil, olive oil, or baby oil into the ear canal. Items that are lodged deep in the ear canal are more difficult to remove because of the possibility of damaging the ear drum. These require emergency treatment from a qualified physician.

### *Airways and stomach*

Mechanical obstruction of the airways, which commonly occurs when food gets lodged in the throat, can be treated by applying the **Heimlich maneuver**. If the object is lodged lower in the airway, a bronchoscope (a special instrument to view the airway and remove obstructions) can be inserted. On other occasions, as when the object is blocking the entrance to the stomach, a fiberoptic endoscope (an illuminated instrument that views the interior of a body cavity) may be used. The physician typically administers a sedative and anesthetizes the throat. The foreign object will then either be pulled out or pushed into the stomach, depending on whether or not the physician

thinks it will pass through the digestive tract on its own. Objects in the digestive tract that are neither irritating, sharp nor large may be followed as they continue on through. Sterile objects that are causing no symptoms may be left in place. Surgical removal of the offending object is necessary if it is causing symptoms.

### **Rectum**

A rectal retractor can remove objects that a physician can feel during **physical examination**. Surgery may be required for objects deeply lodged within the rectum.

### **Prevention**

Using common sense and following safety precautions are the best ways to prevent foreign objects from entering the body. For instance, parents and grandparents should toddler-proof their homes, storing batteries in a locked cabinet and properly disposing of used batteries, so they are not in a location where curious preschoolers can fish them out of a wastebasket. To minimize the chance of youngsters inhaling food, parents should not allow children to eat while walking or playing. Adults should chew food thoroughly and not talk while chewing. Many eye injuries can be prevented by wearing safety glasses while using tools

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For more see **Flesh-eating disease**

47, XXY syndrome see **Klinefelter syndrome**

## **Fracture repair**

### **Definition**

Fracture repair is the process of rejoining and realigning the ends of broken bones. This procedure is usually performed by an orthopedist, general surgeon, or family doctor. In cases of an emergency, first aid measures should be evoked for temporary realignment and **immobilization** until proper medical help is available.

### **Purpose**

Fracture repair is required when there is a need for restoration of the normal position and function of the broken bone. Throughout the stages of fracture healing, the bones must be held firmly in the correct position. In the event the fracture is not properly repaired, malalignment of the bone may occur, resulting in possible physical dysfunction of the bone or joint of that region of the body.

### **Precautions**

Precautions for fracture repair are anything found to be significant with patients' medical diagnosis and history. This would include an individual's tolerance to anesthesia and the presence of bleeding disorders that may be present to complicate surgery.

### **Description**

Fracture repair is applied by means of **traction**, surgery, and/or by immobilization of the bones. The bone fragments are aligned as close as possible to the normal position without injuring the skin. Metal wires or screws may be needed to align smaller bone fragments. Once the

broken ends of the bone are set, the affected area is immobilized for several weeks and kept rigid with a sling, plaster cast, brace or splint. With the use of traction, muscle pull on the fracture site is overcome by weights attached to a series of ropes running over pulleys. Strategically implanted electrical stimulation devices have proven beneficial in healing a fracture site, especially when the fracture is healing poorly and repair by other means is difficult.

## Preparation

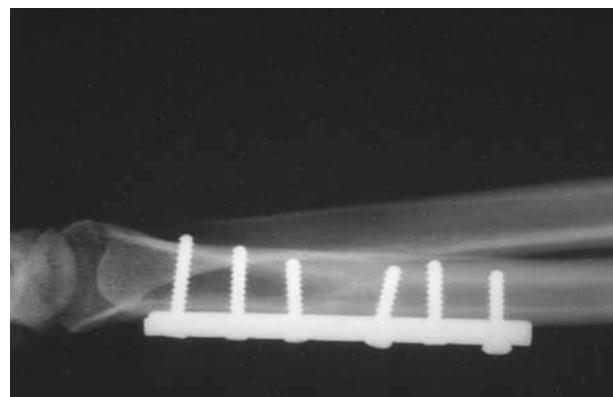
Emergency splinting may be required to immobilize the body part or parts involved. When fracture repair is necessary, the procedure is often performed in a hospital but can also be successfully done in an outpatient surgical facility, doctor's office or emergency room. Before any surgery for fracture repair, blood and urine studies may be taken from the patient. X rays may follow this if not previously acquired. It has been noted however, that not all **fractures** are immediately apparent on an initial x-ray examination. In this case, where a fracture is definitely suspected the extent of the fracture can be properly diagnosed by repeating the x rays 10–14 days later. Depending upon the situation, local or general anesthesia may be used for fracture repair.

## Aftercare

After surgery, x rays may be again taken through the cast or splint to evaluate if rejoined pieces remain in good position for healing. This is usually performed either before the application of the splint or at least before the patient is awakened from the general anesthesia. The patient needs to be cautious not to place excess pressure on any part of the cast until it is completely dry. The patient also should avoid excess pressure on the operative site until complete healing has taken place and the injury has been re-examined by the physician. If the cast becomes exposed to moisture it may soften and require repair. The patient should also be instructed to keep the injured region propped up whenever possible to reduce the possibility of swelling.

## Risks

Surgical risks of fracture repair are greater in patients over 60 years of age because the bones often take longer to heal properly. **Obesity** may place extra stress on the healing site, affecting healing and possibly risking reinjury. **Smoking** may slow the healing process after fracture repair, as well as poor **nutrition**, **alcoholism**, and chronic illness. Some medications may affect the fracture site, causing poor union. Such medications include anti-hypertensives and cortisone.



**An x-ray image of a healing fracture.** (Photograph by Bates, M.D., Custom Medical Stock Photo. Reproduced by permission.)

Possible complications following fracture repair include excessive bleeding, improper fit of joined bone ends, pressure on nearby nerves, delayed healing, and a permanent incomplete healing of the fracture. If there is a poor blood supply to the fractured site with one of the portions of broken bone not properly supplied by the blood, the bony portion will die and healing of the fracture will not take place. This is called aseptic necrosis. Poor immobilization of the fracture from improper casting which permits motion between the bone parts may prevent healing and repair of the bone with possible deformity. Infection can interfere with bone repair. This risk is greater in the case of a compound fracture (a bone fracture causing an open wound) where ideal conditions are present for severe streptococcal and **staphylococcal infections**. Occasionally, fractured bones in the elderly may possibly never heal properly. The risk is increased when nutrition is poor.

## Normal results

Once the procedure for fracture repair is completed, the body begins to produce new tissue to bridge the broken pieces. At first, this tissue (called a callus) is soft and easily injured. Later, the body deposits bone **minerals** until the callus becomes a solid piece of bone. The fracture site is thus strengthened further with extra bone. It usually takes about six weeks for a broken bone to heal together. The exact time required for healing depends on the type of fracture and the extent of damage. Before the use of x rays, fracture repair was not always accurate, resulting in crippling deformities. With modern x-ray technology, the physician can view the extent of the fracture, check the setting following the repair, and be certain after the procedure that the bones have not moved from their intended alignment. Children's bones usually heal relatively rapidly.

## KEY TERMS

**Compound fracture** — A fracture in which the broken end or ends of the bone have torn through the skin. Compound fractures are also known as open fractures

**Staphylococcal infection** — An infection caused by any of several pathogenic species of staphylococcus, commonly characterized by the formation of abscesses of the skin or other organs.

**Streptococcal infection** — An infection caused by a pathogenic bacteria of one of several species of the genus streptococcus or their toxins. Almost any organ in the body may be involved.

### Abnormal results

Abnormal results of fracture repair include damage to nearby nerves or primary blood vessels. Improper alignment causing deformity is also an abnormal outcome, however, with today's medical technology it is relatively rare.

### Resources

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#### OTHER

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## Fractures

### Definition

A fracture is a complete or incomplete break in a bone resulting from the application of excessive force.

### Description

A fracture usually results from traumatic injury to bones causing the continuity of bone tissues or bony cartilage to be disrupted or broken. Fracture classifications include simple, compound, incomplete and complete. Simple fractures (more recently called "closed") are not obvious as the skin has not been ruptured and remains intact. Compound fractures (now commonly called "open") break the skin, exposing bone and causing additional soft tissue injury and possible infection. A single fracture means that one fracture only has occurred and multiple fractures refer to more than one fracture occurring in the same bone. Fractures are termed complete if the break is completely through the bone and described as incomplete or "greenstick" if the fracture occurs partly across a bone shaft. This latter type of fracture is often the result of bending or crushing forces applied to a bone.

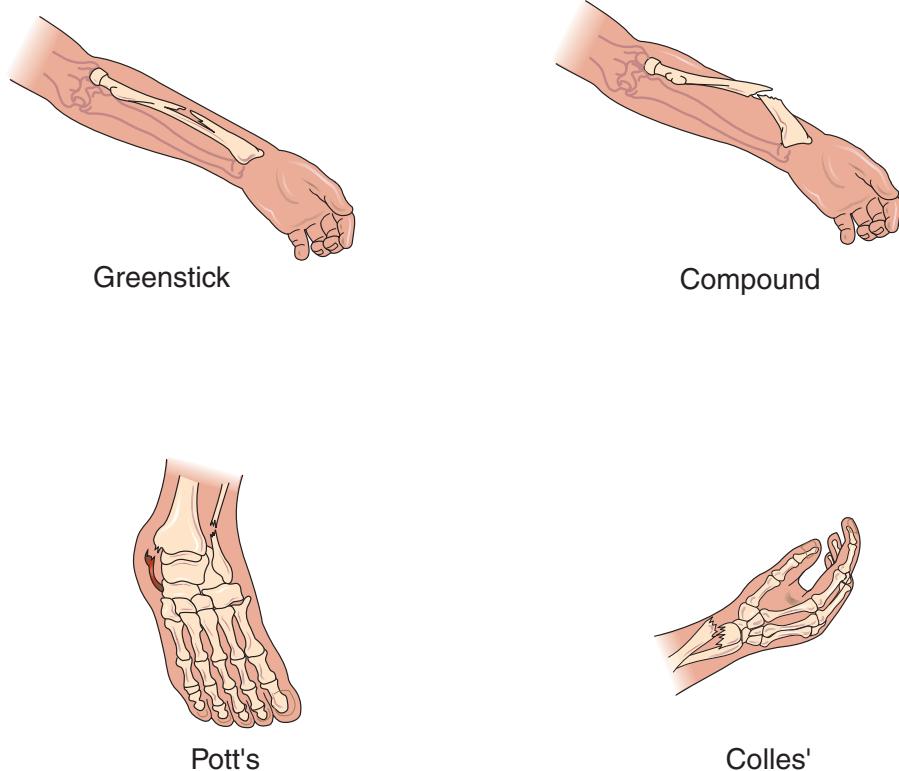
Fractures are also named according to the specific part of the bone involved and the nature of the break. Identification of a fracture line can further classify fractures. Types include linear, oblique, transverse, longitudinal, and spiral fractures. Fractures can be further subdivided by the positions of bony fragments and are described as comminuted, non-displaced, impacted, overriding, angulated, displaced, avulsed, and segmental. Additionally, an injury may be classified as a fracture-dislocation when a fracture involves the bony structures of any joint with associated dislocation of the same joint.

### Fractures line identification

Linear fractures have a break that runs parallel to the bone's main axis or in the direction of the bone's shaft. For example, a linear fracture of the arm bone could extend the entire length of the bone. Oblique and transverse fractures differ in that an oblique fracture crosses a bone at approximately a 45° angle to the bone's axis. In contrast, a transverse fracture crosses a bone's axis at a 90° angle. A longitudinal fracture is similar to a linear fracture. Its fracture line extends along the shaft but is more irregular in shape and does not run parallel to the bone's axis. Spiral fractures are described as crossing a bone at an oblique angle, creating a spiral pattern. This break usually occurs in the long bones of the body such as the upper arm bone (humerus) or the thigh bone (femur).

### Bony fragment position identification

Comminuted fractures have two or more fragments broken into small pieces, in addition to the upper and lower halves of a fractured bone. Fragments of bone that maintain their normal alignment following a fracture are described as being non-displaced. An impacted fracture is characterized as a bone fragment forced into or onto



Fractures usually result from a traumatic injury to a bone where the continuity of bone tissues or bony cartilage is disrupted or broken. The illustrations above feature common sites where fractures occur. (Illustration by Electronic Illustrators Group.)

another fragment resulting from a compressive force. Overriding is a term used to describe bony fragments that overlap and shorten the total length of a bone. Angulated fragments result in pieces of bone being at angles to each other. A displaced bony fragment occurs from disruption of normal bone alignment with deformity of these segments separate from one another. An avulsed fragment occurs when bone fragments are pulled from their normal position by forceful muscle contractions or resistance from ligaments. Segmental fragmented positioning occurs if fractures in two adjacent areas occur, leaving an isolated central segment. An example of segmental alignment is when the arm bone fractures in two separate places, with displacement of the middle section of bone.

### Causes and symptoms

Individuals with high activity levels appear to be at greater risk for fractures. This group includes children and athletes participating in contact sports. Because of an increase in bone brittleness with **aging**, elderly persons are also included in this high-risk population. Up to the

age of 50, more men suffer from fractures than women due to occupational hazards. However, after the age of 50, women are more prone to fractures than men. Specific diseases causing an increased risk for fractures include Paget's disease, rickets, **osteogenesis imperfecta**, **osteoporosis**, bone **cancer** and tumors, and prolonged disuse of a nonfunctional body part such as after a **stroke**.

Symptoms of fractures usually begin with **pain** that increases with attempted movement or use of the area and swelling at the involved site. The skin in the area may be pale and an obvious deformity may be present. In more severe cases, there may be a loss of pulse below the fracture site, such as in the extremities, accompanied by numbness, tingling, or **paralysis** below the fracture. An open or compound fracture is often accompanied by bleeding or bruising. If the lower limbs or pelvis are fractured, pain and resistance to movement usually accompany the injury causing difficulty with weight bearing.

### Diagnosis

Diagnosis begins immediately with an individual's own observation of symptoms. A thorough medical his-

## KEY TERMS

**Avulsion fracture**—A fracture caused by the tearing away of a fragment of bone where a strong ligament or tendon attachment forcibly pulls the fragment away from the bone tissue.

**Axis**—A line that passes through the center of the body or body part.

**Comminuted fracture**—A fracture where there are several breaks in a bone creating numerous fragments.

**Compartment syndrome**—Compartment syndrome is a condition in which a muscle swells but is constricted by the connective tissue around it, which cuts off blood supply to the muscle.

**Contrast hydrotherapy**—A series of hot and cold water applications. A hot compress (as hot as an individual can tolerate) is applied for three minutes followed by an ice cold compress for 30 seconds. These applications are repeated three times each and ending with the cold compress.

**Osteogenesis imperfecta**—A genetic disorder involving defective development of connective tissues, characterized by brittle and fragile bones that are easily fractured by the slightest trauma.

**Osteoporosis**—Literally meaning “porous bones,” this condition occurs when bones lose an excessive amount of their protein and mineral content, partic-

ularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

**Paget's disease**—Chronic disorder of unknown cause, usually affecting middle aged and elderly people, characterized by enlarged and deformed bones. Excessive breakdown and formation of bone tissue occurs with Paget's disease and can cause bone to weaken, resulting in bone pain, arthritis, deformities, and fractures.

**Reduction**—The restoration of a body part to its original position after displacement, such as the reduction of a fractured bone by bringing ends or fragments back into original alignment. The use of local or general anesthesia usually accompanies a fracture reduction. If performed by outside manipulation only, the reduction is described as closed; if surgery is necessary, it is described as open.

**Rickets**—A condition caused by the dietary deficiency of vitamin D, calcium, and usually phosphorus, seen primarily in infancy and childhood, and characterized by abnormal bone formation.

**Traction**—The process of placing a bone, limb, or group of muscles under tension by applying weights and pulleys. The goal is to realign or immobilize the part or to relieve pressure on that particular area to promote healing and restore function.

tory and physical exam by a physician often reveals the presence of a fracture. An x ray of the injured area is the most common test used to determine the presence of a bone fracture. Any x ray series performed involves at least two views of the area to confirm the presence of the fracture because not all fractures are apparent on a single x-ray. Some fractures are often difficult to see and may require several views at different angles to see clear fracture lines. In some cases, CT, MRI or other imaging tests are required to demonstrate fracture. Sometimes, especially with children, the initial x ray may not show any fractures but repeat x rays seven to 14 days later may show changes in the bone(s) of the affected area. If a fracture is open and occurs in conjunction with soft tissue injury, further laboratory studies are often conducted to determine if blood loss has occurred.

In the event of exercise-related **stress** fractures (micro-fractures due to excessive stress), a tuning fork can provide a simple, inexpensive test. The tuning fork is

a metal instrument with a stem and two prongs that vibrate when struck. If an individual has increased pain when the tuning fork is placed on a bone, such as the tibia or shinbone, the likelihood of a stress fracture is high. Bone scans also are helpful in detecting stress fractures. In this diagnostic procedure, a radioactive tracer is injected into the bloodstream and images are taken of specific areas or the entire skeleton by CT or MRI.

### Treatment

Treatment depends on the type of fracture, its severity, the individual's age and general health. The first priority in treating any fracture is to address the entire medical status of the patient. Medical personnel are trained not allow a painful, deformed limb to distract them from potentially life-threatening injury elsewhere or **shock**. If an open fracture is accompanied by serious soft tissue injury, it may be necessary to control bleeding and the shock that can accompany loss of blood.

First aid is the appropriate initial treatment in emergency situations. It includes proper splinting, control of blood loss, and monitoring vital signs such as breathing and circulation.

### **Immobilization**

**Immobilization** of a fracture site can be done internally or externally. The primary goal of immobilization is to maintain the realignment of a bone long enough for healing to start and progress. Immobilization by external fixation uses splints, casts, or braces. This may be the primary and only procedure for fracture treatment. Splinting to immobilize a fracture can be done with or without **traction**. In emergency situations if the injured individual must be moved by someone other than a trained medical person, splinting is a useful form of fracture management. It should be done without causing additional pain and without moving the bone segments. In a clinical environment, plaster of Paris casts are used for immobilization. Braces are useful as they often allow movement above and below a fracture site. Treatments for stress fractures include rest and decreasing or stopping any activity that causes or increases pain.

### **Fracture reduction**

Fracture reduction is the procedure by which a fractured bone is realigned in normal position. It can be either closed or open. Closed reduction refers to realigning bones without breaking the skin. It is performed with manual manipulation and/or traction and is commonly done with some kind of anesthetic. Open reduction primarily refers to surgery that is performed to realign bones or fragments. Fractures with little or no displacement may not require any form of reduction.

Traction is used to help reposition a broken bone. It works by applying pressure to restore proper alignment. The traction device immobilizes the area and maintains realignment as the bone heals. A fractured bone is immobilized by applying opposing force at both ends of the injured area, using an equal amount of traction and countertraction. Weights provide the traction pull needed or the pull is achieved by positioning the individual's body weight appropriately. Traction is a form of closed reduction and is sometimes used as an alternative to surgery. Since it restricts movement of the affected limb or body part, it may confine a person to bed rest for an extended period of time.

A person may need open reduction if there is an open, severe, or comminuted fracture. This procedure allows a physician to examine and surgically correct associated soft tissue damage while reducing the fracture and, if necessary, applying internal or external devices. Internal fixa-

tion involves the use of metallic devices inserted into or through bone to hold the fracture in a set position and alignment while it heals. Devices include plates, nails, screws, and rods. When healing is complete, the surgeon may or may not remove these devices. Virtually any hip fracture requires open reduction and internal fixation so that the bone will be able to support the patient's weight.

### **Alternative treatment**

In addition to the importance of calcium for strong bones, many alternative treatment approaches recommend use of mineral supplements to help build and maintain a healthy, resilient skeleton. Some physical therapists use electro-stimulation over a fractured site to promote and expedite healing. Chinese traditional medicine may be helpful by working to reconnect chi through the meridian lines along the line of a fracture. **Homeopathy** can enhance the body's healing process. Two particularly useful homeopathic remedies are *Arnica* (*Arnica montana*) and *Symphytum* (*Symphytum officinalis*). If possible, applying contrast **hydrotherapy** to an extremity (e.g., a hand or foot) of a fractured area can assist healing by enhancing circulation.

### **Prognosis**

Fractures involving joint surfaces almost always lead to some degree of arthritis of the joint. Fractures can normally be cured with proper first aid and appropriate after-care. If determined necessary by a physician, the fractured site should be manipulated, realigned, and immobilized as soon as possible. Realignment has been shown to be much more difficult after six hours. Healing time varies from person to person with the elderly generally needing more time to heal completely. A non-union fracture may result when a fracture does not heal, such as in the case of an elderly person or an individual with medical complications. Recovery is complete when there is no bone motion at the fracture site, and x rays indicate complete healing. Open fractures may lead to bone infections, which delay the healing process. Another possible complication is compartment syndrome, a painful condition resulting from the expansion of enclosed tissue and that may occur when a body part is immobilized in a cast.

### **Prevention**

Adequate calcium intake is necessary for strong bones and can help decrease the risk of fractures. People who do not get enough calcium in their **diets** can take a calcium supplement. **Exercise** can help strengthen bones by increasing bone density, thereby decreasing the risk of fractures from falls. A University of Southern California study reported that older people who exercised one or

more hours per day had approximately half the incidence of hip fractures as those who exercised fewer than 30 minutes per day or not at all.

Fractures can be prevented if safety measures are taken seriously. These measures include using seat belts in cars and encouraging children to wear protective sports gear. Estrogen replacement for women past the age of 50 has been shown to help prevent osteoporosis and the fractures that may result from this condition. In one study, elderly women on estrogen replacement therapy demonstrated the lowest occurrence of hip fractures when compared to similar women not on estrogen replacement therapy.

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 American College of Sports Medicine, 401 W. Michigan St., Indianapolis, IN 46202. (317) 637-9200, Fax: (317) 634-7817.  
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L. Fleming Fallon, Jr., MD, DrPH

## Fragile X syndrome

### Definition

Fragile X syndrome is the most common form of inherited **mental retardation**. Individuals with this condition have developmental delay, variable levels of mental retardation, and behavioral and emotional difficulties. They may also have characteristic physical traits. Generally, males are affected with moderate mental retardation and females with mild mental retardation.

### Description

Fragile X syndrome is also known as Martin-Bell syndrome, Marker X syndrome, and FRAXA syndrome. It is the most common form of inherited mental retardation. Fragile X syndrome is caused by a mutation in the FMR-1 gene, located on the X chromosome. The role of the gene is unclear, but it is probably important in early development.

In order to understand fragile X syndrome it is important to understand how human genes and chromosomes influence this condition. Normally, each cell in the body contains 46 (23 pairs of) chromosomes. These chromosomes consist of genetic material (DNA) needed for the production of proteins, which lead to growth, development, and physical/intellectual characteristics. The first 22 pairs of chromosomes are the same in males and females. The remaining two chromosomes are called the sex chromosomes (X and Y). The sex chromosomes determine whether a person is male or female. Males have only one X chromosome, which is inherited from the mother at conception, and they receive a Y chromosome from the father. Females inherit two X chromosomes, one from each parent. Fragile X syndrome is caused by a mutation in a gene called FMR-1. This gene is located on the X chromosome. The FMR-1 gene is thought to play an important role in the development of the brain, but the exact way that the gene acts in the body is not fully understood.

Fragile X syndrome affects males and females of all ethnic groups. It is estimated that there are about one in 4,000 to one in 6,250 males affected with fragile X syndrome. There are approximately half as many females with fragile X syndrome as there are males. The carrier frequency in unaffected females is one in 100 to one in 600, with one study finding a carrier frequency of one in 250.

### Causes and symptoms

For reasons not fully understood, the CGG sequence in the FMR-1 gene can expand to contain between 54 and 230 repeats. This stage of expansion is called a pre-

## KEY TERMS

**Amniocentesis**—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman’s abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

**CGG or CGG sequence**—Shorthand for the DNA sequence: cytosine-guanine-guanine. Cytosine and guanine are two of the four molecules, otherwise called nucleic acids, that make up DNA.

**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.

**FMR-1 gene**—A gene found on the X chromosome. Its exact purpose is unknown, but it is suspected that the gene plays a role in brain development.

**Mitral valve prolapse**—A heart defect in which one of the valves of the heart (which normally controls blood flow) becomes floppy. Mitral valve prolapse may be detected as a heart murmur but there are usually no symptoms.

**Premutation**—A change in a gene that precedes a mutation; this change does not alter the function of the gene.

**X chromosome**—One of the two sex chromosomes (the other is Y) containing genetic material that, among other things, determine a person’s gender.

mutation. People who carry a premutation do not usually have symptoms of fragile X syndrome; although there have been reports of individuals with a premutation and subtle intellectual or behavioral symptoms. Individuals who carry a fragile X premutation are at risk to have children or grandchildren with the condition. Female premutation carriers may also be at increased risk for earlier onset of **menopause**; however, premutation carriers may exist through several generations of a family and no symptoms of fragile X syndrome will appear.

The size of the premutation can expand over succeeding generations. Once the size of the premutation exceeds 230 repeats, it becomes a full mutation and the FMR-1 gene is disabled. Individuals who carry the full mutation may have fragile X syndrome. Since the FMR-1 gene is located on the X chromosome, males are more likely to develop symptoms than females. This is because males have only one copy of the X chromosome. Males who inherit the full mutation are expected to have mental impairment. A female’s normal X chromosome may compensate for her chromosome with the fragile X gene mutation. Females who inherit the full mutation have an approximately 50% risk of mental impairment. The phenomenon of an expanding trinucleotide repeat in successive generations is called anticipation. Another unique

aspect fragile X syndrome is that mosaicism is present in 15–20% those affected by the condition. Mosaicism is when there is the presence of cells of two different genetic materials in the same individual.

The mutation involves a short sequence of DNA in the FMR-1 gene. This sequence is designated CGG. Normally, the CGG sequence is repeated between six to 54 times. People who have repeats in this range do not have fragile X syndrome and are not at increased risk to have children with fragile X syndrome. Those affected by fragile X syndrome have expanded CGG repeats (over 200) in the first exon of the FMR1 gene (the full mutation).

Fragile X syndrome inherited in an X-linked dominant manner (characters are transmitted by genes on the X chromosome). When a man carries a premutation on his X chromosome, it tends to be stable and usually will not expand if he passes it on to his daughters (he passes his Y chromosome to his sons). Thus, all of his daughters will be premutation carriers like he is. When a woman carries a premutation, it is unstable and can expand as she passes it on to her children, therefore a man’s grandchildren are at greater risk of developing the syndrome. There is a 50% risk for a premutation carrier female to transmit an abnormal mutation with each **pregnancy**. The likelihood for the premutation to expand is related to the number of

repeats present; the higher the number of repeats, the greater the chance that the premutation will expand to a full mutation in the next generation. All mothers of a child with a full mutation are carriers of an FMR-1 gene expansion. Ninety-nine percent of patients with fragile X syndrome have a CGG expansion, and less than one percent have a point mutation or deletion on the FMR1 gene.

Individuals with fragile X syndrome appear normal at birth but their development is delayed. Most boys with fragile X syndrome have mental impairment. The severity of mental impairment ranges from learning disabilities to severe mental retardation. Behavioral problems include attention deficit and hyperactivity at a young age. Some may show aggressive behavior in adulthood. Short attention span, poor eye contact, delayed and disordered speech and language, emotional instability, and unusual hand mannerisms (hand flapping or hand biting) are also seen frequently. Characteristic physical traits appear later in childhood. These traits include a long and narrow face, prominent jaw, large ears, and enlarged testes. In females who carry a full mutation, the physical and behavioral features and mental retardation tend to be less severe. About 50% of females who have a full mutation are mentally retarded. Other behavioral characteristics include whirling, spinning, and occasionally **autism**.

Children with fragile X syndrome often have frequent ear and sinus infections. Nearsightedness and lazy eye are also common. Many babies with fragile X syndrome may have trouble with sucking and some experience digestive disorders that cause frequent gagging and vomiting. A small percentage of children with fragile X syndrome may experience seizures. Children with fragile X syndrome also tend to have loose joints which may result in joint dislocations. Some children develop a curvature in the spine, flat feet, and a heart condition known as **mitral valve prolapse**.

## Diagnosis

Any child with signs of developmental delay of speech, language, or motor development with no known cause should be considered for fragile X testing, especially if there is a family history of the condition. Behavioral and developmental problems may indicate fragile X syndrome, particularly if there is a family history of mental retardation. Definitive identification of the fragile X syndrome is made by means of a genetic test to assess the number of CGG sequence repeats in the FMR-1 gene. Individuals with the premutation or full mutation may be identified through **genetic testing**. Genetic testing for the fragile X mutation can be done on the developing baby before birth through **amniocentesis** or **chorionic villus sampling (CVS)**, and is 99% effective in

detecting the condition due to trinucleotide repeat expansion. Prenatal testing should only be undertaken after the fragile X carrier status of the parents has been confirmed and the couple has been counseled regarding the risks of recurrence. While prenatal testing is possible to do with CVS, the results can be difficult to interpret and additional testing may be required.

## Treatment

Presently there is no cure for fragile X syndrome. Management includes such approaches as speech therapy, occupational therapy, and physical therapy. The expertise of psychologists, special education teachers, and genetic counselors may also be beneficial. Drugs may be used to treat hyperactivity, seizures, and other problems. Establishing a regular routine, avoiding over stimulation, and using calming techniques may also help in the management of behavioral problems. Children with a troubled heart valve may need to see a heart specialist and take medications before surgery or dental procedures. Children with frequent ear and sinus infections may need to take medications or have special tubes placed in their ears to drain excess fluid. Mainstreaming of children with fragile X syndrome into regular classrooms is encouraged because they do well imitating behavior. Peer tutoring and positive reinforcement are also encouraged.

## Prognosis

Early diagnosis and intensive intervention offer the best prognosis for individuals with fragile X syndrome. Adults with fragile X syndrome may benefit from vocational training and may need to live in a supervised setting. Life span is typically normal.

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Arc of the United States (formerly Association for Retarded Citizens of the US). 500 East Border St., Suite 300, Arlington, TX 76010. (817) 261-6003. <<http://thearc.org>>.

National Fragile X Foundation. PO Box 190488, San Francisco, CA 94119-0988. (800) 688-8765 or (510) 763-6030. Fax: (510) 763-6223. natlfx@sprintmail.com. <<http://nfxf.org>>. National Fragile X Syndrome Support Group. 206 Sherman Rd., Glenview, IL 60025. (708) 724-8626.

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Frambesia see **Yaws**

*Francisella tularensis* infection see **Tularemia**

Fresh cell therapy see **Cell therapy**

## Friedreich's ataxia

### Definition

Friedreich's ataxia (FA) is an inherited, progressive nervous system disorder causing loss of balance and coordination.

### Description

Ataxia is a condition marked by impaired coordination. Friedreich's ataxia is the most common inherited ataxia, affecting between 3,000–5,000 people in the United States. FA is an autosomal recessive disease, which means that two defective gene copies must be inherited to develop symptoms, one from each parent. A person with only one defective gene copy will not show signs of FA, but may pass along the gene to offspring. Couples with one child affected by FA have a 25% chance in each pregnancy of conceiving another affected child.

### Causes and symptoms

#### Causes

The gene for FA codes for a protein called frataxin. Normal frataxin is found in the cellular energy structures known as mitochondria, where it is thought to be involved in regulating the transport of iron. In FA, the frataxin gene on chromosome 9 is expanded with non-sense information known as a “triple repeat.” This extra DNA interferes with normal production of frataxin,

thereby impairing iron transport. Normally, there are 10–21 repeats of the frataxin gene. In FA, this sequence may be repeated between 200–900 times. The types of symptoms and severity of FA seems to be associated with the number of repetitions. Patients with more copies have more severe symptomatology. Researchers are still wrestling with how frataxin and the repeats on chromosome 9 are involved in causing FA. One theory suggests that FA develops in part because defects in iron transport prevent efficient use of cellular energy supplies.

The nerve cells most affected by FA are those in the spinal cord involved in relaying information between muscles and the brain. Tight control of movement requires complex feedback between the muscles promoting a movement, those restraining it, and the brain. Without this control, movements become uncoordinated, jerky, and inappropriate to the desired action.

#### Symptoms

Symptoms of FA usually first appear between the ages of 8 and 15, although onset as early as 18 months or as late as age 25 is possible. The first symptom is usually gait incoordination. A child with FA may graze doorways when passing through, for instance, or trip over low obstacles. Unsteadiness when standing still and deterioration of position sense is common. Foot deformities and walking up off the heels often results from uneven muscle weakness in the legs. **Muscle spasms and cramps** may occur, especially at night.

Ataxia in the arms follows, usually within several years, leading to decreased hand-eye coordination. Arm weakness does not usually occur until much later. Speech and swallowing difficulties are common. **Diabetes mellitus** may also occur. **Nystagmus**, or eye tremor, is common, along with some loss of visual acuity. **Hearing loss** may also occur. A side-to-side curvature of the spine (**scoliosis**) occurs in many cases, and may become severe.

Heartbeat abnormalities occur in about two thirds of FA patients, leading to **shortness of breath** after exertion, swelling in the lower limbs, and frequent complaints of cold feet.

### Diagnosis

Diagnosis of FA involves a careful medical history and thorough neurological exam. Lab tests include **electromyography**, an electrical test of muscle, and a nerve conduction velocity test. An electrocardiogram may be performed to diagnose heart arrhythmia.

Direct DNA testing is available, allowing FA to be more easily distinguished from other types of ataxia. The same test may be used to determine the presence of the genetic defect in unaffected individuals, such as siblings.

## KEY TERMS

**Ataxia**—A condition marked by impaired coordination.

**Scoliosis**—An abnormal, side-to-side curvature of the spine.

### Treatment

There is no cure for FA, nor any treatment that can slow its progress. Amantadine may provide some limited improvement in ataxic symptoms, but is not recommended in patients with cardiac abnormalities. Physical and occupational therapy are used to maintain range of motion in weakened muscles, and to design adaptive techniques and devices to compensate for loss of coordination and strength. Some patients find that using weights on the arms can help dampen the worst of the uncoordinated arm movements.

Heart **arrhythmias** and diabetes are treated with drugs specific to those conditions.

### Prognosis

The rate of progression of FA is highly variable. Most patients lose the ability to walk within 15 years of symptom onset, and 95% require a wheelchair for mobility by age 45. Reduction in lifespan from FA complications is also quite variable. Average age at **death** is in the mid-thirties, but may be as late as the mid-sixties. As of mid-1998, the particular length of the triple repeat has not been correlated strongly enough with disease progression to allow prediction of the course of the disease on this basis.

### Prevention

There is no way to prevent development of FA in a person carrying two defective gene copies.

### Resources

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#### ORGANIZATIONS

Muscular Dystrophy Association. 3300 East Sunrise Drive, Tucson, AZ 85718. (520) 529-2000 or (800) 572-1717. <<http://www.mdausa.org>>.

Rosalyn Carson-DeWitt

## Frostbite and frostnip

### Definition

Frostbite is the term for damage to the skin and other tissues caused by freezing. Frostnip is a mild form of cold injury.

### Description

In North America, frostbite is largely confined to Alaska, Canada, and the northern states. Recent years have witnessed a substantial decline in the number of cases, probably for several reasons, including better winter clothing and footwear and greater public understanding of how to avoid cold-weather dangers. At the same time, the nature of the at-risk population has changed as rising numbers of homeless people have made frostbite an urban as well as a rural public health concern. The growing popularity of outdoor winter activities has also expanded the at-risk population.

### Causes and symptoms

#### Frostbite

Skin exposed to temperatures a little below the freezing mark can take hours to freeze, but very cold skin can freeze in minutes or seconds. Air temperature, wind speed, and moisture all affect how cold the skin becomes. A strong wind can lower skin temperature considerably by dispersing the thin protective layer of warm air that surrounds our bodies. Wet clothing readily draws heat away from the skin because water is a potent conductor of heat. The evaporation of moisture on the skin also produces cooling. For these reasons, wet skin or clothing on a windy day can lead to frostbite even if the air temperature is above the freezing mark.

The extent of permanent injury, however, is determined not by how cold the skin and the underlying tissues become but by how long they remain frozen. Consequently, homeless people and others whose self-preservation instincts may be clouded by alcohol or psychiatric illness face a greater risk of frostbite-related **amputation** because they are more likely to stay out in the cold when prudence dictates seeking shelter or medical attention. Alcohol also affects blood circulation in the extremities in a way that can increase the severity of injury (as does **smoking**). A review of 125 Saskatchewan frostbite cases found a tie to alcohol in 46% and to psychiatric illness in 17%. Other risk factors identified by researchers include inadequate clothing, previous cold injury, **fatigue**, wound infection, **atherosclerosis** (an arterial disease), and diabetes. Driving in poor weather can also be dan-

gerous: vehicular failure was a predisposing factor in 15% of the Saskatchewan cases.

Three nearly simultaneous physiological processes underlie frostbite injury: tissue freezing, tissue hypoxia, and the release of inflammatory mediators. Tissue freezing causes ice crystal formation and other changes that damage and eventually kill cells. Much of this harm occurs because the ice produces pressure changes that cause water (crucial for cell survival) to flow out of the cells. Tissue hypoxia (oxygen deficiency) occurs when the blood vessels in the hands, feet, and other extremities narrow in response to cold. Among its many tasks, blood transfers body heat to the skin, which then dissipates the heat into the environment. Blood vessel narrowing is the body's way of protecting vital internal organs at the expense of the extremities by reducing heat flow away from the core. However, blood also carries life-sustaining oxygen to the skin and other tissues, and narrowed vessels result in oxygen **starvation**. Narrowing also causes acidosis (an increase in tissue acidity) and increases blood viscosity (thickness). Ultimately, blood stops flowing through the capillaries (the tiny blood vessels that connect the arteries and veins) and blood clots form in the arterioles and venules (the smallest arteries and veins). Damage also occurs to the endothelial cells that line the blood vessels. Hypoxia, blood clots, and endothelial damage lead, in turn, to the release of inflammatory mediators (substances that act as links in the inflammatory process), which promote further endothelial damage, hypoxia, and cell destruction.

Frostbite is classified by degree of injury (first, second, third, or fourth), or simply divided into two types, superficial (corresponding to first- or second-degree injury) and deep (corresponding to third- or fourth-degree injury). Most frostbite injuries affect the feet or hands. The remaining 10% of cases typically involve the ears, nose, cheeks, or penis. Once frostbite sets in, the affected part begins to feel cold and, usually, numb; this is followed by a feeling of clumsiness. The skin turns white or yellowish. Many patients experience severe **pain** in the affected part during rewarming treatment and an intense throbbing pain that arises two or three days later and can last days or weeks. As the skin begins to thaw during treatment, **edema** (excess tissue fluid) often accumulates, causing swelling. In second- and higher-degree frostbite, blisters appear. Third-degree cases produce deep, blood-filled blisters and, during the second week, a hard black eschar (scab). Fourth-degree frostbite penetrates below the skin to the muscles, tendons, nerves, and bones. In severe cases of frostbite the dead tissue can mummify and drop off. Infection is also a possibility.

### **Frotnip**

Like frostbite, frotnip is associated with ice crystal formation in the tissues, but no tissue destruction occurs and



**A human hand with frostbite.** (Photo Researchers, Inc. Reproduced by permission.)

the crystals dissolve as soon as the skin is warmed. Frotnip affects areas such as the earlobes, cheeks, nose, fingers, and toes. The skin turns pale and one experiences numbness or tingling in the affected part until warming begins.

### **Diagnosis**

Frostbite diagnosis relies on a **physical examination** and may also include conventional radiography (x rays), **angiography** (x-ray examination of the blood vessels using an injected dye to provide contrast), thermography (use of a heat-sensitive device for measuring blood flow), and other techniques for predicting the course of injury and identifying tissue that requires surgical removal. During the initial treatment period, however, a physician cannot judge how a case will progress. Diagnostic tests only become useful three to five days after rewarming, once the blood vessels have stabilized.

### **Treatment**

#### **Frostbite**

Emergency medical help should always be summoned whenever frostbite is suspected. While waiting for help to arrive, one should, if possible, remove wet or tight clothing and put on dry, loose clothing or wraps. A splint and padding are used to protect the injured area. Rubbing the area with snow or anything else is dangerous. The key to prehospital treatment is to avoid partial thawing and refreezing, which releases more inflammatory mediators and makes the injury substantially worse. For this reason, the affected part must be kept away from heat sources such as campfires and car heaters. Experts advise rewarming in the field only when emergency help will take more than two hours to arrive and refreezing can be prevented.

Because the outcome of a frostbite injury cannot be predicted at first, all hospital treatment follows the same

route. Treatment begins by rewarming the affected part for 15–30 minutes in water at a temperature of 104–108°F (40–42.2°C). This rapid rewarming halts ice crystal formation and dilates narrowed blood vessels. Aloe vera (which acts against inflammatory mediators) is applied to the affected part, which is then splinted, elevated, and wrapped in a dressing. Depending on the extent of injury, blisters may be debrided (cleaned by removing foreign material) or simply covered with aloe vera. A **tetanus** shot and, possibly, penicillin, are used to prevent infection, and the patient is given ibuprofen to combat inflammation. Narcotics are needed in most cases to reduce the excruciating pain that occurs as sensation returns during rewarming. Except when injury is minimal, treatment generally requires a hospital stay of several days, during which **hydrotherapy** and physical therapy are used to restore the affected part to health. Experts recommend a cautious approach to tissue removal, and advise that 22–45 days must pass before a decision on amputation can safely be made.

### Frostnip

Frostsinned fingers are helped by blowing warm air on them or holding them under one's armpits. Other frostsinned areas can be covered with warm hands. The injured areas should never be rubbed.

### Alternative treatment

Alternative practitioners suggest several kinds of treatment to speed recovery from frostbite after leaving the hospital. Bathing the affected part in warm water or using contrast hydrotherapy can help enhance circulation. Contrast hydrotherapy involves a series of hot and cold water applications. A hot compress (as hot as the patient can stand) is applied to the affected area for three minutes followed by an ice cold compress for 30 seconds. These applications are repeated three times each, ending with the cold compress. Nutritional therapy to promote tissue growth in damaged areas may also be helpful. Homeopathic and botanical therapies may also assist recovery from frostbite. Homeopathic *Hypericum (Hypericum perforatum)* is recommended when nerve endings are affected (especially in the fingers and toes) and *Arnica (Arnica montana)* is prescribed for **shock**. Cayenne pepper (*Capsicum frutescens*) can enhance circulation and relieve pain. Drinking hot ginger (*Zingiber officinale*) tea also aids circulation. Other possible approaches include **acupuncture** to avoid permanent nerve damage and oxygen therapy.

### Prognosis

The rapid rewarming approach to frostbite treatment, pioneered in the 1980s, has proved to be much more

effective than older methods in preventing tissue loss and amputation. A study of 56 first-, second-, and third-degree frostbite patients treated with rapid rewarming in 1982–85 found that 68% recovered without tissue loss, 25% experienced some tissue loss, and 7% needed amputation. In a comparison group of 98 patients, treatment using older methods resulted in a tissue loss rate of nearly 35% and an amputation rate of nearly 33%. Although the comparison group included a higher proportion of second- and third-degree cases, the difference in treatment results was determined to be statistically significant.

The extreme throbbing pain that many frostbite sufferers endure for days or weeks after rewarming is not the only prolonged symptom of frostbite. During the first weeks or months, people often experience tingling, a burning sensation, or a sensation resembling shocks from an electric current. Other possible consequences of frostbite include skin—color changes, nail deformation or loss, joint stiffness and pain, **hyperhidrosis** (excessive sweating), and heightened sensitivity to cold. For everyone, a degree of sensory loss lasting at least four years—and sometimes a lifetime—is inevitable.

### Prevention

With the appropriate knowledge and precautions, frostbite can be prevented even in the coldest and most challenging environments. Appropriate clothing and footwear are essential. To prevent heat loss and keep the blood circulating properly, clothing should be worn loosely and in layers. Covering the hands, feet, and head is also crucial for preventing heat loss. Outer garments need to be wind and water resistant, and wet clothing and footwear must be replaced as quickly as possible. Alcohol and drugs should be avoided because of their harmful effects on judgment and reasoning. Experts also warn against alcohol use and smoking in the cold because of the circulatory changes they produce. Paying close attention to the weather report before venturing outdoors and avoiding unnecessary risks such as driving in isolated areas during a blizzard are also important.

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Howard Baker

Frostnip see **Frostbite and frostnip**

FSH test see **Follicle-stimulating hormone test**

Fugue see **Dissociative disorders**

FUO see **Fever of unknown origin**

Furosemide see **Diuretics**

Furunculosis see **Boils**

Fusobacterium infection see **Anaerobic infections**